**TÍTULO / TITLE:** - A Phase II multicenter, open-label, clinical and pharmokinetic trial of PM00104 in patients with advanced Ewing Family of Tumors.

**RESUMEN / SUMMARY:** - A Phase II multicenter, open-label, clinical and pharmokinetic trial of PM00104 in patients with advanced Ewing Family of Tumors.

**REVISTA / JOURNAL:** - Invest New Drugs. 2013 Nov 1.

**AUTORES / AUTHORS:** - Jones RL; Ferrari S; Blay JY; Navid F; Lardelli P; Alfaro V; Siguero M; Soman N; Chawla SP

**INSTITUCIÓN / INSTITUTION:** - University of Washington/Fred Hutchinson Cancer Research Center, Seattle, WA, USA, rjones@seattlecca.org.

**RESUMEN / SUMMARY:** - Ewing sarcoma is a rare connective tissue tumor characterized by the translocation of the EWS gene, mainly between chromosome 11 and 22, giving rise to gene re-arrangements between the EWS gene and various members of the ETS gene family. Multi-agent chemotherapy has improved the outcome for patients with localized Ewing sarcoma, but survival of patients with recurrent/metastatic disease remains poor. An exploratory two-stage, single-arm Phase II multicenter trial of the synthetic alkaloid, PM00104, was conducted in patients with recurrent Ewing sarcoma. The primary end point of the trial was objective response rate. PM00104 was administered at a dose of 2 mg/m2 on Days 1, 8 and 15 of a 4 week cycle. Seventeen patients were recruited. No objective responses were reported in the 16 patients evaluable for efficacy. Recruitment was closed without proceeding to the second stage of the trial. Four patients achieved stable disease as best response, and in two of these patients the stabilization was longer than 4 months. The median progression-free survival was 1.8 months (95 % CI, 0.9-3.5 months) and median overall survival was not reached (95%CI, 56.2 % at censored data). Pharmacokinetics
in patients with Ewing sarcoma was similar to that previously reported in other patient populations. PM00104 showed modest activity in Ewing sarcoma at 2 mg/m² on a weekly schedule. There remains an unmet need for effective therapies for patients with advanced/metastatic Ewing sarcoma.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Behjati S; Tarpey PS; Presneau N; Scheipl S; Pillay N; Van Loo P; Wedge DC; Cooke SL; Gundem G; Davies H; Nik-Zainal S; Martin S; McLaren S; Goodie V; Robinson B; Butler A; Teague JW; Halai D; Khatri B; Myklebost O; Baumhoer D; Jundt G; Hamoudi R; Tirabosco R; Amary MF; Futreal PA; Stratton MR; Campbell PJ; Flanagan AM
RESUMEN / SUMMARY: It is recognized that some mutated cancer genes contribute to the development of many cancer types, whereas others are cancer type specific. For genes that are mutated in multiple cancer classes, mutations are usually similar in the different affected cancer types. Here, however, we report exquisite tumor type specificity for different histone H3.3 driver alterations. In 73 of 77 cases of chondroblastoma (95%), we found p.Lys36Met alterations predominantly encoded in H3F3B, which is one of two genes for histone H3.3. In contrast, in 92% (49/53) of giant cell tumors of bone, we found histone H3.3 alterations exclusively in H3F3A, leading to p.Gly34Trp or, in one case, p.Gly34Leu alterations. The mutations were restricted to the stromal cell population and were not detected in osteoclasts or their precursors. In the context of previously reported H3F3A mutations encoding p.Lys27Met and p.Gly34Arg or p.Gly34Val alterations in childhood brain tumors, a remarkable picture of tumor type specificity for histone H3.3 driver alterations emerges, indicating that histone H3.3 residues, mutations and genes have distinct functions.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: George S; Feng Y; Manola J; Nucci MR; Butrynski JE; Morgan JA; Ramaiya N; Quek R; Penson RT; Wagner AJ; Harmon D; Demetri GD; Krasner C
INSTITUCIÓN / INSTITUTION: Center for Sarcoma and Bone Oncology, Dana-Farber Cancer Institute, Harvard Medical School, Boston, Massachusetts.
RESUMEN / SUMMARY: - BACKGROUND: Advanced uterine leiomyosarcoma (ULMS) is an incurable disease. A significant percentage of cases of ULMS express estrogen and/or progesterone receptors (ER and/or PR). To the authors’ knowledge, the role of estrogen suppression in disease management is not known. METHODS: The authors performed a single-arm phase 2 study of the aromatase inhibitor letrozole at a dose of 2.5 mg daily in patients with unresectable ULMS with ER and/or PR expression confirmed by immunohistochemistry. Tumor assessments were performed at baseline, 6 weeks, 12 weeks, and every 8 weeks thereafter. Toxicity was monitored throughout treatment. The primary endpoint was the progression-free survival at 12 weeks.

RESULTS: A total of 27 patients was accrued, with a median of 2 prior treatment regimens (range, 0-9 treatment regimens). The median duration of protocol treatment was 2.2 months (range, 0.4 months-9.9 months). The 12-week progression-free survival rate was 50% (90% confidence interval, 30%-67%). The best response was stable disease in 14 patients (54%; 90% CI, 36%-71%). Three patients, all of whom had tumors expressing ER and PR in > 90% of tumor cells, continued to receive letrozole for > 24 weeks. The most common reason for treatment discontinuation was disease progression (85%). Letrozole was found to be well tolerated. CONCLUSIONS: Letrozole met protocol-defined criteria as an agent with activity in patients with advanced ULMS. Patients with the longest progression-free survival rate were those whose tumors strongly and diffusely expressed ER and PR. Cancer 2013. © 2013 American Cancer Society.

[4]

TÍTULO / TITLE: - Neo-adjuvant imatinib in advanced primary or locally recurrent dermatofibrosarcoma protuberans: a multicenter phase-II DeCOG trial with long-term follow-up.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Ugurel S; Mentzel T; Utikal J; Helmbold P; Mohr P; Pföhler C; Schiller M; Hauschild A; Hein R; Kampgen E; Kellner I; Leverkus M; Becker JC; Strobel P; Schadendorf D

INSTITUCIÓN / INSTITUTION: - Dermatology, University of Wuerzburg.

RESUMEN / SUMMARY: - PURPOSE: Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous tumor. COL1A1-PDGFB gene fusion is frequent in DFSP, rendering tumor cell proliferation and survival dependent on PDGFRB signaling. This trial investigated imatinib as neo-adjuvant treatment of DFSP including long-term follow-up.

EXPERIMENTAL DESIGN: The primary endpoint of this multicenter phase-II trial was response; secondary endpoints were safety, tumor relapse, and response biomarkers. Patients with advanced primary or locally recurrent DFSP and measurable disease by RECIST were eligible and received imatinib 600 mg/d until definitive surgery with histopathological proof of tumor-free margins. RESULTS: 16 patients received imatinib; 14 patients were evaluable for all endpoints. Median treatment duration was 3.1 months; median tumor shrinkage was 31.5%. Best overall response was 7.1% CR, 50.0% PR, 35.7% SD, and 7.1% PD. Toxicity was moderate with 25.0% grade 3-4 events. During a median follow-up of 6.4 years, one patient developed secondary
resistance to imatinib but responded to second-line sunitinib. This patient also presented local recurrence, distant metastasis and death from DFSP. Exploratory analysis showed that response to imatinib was associated with decreased tumor cellularity and formation of strong hyaline fibrosis. Weak PDGFRB phosphorylation and pigmented-type DFSP were associated with non-response. Additional to PDGFRB, the kinases EGFR and insulin receptor were found activated in a high percentage of DFSPs. CONCLUSION: The neo-adjuvant use of imatinib 600 mg/d in DFSP is efficacious and well-tolerated. Long-term follow-up results do not definitely support smaller surgical margins after successful imatinib pre-treatment, and presume that secondary resistance to imatinib might promote accelerated disease progression.

[5]
**TÍTULO / TITLE:** Human and viral interleukin-6 and other cytokines in Kaposi sarcoma herpesvirus-associated multicentric Castleman disease.
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary
**REVISTA / JOURNAL:** Blood. 2013 Oct 30.
**AUTORES / AUTHORS:** Polizzotto MN; Uldrick TS; Wang V; Aleman K; Wyvill KM; Marshall V; Pittaluga S; O’Mahony D; Whitby D; Tosato G; Steinberg SM; Little RF; Yarchoan R
**INSTITUCIÓN / INSTITUTION:** HIV/AIDS Malignancy Branch, Center for Cancer Research, National Cancer Institute, Bethesda, MD, United States;
**RESUMEN / SUMMARY:** Kaposi sarcoma herpesvirus (KSHV)-associated multicentric Castleman disease (MCD) is a polyclonal B-cell lymphoproliferative disorder. Human IL-6 and a KSHV-encoded homolog, viral IL-6, have been hypothesized to contribute to its pathogenesis but their relative contributions to disease activity is not well understood. We prospectively characterized KSHV viral load (VL), viral (v) and human (h) IL-6, and other cytokines during KSHV-MCD flare and remission in 21 patients with 34 flares and 20 remissions. KSHV-VL, vIL-6, hIL-6, IL-10, and to a lesser extent TNF-alpha, and IL-1beta were each elevated during initial flares compared with remission. Flares fell into three distinct IL-6 profiles: those associated with elevations of vIL-6 only (2 flares, 6%), hIL-6 elevations only (17 flares, 50%), and elevations in both hIL-6 and vIL-6 (13 flares, 38%). Compared with hIL-6-only flares, flares with elevated hIL-6 plus vIL-6 exhibited higher CRP (P=0.0009); worse hyponatremia (P=0.02); higher KSHV VL (P=0.016) and higher IL-10 (P=0.012). This analysis shows vIL-6 and hIL-6 can independently or together lead KSHV-MCD flares, and suggests that vIL-6 and hIL-6 may jointly contribute to disease severity. These findings have implications for the development of novel KSHV-MCD therapies targeting IL-6 and its downstream signaling. This study was registered at clinicaltrials.gov as NCT099073.

[6]
**TÍTULO / TITLE:** Menetrier’s disease and Kaposi’s sarcoma in a HIV-positive patient.
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** Baena-Del-Valle J; Villareal-Velasquez T; Bello-Espinosa A; Marquez G; Posada-Viana J; Segovia-Fuentes J; Redondo-Bermudez C
TÍTULO / TITLE: - Brostallicin versus doxorubicin as first-line chemotherapy in patients with advanced or metastatic soft tissue sarcoma: An European Organisation for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group randomised phase II and pharmacogenetic study.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


●● Enlace al texto completo (gratuito o de pago) 1016/j.ejca.2013.10.002

AUTORES / AUTHORS: - Gelderblom H; Blay JY; Seddon BM; Leahy M; Ray-Coquard I; Sleijfer S; Kerst JM; Rutkowski P; Bauer S; Ouali M; Marreaud S; van der Straaten RJ; Guchelaar HJ; Weitman SD; Hogendoorn PC; Hohenberger P

INSTITUCIÓN / INSTITUTION: - Department of Clinical Oncology, Leiden University Medical Center, Leiden, The Netherlands. Electronic address: a.j.gelderblom@lumc.nl.

RESUMEN / SUMMARY: - AIM: Brostallicin is a DNA minor groove binder that has shown activity in patients with soft tissue sarcoma (STS) failing first-line therapy. The present study assessed the safety and efficacy of first-line brostallicin in patients with advanced or metastatic STS>60years or not fit enough to receive combination chemotherapy. A prospective explorative pharmacogenetic analysis was undertaken in parallel. METHODS: Patients were randomised in a 2:1 ratio between IV brostallicin 10mg/m2 and doxorubicin 75mg/m2 once every 3weeks for a maximum of six cycles. Disease stabilisation at 26weeks (primary end-point) was considered a ‘success’. Further testing of brostallicin was warranted if 35 ‘successes’ were observed in the first 72 eligible patients treated with brostallicin. In addition, patients were genotyped for glutathione S transferase (GST) polymorphisms. RESULTS: One hundred and eighteen patients were included (79 brostallicin and 39 doxorubicin). Brostallicin was well tolerated in comparison to doxorubicin with less grade 3-4 neutropenia (67% versus 95%), grade 2-3 systolic dysfunction (0% versus 11%), alopecia (17% versus 61%) and grade 2-3 mucositis (0% versus 18%). For brostallicin versus doxorubicin, ‘successes’ were observed in 5/77 versus 10/36, progression free survival at 1year was 6.5% versus 15.6%, objective response rate was 3.9% versus 22.2% and overall survival at 1year was 50.5% versus 57.9%, respectively. Only GSTA1 genotype was significantly associated with success rate of doxorubicin treatment. CONCLUSION: Brostallicin cannot be recommended at this dose and schedule in this patient population as first-line therapy. GSTA1 genotype may be predictive for doxorubicin efficacy but warrants further study.

TÍTULO / TITLE: - Bone marrow metastases by alveolar rhabdomyosarcoma in a 31-year-old patient.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Parmentier S; Richter S
Ganglioside GD2 as a therapeutic target for antibody-mediated therapy in patients with osteosarcoma.

**RESUMEN / SUMMARY:**

**BA**CKGROUND: Survival outcomes for patients with osteosarcoma have remained stagnant over the past 30 years. Targeting of ganglioside GD2, a glycosphingolipid on the cell surface of some tumors, with immunotherapy has resulted in improved outcomes for patients with neuroblastoma. In the current study, the expression pattern of GD2 was examined in osteosarcoma.

**METHODS:** Immunohistochemistry was performed on osteosarcoma samples from patients at the time of initial biopsy, definitive surgery, and disease recurrence. The intensity and location of staining were scored. Cell-based enzyme-linked immunoadsorbent assay was performed on osteosarcoma cell lines to quantitate the level of GD2 expression.

**RESULTS:** Forty-four osteosarcoma samples were evaluated by immunohistochemistry, including 8 samples from the initial biopsy, 28 samples from the definitive surgery, and 8 samples from the time of disease recurrence. GD2 was expressed on all 44 osteosarcoma samples. Osteosarcoma tissue obtained at the time of disease recurrence demonstrated a higher intensity of staining compared with samples obtained at initial biopsy and definitive surgery (P = .016). The majority of osteosarcoma cell lines expressed GD2 at higher levels than the neuroblastoma cell line BE(2)-C.

**CONCLUSIONS:** Ganglioside GD2 is highly expressed on osteosarcomas. Clinical trials are needed to assess the efficacy of targeting GD2 in patients with osteosarcoma.

The elevated pre-operative plasma fibrinogen level is an independent negative prognostic factor for cancer-specific, disease-free and overall survival in soft-tissue sarcoma patients.

**RESUMEN / SUMMARY:**

**BACKGROUND AND OBJECTIVES:** Accumulating evidence indicates an important pathophysiological role of fibrinogen on tumor cell progression and metastases in different types of cancer. The aim of the present study was to evaluate the prognostic relevance of pre-operative fibrinogen levels on clinical outcome...
METHODS: Two hundred ninety-four consecutive STS patients were retrospectively evaluated. Cancer-specific survival (CSS), disease-free survival (DFS), and overall survival (OS) were assessed using the Kaplan-Meier curves and Cox regression models. Finally, we supplemented the well-established Kattan nomogram by the fibrinogen level and evaluated the gain of predictive accuracy of this novel nomogram by Harrell’s concordance index (c-index). RESULTS: An elevated plasma fibrinogen level was significantly associated with established prognostic factors, including age, tumor grade, size, and depth (P < 0.05). Furthermore, in multivariate analysis, increased fibrinogen levels were significantly associated with a poor outcome for CSS (HR = 2.48; 95% CI = 1.28-4.78; P = 0.007), DFS (HR = 2.00; 95% CI = 1.11-3.60; P = 0.021), and OS (HR = 2.20; 95% CI = 1.39-3.47; P < 0.001). The estimated c-index was 0.747 using the original Kattan nomogram and 0.779 when the fibrinogen levels was added. CONCLUSION: The pre-operative plasma fibrinogen level may represent a strong and independent unfavorable prognostic factor for CSS, DFS and OS in STS patients. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.
thrombocytopenia (50%), and neutropenic fever/infection (50%). Of note, 38% developed wound complications requiring surgical intervention. The rate of ≥95% histopathologic tumor necrosis was 44%. Changes in DCE-MRI biomarker DeltaKtrans after 2 weeks of sorafenib correlated with histologic response (R² = 0.67, P = 0.012) at surgery. CONCLUSION: The addition of sorafenib to preoperative chemoradiotherapy is feasible and warrants further investigation in a larger trial. DCE-MRI detected changes in tumor perfusion after 2 weeks of sorafenib and may be a minimally invasive tool for rapid assessment of drug effect in soft tissue sarcoma. Clin Cancer Res; 19(24); 1-10. ©2013 AACR.

[12]
TÍTULO / TITLE: - Breast sarcoma after breast-conserving therapy for breast cancer in a patient with li-fraumeni syndrome presenting as focal nonmasslike enhancement on MRI.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Sun S; Tremblay F; Mesurolle B
INSTITUCIÓN / INSTITUTION: - 1 McGill University Health Center, Royal Victoria Hospital, Montreal, QC, Canada.

[13]
TÍTULO / TITLE: - Paracrine activation of WNT/beta-catenin pathway in uterine leiomyoma stem cells promotes tumor growth.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Ono M; Yin P; Navarro A; Moravek MB; Coon JS 5th; Druschitz SA; Serna VA; Qiang W; Brooks DC; Malpani SS; Ma J; Erkan CM; Mittal N; Monsivais D; Dyson MT; Yemelyanov A; Maruyama T; Chakravarti D; Kim JJ; Kurita T; Gottardi CJ; Bulun SE
INSTITUCIÓN / INSTITUTION: - Departments of Obstetrics and Gynecology and Medicine, Feinberg School of Medicine at Northwestern University, Chicago, IL 60611.
RESUMEN / SUMMARY: - Uterine leiomyomas are extremely common estrogen and progesterone-dependent tumors of the myometrium and cause irregular uterine bleeding, severe anemia, and recurrent pregnancy loss in 15-30% of reproductive-age women. Each leiomyoma is thought to arise from a single mutated myometrial smooth muscle stem cell. Leiomyoma side-population (L MSP ) cells comprising 1% of all tumor cells and displaying tumor-initiating stem cell characteristics are essential for estrogen- and progesterone-dependent in vivo growth of tumors, although they have remarkably lower estrogen/progesterone receptor levels than mature myometrial or leiomyoma cells. However, how estrogen/progesterone regulates the growth of L MSP cells via mature neighboring cells is unknown. Here, we demonstrate a critical paracrine role of the wingless-type (WNT)/beta-catenin pathway in estrogen/progesterone-dependent
tumorigenesis, involving LMSP and differentiated myometrial or leiomyoma cells. Estrogen/progesterone treatment of mature myometrial cells induced expression of WNT11 and WNT16, which remained constitutively elevated in leiomyoma tissues. In LMSP cells cocultured with mature myometrial cells, estrogen-progesterone selectively induced nuclear translocation of beta-catenin and induced transcriptional activity of its heterodimeric partner T-cell factor and their target gene AXIN2, leading to the proliferation of LMSP cells. This effect could be blocked by a WNT antagonist. Ectopic expression of inhibitor of beta-catenin and T-cell factor 4 in LMSP cells, but not in mature leiomyoma cells, blocked the estrogen/progesterone-dependent growth of human tumors in vivo. We uncovered a paracrine role of the WNT/beta-catenin pathway that enables mature myometrial or leiomyoma cells to send mitogenic signals to neighboring tissue stem cells in response to estrogen and progesterone, leading to the growth of uterine leiomyomas.

[14]

TÍTULO / TITLE: - Cancer Patient Pathways shortens waiting times and accelerates the diagnostic process of suspected sarcoma patients in Denmark.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Dyrop HB; Safwat A; Vedsted P; Maretty-Nielsen K; Hansen BH; Jorgensen PH; Baad-Hansen T; Bunger C; Keller J

INSTITUCIÓN / INSTITUTION: - Department of Experimental Clinical Oncology, Aarhus University Hospital, Norrebrogade 44, Bldg. 5, DK-8000 Aarhus C, Denmark; Sarcoma Centre of Aarhus University Hospital, Norrebrogade 44, DK-8000 Aarhus C, Denmark. Electronic address: heidi@oncology.dk.

RESUMEN / SUMMARY: - Cancer Patient Pathways (CPPs) for suspected cancer were implemented in Denmark to reduce waiting times for cancer diagnosis and treatment. Our study describes developments in time intervals and tumour size in a natural experiment before and after implementation of the CPP for sarcomas (January 1st, 2009). Medical files for patients referred with suspected sarcoma from other hospitals to Aarhus Sarcoma Centre during 2007-2010 (n=1126) were reviewed for data on milestones, time intervals, performed diagnostics, and tumour size. Results showed a statistically significant reduction in median number of work days in the phase “referral to first appointment” for all patients. For bone sarcomas, median time was significantly reduced from 11 to five work days in the phase “first appointment to decision of treatment”, for soft tissue sarcomas it was reduced from 28 to 18 work days in the phase “referral to start of treatment”. Passive waiting time was reduced, and delays in the fast-track programme were caused mostly by supplementary diagnostics. Median tumour size for soft tissue sarcomas was reduced from 7.0 to 4.9cm, possibly a secondary effect of increased awareness. CPPs have accelerated the diagnostic process for sarcomas, and our results may aid international development of similar initiatives.
TÍTULO / TITLE: - Escherichia coli Virulence Protein NleH1 Interaction with the v-Crk Sarcoma Virus CT10 Oncogene-like Protein (CRKL) Governs NleH1 Inhibition of the Ribosomal Protein S3 (RPS3)/Nuclear Factor kappaB (NF-kappaB) Pathway.

RESUMEN / SUMMARY: - Enterohemorrhagic Escherichia coli and other attaching/effacing bacterial pathogens cause diarrhea in humans. These pathogens use a type III secretion system to inject virulence proteins (effectors) into host cells, some of which inhibit the innate immune system. The enterohemorrhagic E. coli NleH1 effector prevents the nuclear translocation of RPS3 (ribosomal protein S3) to inhibit its participation as a nuclear “specifier” of NF-kappaB binding to target gene promoters. NleH1 binds to RPS3 and inhibits its phosphorylation on Ser-209 by IkappaB kinase-beta (IKKbeta). However, the precise mechanism of this inhibition is unclear. NleH1 possesses a Ser/Thr protein kinase activity that is essential both for its ability to inhibit the RPS3/NF-kappaB pathway and for full virulence of the attaching/effacing mouse pathogen Citrobacter rodentium. However, neither RPS3 nor IKKbeta is a substrate of NleH1 kinase activity. We therefore screened approximately 9,000 human proteins to identify NleH1 kinase substrates and identified CRKL (v-Crk sarcoma virus CT10 oncogene-like protein), a substrate of the BCR/ABL kinase. Knockdown of CRKL abundance prevented NleH1 from inhibiting RPS3 nuclear translocation and NF-kappaB activity. CRKL residues Tyr-198 and Tyr-207 were required for interaction with NleH1. Lys-159, the kinase-active site of NleH1, was necessary for its interaction with CRKL. We also identified CRKL as an IKKbeta interaction partner, mediated by CRKL Tyr-198. We propose that the CRKL interaction with IKKbeta recruits NleH1 to the IKKbeta complex, where NleH1 then inhibits the RPS3/NF-kappaB pathway.

[16]

TÍTULO / TITLE: - Immune response to RB1-regulated senescence limits radiation-induced osteosarcoma formation.

RESUMEN / SUMMARY: - Ionizing radiation (IR) and germline mutations in the retinoblastoma tumor suppressor gene (RB1) are the strongest risk factors for developing osteosarcoma. Recapitulating the human predisposition, we found that Rb1+/− mice exhibited accelerated development of IR-induced osteosarcoma, with a latency of 39 weeks. Initial exposure of osteoblasts to carcinogenic doses of IR in vitro and in vivo induced RB1-dependent senescence and the expression of a panel of
proteins known as senescence-associated secretory phenotype (SASP), dominated by IL-6. RB1 expression closely correlated with that of the SASP cassette in human osteosarcomas, and low expression of both RB1 and the SASP genes was associated with poor prognosis. In vivo, IL-6 was required for IR-induced senescence, which elicited NKT cell infiltration and a host inflammatory response. Mice lacking IL-6 or NKT cells had accelerated development of IR-induced osteosarcomas. These data elucidate an important link between senescence, which is a cell-autonomous tumor suppressor response, and the activation of host-dependent cancer immunosurveillance. Our findings indicate that overcoming the immune response to senescence is a rate-limiting step in the formation of IR-induced osteosarcoma.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Ruocco E; Valenzano F; Brunetti G; Schwartz RA; Ruocco V
INSTITUCIÓN / INSTITUTION: *Department of Dermatology, Second University of Naples, Naples, Italy; daggerDepartment of Dermatology, Catholic University, Rome, Italy; and Departments of double daggerDermatology, section signPathology, and paragraph signPreventive Medicine and Community Health, Rutgers University, New Jersey Medical School, Newark, NJ.
RESUMEN / SUMMARY: Besides the well-known systemic immune deficiency, also a regional immune deficiency, labeled as “immunocompromised district” (ICD), has been documented and focused in the recent years. The objective of the study is to gain more insights into the mechanisms involved in systemic and local immune destabilization. A 35-year-old, homosexual, and drug-addicted HIV+ man presented with a single nodule of Kaposi sarcoma (KS) located on the penis, where a slow to heal herpes zoster had appeared 2 months before. It has been assumed that the unusual penile location of herpes zoster facilitated the outbreak of KS in the affected dermatome because of a viral damage to sensory nerve fibers of the same dermatome. This damage, by interfering with the immunoregulatory function of neuropeptides released by nerve endings in that area, may have caused a regional alteration of the immune control favoring the local onset of the “opportunistic” angiogenic tumor (KS). In a few words, an ICD took place in an immunocompromised patient, thus introducing a more vulnerable site in an already vulnerable subject. The present case is the second one in the literature to document an ICD in the setting of preexisting systemic immune deficiency.

[18] TÍTULO / TITLE: Particle Therapy Using Carbon Ions or Protons as a Definitive Therapy for Patients with Primary Sacral Chordoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

  ●● Enlace al texto completo (gratuito o de pago) 1259/bjr.20130512

AUTORES / AUTHORS: - Mima M; Demizu Y; Jin D; Hashimoto N; Takagi M; Terashima K; Fujii O; Niwa Y; Akagi T; Daimon T; Hishikawa Y; Abe M; Murakami M; Sasaki R; Fuwa N

INSTITUCIÓN / INSTITUTION: - Departments of Radiology and.

RESUMEN / SUMMARY: - Objectives: This study retrospectively evaluated the efficacy and toxicity of particle therapy using carbon ions or protons for primary sacral chordomas. Methods: We evaluated 23 patients with primary sacral chordoma treated with carbon ion therapy (CIT) or proton therapy (PT) between July 2005 and June 2011 at the Hyogo Ion Beam Medical Center. The median patient age was 72 years. Fourteen patients were treated with 70.4 GyE in 16 fractions, and 9 were treated with 70.4 GyE in 32 fractions. CIT was used for 16 patients, and PT was used for 7 patients. Results: The median follow-up period was 38 months. At 3 years, local control (LC), overall survival (OS), and progression-free survival (PFS) for all patients were 94%, 83%, and 68%, respectively. The log-rank test revealed that male sex was significantly related to better PFS (p = 0.029). No other factors, including dose-fractionation and ion type, were significant for LC, OS, or PFS. Grade 3 acute dermatitis was observed in 9 patients, and grade 3 late toxicities were observed in 9 patients. The 32-fraction protocol reduced severe toxicities in both the acute and late phases compared to the 16-fraction protocol. Conclusions: Particle therapy for patients with sacral chordoma showed favourable LC and OS. Severe toxicities were successfully reduced by modifying the dose-fractionation and treatment planning in the later treatment era. Thus, this therapeutic modality should be considered useful and safe. Advances in knowledge: This is the first study including both CIT and PT for sacral chordomas.

[19]

TÍTULO / TITLE: - Validation of the prognostic relevance of plasma C-reactive protein levels in soft-tissue sarcoma patients.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

  ●● Enlace al texto completo (gratuito o de pago) 1038/bjc.2013.595

AUTORES / AUTHORS: - Szkandera J; Gerger A; Liegl-Atzwanger B; Absenger G; Stotz M; Samonigg H; Maurer-Ertl W; Stojakovic T; Ploner F; Leithner A; Pichler M

INSTITUCIÓN / INSTITUTION: - Division of Clinical Oncology, Department of Medicine, Medical University of Graz, Graz, Austria.

RESUMEN / SUMMARY: - Background: The concept of the involvement of systemic inflammation in cancer progression and metastases has gained attraction within the past decade. C-reactive protein (CRP), a non-specific blood-based marker of the systemic inflammatory response, has been associated with decreased survival in several cancer types. The aim of the present study was to validate the prognostic value of pre-operative plasma CRP levels on clinical outcome in a large cohort of soft-tissue sarcoma (STS) patients. Methods: Three hundred and four STS patients, operated between 1998 and 2010, were retrospectively evaluated. CRP levels and the impact on
cancer-specific survival (CSS), disease-free survival (DFS) and overall survival (OS) were assessed using Kaplan-Meier curves and univariate as well as multivariate Cox proportional models. Additionally, we developed a nomogram by supplementing the plasma CRP level to the well-established Kattan nomogram and evaluated the improvement of predictive accuracy of this novel nomogram by applying calibration and Harrell's concordance index (c-index).

Results: An elevated plasma CRP level was significantly associated with established prognostic factors, including age, tumour grade, size and depth (P<0.05). In multivariate analysis, increased CRP levels were significantly associated with a poor outcome for CSS (HR=2.05; 95% CI=1.13-3.74; P=0.019) and DFS (HR=1.88; 95% CI=1.07-3.34; P=0.029). The estimated c-index was 0.74 using the original Kattan nomogram and 0.77 when the plasma CRP level was added.

Conclusion: An elevated pre-operative CRP level represents an independent prognostic factor that predicts poor prognosis and improves the predictive ability of the Kattan nomogram in STS patients. Our data suggest to further prospectively validate its potential utility for individual risk stratification and clinical management of STS patients.

[20]


RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Baresova P; Musilova J; Pitha PM; Lubyova B

INSTITUCIÓN / INSTITUTION: Institute of Immunology and Microbiology, First Faculty of Medicine, Charles University, Prague, Czech Republic.

RESUMEN / SUMMARY: Viruses have developed numerous strategies to counteract the host cell defense. Kaposi’s sarcoma-associated herpesvirus (KSHV) is a DNA tumor virus linked to the development of Kaposi’s sarcoma, Castleman’s disease and primary effusion lymphoma. The virus-encoded viral interferon regulatory factor-3 (vIRF-3) is a latent gene which is involved in the regulation of apoptosis, cell cycle, antiviral immunity and tumorigenesis. vIRF-3 was shown to interact with p53 and inhibit p53-mediated apoptosis. However, the molecular mechanism underlying this phenomenon has not been established. Here, we show that vIRF-3 associates with the DNA-binding domain of p53, inhibits p53 phosphorylation on serine residues S15 and S20, and antagonizes p53 oligomerization and the DNA-binding affinity. Furthermore, vIRF-3 de-stabilizes p53 protein by increasing the levels of p53 polyubiquitination and targeting p53 for proteasome-mediated degradation. Consequently, vIRF-3 attenuates p53-mediated transcription of the growth regulatory gene p21. These effects of vIRF-3 are of biological relevance, since the knock-down of vIRF-3 expression in KSHV-positive BC-3 cells, derived from PEL lymphoma, leads to an increase in p53 phosphorylation, enhancement of p53 stability and activation of p21 gene transcription. Collectively, these data suggest that KSHV evolved an efficient mechanism to down-regulate p53 function and thus facilitate uncontrolled cell proliferation and tumor growth.

[21]
TÍTULO / TITLE: Pulmonary vascular shunts in exercise-intolerant patients with lymphangioleiomyomatosis.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: Am J Respir Crit Care Med. 2013 Nov 1;188(9):1167-70. doi: 10.1164/rccm.201304-0618LE.

AUTORES / AUTHORS: Zafar MA; McCormack FX; Rahman S; Tencza C; Wikenheiser-Brokamp KA; Young LR; Shizukuda Y; Elwing JM

INSTITUCIÓN / INSTITUTION: 1 University of Cincinnati Medical Center Cincinnati, Ohio.

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TÍTULO / TITLE: Subgroups of patients with very large gastrointestinal stromal tumors with distinct prognoses: A multicenter study.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: Wada N; Kurokawa Y; Nishida T; Takahashi T; Toyokawa T; Kusanagi H; Hirota S; Tsujinaka T; Mori M; Doki Y

INSTITUCIÓN / INSTITUTION: Department of Gastroenterological Surgery, Osaka University Graduate School of Medicine, Osaka, Japan.

RESUMEN / SUMMARY: BACKGROUND AND OBJECTIVES: Any gastrointestinal stromal tumors (GISTs) larger than 10 cm are classified as “high risk” according to the modified National Institutes of Health consensus criteria. We conducted a multicenter study to identify a subgroup with moderate prognosis even within the “high-risk” group. METHODS: We retrospectively collected data on 107 patients with tumors >/=10 cm from a multicenter database of GIST patients. Patients with macroscopic residual lesions or tumor rupture were excluded. The relationship between recurrence-free survival (RFS) and clinicopathological factors was analyzed. RESULTS: The median tumor size and mitotic count were 12.5 cm and 8/50 HPF. The RFS rate was 58.5% at 3 years, 52.1% at 5 years. Only mitotic count was an independent prognostic factor of RFS in the multivariate analysis (P = 0.001). The hazard ratio for recurrence in the subgroup with mitotic count >5/50 HPF was 2.91 (95% confidence interval, 1.53 to 5.56). The subgroup with mitotic count </=5/50 HPF showed significantly better RFS than the mitotic count >5/50 HPF subgroup (P < 0.001). CONCLUSIONS: Mitotic count is closely associated with outcome in patients with large GISTs. This suggests that the subset of large GISTs with low mitotic counts may be considered as “intermediate-risk” lesions. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.

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TÍTULO / TITLE: Phase II study of dovitinib in patients with metastatic and/or unresectable gastrointestinal stromal tumours after failure of imatinib and sunitinib.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: -

INSTITUCIÓN / INSTITUTION: -
Background: This prospective, phase II trial evaluated the efficacy and safety of dovitinib in patients with metastatic and/or unresectable gastrointestinal stromal tumours (GISTs) after failure of at least imatinib and sunitinib.

Methods: Patients received oral dovitinib, 500 mg once daily, for 5 consecutive days, followed by a 2-day rest, every 28 days. The primary endpoint was disease control rate (DCR; objective response + stable disease (SD)) at 24 weeks, assessed by computed tomography (CT) scan according to RECIST v1.0. Metabolic response was evaluated by positron emission tomography (PET)-CT scans performed at baseline and after 4 weeks of treatment.

Results: Between September 2011 and April 2012, 30 patients were enrolled. DCR at 24 weeks by RECIST v1.0 was 13% and one patient (3%) had a partial response. Based on the European Organization for Research and Treatment of Cancer PET response criteria, four patients (13%) had a metabolic partial response after 4 weeks of treatment. At a median follow-up of 8.3 months (range, 6.3-12.2 months), median progression-free survival (PFS) was 3.6 months (95% confidence interval (CI), 3.5-3.7 months) and median overall survival was 9.7 months (95% CI, 6.0-13.4 months). Metabolic progressive disease at Week 4 was significantly associated with shorter PFS (P=0.03). Grade ¾ adverse events included asthenia (20%), neutropenia (13%), thrombocytopenia (10%), and hypertriglyceridaemia (10%). Most toxicities were manageable by dose modification.

Conclusion: Dovitinib showed modest antitumour activity with manageable toxicities in heavily pretreated patients with advanced GISTs.
regarding toxicity, progression-free survival (PFS) and overall survival (OS) after allo-SCT. Twenty patients were conditioned with reduced intensity and ten with high-dose chemotherapy. Twenty-three patients were transplanted with HLA-matched and seven with HLA-mismatched grafts. Three patients additionally received donor lymphocyte infusions (DLIs). Median follow-up was 9 months. Results: Three-year OS was 20% (s.e.+/-8%) with a median survival time of 12 months. Cumulative risk of progression was 67% (s.e.+/-10%) and 11% (s.e.+/-6%) for death of complications. Thirteen patients developed acute graft-vs-host disease (GvHD) and five developed chronic GvHD. Eighteen patients died of disease and four of complications. Eight patients survived in complete remission (CR) (median: 44 months). No patients with residual disease before allo-SCT were converted to CR. Conclusion: The use of allo-SCT in patients with advanced RMS is currently experimental. In a subset of patients, it may constitute a valuable approach for consolidating CR, but this needs to be validated in prospective trials.

[25]

**TITULO / TITLE:** - Generation of a patient-derived chordoma xenograft and characterization of the phosphoproteome in a recurrent chordoma.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - J Neurosurg. 2013 Nov 29.

**AUTORES / AUTHORS:** - Davies JM; Robinson AE; Cowdrey C; Mummaneni PV; Ducker GS; Shokat KM; Bollen A; Hann B; Phillips JJ

**INSTITUCIÓN / INSTITUTION:** - Department of Neurological Surgery.

**RESUMEN / SUMMARY:** - Object The management of patients with locally recurrent or metastatic chordoma is a challenge. Preclinical disease models would greatly accelerate the development of novel therapeutic options for chordoma. The authors sought to establish and characterize a primary xenograft model for chordoma that faithfully recapitulates the molecular features of human chordoma. Methods Chordoma tissue from a recurrent clival tumor was obtained at the time of surgery and implanted subcutaneously into NOD-SCID interleukin-2 receptor gamma (IL-2Rgamma) null (NSG) mouse hosts. Successful xenografts were established and passaged in the NSG mice. The recurrent chordoma and the derived human chordoma xenograft were compared by histology, immunohistochemistry, and phospho-specific immunohistochemistry. Based on these results, mice harboring subcutaneous chordoma xenografts were treated with the mTOR inhibitor MLN0128, and tumors were subjected to phosphoproteome profiling using Luminex technology and immunohistochemistry. Results SF8894 is a novel chordoma xenograft established from a recurrent clival chordoma that faithfully recapitulates the histopathological, immunohistological, and phosphoproteomic features of the human tumor. The PI3K/Akt/mTOR pathway was activated, as evidenced by diffuse immunopositivity for phospho-epitopes, in the recurrent chordoma and in the established xenograft. Treatment of mice harboring chordoma xenografts with MLN0128 resulted in decreased activity of the PI3K/Akt/mTOR signaling pathway as indicated by decreased phospho-mTOR levels (p = 0.019, n = 3 tumors per group). Conclusions The authors report the establishment of SF8894, a recurrent clival chordoma xenograft that mimics
many of the features of the original tumor and that should be a useful preclinical model for recurrent chordoma.
BACKGROUND: Few treatment options remain for patients with metastatic or unresectable gastrointestinal stromal tumours (GIST) after objective progression on approved tyrosine-kinase inhibitors. We aimed to assess efficacy of imatinib rechallenge in these patients. METHODS: In our prospective, randomised, double-blind trial, we enrolled adults (≥18 years) who had previously benefited from first-line imatinib (initial response or stable disease for ≥6 months) but whose metastatic or unresectable GIST had progressed on at least imatinib and sunitinib. We randomly allocated participants in a 1:1 ratio, with a centralised computer-generated allocation procedure (random permuted blocks of two, four, and six) and stratified by previous treatment and Eastern Cooperative Oncology Group performance status, to receive best supportive care with imatinib 400 mg per day or matched placebo. Crossover to open-label imatinib was allowed after investigator-adjudicated disease progression. The primary endpoint was progression-free survival (PFS), as determined by a masked external radiological review. All analyses were done for all patients who received at least one dose of study drug. This study is registered with ClinicalTrials.gov, number NCT01151852. FINDINGS: Between July 20, 2010, and Jan 17, 2013, we randomly allocated 41 patients to the imatinib group and 40 patients to the placebo group. After a median follow-up of 5.2 months (IQR 3.4-9.4), median PFS was 1.8 months (95% CI 1.7-3.6) with imatinib compared with 0.9 months (0.9-1.7) with placebo (hazard ratio for progression or death 0.46, 95% CI 0.27-0.78; p=0.005). 37 (93%) patients in the placebo group crossed over to open-label imatinib after progression. The most common grade 3 or worse adverse events were anaemia (12 [29%] of 41 patients in the imatinib group vs three [8%] of 40 in the placebo group), fatigue (four [10%] vs none), and hyperbilirubinaemia (three [7%] vs one [3%]). INTERPRETATION: In patients with GIST that is refractory to treatment with all standard tyrosine-kinase inhibitors, the disease continues to harbour many clones that are sensitive to kinase inhibitors. Continued kinase suppression might slow, although not halt, disease progression. FUNDING: Novartis Oncology, Ludwig Center at Dana-Farber/Harvard.

[28] TÍTULO / TITLE: PAX3-NCOA2 fusion gene has a dual role in promoting the proliferation and inhibiting the myogenic differentiation of rhabdomyosarcoma cells.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Yoshida H; Miyachi M; Sakamoto K; Ouchi K; Yagyu S; Kikuchi K; Kuwahara Y; Tsuchiya K; Imamura T; Iehara T; Kakazu N; Hojo H; Hosoi H

INSTITUCIÓN / INSTITUTION: Department of Pediatrics, Graduate School of Medical Science, Kyoto Prefectural University of Medicine, Kyoto, Japan.

RESUMEN / SUMMARY: We analyzed a complex chromosomal translocation in a case of embryonal rhabdomyosarcoma (RMS) and showed that it generates the fusion gene PAX3 (paired box 3)-NCOA2 (nuclear receptor coactivator 2). To understand the role of this translocation in RMS tumorigenesis, we established two types of stable mouse...
myoblast C2C12 cell lines expressing PAX3-NCOA2 and PAX3-FOXO1A (forkhead box O1A), respectively. Compared with control cells, PAX3-NCOA2 cells grew faster, were more motile, were less anchorage dependent, progressed more quickly through the G1/S phase of cell cycle and showed greater transcriptional activation of the PAX3 consensus-binding site. However, PAX3-NCOA2 cells proliferated more slowly and differentiated more weakly than did PAX3-FOXO1A cells. Both PAX3-NCOA2 cells and PAX3-FOXO1A cells formed tumors in nude mice, although the PAX3-NCOA2-induced tumors grew more slowly. Our results may explain why NCOA2 rearrangement is mainly found in embryonal rhabdomyosarcoma, which has a better prognosis than alveolar rhabdomyosarcoma, which expresses the PAX3-FOXO1A fusion gene. These results indicate that the PAX3-NCOA2 fusion gene has a dual role in the tumorigenesis of RMS: promotion of the proliferation and inhibition of the myogenic differentiation of RMS cells. Oncogene advance online publication, 11 November 2013; doi:10.1038/onc.2013.491.

[29]


**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Italiano A; Le Cesne A; Bellera C; Piperno-Neumann S; Duffaud F; Penel N; Cassier P; Domont J; Takebe N; Kind M; Coindre JM; Blay JY; Bui B

**INSTITUCIÓN / INSTITUTION:** Department of Medical Oncology, Institut Bergonie, Bordeaux.

**RESUMEN / SUMMARY:** BACKGROUND: Pre-clinical data have suggested a therapeutic role of Hedgehog (Hh) pathway inhibitors in chondrosarcoma. METHODS: This phase II trial included patients with progressive advanced chondrosarcoma. They received GDC-0449 150 mg/day (days 1-28, 28-day cycle). The primary end point was the 6-month clinical benefit rate (CBR) defined as the proportion of patients with non-progressive disease at 6 months. A 6-month CBR of 40% was considered as a reasonable objective to claim drug efficacy. RESULTS: Between February 2011 and February 2012, 45 patients were included. Twenty had received prior chemotherapy. Thirty-nine were assessable for efficacy. The 6-month CBR was 25.6% (95% confidence interval 13.0-42.1). All stable patients had grade 1 or 2 conventional chondrosarcoma with documented progression within the 6 months before inclusion. All but one with available data also had overexpression of the Hh ligand. Median progression-free and overall survivals were 3.5 and 12.4 months, respectively. The most frequent adverse events were grade 1 or 2 myalgia, dysgeusia and alopecia. CONCLUSIONS: GDC-0449 did not meet the primary end point of this trial. Results suggest some activity in a subset of patients with progressive grade 1 or 2 conventional chondrosarcoma. Further studies assessing its role in combination with chemotherapy are warranted. CLINICALTRIALSGOV IDENTIFIER: NCT01267955.
Mitochondrial dysfunction and permeability transition in osteosarcoma cells showing the warburg effect.

Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Giang AH; Raymond T; Brookes P; de Mesy Bentley K; Schwarz E; O'Keefe R; Eliseev R

INSTITUCIÓN / INSTITUTION: From the Center for Musculoskeletal Research and.

RESUMEN / SUMMARY: Metabolic reprogramming in cancer is manifested by persistent aerobic glycolysis and suppression of mitochondrial function and is known as the Warburg effect. The Warburg effect contributes to cancer progression and is considered to be a promising therapeutic target. Understanding the mechanisms used by cancer cells to suppress their mitochondria may lead to development of new approaches to reverse metabolic reprogramming. We have evaluated mitochondrial function and morphology in poorly respiring LM7 and 143B osteosarcoma (OS) cell lines showing the Warburg effect in comparison with actively respiring Saos2 and HOS OS cells and noncancerous osteoblastic hFOB cells. In LM7 and 143B cells, we detected markers of the mitochondrial permeability transition (MPT), such as mitochondrial swelling, depolarization, and membrane permeabilization. In addition, we detected mitochondrial swelling in human OS xenografts in mice and archival human OS specimens using electron microscopy. The MPT inhibitor sanglifehrin A reversed MPT markers and increased respiration in LM7 and 143B cells. Our data suggest that the MPT may play a role in suppression of mitochondrial function, contributing to the Warburg effect in cancer.

SS18-SSX fusion protein-induced Wnt/beta-catenin signaling is a therapeutic target in synovial sarcoma.

Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Trautmann M; Sievers E; Aretz S; Kindler D; Michels S; Friedrichs N; Renner M; Kirfel J; Steiner S; Huss S; Koch A; Penzel R; Larsson O; Kawai A; Tanaka S; Sonobe H; Waha A; Schirmacher P; Mechtersheimer G; Wardemann E; Buttnar R; Hartmann W

INSTITUCIÓN / INSTITUTION: 1] Department of Pathology, University Hospital Cologne, Cologne, Germany [2] Department of Pathology, University Hospital Bonn, Bonn, Germany.

RESUMEN / SUMMARY: Synovial sarcoma is a high-grade soft tissue malignancy characterized by a specific reciprocal translocation t(X;18), which leads to the fusion of the SS18 (SYT) gene to one of three SSX genes (SSX1, SSX2 or SSX4). The resulting chimeric SS18-SSX protein is suggested to act as an oncogenic transcriptional regulator. Despite multimodal therapeutic approaches, metastatic disease is often lethal and the development of novel targeted therapeutic strategies is
required. Several expression-profiling studies identified distinct gene expression signatures, implying a consistent role of Wnt/beta-catenin signaling in synovial sarcoma tumorigenesis. Here we investigate the functional and therapeutic relevance of Wnt/beta-catenin pathway activation in vitro and in vivo. Immunohistochemical analyses of nuclear beta-catenin and Wnt downstream targets revealed activation of canonical Wnt signaling in a significant subset of 30 primary synovial sarcoma specimens. Functional aspects of Wnt signaling including dependence of Tcf/beta-catenin complex activity on the SS18-SSX fusion proteins were analyzed. Efficient SS18-SSX-dependent activation of the Tcf/beta-catenin transcriptional complex was confirmed by TOPflash reporter luciferase assays and immunoblotting. In five human synovial sarcoma cell lines, inhibition of the Tcf/beta-catenin protein-protein interaction significantly blocked the canonical Wnt/beta-catenin signaling cascade, accompanied by the effective downregulation of Wnt targets (AXIN2, CDC25A, c-MYC, DKK1, CyclinD1 and Survivin) and the specific suppression of cell viability associated with the induction of apoptosis. In SYO-1 synovial sarcoma xenografts, administration of small molecule Tcf/beta-catenin complex inhibitors significantly reduced tumor growth, associated with diminished AXIN2 protein levels. In summary, SS18-SSX-induced Wnt/beta-catenin signaling appears to be of crucial biological importance in synovial sarcoma tumorigenesis and progression, representing a potential molecular target for the development of novel therapeutic strategies.

Oncogene advance online publication, 28 October 2013; doi:10.1038/onc.2013.443.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o de pago) 1016/j.ijrobp.2013.07.003
AUTORES / AUTHORS: - Spalding AC; Hawkins DS; Donaldson SS; Anderson JR; Lyden E; Laurie F; Wolden SL; Arndt CA; Michalski JM
INSTITUCIÓN / INSTITUTION: - Kosair Children’s Hospital and Brain Tumor Center, Louisville, Kentucky. Electronic address: Aaron.Spalding@nortonhealthcare.org.
RESUMEN / SUMMARY: - PURPOSE: Radiation therapy remains an essential treatment for patients with parameningeal rhabdomyosarcoma (PMRMS), and early radiation therapy may improve local control for patients with intracranial extension (ICE). METHODS AND MATERIALS: To address the role of radiation therapy timing in PMRMS in the current era, we reviewed the outcome from 2 recent clinical trials for intermediate-risk RMS: Intergroup Rhabdomyosarcoma Study (IRS)-IV and Children’s Oncology Group (COG) D9803. The PMRMS patients on IRS-IV with any high-risk features (cranial nerve palsy [CNP], cranial base bony erosion [CBBE], or ICE) were treated immediately at day 0, and PMRMS patients without any of these 3 features received week 6-9 radiation therapy. The D9803 PMRMS patients with ICE received day 0 X-Ray Therapy (XRT) as well; however, those with either CNP or CBBE had XRT at week 12. RESULTS: Compared with the 198 PMRMS patients from IRS-IV, the 192 PMRMS patients from D9803 had no difference (P<.05) in 5-year local failure (19% vs 19%), failure-free-survival (70% vs 67%), or overall survival (75% vs 73%) in
aggregate. The 5-year local failure rates by subset did not differ when patients were classified as having no risk features (None, 15% vs 19%, P=.25), cranial nerve palsy/cranial base of skull erosion (CNP/CBBE, 15% vs 28%, P=.22), or intracranial extension (ICE, 21% vs 15%, P=.27). The D9083 patients were more likely to have received initial staging by magnetic resonance imaging (71% vs 53%).

CONCLUSIONS: These data support that a delay in radiation therapy for high-risk PMRMS features of CNP/CBBE does not compromise clinical outcomes.

[33]
TÍTULO / TITLE: - Sun exposure causes somatic second hit mutations and angiofibroma development in Tuberous Sclerosis Complex.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Tyburczy ME; Wang JA; Li S; Thangapazham R; Chekaluk Y; Moss J; Kwiatkowski DJ; Darling TN
INSTITUCIÓN / INSTITUTION: - Department of Medicine, Brigham and Women’s Hospital, Harvard Medical School, Boston, MA 02115, USA.
RESUMEN / SUMMARY: - Tuberous Sclerosis Complex (TSC) is characterized by the formation of tumors in multiple organs and is due to germline mutation in one of two tumor suppressor genes, TSC1 and TSC2. As for other tumor suppressor gene syndromes, the mechanism of somatic second-hit events in TSC tumors is unknown. We grew fibroblast-like cells from 29 TSC skin tumors from 22 TSC subjects, and identified germline and second-hit mutations in TSC1/TSC2 using next-generation sequencing. Eighteen of 22 (82%) subjects had a mutation identified, and 8 of the 18 (44%) subjects were mosaic with mutant allele frequencies of 0 to 19% in normal tissue DNA. Multiple tumors were available from 4 patients, and in each case second-hit mutations in TSC2 were distinct indicating they arose independently. Most remarkably, 7 (50%) of the 14 somatic point mutations were CC>TT ultraviolet "signature" mutations, never seen as a TSC germline mutation. These occurred exclusively in facial angiofibroma tumors from sun-exposed sites. These results implicate UV-induced DNA damage as a cause of second-hit mutations and development of TSC facial angiofibromas, and suggest that measures to limit UV exposure in TSC children and adults should reduce the frequency and severity of these lesions.

[34]
TÍTULO / TITLE: - The Multifunctional Protein Fused in Sarcoma (FUS) is a Coactivator of Microphthalmia associated Transcription Factor (MITF).
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Bronisz A; Carey HA; Godlewski J; Sif S; Ostrowski MC; Sharma SM
INSTITUCIÓN / INSTITUTION: - Brigham and Women’s Hospital, United States;
The microphthalmia-associated transcription factor (MITF) is required for terminal osteoclast differentiation and is a signaling effector engaged by macrophage colony stimulating factor 1 (CSF-1) and receptor activator of nuclear factor-kappa B ligand (RANKL). MITF exerts its regulatory functions through its association with cofactors. Discovering the identity of its various partners will provide insights into the mechanisms governing gene expression during osteoclastogenesis. Here we demonstrate that the proto-oncogene FUS, the chromatin remodeling ATPase BRG1 and MITF form a trimeric complex that is regulated by phosphorylation of MITF at Ser307 by p38 MAPK during osteoclast differentiation. FUS was recruited to MITF target gene promoters Acp5 and Ctsk during osteoclast differentiation and FUS knockdown abolished efficient transcription of Acp5 and Ctsk. Furthermore, sumoylation of MITF at Lys316, known to negatively regulate MITF transcriptional activity, inhibited MITF interactions with FUS and BRG1 in a p38 MAPK phosphorylation dependent manner. These results demonstrate that FUS is a coregulator of MITF activity and provide new insights into how the RANKL/p38 MAPK signaling nexus controls gene expression in osteoclasts.

[35]

**TITULO / TITLE:** Clinical and pathological characteristics, pathological reevaluation and recurrence patterns of cellular leiomyomas: a retrospective study in 76 patients.

**RESUMEN / SUMMARY:** To analyze clinical and pathologic features as well as recurrence patterns of cellular leiomyomas (CL) in women who underwent surgical therapy for symptomatic disease. STUDY DESIGN: This retrospective study was conducted at the Department of Obstetrics and Gynecology, University Women's Clinic, Tuebingen, Germany. We identified all women who had CL on final diagnosis after surgery between January 1, 2000, and December 31, 2010. RESULTS: Our study sample comprised 76 women with a diagnosis of CL. A single uterine mass was present in 51.3% of the cases; in uteri with both CL and uterine leiomyomas (UL), the CL constituted the largest uterine mass in 20 of 21 (95.2%) cases. Additionally, in 98% of the uteri, CL were either the largest or the only uterine mass. Five women (6.6%; 5/76) had reported surgical procedures for symptomatic leiomyoma before the index surgery in our analysis. Three women underwent hysteroscopic resection of the leiomyomas and 2 women underwent abdominal myomectomy. Mean time to recurrence was 14.0 months (median 6.0; range, 4.0-52.0). Over the follow-up period, 6 women who underwent uterus-conserving surgery (12.0%; 6/50) with CL had leiomyoma recurrence. Five women underwent abdominal myomectomy and one underwent hysteroscopic resection of the CL. One patient had recurrence of a CL 43 months after abdominal myomectomy and underwent vaginal hysterectomy; the other five women had recurrences of UL. Mean time to recurrence was 28.6 months (median...
12.5; range, 4.0-83.0). CONCLUSIONS: Recurrence rates of CL in our study group resemble recurrence rates of UL.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Halder SK; Osteen KG; Al-Hendy A
RESUMEN / SUMMARY: - Uterine fibroids (leiomyoma) are the most common benign tumors associated with excessive deposition of extracellular matrix (ECM) that increase fibroid tumorigenicity. Herein, we determine the expression levels of vitamin D receptor (VDR) protein in human uterine fibroids and compared to adjacent normal myometrium. Using western blot analyses we found that at least 60% of uterine fibroids analyzed (25 out of 40) expressed low levels of VDR. The biologically active 1,25-dihydroxyvitamin D3 \[\text{1,25(OH)}_2\text{D3}\] function via binding to its nuclear VDR. We also found that 1,25(OH)2D3 induced VDR in a concentration-dependent manner, and reduced ECM-associated fibrotic and proteoglycans expressions in immortalized human uterine fibroid cell line (HuLM). At 1-10 nM concentrations, 1,25(OH)2D3 significantly induced (P<0.05) nuclear VDR, which was further stimulated by higher concentrations of 1,25(OH)2D3 in HuLM cells. 1,25(OH)2D3 at 10 nM concentration also significantly reduced (P<0.05) the protein expression of ECM-associated collagen type 1, fibronectin and plasminogen activator inhibitor (PAI-1) in HuLM cells. We also found that 1,25(OH)2D3 reduced mRNA and protein expressions of proteoglycans such as fibromodulin, biglycan and versican in HuLM cells. Moreover, the aberrant expression of structural smooth muscle actin fibers was reduced by 1,25(OH)2D3 treatment in a concentration-dependent manner in HuLM cells. Together, our results suggest that human uterine fibroids express reduced levels of VDR than the adjacent normal myometrium, and treatment with 1,25(OH)2D3 can potentially reduce the aberrant expression of major ECM-associated proteins in HuLM cells. Thus 1,25(OH)2D3 might be an effective, safe non-surgical treatment option for human uterine fibroids.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Vadakara J; von Mehren M
INSTITUCIÓN / INSTITUTION: - Department of Medical Oncology, Fox Chase Cancer Center, 333 Cottman Avenue, Philadelphia, PA 19111, USA.
RESUMEN / SUMMARY: - Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract. Before the advent of tyrosine kinase inhibitors (TKIs) there were few treatment options available to patients with metastatic
GIST. Surgery was the mainstay of treatment and the prognosis was dismal. With the advent of imatinib and second-line TKIs the prognosis of metastatic GIST has improved dramatically; however, there is still a need for therapies for patients with disease refractory to TKI therapy. Newer agents are under investigation and may have promise. This article discusses the current standard of care in terms of standard and investigational pharmacotherapy in the management of metastatic GIST.

[38]
**TITULO / TITLE:** - Osteosarcoma cells promote the production of pro-tumor cytokines in mesenchymal stem cells by inhibiting their osteogenic differentiation through the TGF-beta/Smad2/3 pathway.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Tu B; Peng ZX; Fan QM; Du L; Yan W; Tang TT

**INSTITUCION / INSTITUTION:** - Shanghai Key Laboratory of Orthopedic Implant, Department of Orthopedic Surgery, Shanghai Ninth People’s Hospital, Shanghai Jiao Tong University School of Medicine, Zhizaoju Road 639, Shanghai 200011, China.

**RESUMEN / SUMMARY:** - Mesenchymal stem cells (MSCs) are among the most important components of the osteosarcoma microenvironment and are reported to promote tumor progression. However, the means by which osteosarcoma cells modulate MSC behavior remains unclear. The aim of this study was to determine the effects of osteosarcoma cells on both the production of pro-tumor cytokines by mesenchymal stem cells (MSCs) and the osteogenic differentiation of MSCs. High level of transforming growth factor-beta (TGF-beta) was detected in three osteosarcoma cell lines. Conditioned media (CM) from the osteosarcoma cell lines Saos-2 and U2-OS were used to stimulate the cultured MSCs. We found that osteosarcoma cells promoted the production of IL-6 and VEGF in MSCs by inhibiting their osteogenic differentiation. Furthermore, TGF-beta in tumor CM was proved to be an important factor. The TGF-beta neutralizing antibody antagonized the effects induced by osteosarcoma CM. The inhibition of Smad2/3 by siRNA significantly decreased the production of IL-6 and VEGF in MSCs and induced their osteogenic differentiation. We also found that Smad2/3 enhanced the expression of beta-catenin in MSCs by decreasing the level of Dickkopf-1 (DKK1). Although the inhibition of beta-catenin did not affect the production of IL-6 or VEGF, or the gene expression of the early osteogenic markers Runx2 and ALP, it did enhance the gene expression of osteocalcin. Taken together, our data indicate that osteosarcoma cells secrete TGF-beta to maintain the stemness of MSCs and promote the production of pro-tumor cytokines by these cells.

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[39]
**TITULO / TITLE:** - Chemotherapy-related toxicity in patients with non-metastatic Ewing sarcoma: influence of sex and age.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

Influence of age and sex on chemotherapy-related toxicity was evaluated in children (3-9 years), adolescents (10-17 years), and adults (up to 40 years) with localized Ewing sarcoma (ES) enrolled in the ISG/SSG III protocol. Treatment was based on vincristine, doxorubicin, cyclophosphamide, ifosfamide, dactinomycin, and etoposide. High-dose chemotherapy with busulfan and melphalan was given in poor responder patients. The analysis was based on 2191 courses of standard chemotherapy and 230 patients. A lower risk of G4 leukopenia and thrombocytopenia, hospitalization, febrile neutropenia, and red blood cell (RBC) transfusions was observed in males. Use of granulocyte colony-stimulating factor (G-CSF) was more frequent in adults, while children more often received RBC transfusions. A significant correlation between sex and chemotherapy-related toxicity was observed in the study, whereas no significant differences in terms of bone marrow toxicity can be expected according to patient age. Further studies should analyse the role of pharmacokinetics, pharmacogenomics, and clinical characteristics.

[40]

TITULO / TITLE: - The economic impact of cytoreductive surgery and tyrosine kinase inhibitor therapy in the treatment of advanced gastrointestinal stromal tumours: A Markov chain decision analysis.

RESUMEN / SUMMARY: - The current first-line treatment for patients with recurrent or metastatic gastrointestinal stromal tumours (GIST) is management with tyrosine kinase inhibition (TKI). There is an undefined role for surgery in the management of these patients. This study uses a cost analysis to examine the economic impact of treating patients with TKI in combination with surgery at different time-points in their treatment trajectories. METHODS: A Markov chain decision analysis was modelled over a 2-year time horizon to determine costs associated with surgery in combination with imatinib mesylate (IM) or sunitinib malate (SU) in seven scenarios varied by TKI agent, dose and disease status (stable versus localised progressive disease). Rates of disease progression, surgical morbidity, mortality and adverse drug reactions were extracted from the existing literature. Deterministic sensitivity analyses were performed to examine changes in cost due to variations in key variables. RESULTS: The least-costly scenario was to perform no surgery. The most costly scenario was to perform surgery on patients with localised progressive disease on IM 800mg. The overall range of costs clustered within approximately...
$47,000 (USD). Variations in surgical cost, surgical mortality and cost of IM demonstrated thresholds for changing the least-costly scenario within plausible tested ranges. CONCLUSION: Costs of surgical intervention at different time-points within the treatment course of patients with advanced GIST fluctuate within a relatively narrow range, suggesting that costs arise primarily from the administration of TKI. The decision to pursue cytoreductive surgery should not be based on cost alone. Future studies should incorporate health-state utilities when available.

[41] TÍTULO / TITLE: - The case of a patient affected by primary gliosarcoma and neuroendocrine pancreatic cancer with prolonged survival.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

[43] TÍTULO / TITLE: - The case of a patient affected by primary glisarcoma and neuroendocrine pancreatic cancer with prolonged survival.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
patients with localized STS who visited 11 institutions participating in Japanese Musculoskeletal Oncology Group between 1995 and 2006 and were treated by surgical resection. Univariate and multivariate analyses were performed to identify prognostic factors. RESULTS: Median follow-up period was 38 months. Histologically high-grade tumors were detected in 71% of the patients. Wide resection with adequate margins was performed in 66% of the cases. Systemic chemotherapy was performed in only 5 patients. Univariate analysis identified histological grade and gender as statistically significant prognostic factors for sarcoma-specific survival. Multivariate analysis did not identify significant prognostic factors for sarcoma-specific survival, although high grade sarcoma emerged as a potentially significant prognostic factor (P = 0.050). Local recurrence was detected in 19% of the patients. Multivariate analysis of local recurrence-free survival showed that tumor site and surgical margins were statistically significant prognostic factors. CONCLUSIONS: Older age was not identified as a prognostic factor for sarcoma-specific survival, which is not consistent with the findings of previous studies showing that older age was associated with decreased sarcoma-specific survival. Complete resection should be indicated and can lead to optimal treatment outcome for properly selected elderly patients.

[43]

**TÍTULO / TITLE:** - Multiple tumor types including leiomyoma and Wilms tumor in a patient with Gorlin syndrome due to 9q22.3 microdeletion encompassing the PTCH1 and FANC-C loci.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Garavelli L; Piemontese MR; Cavazza A; Rosato S; Wischmeijer A; Gelmini C; Albertini E; Albertini G; Forzano F; Franchi F; Carella M; Zelante L; Superti-Furga A

**INSTITUCIÓN / INSTITUTION:** - Clinical Genetics Unit, Obstetric and Paediatric Department, Istituto di Ricovero e Cura a Carattere Scientifico, Arcispedale S Maria Nuova, Reggio Emilia, Italy.

**RESUMEN / SUMMARY:** - Gorlin syndrome or nevoid basal cell carcinoma syndrome (NBCCS) is an autosomal dominant condition mainly characterized by the development of mandibular keratocysts which often have their onset during the second decade of life and/or multiple basal cell carcinoma (BCC) normally arising during the third decade. Cardiac and ovarian fibromas can be found. Patients with NBCCS develop the childhood brain malignancy medulloblastoma (now often called primitive neuro-ectodermal tumor [PNET]) in 5% of cases. The risk of other malignant neoplasms is not clearly increased, although lymphoma and meningioma can occur in this condition. Wilms tumor has been mentioned in the literature four times. We describe a patient with a 10.9 Mb 9q22.3 deletion spanning 9q22.2 through 9q31.1 that includes the entire codifying sequence of the gene PTCH1, with Wilms tumor, multiple neoplasms (lung, liver, mesenteric, gastric and renal leiomyomas, lung typical carcinoid tumor, adenomatoid tumor of the pleura) and a severe clinical presentation. We propose including leiomyomas among minor criteria of the NBCCS. © 2013 Wiley Periodicals, Inc.
Distinct cellular origin and genetic requirement of Hedgehog-Gli in postnatal rhabdomyosarcoma genesis.

Dysregulation of the Hedgehog (Hh)-Gli signaling pathway is implicated in a variety of human cancers, including basal cell carcinoma (BCC), medulloblastoma (MB) and embryonal rhabdomyosarcoma (eRMS), three principle tumors associated with human Gorlin syndrome. However, the cells of origin of these tumors, including eRMS, remain poorly understood. In this study, we explore the cell populations that give rise to Hh-related tumors by specifically activating Smoothened (Smo) in both Hh-producing and -responsive cell lineages in postnatal mice. Interestingly, we find that unlike BCC and MB, eRMS originates from the stem/progenitor populations that do not normally receive active Hh signaling. Furthermore, we find that the myogenic lineage in postnatal mice is largely Hh quiescent and that Pax7-expressing muscle satellite cells are not able to give rise to eRMS upon Smo or Gli1/2 overactivation in vivo, suggesting that Hh-induced skeletal muscle eRMS arises from Hh/Gli quiescent non-myogenic cells. In addition, using the Gli1 null allele and a Gli3 repressor allele, we reveal a specific genetic requirement for Gli proteins in Hh-induced eRMS formation and provide molecular evidence for the involvement of Sox4/11 in eRMS cell survival and differentiation. Oncogene advance online publication, 25 November 2013; doi:10.1038/onc.2013.480.

Determinants of quality of life in patients with skull base chordoma.

Object Skull base chordomas can be managed by surgical intervention and adjuvant radiotherapy. As survival for this disease increases, identification of determinants of quality of life becomes an important focus for guiding comprehensive patient care. In this study the authors sought to measure functional outcome and quality of life in patients with skull base chordomas and to identify determinants of quality of life in these patients. Methods The authors carried out an internet-based cross-sectional survey, collecting detailed data for 83 individual patients. Demographic and clinical variables were evaluated. Functional outcomes were determined by Karnofsky Performance Scale (KPS) and Glasgow Outcome Scale.
Extended (GOSE), quality of life was measured using the 36-Item Short Form Health Survey (SF-36), and depression was assessed using Patient Health Questions-9 (PHQ-9) instrument. Caregiver burden was assessed using the Zarit Burden Interview (ZBI). Univariate and multivariate analysis was performed to identify determinants of the physical and mental components of the SF-36. Results Patients with skull base chordomas who have undergone surgery and/or radiation treatment had a median KPS score of 90 (range 10-100, IQR 10) and a median GOSE score of 8 (range 2-8, IQR 3). The mean SF-36 Physical Component Summary score (+/- SD) was 43.6 +/- 11.8, the mean Mental Component Summary score was 44.2 +/-12.6, and both were significantly lower than norms for the general US population (p < 0.001). The median PHQ-9 score was 5 (range 0-27, IQR 8). A PHQ-9 score of 10 or greater, indicating moderate to severe depression, was observed in 29% of patients. The median ZBI score was 12 (range 0-27, IQR 11), indicating a low burden. Neurological deficit, use of pain medication, and requirement for corticosteroids were found to be associated with worse SF-36 Physical Component Summary score, while higher levels of depression (higher PHQ-9 score) correlated with worse SF-36 Mental Component Summary score. Conclusions Patients with skull base chordomas have a lower quality of life than the general US population. The most significant determinants of quality of life in the posttreatment phase in this patient population were neurological deficits (sensory deficit and bowel/bladder dysfunction), pain medication use, corticosteroid use, and levels of depression as scored by PHQ-9.

[46]

**TÍTULO / TITLE:** - The Polycomb group (PcG) protein EZH2 supports the survival of PAX3-FOXO1 alveolar rhabdomyosarcoma by repressing FBXO32 (Atrogin1/MAFbx).

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Ciarapica R; De Salvo M; Carcarino E; Bracaglia G; Adesso L; Leoncini PP; Dall’agnese A; Walters ZS; Verginelli F; De Sio L; Boldrini R; Inserra A; Bisogno G; Rosolen A; Alaggio R; Ferrari A; Collini P; Locatelli M; Stifani S; Screpanti I; Rutella S; Yu Q; Marquez VE; Shipley J; Valente S; Mai A; Miele L; Puri PL; Locatelli F; Palacios D; Rota R

**INSTITUCIÓN / INSTITUTION:** - Department of Oncohematology, Ospedale Pediatrico Bambino Gesu, IRCCS, Roma, Italy.

**RESUMEN / SUMMARY:** - The Polycomb group (PcG) proteins regulate stem cell differentiation via the repression of gene transcription, and their deregulation has been widely implicated in cancer development. The PcG protein Enhancer of Zeste Homolog 2 (EZH2) works as a catalytic subunit of the Polycomb Repressive Complex 2 (PRC2) by methylating lysine 27 on histone H3 (H3K27me3), a hallmark of PRC2-mediated gene repression. In skeletal muscle progenitors, EZH2 prevents an unscheduled differentiation by repressing muscle-specific gene expression and is downregulated during the course of differentiation. In rhabdomyosarcoma (RMS), a pediatric soft-tissue sarcoma thought to arise from myogenic precursors, EZH2 is abnormally expressed and its downregulation in vitro leads to muscle-like differentiation of RMS cells of the embryonal variant. However, the role of EZH2 in the clinically aggressive subgroup of alveolar RMS, characterized by the expression of PAX3-FOXO1
oncoprotein, remains unknown. We show here that EZH2 depletion in these cells leads to programmed cell death. Transcriptional derepression of F-box protein 32 (FBXO32) (Atrogin1/MAFbx), a gene associated with muscle homeostasis, was evidenced in PAX3-FOXO1 RMS cells silenced for EZH2. This phenomenon was associated with reduced EZH2 occupancy and H3K27me3 levels at the FBXO32 promoter. Simultaneous knockdown of FBXO32 and EZH2 in PAX3-FOXO1 RMS cells impaired the pro-apoptotic response, whereas the overexpression of FBXO32 facilitated programmed cell death in EZH2-depleted cells. Pharmacological inhibition of EZH2 by either 3-Deazaneplanocin A or a catalytic EZH2 inhibitor mirrored the phenotypic and molecular effects of EZH2 knockdown in vitro and prevented tumor growth in vivo. Collectively, these results indicate that EZH2 is a key factor in the proliferation and survival of PAX3-FOXO1 alveolar RMS cells working, at least in part, by repressing FBXO32. They also suggest that the reducing activity of EZH2 could represent a novel adjuvant strategy to eradicate high-risk PAX3-FOXO1 alveolar RMS. Oncogene advance online publication, 11 November 2013; doi:10.1038/onc.2013.471.

[47] TÍTULO / TITLE: - Primary pulmonary artery Rosai-Dorfman disease mimicking sarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   - Enlace al texto completo (gratuito o de pago) 1016/j.jtcvs.2013.08.001
AUTORES / AUTHORS: - Morsolini M; Nicola M; Paulli M; D'Armini AM
INSTITUCIÓN / INSTITUTION: - Division of Cardiac Surgery, University of Pavia School of Medicine, Foundation I.R.C.C.S. Policlinico San Matteo, Pavia, Italy.

[48] TÍTULO / TITLE: - Desmoid-type fibromatosis of the head and neck region in the pediatric population: A clinicopathological and genetic study of 7 cases.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   - Enlace al texto completo (gratuito o de pago) 1111/his.12323
AUTORES / AUTHORS: - Flucke U; Tops BB; van Diest PJ; Slootweg PJ
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Radboud University Nijmegen Medical Center, Nijmegen, The Netherlands.
RESUMEN / SUMMARY: - AIMS: Desmoid-type fibromatosis (desmoids) is a locally aggressive (myo)fibroblastic lesion. It represents one of the more common fibrous tumours in children and adolescents. The head and neck region is more often involved when compared to adults. METHODS AND RESULTS: We investigated the clinicopathological and genetic characteristics of seven pediatric desmoids at this anatomic site, including two cases of desmoplastic fibroma, located in the mandible. There were two females and five males with an age range from 1.5 - 8 years. Sites of the soft tissue lesions were sinonasal (n=4), and paramandibular (n=1). All cases showed typical morphology and nuclear beta-catenin expression. CTNNB1 gene sequencing, successfully performed in five cases, revealed mutations in three cases
with one p.T41A (bone lesion), one p.S37A and one novel mutation, p.D32V (sinonasal soft tissue lesion each). Seven patients were treated by excision with positive margins in five cases. Follow-up, available for six patients (median, 4 years) showed no evidence of disease in four cases, slow progression in one case and recurrence with stable disease in the last case. CONCLUSION: Our study provides evidence of genetic similarities in desmoid and desmoplastic fibroma. Additionally, we expanded the spectrum of mutations in CTNNB1 with one for desmoid novel mutation. This article is protected by copyright. All rights reserved.

[49]

**Título / Title:** Myxoinflammatory Fibroblastic Sarcoma: A Clinicopathologic Analysis of 104 Cases, With Emphasis on Predictors of Outcome.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


**Autores / Authors:** Laskin WB; Fetsch JF; Miettinen M

**Institución / Institution:** *Department of Pathology, Feinberg School of Medicine, Northwestern University, Chicago, IL daggerDepartment of Soft Tissue Pathology, Joint Pathology Center, Silver Spring double daggerLaboratory of Pathology, National Institutes of Health/National Cancer Institute, Bethesda, MD.

**Resumen / Summary:** In this study, we examine the clinicopathologic features of 104 cases of myxoinflammatory fibroblastic sarcoma (MIFS), a low-grade, inflammatory fibromyxoid tumor with a predilection to distal extremity soft tissue, and attempt to identify factors predictive of aggressive behavior. The study cohort consisted of 49 male and 55 female patients ranging in age from 17 to 83 (mean, 42; median, 39) years. The tumor arose primarily on the dorsal aspect of the distal extremities as a solitary and usually painless mass. Tumors ranged in size from 0.5 to 15 (mean, 3.2; median; 2.4) cm. Microscopically, tumors consisted of variably cellular and inflamed fibromyxoid tissue growing as a lobulated mass or as multiple nodules within subcutaneous tissue or along tendinofascial planes. Tumor cells ranged from plump spindled to more epithelioid cells with enlarged, vesicular nuclei. Characteristic of the process was a strikingly bizarre cell with an inclusion body-like nucleolus (85% of cases) and/or a smudgy hyperchromatic nucleus (51%) present in all but 7 cases. The mitotic rate per 50 high-power field ranged from 0 to 13 (mean, 2.9; median, 2) mitoses. Twenty-two tumors demonstrated 1 or more of the following atypical features: (1) foci with complex sarcoma-like vasculature; (2) hypercellular areas; and (3) increased mitotic activity or atypical mitotic figures. Immunohistochemically, tumor cells demonstrated immunoreactivity for vimentin (100%), D2-40 (86%), CD34 (50%), keratin(s) (33%), CD68 (27%), actin(s) (26%), desmin (9%), S-100 protein (7%), and epithelial membrane antigen (6%). Thirty of 59 patients (51%) with follow-up data suffered (at least) 1 local recurrence, and 1 patient developed metastatic disease after multiple local recurrences. Completeness of initial surgical excision was the only clinicopathologic parameter that statistically correlated with a lower incidence of recurrence (P=0.004). Histologically atypical MIFS recurred more often than conventional tumors (67% vs. 47%), but the difference was not statistically significant (P=0.35). Our study shows that histologic features often associated with more
aggressive sarcomas do not substantially impact the morbidity of MIFS, and complete surgical excision provides the best chance for disease-free survival.

[50]
**TÍTULO / TITLE:** - Complete regression of primary epicardial leiomyosarcoma with single agent doxorubicin in an elderly patient.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Karavelioglu Y; Arisoy A; Sen F

[51]
**TÍTULO / TITLE:** - Sentinel lymph node biopsy in pediatric soft tissue sarcoma patients: utility and concordance with imaging.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Alcorn KM; Deans KJ; Congeni A; Sulkowski JP; Bagatell R; Mattei P; Minneci PC
**INSTITUCIÓN / INSTITUTION:** - Perelman School of Medicine, University of Pennsylvania, USA.

**RESUMEN / SUMMARY:** - BACKGROUND: The purpose of this study was to report our experience with sentinel lymph node biopsy (SLNB) for pediatric soft tissue sarcomas to add to the limited literature about its feasibility, utility, and concordance with pre-operative imaging, including CT and (18)F-FDG PET (PET) scanning. METHODS: Medical records of patients with a sarcoma who underwent SLNB as part of their treatment for a soft tissue sarcoma at our institution from 2000 to 2011 were identified and reviewed. RESULTS: Eight patients underwent SLNB for soft tissue sarcoma during the study period. Two patients had positive SLNBs; both of these patients had rhabdomyosarcoma. Three patients with pathologically enlarged lymph nodes on CT scan underwent PET functional imaging prior to SLNB. The PET suggested the presence of nodal disease in all three patients; however, only one of these patients had a positive SLNB. CONCLUSIONS: Our series confirms that SLNB is feasible in pediatric sarcoma patients. Small numbers preclude definitive conclusions regarding the utility of SLNB compared with PET, however our data suggest functional imaging alone may not be sufficient to definitively determine lymph node status in these patients. Surgical lymph node sampling may still need to be performed to accurately identify nodal status in pediatric patients with soft tissue sarcoma.

[52]
**TÍTULO / TITLE:** - DeltaNp63 promotes pediatric neuroblastoma and osteosarcoma by regulating tumor angiogenesis.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: Bid HK; Roberts RD; Cam M; Audino A; Kurmasheva RT; Lin J; Houghton PJ; Cam H

INSTITUCIÓN / INSTITUTION: Center for Childhood Cancer, Nationwide Children's Hospital.

RESUMEN / SUMMARY: The tumor suppressor gene p53 and its family members p63/p73 are critical determinants of tumorigenesis. DeltaNp63 is a splice variant of p63 which lacks the N-terminal transactivation domain. It is thought to antagonize p53-, p63- and p73-dependent translation, thus blocking their tumor suppressor activity. In our studies of the pediatric solid tumors neuroblastoma and osteosarcoma, we find overexpression of DeltaNp63; however, there is no correlation of DeltaNp63 expression with p53 mutation status. Our data suggest that DeltaNp63 itself endows cells with a gain of function that leads to malignant transformation, a function independent of any p53 antagonism. Here, we demonstrate that DeltaNp63 overexpression, independent of p53, increases secretion of interleukin-6 (IL-6) and interleukin-8 (IL-8), leading to elevated phosphorylation of STAT-3 (Tyr-705). We show that elevated phosphorylation of STAT-3 leads to stabilization of HIF-1alpha protein, resulting in VEGF secretion. We also show human clinical data suggesting a mechanistic role for DeltaNp63 in osteosarcoma metastasis. In summary, our study reveals the mechanism by which DeltaNp63, as a master transcription factor, modulates tumor angiogenesis.

[53] TÍTULO / TITLE: From image statistics to scene gist: evoked neural activity reveals transition from low-level natural image structure to scene category.

AUTORES / AUTHORS: Groen II; Ghebreab S; Prins H; Lamme VA; Scholte HS

INSTITUCIÓN / INSTITUTION: Cognitive Neuroscience Group, Department of Psychology, Amsterdam Center for Brain and Cognition, Institute for Interdisciplinary Studies, and Intelligent Systems Laboratory Amsterdam, Institute of Informatics, University of Amsterdam. 1018 WS, Amsterdam, The Netherlands.

RESUMEN / SUMMARY: The visual system processes natural scenes in a split second. Part of this process is the extraction of "gist," a global first impression. It is unclear, however, how the human visual system computes this information. Here, we show that, when human observers categorize global information in real-world scenes, the brain exhibits strong sensitivity to low-level summary statistics. Subjects rated a specific instance of a global scene property, naturalness, for a large set of natural scenes while EEG was recorded. For each individual scene, we derived two physiologically plausible summary statistics by spatially pooling local contrast filter outputs: contrast energy (CE), indexing contrast strength, and spatial coherence (SC), indexing scene fragmentation. We show that behavioral performance is directly related to these statistics, with naturalness rating being influenced in particular by SC. At the
neural level, both statistics parametrically modulated single-trial event-related potential amplitudes during an early, transient window (100-150 ms), but SC continued to influence activity levels later in time (up to 250 ms). In addition, the magnitude of neural activity that discriminated between man-made versus natural ratings of individual trials was related to SC, but not CE. These results suggest that global scene information may be computed by spatial pooling of responses from early visual areas (e.g., LGN or V1). The increased sensitivity over time to SC in particular, which reflects scene fragmentation, suggests that this statistic is actively exploited to estimate scene naturalness.

[54]

**TITULO / TITLE:** Endobronchial lipomatous tumors: clinicopathologic analysis of 12 cases with molecular cytogenetic evidence supporting classification as "lipoma".

**RESUMEN / SUMMARY:** Lipomatous lesions rarely involve the bronchial tree, and detailed morphologic and molecular cytogenetic analysis of these tumors is lacking. The clinicopathologic features of 12 endobronchial lipomatous neoplasms were studied, with ancillary fluorescence in situ hybridization performed in subsets of cases for CPM, which is amplified in atypical lipomatous tumors/well-differentiated liposarcomas (ALT/WDL), and HMGA1 and HMGA2, which are often rearranged in lipomas. The cases occurred predominately in older men (91%) (age range 44 to 80 y, mean 65 y). Most patients (80%) had a former or current history of heavy smoking (20 to 100 pack-years). Three patients had concurrent pulmonary squamous cell carcinoma, and 1 had a history of multiple lung cancers. Most lesions were small (<2.5 cm) and discovered incidentally. A subset of tumors showed atypical morphologic features that would be suggestive of ALT/WDL in soft tissue sites, including regions of fibrosis and scattered hyperchromatic stromal cells. However, all cases with atypia were CPM negative and behaved in a clinically benign manner. Seven cases were tested for HMGA1 and HMGA2 rearrangement; 4 showed HMGA2 rearrangement, and 1 showed HMGA1 rearrangement, consistent with lipomas. Two cases were negative for HMGA1/2 rearrangements. We conclude that endobronchial lipomatous neoplasms represent lipomas, even in the presence of morphologic features suggestive of ALT/WDL. Ancillary fluorescence in situ hybridization testing may be very valuable in the analysis of these rare tumors, as true ALT/WDL seem to be very rare or nonexistent at this anatomic site.

[55]

RESUMEN / SUMMARY: Giant cell lesions of bone share similar clinical, radiological, and histological features. The most challenging differential diagnosis is between giant cell tumor (GCT) and brown tumor (BT) secondary to hyperparathyroidism. Differential diagnosis is based on determining serum calcium concentration and other markers of calcium metabolism. The authors present the unusual case of a 37-year-old Caucasian woman affected by a GCT of the proximal left tibia and concomitant asymptomatic primary hyperparathyroidism (PHPT) due to a parathyroid adenoma. The presence of two concurrent diseases complicated diagnosis and relative treatment. The patient was first treated for the adenoma, then after 9 months, she underwent curettage of tibial GCT. Denosumab treatment was administered for 12 months to control a relapse occurring at 15 months post-curettage. At 32-month follow-up from primary tibial surgery, the patient was free from tumor disease. To our knowledge, this is the first case in the literature reporting the concomitant presence of asymptomatic PHPT and GCT. The possibility of concomitant finding these two diseases has to be considered during the decision-making process.

[56]

TÍTULO / TITLE: Hibernoma: a possible clinical model for exploring the role of brown adipose tissue in the regulation of body weight?

RESUMEN / SUMMARY: Hibernoma is a rare benign tumor histologically similar to brown adipose tissue. Some studies reported weight loss in patients with this tumor; however the mechanisms have never been investigated.OBJECTIVE:The purpose of this study is to explore the impact of hibernoma resection on the whole body metabolism.PATIENT AND METHODS:A 68-year-old woman was examined following a weight loss of 10 kg in 6 months. Body composition, food intake, physical activity, blood levels of thyroid hormones and lipid profile were assessed before and during one year after surgery. Patient’s resting energy expenditure (REE) over time was compared to a control group of 18 matched healthy volunteers.RESULTS:Within one year after hibernoma resection, the patient gained +15kg of body weight. This was associated with fat mass gain (+41%), mainly in the abdominal region (+48%). The patient also developed hepatic non-alcoholic steatosis, mild hypertriglyceridemia, and reduced...
levels of high-density lipoproteins. REE increased during the dynamic phase of weight gain, compared to the pre-surgery measurement, and returned to baseline after one year. Food intake was increased by 37.5% six weeks after resection of the hibernoma, and returned to baseline values within 6 months. CONCLUSIONS: In our study conditions, hibernoma did not alter REE, but weight gain did. Specific physical activities and dietetic follow-ups are suggested for those patients, in order to prevent excess fat mass gain and metabolic disorders after hibernoma resection. More studies should focus on hibernoma mechanisms inducing weight loss.
PROGNOSTIC FACTORS AND OUTCOMES IN ENDOMETRIAL STROMAL SARCOMA WITH THE 2009 FIGO STAGING SYSTEM: A MULTICENTER REVIEW OF 114 CASES.

OBJECTIVE: To assess prognostic factors associated with disease-related survival in endometrial stromal sarcoma (ESS) using the 2009 FIGO staging system. METHODS: From January 1990 to January 2012, 114 patients with ESS were identified at the Samsung and Asan Medical Center and data were retrospectively analyzed. RESULTS: Ten (8.7%) patients died of the disease and 33 (28.9%) patients relapsed. The 5- and 10-year overall survival (OS) rates for the entire cohort were 92.6% and 87.1%, respectively, and the 5- and 10-year recurrence-free survival (RFS) rates were 71.8% and 52.1%, respectively. The estimated median survival after recurrence for the 33 patients whose tumors relapsed was 133 months (95% CI, 7.7-258.4), and 5-year survival after recurrence was 68.9%. Stage I (P=0.006), estrogen and/or progesterone receptor (ER/PR) positivity (P=0.0027), and no nodal metastasis (P=0.033) were associated with a good prognosis for OS in the univariate analysis. Ovarian preservation was revealed to be an independent predictor for poorer RFS (HR, 6.5; 95% CI, 1.23-34.19; P=0.027). Positivity for ER/PR (HR, 0.05; 95% CI, 0.006-0.4; P=0.006) and cytoreductive resection of recurrent lesions (HR, 0.14; 95% CI, 0.02-0.93; P=0.042) were independent predictors of better survival after recurrence. CONCLUSIONS: Stage, expression of ER/PR, and nodal metastasis are significantly associated with OS in ESS. Bilateral salpingo-oophorectomy (BSO) as the primary treatment and cytoreductive resection of recurrent lesions should be considered for improving survival of patients with ESS.

ENDOMETRIOID STROMAL SARCOMA: A CLINICOPATHOLOGIC STUDY OF 63 CASES.

CONCLUSIONS: Stage, expression of ER/PR, and nodal metastasis are significantly associated with OS in ESS. Bilateral salpingo-oophorectomy (BSO) as the primary treatment and cytoreductive resection of recurrent lesions should be considered for improving survival of patients with ESS.

ENDOMETRIOID STROMAL SARCOMA (ALSO KNOWN AS EXTRAUTERINE ENDOMETRIAL STROMAL SARCOMA [EESS]) IS AN UNCOMMON TUMOR THAT OCCURS IN WOMEN OVER A WIDE AGE RANGE. THE EXTRAUTERINE LOCATION, NON-GYNECOLOGIC SYMPTOMS AND
signs at presentation, and confounding histologic features can pose a diagnostic challenge. In this study, we present the clinicopathologic features of 63 cases of EESS seen during a period of 21 years at our institution. Clinical information and pathology material were reviewed. Ages ranged from 27 to 87 years (median: 50 years). The most common symptoms and signs were an abdominal or pelvic mass, pain, vaginal bleeding, and gastrointestinal symptoms. The tumor size ranged from 1.2 to 24.5 cm. The most common sites of involvement were the ovaries (25), bowel wall (28), abdomen/peritoneum (37), pelvis (20), and vagina (6). Multiple sites were involved in 40 cases. Forty-six of 49 tumors had a classic microscopic appearance, and 3 had dedifferentiation; in 20 cases, there was vascular invasion. Fibroma-like stroma was seen in 30, hyaline plaques in 23, sex cord elements in 11, smooth muscle differentiation in 4, and myxoid change in 4 cases. Endometriosis was noted in 30 cases. Immunohistochemical results included: CD10 positivity in 31, desmin positivity in 9 (focal), estrogen receptor positivity in 28, and progesterone receptor positivity in 33 cases. In 25% of cases, an initial diagnosis other than EESS was made: sex cord-stromal tumors (4), gastrointestinal stromal tumor (3), leiomyosarcoma (3), liposarcoma (1), mullerian adenosarcoma (1), synovial sarcoma (1), malignant peripheral nerve sheath tumor (1), small round blue cell tumor (1), and atypical stromal endometriosis (1). Primary treatment was cytoreductive surgery for 61 patients and hormonal therapy for 2 patients. Adjuvant treatment included hormonal therapy, chemotherapy, and radiation therapy. Follow-up (5 to 336 months) information was available for 53 patients: alive with no evidence of disease, 29; alive with disease, 15; and dead of disease, 9 (median period of 70 months from diagnosis to death). Thirty-three patients had recurrent disease, and 10 patients were lost to follow-up. EESS is commonly associated with endometriosis and tends to be indolent with a propensity for recurrence. Seven of 9 patients who died of the disease had bowel involvement, and 3 had tumors with dedifferentiation. Besides the latter, no other histologic finding correlated with the clinical behavior of these tumors.

[60]

**Título / Title:** Transactivating mutation of the MYOD1 gene is a frequent event in adult spindle cell rhabdomyosarcoma.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


**Autores / Authors:** Szuhai K; de Jong D; Leung WY; Fletcher CD; Hogendoorn PC

**Institución / Institution:** Department of Molecular Cell Biology, Leiden University Medical Center, Leiden, The Netherlands.

**Resumen / Summary:** Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children and adolescents, being characterised by expression of genes, morphological and ultrastructural features of sarcomeric differentiation. The spindle cell variant of rhabdomyosarcoma (spindle cell RMS) in adults has been defined as an entity, separated from embryonal rhabdomyosarcoma (ERMS) with unfavourable clinical outcome. So far no recurrent genetic alteration has been identified in the adult form of spindle cell RMS. We studied a case of adult spindle cell RMS using next generation sequencing (NGS) after exome capturing. Using this approach,
we identified 31 tumour specific somatic alterations and selected 4 genes with predicted functional relevance to muscle differentiation and growth. MYOD1, KIF18A, NOTCH1, and EML5 were further tested for mutations using Sanger sequencing on DNA from FFPE samples from 16 additional, adult spindle cell RMS samples. The highly conserved sequence homology of MYOD1 with other myogenic transcription factors prompted us to screen the basic DNA-binding domains of MYF5, MYF6 and MYOG for mutations. From the investigated 17 samples, 7 (41%) showed homozygous mutation of MYOD1 indicating a critical role in this rare subtype of adult spindle cell RMS while no mutation were found in any of the other genes involved in myogenic differentiation. The p.L122R mutation occurs in the conserved DNA binding domain in MYOD1 and leads to transactivation and MYC-like functions. MYOD1 homozygous mutations are frequent, recurrent and pathognomonic events in adult type spindle cell RMS.

[61] TÍTULO / TITLE: - What are the current outcomes of advanced gastrointestinal stromal tumors: who are the long-term survivors treated initially with imatinib?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Rutkowski P; Andrzejuk J; Bylina E; Osuch C; Switaj T; Jerzak Vel Dobosz A; Grzesiakowska U; Jurkowska M; Wozniak A; Limon J; Debiec-Rychter M; Siedlecki JA
INSTITUCIÓN / INSTITUTION: - Department of Soft Tissue/Bone Sarcoma and Melanoma, Maria Sklodowska-Curie Memorial Cancer Center and Institute of Oncology, Roentgena 5, 02-781, Warsaw, Poland, rutkowskip@coi.waw.pl.
RESUMEN / SUMMARY: - The introduction of imatinib to clinical practice revolutionized therapy of advanced gastrointestinal stromal tumors (GIST), but its long-term results have been only just collected. We have attempted to identify factors related to the long-term survival. We have analyzed the data of 430 inoperable/metastatic/recurrent GIST patients treated with imatinib in reference centers, assessed the factors influencing the long-term overall survival (OS), and compared the outcomes in three periods of initiation of imatinib therapy during one decade (2001-2003, 2004-2006, 2007-2010). During analyzed time periods, we have found decrease in median largest tumor size at the start of imatinib therapy: 90.5 mm (2001-2003) versus 74 mm (2004-2006) versus 58 mm (2007-2010) (p = 0.002). Median progression-free survival (PFS) on 1st line imatinib was 37.5 months, without differences in PFS between three groups. Median OS was 5.8 years, 8-year OS rate was 43 %, and no difference in OS was demonstrated for patients treated in analyzed time periods. Independent good prognostic factors for longer OS were as follows: surgery of residual disease, initial WHO performance status 0/1, normal baseline albumin level, and the presence of exon 11 KIT mutations. Current median OS in advanced GIST reaches 6 years. The long-term survivors were characterized by smaller maximal tumors at imatinib start, better blood tests results, better performance status, and the surgical removal of residual disease. The latter might reduce the impact of tumor size and equalize the long-term results of therapy during last decade from introduction of imatinib. After introduction of
subsequent lines of therapy (as sunitinib), the effect of primary mutational status on the long-term OS is also less visible.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Froeling V; Meckelburg K; Schreiter NF; Scheurig-Muenkler C; Kamp J; Maurer MH; Beck A; Hamm B; Kroencke TJ
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Charite - Universitätsmedizin Berlin, Campus Virchow, Augustenburger Platz 1, 13353 Berlin, Germany. Electronic address: Vera.Froeling@charite.de.
RESUMEN / SUMMARY: - OBJECTIVES: To compare the long-term outcome after uterine artery embolization (UAE) versus magnetic resonance-guided high-intensity focused ultrasound (MR-g HIFU) for symptomatic uterine fibroids. METHODS: Seventy-seven women (median age, 39.3 years; range, 29.2-52.2 years) with symptomatic uterine fibroids, equally eligible for UAE and MR-g HIFU based on our exclusion criteria underwent treatment (UAE, N=41; MR-g HIFU, N=36) from 2002 to 2009 at our institution. Symptom severity (SS) and total health-related quality of life (Total HRQoL) scores were assessed by the uterine fibroid symptom and quality of life (UFS-QoL) questionnaire before treatment and at long-term follow-up after UAE (median 61.9 months) and after MR-g HIFU (median: 60.7 months). Re-intervention rates were assessed for each therapy and compared. RESULTS: Re-intervention was significantly lower after UAE (12.2%) than after MR-g HIFU (66.7%) at long-term follow-up (p<0.001). After UAE changes in SS (50 pre-treatment vs. 6.3 post-treatment) and Total HRQoL (57.8 pre-treatment vs. 100 post-treatment) were significantly better than changes in SS (42.2 pre-treatment vs. 26.6 post-treatment) and Total HRQoL score (66.4 pre-treatment vs. 87.9 post-treatment) after MR-g HIFU (p=0.019 and 0.049 respectively). CONCLUSIONS: Improvement of SS and Total HRQoL scores was significantly better after UAE resulting in a significant lower re-intervention rate compared to MR-g HIFU.

[63] TÍTULO / TITLE: - Efficient infection of a human B cell line with cell-free Kaposi's sarcoma-associated herpesvirus.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Dollery SJ; Santiago-Crespo RJ; Kardava L; Moir S; Berger EA
INSTITUCIÓN / INSTITUTION: - Laboratory of Viral Diseases.
RESUMEN / SUMMARY: - Kaposi's sarcoma-associated herpesvirus (KSHV) is causatively linked to two B cell lymphoproliferative disorders, multicentric Castleman's
disease and primary effusion lymphoma. Latently infected B cells are a major KSHV reservoir, and virus activation from tonsillar B cells can result in salivary shedding and virus transmission. Paradoxically, human B cells (primary and continuous) are notoriously refractory to infection, thus posing a major obstacle to the study of KSHV in this cell type. By performing a strategic search of human B cell lymphoma lines, we found that MC116 cells were efficiently infected by cell-free KSHV. Upon exposure to recombinant KSHV.219, EGFP reporter expression was detected in 17-20% of MC116 cells. Latent phase transcription and protein synthesis were detected by RT-PCR and latency-associated nuclear antigen expression in cell lysates and individual cells. Selection based on the puromycin-resistance gene in KSHV.219 yielded cultures with all cells infected. After repeated passaging of the selected KSHV-infected cells without puromycin, latent KSHV was maintained in a small fraction of cells. Infected MC116 cells could be induced into lytic phase with histone deacetylase inhibitors as known for latently infected non-B cell lines, and also selectively by the B cell-specific pathway involving B cell receptor crosslinking. Lytic phase transition was documented by RFP reporter expression, late structural glycoprotein detection (K8.1A, gH), and infectious KSHV production. MC116 cells were CD27-/CD10+, characteristic of transitional B cells. These findings represent an important step in the establishment of an efficient continuous B cell line model to study the biologically relevant steps of KSHV infection.
is coordinated by IKKbeta and IKKepsilon that sequentially phosphorylate RelA in a site-specific manner to enable latent infection after KSHV de novo infection.

[65] **TÍTULO / TITLE:** miR-203, a tumor suppressor frequently down-regulated by promoter hypermethylation in Rhabdomysarcoma.  
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary  
**AUTORES / AUTHORS:** Diao Y; Guo X; Jiang L; Wang G; Zhang C; Wan J; Jin Y; Wu Z  
**INSTITUCIÓN / INSTITUTION:** Shenzhen-PKU-HKUST Medical Center, China;  
**RESUMEN / SUMMARY:** Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma found in children and young adults. It is characterized by the expression of a number of skeletal muscle-specific proteins including MyoD and muscle alpha-actin. However, unlike normal myoblasts, RMS cells differentiate poorly both in vivo and in culture. As microRNAs are known to regulate tumorigenesis, intensive efforts have been made to identify microRNAs that are involved in RMS development. In the current study, we found that miR-203 was frequently down-regulated by promoter hypermethylation in both RMS cell lines and RMS biopsies and could be reactivated by DNA-demethylating agents. Re-expression of miR-203 in RMS cells inhibited their migration and proliferation, and promoted terminal myogenic differentiation. Mechanistically, miR-203 exerts its tumor suppressive effect by directly targeting p63 and leukemia inhibitory factor receptor in RMS cells, which promotes myogenic differentiation by inhibiting the Notch and the JAK1/STAT1/STAT3 pathways, respectively. Our work reveals that miR-203 functions as a tumor suppressor in RMS development.

[66] **TÍTULO / TITLE:** Prenatal Diethylstilbestrol Exposure and Risk of Uterine Leiomyomata in the Nurses’ Health Study II.  
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary  
**AUTORES / AUTHORS:** Mahalingaiah S; Hart JE; Wise LA; Terry KL; Boynton-Jarrett R; Missmer SA  
**RESUMEN / SUMMARY:** Previous studies evaluating the association of prenatal exposure to diethylstilbestrol (DES), a potent endocrine disruptor, with incidence of uterine leiomyomata (UL) have had conflicting results. We evaluated the association between prenatal DES exposure and incident UL in women in the Nurses’ Health Study II from 1989 to 2009. Women were aged 25-42 years at enrollment and had a prenatal exposure window corresponding to DES use. The analytical sample was larger than previous studies and included 102,164 premenopausal women with intact uteri, no prior history of UL or cancer, and prenatal DES exposure. Multivariable-adjusted Cox proportional hazard models were used to estimate the relationship between DES exposure and UL risk. During 1,273,342 person-years of follow-up, there were 11,831
incident cases of UL. Women with prenatal exposure to DES had a higher incidence of UL compared with unexposed women, with an adjusted hazard ratio of 1.12 (95% confidence interval: 0.98, 1.27). Risk was strongest for women exposed to DES in the first trimester, when exposure corresponds to early stages of fetal Mullerian development (adjusted hazard ratio = 1.21, 95% confidence interval: 1.02, 1.43). These results suggest that first-trimester DES exposure may be associated with an increased risk of UL, but they must be interpreted with concern for detection and recall biases.
Leiomyomas (LMs) of the gastrointestinal tract arise within the muscularis mucosae (superficial) and muscularis propria (deep). There are isolated reports of KIT-positive cells, presumed interstitial cells of Cajal (ICCs), within gastrointestinal LMs. We have encountered esophageal LMs with a high proportion of KIT-positive and DOG1-positive spindle-shaped cells, an appearance that mimicked gastrointestinal stromal tumor. Our aim was to explore the prevalence of ICCs in LMs of the gastrointestinal tract and the etiopathogenic significance of these cells in this benign neoplasm. We identified 34 esophageal LMs (28 deep, 6 superficial), 8 gastric LMs, and 5 small-bowel LMs (all lesions in muscularis propria). We performed immunohistochemical staining studies for desmin, DOG1, and KIT on these neoplasms. We also evaluated 12 superficial colonic LMs. ICCs were distinguished from mast cells on the basis of morphology (elongated and occasionally branching spindle-shaped cells) and the presence of DOG1 reactivity. Four cases were screened for mutations in PDGFRA exons 12, 14, and 18 and KIT exons 9, 11, 13, and 17. ICCs were identified in all deep esophageal LMs and constituted an average of 20% of the lesional cells; focally, these cells comprised >50% of cells. The density of these cells was significantly higher than the background muscularis propria, and hyperplasia of ICCs was not identified in the adjacent muscle. ICCs were identified in 6 of 8 gastric LMs and 1 of 5 small-bowel LMs and were entirely absent in all superficial esophageal and colonic/rectal LMs. There were no mutations in KIT or PDGFRA. ICCs are universally present in deep esophageal LMs, and thus these neoplasms could be mistaken for gastrointestinal stromal tumors, particularly on biopsy samples, an error associated with adverse clinical consequences. ICCs are also identified in gastric and intestinal LMs, albeit in a smaller proportion of cases. Colonization and hyperplasia by non-neoplastic ICCs likely account for this phenomenon.
thromboembolic events. The aim of this study was to evaluate the relation between MPV level and CM. We compared the MPV levels between patients with CM and control subjects, and also evaluated the differences in the MPV levels in patients with CM before and after a successful surgical treatment. Furthermore, we compared the MPV levels in patients with and without cerebrovascular embolic symptoms. Fifty-one consecutive patients (13 men, 38 women, mean age 51.1 +/- 16.9 years) who underwent excision of CM in our hospital in the last 13 years and 50 normal subjects as the control group were included in this study. The patients with CM were divided into two groups according to the presence or absence of cerebrovascular embolic symptoms. The preoperative and postoperative MPV levels of each CM patient was evaluated. MPV levels were found to be significantly higher in patients with CM than in control subjects (9.86 +/- 1.30 fL vs. 7.63 +/- 0.78 fL, p < 0.001). Moreover, the MPV levels significantly decreased after the surgical removal of CM (9.86 +/- 1.30 fL vs. 8.68 +/- 1.20 fL, p < 0.001). We also found that the MPV values were significantly higher in patients with neurological embolic events (10.55 +/- 1.29 fL vs. 9.59 +/- 0.78 fL, p = 0.016). We conclude that increased MPV levels might be considered as a marker of increased thromboembolic risk in patients with CM.

[70]
TITULO / TITLE: - Granulocytic sarcoma of the ileum observed by double-balloon endoscopy before treatment (with video).
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Hotta K; Kunieda K
INSTITUCIÓN / INSTITUTION: - Division of Endoscopy and Gastrointestinal Oncology, Shizuoka Cancer Center, Japan Department of Gastroenterology, Saku Central Hospital, Japan.

[71]
TITULO / TITLE: - Multiple Metachronous Osteosarcomas in a Patient with Li-Fraumeni Syndrome.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Zils K; Wirth T; Lofst S; Biskup S; von Kalle T; Bielack S
INSTITUCIÓN / INSTITUTION: - 1 Pediatrics 5 (Oncology, Hematology, Immunology; Gastroenterology, Rheumatology, General Pediatrics), Klinikum Stuttgart -Olghospital, Germany.

[72]
TITULO / TITLE: - Progression of carcinogen-induced fibrosarcomas is associated with the accumulation of naive CD4+ T cells via blood vessels and lymphatics.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

**Autores / Authors:** Ondondo B; Jones E; Hindley J; Cutting S; Smart K; Bridgeman H; Matthews KK; Ladell K; Price DA; Jackson DG; Godkin A; Ager A; Gallimore A

**Institución / Institution:** Institute of Infection and Immunity, School of Medicine, Cardiff University, Cardiff, United Kingdom.

**Resumen / Summary:** The tumor microenvironment comprises newly formed blood and lymphatic vessels which shape the influx, retention and departure of lymphocytes within the tumor mass. Thus, by influencing the intratumoral composition of lymphocytes, these vessels affect the manner in which the adaptive immune system responds to the tumor, either promoting or impairing effective antitumor immunity. In our study, we utilized a mouse model of carcinogen-induced fibrosarcoma to examine the composition of tumor-infiltrating lymphocytes during tumor progression. In particular, we sought to determine whether CD4+ Foxp3+ regulatory T cells (Tregs) became enriched during tumor progression thereby contributing to tumor-driven immunosuppression. This was not the case as the proportion of Tregs and effector CD4+ T cells actually declined within the tumor owing to the unexpected accumulation of naive T cells. However, we found no evidence for antigen-driven migration of these T cells or for their participation in an antitumor immune response. Our data support the notion that lymphocytes can enter tumors via aberrantly formed blood and lymphatic vessels. Such findings suggest that targeting both the tumor vasculature and lymphatics will alter the balance of lymphocyte subpopulations that enter the tumor mass. A consideration of this aspect of tumor immunology may be critical to the success of solid cancer immunotherapies.

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**Título / Title:** Long-term Survival (>13 Years) in a Child With Recurrent Diffuse Pontine Gliosarcoma: A Case Report.

**Resumen / Summary:** Pediatric gliosarcoma (GS) is a rare variant of glioblastoma multiforme. The authors describe the case of an unusual pontine location of GS in a 9-year-old boy who was initially diagnosed with low-grade astrocytoma (LGA) that was successfully controlled for 4 years. Subsequently, his brain tumor transformed into a GS. Prior treatment of his LGA included subtotal tumor resection 3 times, standard radiation therapy, and Gamma Knife procedure twice. His LGA was also treated with a standard chemotherapy regimen of carboplatin and vincristine, and his GS with subtotal resection, high-dose cyclophosphamide, and thiotepa with stem cell rescue and temozolomide. Unfortunately, he developed disseminated disease with multiple lesions and leptomeningeal involvement including a tumor occupying 80% of the pons. Upon presentation at our clinic, he had rapidly progressing disease. He received treatment with antineoplastons (ANP) A10 and AS2-1 for 6 years and 10 months under
special exception to our phase II protocol BT-22. During his treatment with ANP his tumor stabilized, then decreased, and, ultimately, did not show any metabolic activity. The patient’s response was evaluated by magnetic resonance imaging and positron emission tomography scans. His pathology diagnosis was confirmed by external neuropathologists, and his response to the treatment was determined by central radiology review. He experienced the following treatment-related, reversible toxicities with ANP: fatigue, xerostomia and urinary frequency (grade 1), diarrhea, incontinence and urine color change (grade 2), and grade 4 hypernatremia. His condition continued to improve after treatment with ANP and, currently, he complains only of residual neurological deficit from his previous surgery. He achieved a complete response, and his overall and progression-free survival is in excess of 13 years. This report indicates that it is possible to obtain long-term survival of a child with a highly aggressive recurrent GS with diffuse pontine involvement with a currently available investigational treatment. This is an open-access article distributed under the terms of the Creative Commons Attribution-NonCommercial-NoDerivatives 3.0 License, where it is permissible to download and share the work provided it is properly cited. The work cannot be changed in any way or used commercially.

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RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Van Raak SM; Meuffels DE; Van Leenders GJ; Oei EH
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Albert Schweitzer Hospital, Dordrecht, The Netherlands.
RESUMEN / SUMMARY: - Hyaline fibromatosis syndrome (HFS) is a rare, homozygous, autosomal recessive disease, characterized by deposition of hyaline material in skin and other organs, resulting in esthetic problems, disability, and potential life-threatening complications. Most patients become clinically apparent in the first few years of life, and the disorder typically progresses with the appearance of new lesions. We describe a rare case of a 20-year-old patient with juvenile-onset mild HFS who presented with a history of progressive anterior knee pain. Detailed magnetic resonance (MR) imaging findings with histopathological correlation are presented of hyaline fibromatosis of Hoffa’s fat pad, including differential diagnosis. The diagnosis of HFS is generally made on basis of clinical and histopathological findings. Imaging findings, however, may contribute to the correct diagnosis in patients who present with a less typical clinical course of HFS.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
It is well established that osteoblasts, the key cells involved in bone formation during development and in adult life, secrete a number of glycoproteins harboring autocrine and paracrine functions. Thus, investigating the osteoblastic secretome could yield important information for the pathophysiology of bone. In the present study, we characterized for the first time the secretome of human Hobit osteoblastic cells. We discovered that the secretome comprised 89 protein species including the powerful growth factor progranulin. Recombinant human progranulin (6nM) induced phosphorylation of mitogen-activated protein kinase in both Hobit and osteocytic cells and induced cell proliferation and survival. Notably, risedronate, a nitrogen-containing bisphosphonate widely used in the treatment of osteoporosis, induced the expression and secretion of progranulin in the Hobit secretome. In addition, our proteomic study of the Hobit secretome revealed that risedronate induced the expression of ERp57, HSP60 and HSC70, three proteins already shown to be associated with the prevention of bone loss in osteoporosis. Collectively, our findings unveil novel targets of risedronate-evoked biological effects on osteoblast-like cells and further our understanding of the mechanisms of action of this currently used compound.
their ability to consecutively generate tumorigenesis in NOD/SCID mice. The properties of xenograft tumor and explants cells were investigated by immunohistochemistry, cytogenetic, and FACS analysis. Anticancer drug susceptibility of primary CS was analyzed using CCK-8. RESULTS: Primary CS cells greater than 27 passages in vitro showed an ability of a series of xenograft tumorigenesis in vivo having the same marker expression and cytogenetic character as that of original tumor. In addition, explants of xenograft tumors retained their original characteristics in the in vitro culture system. Finally, the analysis of the susceptibility to anticancer drug revealed that primary CS cells were susceptible to both doxorubicin and nilotinib, which are tyrosine kinase inhibitors. CONCLUSIONS: The primary CS cells and the primary CS xenograft tumorigenesis introduce a new therapeutic model for targeting cancer and also explore a deeper understanding of generation of the tumor itself.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Bodega-Quiroga I; Tejedor-Togores P; Saez-Garcia MA; Peraza-Casajus JM; Rosado-Dawid N; Serrano-Munoz A

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Abdou M; Hayek S; Williams BR 3rd
INSTITUCIÓN / INSTITUTION: - J. Willis Hurst Internal Medicine Residency Program (Drs. Abdou and Hayek) and Division of Cardiology, Department of Medicine (Dr. Williams), Emory University School of Medicine, Atlanta, Georgia 30322.
RESUMEN / SUMMARY: - Atrial myxoma is the most common primary cardiac tumor. Patients with atrial myxoma typically present with obstructive, embolic, or systemic symptoms; asymptomatic presentation is very rare. To our knowledge, isolated association of atrial myxoma with hypertrophic cardiomyopathy has been reported only once in the English-language medical literature. We report the case of an asymptomatic 71-year-old woman with known hypertrophic cardiomyopathy in whom a left atrial mass was incidentally identified on cardiac magnetic resonance images. After surgical excision of the mass and partial excision of the left atrial septum, histopathologic analysis confirmed the diagnosis of atrial myxoma. The patient was placed on preventive implantable cardioverter-defibrillator therapy and remained asymptomatic. The management of asymptomatic cardiac myxoma is a topic of debate, because no reports definitively favor either conservative or surgical measures.

[79] TÍTULO / TITLE: - Major Efficacy of Trabectedin in 2 Metastatic Osteosarcoma Patients with Wild-Type Asp1104 ERCC5 Tumor Status.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Background: Treatment of osteosarcoma of the extremities consists of surgical resection preceded and followed by chemotherapy, including high-dose methotrexate or adriamycin-based protocols. When distant relapse occurs, therapeutic options are scarce. Trabectedin, a DNA-binding agent, is indicated for the treatment of patients with advanced soft tissue sarcomas after failure of anthracyclines and ifosfamide. In this indication, the 6-month progression-free survival is about 35-40%. Recent reports showed that some specific single nucleotide polymorphisms (SNPs) from DNA repair genes could be associated with sensitivity to trabectedin in soft tissue sarcomas. Case Reports: We report our experience of 2 metastatic, heavily pretreated osteosarcoma patients who were treated with trabectedin. Pyrosequencing analyses of tumors from both patients for several SNPs of the ERCC1, ERCC5 and BRAC1 genes were performed. Both patients showed major response to trabectedin, which was interestingly related with homozygoty of the common guanine allele of ERCC5 (G/G genotype; Asp/Asp) after pyrosequencing analysis of tumors from both patients. This polymorphism was previously shown to be associated with better outcome in soft tissue sarcoma patients treated with trabectedin. Conclusion: Homozygoty for the wild-type Asp1104 SNP of the ERCC5 gene was found in 2 cases of relapsed osteosarcoma, who responded to trabectedin. © 2013 S. Karger GmbH, Freiburg.
**TÍTULO / TITLE:** - Novel ZC3H7B-BCOR, MEAF6-PHF1, and EPC1-PHF1 fusions in ossifying fibromyxoid tumors-molecular characterization shows genetic overlap with endometrial stromal sarcoma.

**RESUMEN / SUMMARY:** - PHF1 gene rearrangements have been recently described in around 50% of ossifying fibromyxoid tumors (OFMT) including benign and malignant cases, with a small subset showing EP400-PHF1 fusions. In the remaining cases no alternative gene fusions have been identified. PHF1-negative OFMT, especially if lacking S100 protein staining or peripheral ossification, are difficult to diagnose and distinguish from other soft tissue mimics. In seeking more comprehensive molecular characterization, we investigated a large cohort of 39 OFMT of various anatomic sites, immunoprofiles and grades of malignancy. Tumors were screened for PHF1 and EP400 rearrangements by FISH. RNA sequencing was performed in two index cases (OFMT1, OFMT3), negative for EP400-PHF1 fusions, followed by FusionSeq data analysis, a modular computational tool developed to discover gene fusions from paired-end RNA-seq data. Two novel fusions were identified ZC3H7B-BCOR in OFMT1 and MEAF6-PHF1 in OFMT3. After being validated by FISH and RT-PCR, these abnormalities were screened on the remaining cases. With these additional gene fusions, 33/39 (85%) of OFMTs demonstrated recurrent gene rearrangements, which can be used as molecular markers in challenging cases. The most common abnormality is PHF1 gene rearrangement (80%), being present in benign, atypical and malignant lesions, with fusion to EP400 in 44% of cases. ZC3H7B-BCOR and MEAF6-PHF1 fusions occurred predominantly in S100 protein-negative and malignant OFMT. As similar gene fusions were reported in endometrial stromal sarcomas, we screened for potential gene abnormalities in JAZF1 and EPC1 by FISH and found two additional cases with EPC1-PHF1 fusions. © 2013 Wiley Periodicals, Inc.

**AUTORES / AUTHORS:** - Antonescu CR; Sung YS; Chen CL; Zhang L; Chen HW; Singer S; Agaram NP; Sboner A; Fletcher CD

**INSTITUCIÓN / INSTITUTION:** - Department of Pathology, Memorial Sloan-Kettering Cancer Center, New York, NY.


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**TÍTULO / TITLE:** - Small gastrointestinal stromal tumor in the stomach: identification of precursor for clinical gastrointestinal stromal tumor using c-kit and alpha-smooth muscle actin expression.

**RESUMEN / SUMMARY:** - PHF1 gene rearrangements have been recently described in around 50% of ossifying fibromyxoid tumors (OFMT) including benign and malignant cases, with a small subset showing EP400-PHF1 fusions. In the remaining cases no alternative gene fusions have been identified. PHF1-negative OFMT, especially if lacking S100 protein staining or peripheral ossification, are difficult to diagnose and distinguish from other soft tissue mimics. In seeking more comprehensive molecular characterization, we investigated a large cohort of 39 OFMT of various anatomic sites, immunoprofiles and grades of malignancy. Tumors were screened for PHF1 and EP400 rearrangements by FISH. RNA sequencing was performed in two index cases (OFMT1, OFMT3), negative for EP400-PHF1 fusions, followed by FusionSeq data analysis, a modular computational tool developed to discover gene fusions from paired-end RNA-seq data. Two novel fusions were identified ZC3H7B-BCOR in OFMT1 and MEAF6-PHF1 in OFMT3. After being validated by FISH and RT-PCR, these abnormalities were screened on the remaining cases. With these additional gene fusions, 33/39 (85%) of OFMTs demonstrated recurrent gene rearrangements, which can be used as molecular markers in challenging cases. The most common abnormality is PHF1 gene rearrangement (80%), being present in benign, atypical and malignant lesions, with fusion to EP400 in 44% of cases. ZC3H7B-BCOR and MEAF6-PHF1 fusions occurred predominantly in S100 protein-negative and malignant OFMT. As similar gene fusions were reported in endometrial stromal sarcomas, we screened for potential gene abnormalities in JAZF1 and EPC1 by FISH and found two additional cases with EPC1-PHF1 fusions. © 2013 Wiley Periodicals, Inc.

**AUTORES / AUTHORS:** - Mikami T; Nemoto Y; Numata Y; Hana K; Nakada N; Ichinoe M; Murakumo Y; Okayasu I

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the digestive tract. To find precursors for clinical GISTs of the stomach, small gastric stromal tumors of less than 3 cm were collected and examined immunohistochemically with analysis of the KIT mutation. Sixty-eight of 74 lesions were classified into 4 representative groups according to the expression of c-kit and alpha-smooth muscle actin (alphaSMA): group A, c-kit diffusely positive and alphaSMA negative (18 cases); group B, c-kit diffusely positive and alphaSMA focally positive (13); group C, c-kit focally positive and alphaSMA diffusely positive (27); and group D, c-kit negative and alphaSMA diffusely positive (10). Of the 4 groups, groups A and B of c-kit diffuse expression showed higher cellularity and labeling indices of p27(Kip1) and Ki-67 than did groups C and D of diffuse alphaSMA expression. Incidence of KIT exon 11 mutation in groups A and B was 86% (25/29), whereas that in groups C and D was 0% (0/20). Small gastric stromal tumors with c-kit diffuse expression were considered precursors for clinical GIST because they were significantly different from c-kit focally positive or negative tumors. The mutation of KIT is considered as an early event in tumorigenesis of GIST.
Discrepancy of uterine leiomyoma and myometrium to hypoxia-induced endoplasmic reticulum stress after uterine occlusion therapy accounts for therapeutic effect.

TÍTULO / TITLE: - Discrepancy of uterine leiomyoma and myometrium to hypoxia-induced endoplasmic reticulum stress after uterine occlusion therapy accounts for therapeutic effect.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


●● Enlace al texto completo (gratuito o de pago) 1007/s00404-013-3100-9

AUTORES / AUTHORS: - Xie Y; Tao X; Cheng Z; Guan Q; Yang W; Zhu Y

INSTITUCIÓN / INSTITUTION: - Department of Obstetrics and Gynecology, Yangpu District Central Hospital, 450, Tengyue Road, Yangpu District, Shanghai, 200090, China.

RESUMEN / SUMMARY: - PURPOSE: Uterine artery occlusion (UAO) is a promising method for the treatment of leiomyoma. This study is intended to demonstrate the discrepancy of ER stress-induced apoptosis in leiomyoma and myometrium as a result of UAO therapy. METHODS: Primary cultured leiomyoma and myometrial cells were incubated in low oxygen supply (1 % O2). Then, real time RT-PCR and Western blotting were performed to analyze the mRNA and protein levels of ER stress-related molecules including GRP78, CHOP, JNK, Bax, Bcl-2 and Caspase4. Furthermore, the activity of Caspase4 was detected. Tissues of leiomyoma and myometria were also collected before and 30 min after UAO during surgery and evaluated. RESULTS: The leiomyoma cells and tissues expressed higher ER stress-related molecules compared to myometrial cells or tissues, while the levels of Bcl-2, an anti-apoptotic protein, declined. In myometrial cells, an elevated level of Caspase4 activation as well as its expression was not significant during the first 12 h, suggesting that hypoxia might not intensely affect the myometrium compared with leiomyoma. CONCLUSION: ER stress-related apoptosis partly accounts for the effects of UAO therapy on uterine leiomyoma, which leads to the death of leiomyoma while maintaining the survival of the uterus itself.

Gastrointestinal stromal tumors: risk assessment and adjuvant therapy.

TÍTULO / TITLE: - Gastrointestinal stromal tumors: risk assessment and adjuvant therapy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


●● Enlace al texto completo (gratuito o de pago) 1016/j.hoc.2013.07.004

AUTORES / AUTHORS: - Joensuu H

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RESUMEN / SUMMARY: - Adjuvant imatinib prolongs recurrence-free survival and probably overall survival of patients who have undergone surgery for gastrointestinal stromal tumor (GIST). Estimation of the risk of recurrence with a prognostication tool and tumor mutation analysis is essential before imatinib initiation, because approximately 60% of patients with GIST with operable tumor are cured by surgery.
alone and some mutated tyrosine kinases are insensitive to imatinib. Adjuvant imatinib is usually administered for 3 years at the dose of 400 mg once daily. Early detection of tumors that recur despite adjuvant therapy with longitudinal imaging of the abdomen is likely beneficial.

[86]
**Título / Title:** A non-comparative phase II study of dose intensive chemotherapy with doxorubicin and ifosfamide followed by high dose ICE consolidation with PBSCT in non-resectable, high grade, adult type soft tissue sarcomas.
**Resumen / Summary:** The objective was to determine the role of dose intensive induction chemotherapy in patients with soft tissue sarcomas (STS) that were considered unresectable. Treatment consisted of 2-3 cycles of doxorubicin (Dox) and ifosfamide (Ifo) followed by high dose chemotherapy with ifosfamide, carboplatin, etoposide (HD-ICE) plus peripheral blood stem cell transplantation (PBSCT). 30 out of 631 consecutive patients, median age 46 years (21-62), with high grade STS were included. 29 patients completed at least 2 cycles of Dox/Ifo. HD-ICE was withheld because of progressive disease (PD) in 5 patients, neurotoxicity in 6 cases, insufficient peripheral blood stem cell (PBSC) mobilization, complete remission (CR) and refusal in 1 patient each. HD-ICE was associated with non-haematological grade III toxicity including emesis, mucositis, fever, neurotoxicity, and transaminase level elevation. Two additional patients attained a partial response after HD-ICE. Overall, 24 of 30 (80 %) patients underwent surgery, with complete tumor resections in 19 patients (63 % of all patients, 79 % of the operated subgroup); however, 2 of these required amputation. After a median follow up period of 50 months in surviving patients (range, 26-120), 5-year PFS and OS rates were 39 % and 48 %, respectively. Induction chemotherapy plus consolidation HD-ICE is generally feasible, but is associated with significant neurotoxicity. The advantage of HD-ICE over conventional dose chemotherapy plus external beam radiation therapy (EBRT) in non-resectable disease remains unproven.

[87]
**Título / Title:** Vascular-targeted agents for the treatment of angiosarcoma.
**Resumen / Summary:** The objective was to determine the role of dose intensive induction chemotherapy in patients with soft tissue sarcomas (STS) that were considered unresectable. Treatment consisted of 2-3 cycles of doxorubicin (Dox) and ifosfamide (Ifo) followed by high dose chemotherapy with ifosfamide, carboplatin, etoposide (HD-ICE) plus peripheral blood stem cell transplantation (PBSCT). 30 out of 631 consecutive patients, median age 46 years (21-62), with high grade STS were included. 29 patients completed at least 2 cycles of Dox/Ifo. HD-ICE was withheld because of progressive disease (PD) in 5 patients, neurotoxicity in 6 cases, insufficient peripheral blood stem cell (PBSC) mobilization, complete remission (CR) and refusal in 1 patient each. HD-ICE was associated with non-haematological grade III toxicity including emesis, mucositis, fever, neurotoxicity, and transaminase level elevation. Two additional patients attained a partial response after HD-ICE. Overall, 24 of 30 (80 %) patients underwent surgery, with complete tumor resections in 19 patients (63 % of all patients, 79 % of the operated subgroup); however, 2 of these required amputation. After a median follow up period of 50 months in surviving patients (range, 26-120), 5-year PFS and OS rates were 39 % and 48 %, respectively. Induction chemotherapy plus consolidation HD-ICE is generally feasible, but is associated with significant neurotoxicity. The advantage of HD-ICE over conventional dose chemotherapy plus external beam radiation therapy (EBRT) in non-resectable disease remains unproven.
PURPOSE: Angiosarcomas are rare, aggressive vascular tumours known to express vascular endothelial growth factor (VEGF), a key pro-angiogenic growth factor. The aim of this study was to determine the potential effects of vascular-targeted agents for the treatment of angiosarcoma, using two human cutaneous angiosarcoma cell lines (ASM and ISO-HAS), and human dermal microvascular endothelial cells (HuDMECs) for comparison. METHODS: Protein arrays were used to assess the expression of angiogenesis-related proteins, and potential drug targets were assessed by ELISA and Western blotting. Response to vascular-targeted agents, including bevacizumab an anti-VEGF antibody, axitinib a VEGF-receptor tyrosine kinase inhibitor, everolimus an mTOR inhibitor, selumetinib a MEK inhibitor and vadimezan a vascular-disrupting agent were compared in functional in vitro cellular assays, including viability, differentiation and migration assays. RESULTS: ASM and ISO-HAS cells expressed a broad range of pro-angiogenic growth factors. ASM and ISO-HAS VEGF expression was significantly increased (p = 0.029) compared with HuDMECs. Striking responses were seen to vadimezan with an IC50 of 90 and 150 mug/ml for ASM and ISO-HAS cells, respectively. Selumetinib inhibited ASM with an IC50 of 1,750 ng/ml, but was not effective in ISO-HAS. Everolimus reduced both ASM and ISO-HAS viable cell counts by 20 % (p < 0.001). Minimal responses were observed to bevacizumab and axitinib in assays with ASM and ISO-HAS cells. CONCLUSIONS: Further studies are warranted to investigate mTOR inhibitors, MEK inhibitors and vascular-disrupting agents for the treatment of angiosarcoma.
RESULTS: Median tumor size was 4.0 cm (range 2-12). Objective response, assessed on 71 target lesions, was 92.2 % (complete 32.3, 95 % CI 28-64). A total of 15 patients received up to four cycles due to incomplete response, but re-treatment did not significantly improve outcome (p = 0.205). After a median follow-up of 19.3 months, 2-year local control rate was 72.5 %. Median time to local failure (N = 11 patients) was 5.1 months. Tumor response (p = 0.041) and control (p = 0.047) correlated with histological grading. Relevant toxicity consisted of G3 skin ulceration and soft tissue necrosis (35 and 23 % of patients, respectively), although this was manageable on an outpatient basis. The accuracy of electrode placement was 47.1 %, and the adequacy of electroporative current 85.3 %. CONCLUSIONS: ECT may represent an active and safe treatment to achieve local control in advanced STS patients with symptomatic disease. Future research challenges include the improvement of electrode placement and voltage delivery together with the containment of soft tissue toxicity.

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TÍTULO / TITLE: ESET histone methyltransferase regulates osteoblastic differentiation of mesenchymal stem cells during postnatal bone development.
RESUMEN / SUMMARY: To investigate the effects of histone methyltransferase ESET (also known as SETDB1) on bone metabolism, we analyzed osteoblasts and osteoclasts in ESET knockout animals, and performed osteogenesis assays using ESET-null mesenchymal stem cells. We found that ESET deletion severely impairs osteoblast differentiation but has no effect on osteoclastogenesis, that co-transfection of ESET represses Runx2-mediated luciferase reporter while siRNA knockdown of ESET activates the luciferase reporter in mesenchymal cells, and that ESET is required for postnatal expression of Indian hedgehog protein in the growth plate. As the bone phenotype in ESET-null mice is 100% penetrant, these results support ESET as a critical regulator of osteoblast differentiation during bone development.

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TÍTULO / TITLE: Surgical treatment of primary pulmonary artery sarcoma.
RESUMEN / SUMMARY: To investigate the effects of histone methyltransferase ESET (also known as SETDB1) on bone metabolism, we analyzed osteoblasts and osteoclasts in ESET knockout animals, and performed osteogenesis assays using ESET-null mesenchymal stem cells. We found that ESET deletion severely impairs osteoblast differentiation but has no effect on osteoclastogenesis, that co-transfection of ESET represses Runx2-mediated luciferase reporter while siRNA knockdown of ESET activates the luciferase reporter in mesenchymal cells, and that ESET is required for postnatal expression of Indian hedgehog protein in the growth plate. As the bone phenotype in ESET-null mice is 100% penetrant, these results support ESET as a critical regulator of osteoblast differentiation during bone development.
RESUMEN / SUMMARY: - OBJETIVO: El sarcoma de la arteria pulmonar es una enfermedad severa y subdiagnosticada, con el enfoque clínico y quirúrgico no claramente establecido. Sólo un puñado de casos individuales o pequeñas series sobre este tema han sido publicados. El objetivo del presente estudio fue informar nuestra experiencia quirúrgica en este campo. 

MÉTODOS: De marzo de 2004 a diciembre de 2012, 13 pacientes fueron sometidos a cirugía para sarcoma de la arteria pulmonar en nuestro instituto. En 7 pacientes, el sarcoma fue unilateral (53.8%), y en 6 (46.2%), el tumor había ya extendido a ambos pulmones. La estrategia quirúrgica se evolucionó a lo largo de los años, pero las dos técnicas utilizadas siempre fueron las mismas: neumonectomía en 5 pacientes y endarterectomía pulmonar en 8.

RESULTADOS: Dos pacientes murieron en el hospital, ambos en la grupo de neumonectomía. La mediana de la estadía en el servicio de cuidados intensivos y hospital fue de 1 día (rango, 1-10) y 14 días (rango, 11-17) para el grupo de neumonectomía y 6 días (rango, 3-23) y 19 días (rango, 10-32) para el grupo de endarterectomía pulmonar, respectivamente. La mediana de supervivencia fue de 26.8 meses después de neumonectomía y 6.6 meses después de endarterectomía pulmonar. 

CONCLUSIONES: El sarcoma de la arteria pulmonar tiene un mal pronóstico. La estrategia quirúrgica en nuestro instituto incluyó neumonectomía, para posibles resecciones radicales, y endarterectomía pulmonar, para reducir los síntomas y aumentar la expectativa de vida. La correcta estrategia quirúrgica debe ser evaluada individualmente, según la presentación del tumor, la presencia de hipertensión pulmonar, y el estado clínico del paciente.
results show the main mechanisms of the deleterious effects of VOsil in the osteosarcoma cell line, demonstrating that this complex is a promising compound for cancer treatments.

[92] TÍTULO / TITLE: - Multimodal therapy including liver transplantation for hepatic undifferentiated embryonal sarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Walther A; Geller J; Coots A; Towbin A; Nathan J; Alonso M; Sheridan R; Tiao G
INSTITUCIÓN / INSTITUTION: - Departments of Pediatric Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, OH.
RESUMEN / SUMMARY: - The outcomes of hepatic undifferentiated embryonal sarcoma (HUES) have historically been limited by persistent, unresectable disease and the subsequent development of disease resistance and dissemination. We present our institutional experience with HUES and assess current treatment trends and outcomes in the era of liver transplantation. We conducted a retrospective chart review of cases presenting with HUES at our institution over the past 10 years. The collected data included age, sex, presenting symptoms, imaging and the associated Pretreatment Extent of Disease (PRETEXT) score, pathology, chemotherapy, surgical interventions, and outcomes. Approval was obtained from the institutional review board of the Cincinnati Children's Hospital Medical Center. HUES was identified in 6 patients (4 males and 2 females) with a median age at diagnosis of 11 years (range = 7-13 years). Initial imaging was available for all but 1 patient. The PRETEXT stage for these patients ranged from II to III. One patient was diagnosed with lung metastases. Two patients underwent upfront resection, and 1 patient received neoadjuvant therapy and then conventional resection. Three patients were treated with orthotopic liver transplantation (OLT) after neoadjuvant chemotherapy (primary OLT in 2 cases and salvage OLT for local recurrence in 1 case). Two patients received posttransplant adjuvant chemotherapy. All 6 patients remained in clinical remission with a mean follow-up of 35 months (range = 12-84 months). In conclusion, OLT has rarely been reported as a treatment option for HUES. The addition of liver transplantation as a surgical option for treating patients with HUES can result in improved survival for patients whose tumors are initially unresectable or recur. Liver Transpl, 2013. © 2013 AASLD.

[93] TÍTULO / TITLE: - Circumvention of resistance to photodynamic therapy in doxorubicin-resistant sarcoma by photochemical internalization of gelonin.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Enlace al texto completo (gratuito o de pago) 1016/j.freeradbiomed.2013.09.010
AUTORES / AUTHORS: Olsen CE; Berg K; Selbo PK; Weyergang A
INSTITUCIÓN / INSTITUTION: Department of Radiation Biology, Institute for Cancer Research, The Norwegian Radium Hospital, Oslo University Hospital, Norway. Electronic address: cathrine.elisabeth.olsen@rr-research.no.
RESUMEN / SUMMARY: A wide range of anti-cancer therapies have been shown to induce resistance upon repetitive treatment and such adapted resistance may also cause cross-resistance to other treatment modalities. We here show that MES-SA/Dx5 cells with adapted resistance to doxorubicin (DOX) are cross-resistant to photodynamic therapy (PDT). A DOX-induced increased expression of the reactive oxygen species (ROS)-scavenging proteins glutathione peroxidase (GPx) 1 and GPx4 in MES-SA/Dx5 cells was indicated as the mechanism of resistance to PDT in line with the reduction in PDT-generated ROS observed in this cell line. ROS-induced p38 activation was, in addition, shown to be reduced to one-third of the signal of the parental MES-SA cells 2h after PDT, and addition of the p38 inhibitor SB203580 confirmed p38 activation as a death signal after PDT in the MES-SA cells. The MES-SA/Dx5 cells were also cross-resistant to ionizing radiation in agreement with the increased GPx1 and GPx4 expression. Surprisingly, PDT-induced endo/lysosomal release of the ribosome-inactivating protein gelonin (photochemical internalization (PCI)) was more effective in the PDT-resistant MES-SA/Dx5 cells, as measured by synergy calculations in both cell lines. Analysis of death-inducing signaling indicated a low activation of caspase-3 and a strong PARP I cleavage after PDT and PCI in both cell lines. The PARP I activation was, however, stronger after PCI than after PDT in the MES-SA cells, but not in the MES-SA/Dx5 cells, and therefore cannot explain the strong PCI effect in the MES-SA/Dx5 cells. In conclusion PCI of recombinant gelonin circumvents ROS resistance in an apoptosis-independent manner.

[94] TÍTULO / TITLE: The cellular peptidyl-prolyl cis/trans isomerase Pin1 regulates reactivation of Kaposi’s sarcoma-associated herpesvirus from latency.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Guito J; Gavina A; Palmeri D; Lukac DM
INSTITUCIÓN / INSTITUTION: Dept. of Microbiology and Molecular Genetics, New Jersey Medical School and Graduate School of Biomedical Sciences, Rutgers Biomedical and Health Sciences, Rutgers University, Newark, NJ 07103.
RESUMEN / SUMMARY: Kaposi’s sarcoma-associated herpesvirus (KSHV) causes Kaposi’s sarcoma and primary effusion lymphoma. KSHV-infected cells are predominantly latent, with a subset undergoing lytic reactivation. Rta is the essential, lytic switch protein that reactivates virus by forming transactivation-competent complexes with the Notch effector protein RBP-JK and promoter DNA. Strikingly, Rta homolog analysis reveals that prolines constitute 18% of conserved residues. Rta is also highly phosphorylated in vivo. We previously demonstrated that proline content determines Rta homo-tetramerization and function. We hypothesize that proline-directed modifications regulate Rta function by controlling binding to peptidyl-prolyl cis/trans isomerases (PPlases). Cellular PPlase Pin1 binds specifically to phosphoserine- or phosphothreonine-proline (pS/T-P) motifs in target proteins. Pin1
dysregulation is implicated in myriad human cancers and can be subverted by viruses. Our data show that KSHV Rta protein contains potential pS/T-P motifs and binds directly to Pin1. Rta transactivation is enhanced by Pin1 at two delayed early viral promoters in uninfected cells. Pin1’s effect, however, suggests a rheostat-like influence on Rta function. In infected cells, we show that endogenous Pin1 is active during reactivation and enhances Rta-dependent early protein expression induced by multiple signals, as well as DNA replication. Surprisingly, ablation of Pin1 activity by the chemical juglone or dominant negative Pin1 enhanced late gene expression and production of infectious virus, while ectopic Pin1 showed inhibitory effects. Our data thus suggest that Pin1 is a unique, dose-dependent molecular timer that enhances Rta protein function, but inhibits late gene synthesis and virion production, during KSHV lytic reactivation.

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[TITULO / TITLE: - Frequent expression of KIT in endometrial stromal sarcoma with YWHAE genetic rearrangement.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Lee CH; Hoang LN; Yip S; Reyes C; Marino-Enriquez A; Eilers G; Tao D; Chiang S; Fletcher JA; Soslow RA; Nucci MR; Oliva E
INSTITUCIÓN / INSTITUTION: - Department of Laboratory Medicine and Pathology, University of Alberta and Royal Alexandra Hospital, Edmonton, AB, Canada.
RESUMEN / SUMMARY: - Endometrial stromal sarcomas with the YWHAE-NUTM2A/B genetic fusion characteristically contain high-grade round to epithelioid cell component that is strongly and diffusely cyclin D1-positive and it may or may not show an associated low-grade fibroblastic/myxoid cell component. They are clinically more aggressive than endometrial stromal sarcomas with the JAZF1-SUZ12 genetic fusion and frequently demonstrate extrauterine extension at initial clinical presentation. In this setting, the tumor may be misdiagnosed as gastrointestinal stromal tumor. This study examines the expression of KIT and ANO1 in 14 YWHAE-NUTM2A/B tumors by immunohistochemistry. Staining localization was determined as membranous and/or cytoplasmic, and the staining intensity was assessed (negative, weak, moderate and strong). Of the 14 tumors, 6 contained only a high-grade round cell component, 2 only a low-grade fibroblastic component and 6 had both components in the slides evaluated. The high-grade round cell component displayed moderate to strong membranous/cytoplasmic KIT staining in all tumors (12 of 12). The low-grade fibroblastic cell component showed only weak cytoplasmic KIT staining in 3 of 8 tumors. In contrast, ANO1 was negative in all 14 neoplasms, irrespective of the component evaluated. Sanger sequencing analysis (exons 9, 11, 13 and 17) and Ampliseq Cancer Panel mutation screen (Ion Torrent) demonstrated no KIT mutations in three KIT-positive YWHAE-NUTM2A/B tumors. This study shows that the high-grade round cell component of YWHAE-NUTM2A/B endometrial stromal sarcoma consistently expresses KIT but lacks KIT hotspot mutations. KIT expression may represent a potential diagnostic pitfall in the evaluation of YWHAE-NUTM2A/B endometrial stromal sarcoma presenting with pelvic/abdominal mass, particularly in situations where its uterine origin is not definitive, and thus a panel of antibodies that
includes ANO1 and cyclin D1 is necessary. Modern Pathology advance online publication, 1 November 2013; doi:10.1038/modpathol.2013.199.

[96]

**TÍTULO / TITLE:** Treatment Outcomes in Completely Resected Stage I to Stage IV Uterine Carcinosarcoma With Rhabdomyosarcoma Differentiation.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** Int J Gynecol Cancer. 2013 Nov;23(9):1635-41. doi: 10.1097/IGC.0000000000000001.

**AUTORES / AUTHORS:** Makker V; Kravetz SJ; Gallagher J; Orodel OP; Zhou Q; Iasonos A; Delair D; Aghajanian C; Hensley ML

**INSTITUCIÓN / INSTITUTION:** *Gynecologic Medical Oncology Service, Department of Medicine, Memorial Sloan-Kettering Cancer Center; daggerDepartment of Medicine, Weill Cornell Medical College; double daggerDepartment of Epidemiology and Biostatistics, and section signDepartment of Gynecologic Pathology, Memorial Sloan-Kettering Cancer Center, New York, NY.

**RESUMEN / SUMMARY:** OBJECTIVE: To evaluate overall survival (OS) and progression-free survival (PFS) after adjuvant therapy in stage I to stage IV uterine carcinosarcoma with rhabdomyosarcoma differentiation. METHODS: Memorial Sloan-Kettering Cancer Center medical records from 1990 to 2012 were reviewed. Patients who received chemotherapy with or without radiation therapy (RT), or RT alone, for completely resected stage I to stage IV uterine carcinosarcoma with rhabdomyosarcoma differentiation were included. RESULTS: Of 53 patients, International Federation of Gynecology and Obstetrics stage distribution was as follows: I, 13 (24.5%); II, 8 (15.1%); III, 13 (24.5%); and IV, 19 (35.9%). Forty-one (77.4%) of 53 patients received adjuvant chemotherapy, and 34% of the patients who received chemotherapy also received pelvic RT or intravaginal brachytherapy (IVRT). Twelve (22.6%) of the 53 patients received only pelvic RT with/without IVRT. Paclitaxel-carboplatin was the most commonly used adjuvant chemotherapy treatment. The median PFS for the entire cohort was 13.4 months (95% confidence interval [CI], 10.5-17.0). The median OS for the entire cohort was 23.0 months (95% CI, 16.9-34.3). The median PFS periods by stage were 15.9 months for stages I/II versus 11.2 months for stages III/IV (P = 0.012). Median OS was not reached in the early-stage cohort. The median OS for the late-stage cohort was 20.9 months (P = 0.004). The median PFS periods by treatment were 10.4 months for pelvic RT with/without IVRT group versus 13.1 months for chemotherapy with/without pelvic RT with/without IVRT group (P = 0.498). The median OS periods by treatment were 23.6 months for chemotherapy with/without pelvic RT with/without IVRT group versus 16.9 months for pelvic RT with/without IVRT group (P = 0.501). CONCLUSION: The results suggest that chemotherapy alone or in combination with RT is associated with longer PFS and OS compared to RT alone. Only the stage of disease significantly affected PFS and OS.

[97]
Histone deacetylase classes I and II regulate Kaposi’s sarcoma-associated herpesvirus reactivation.

In KSHV latency in primary effusion lymphoma (PEL) cells, the promoter of the viral lytic switch gene, Rta, is organized into bivalent chromatin, similar to cellular developmental switch genes. Histone deacetylase inhibitors (HDACi) reactivate latent KSHV, and dramatically remodel viral genome topology and chromatin architecture. However, reactivation is not uniform across a population of infected cells. We sought to identify an HDACi cocktail that would uniformly reactivate KSHV and reveal the regulatory HDACs. Using HDACi with varying specificities, we found that Class I HDACi were sufficient to reactivate the virus, but differed in potency. Valproic acid (VPA) was the most effective HDACi, inducing lytic cycle gene expression in 75% of cells, while trichostatin A (TSA) induced less widespread lytic gene expression and inhibited VPA-stimulated reactivation. VPA was only slightly superior to TSA in inducing histone acetylation of Rta’s promoter, but only VPA induced significant production of infectious virus, suggesting that HDAC regulation post-Rta expression has a dramatic effect on reactivation progression. Ectopic HDACs 1, 3, and 6 inhibited TPA-stimulated KSHV reactivation. Surprisingly, ectopic HDACs 1 and 6 stimulated reactivation independently, suggesting that HDAC-complex stoichiometry is critical for the switch. Tubacin, a specific inhibitor of the ubiquitin-binding, pro-autophagic HDAC6, also inhibited VPA-stimulated reactivation. Immunofluorescence indicated that HDAC6 is expressed diffusely throughout latently-infected cells, but is found in the cytoplasm and nucleus during reactivation. Overall, our data suggest that inhibition of HDAC classes I and IIa, and maintenance of HDAC 6 (IIb) activity, are required for optimal KSHV reactivation.

Next-Generation Sequence Analysis of the Genome of RFHVMn, the Macaque Homolog of Kaposi’s Sarcoma (KS)-Associated Herpesvirus, from a KS-Like Tumor of a Pig-Tailed Macaque.

The complete sequence of retroperitoneal fibromatosis-associated herpesvirus Macaca nemestrina (RFHVMn), the pig-tailed macaque homolog of Kaposi’s sarcoma-associated herpesvirus (KSHV), was determined by
next-generation sequence analysis of a Kaposi’s sarcoma (KS)-like macaque tumor. Colinearity of genes was observed with the KSHV genome, and the core herpesvirus genes had strong sequence homology to the corresponding KSHV genes. RFHVMn lacked homologs of open reading frame 11 (ORF11) and KSHV ORFs K5 and K6, which appear to have been generated by duplication of ORFs K3 and K4 after the divergence of KSHV and RFHV. RFHVMn contained positional homologs of all other unique KSHV genes, although some showed limited sequence similarity. RFHVMn contained a number of candidate microRNA genes. Although there was little sequence similarity with KSHV microRNAs, one candidate contained the same seed sequence as the positional homolog, kshv-miR-K12-10a, suggesting functional overlap. RNA transcript splicing was highly conserved between RFHVMn and KSHV, and strong sequence conservation was noted in specific promoters and putative origins of replication, predicting important functional similarities. Sequence comparisons indicated that RFHVMn and KSHV developed in long-term synchrony with the evolution of their hosts, and both viruses phylogenetically group within the RV1 lineage of Old World primate rhadinoviruses. RFHVMn is the closest homolog of KSHV to be completely sequenced and the first sequenced RV1 rhadinovirus homolog of KSHV from a nonhuman Old World primate. The strong genetic and sequence similarity between RFHVMn and KSHV, coupled with similarities in biology and pathology, demonstrate that RFHVMn infection in macaques offers an important and relevant model for the study of KSHV in humans.

[99]

**TITULO / TITLE:** Curcumin-loaded nanoparticles enhance apoptotic cell death of U2OS human osteosarcoma cells through the Akt-Bad signaling pathway.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Peng SF; Lee CY; Hour MJ; Tsai SC; Kuo DH; Chen FA; Shieh PC; Yang JS

**INSTITUCIÓN / INSTITUTION:** Department of Biological Science and Technology, China Medical University, Taichung 404, Taiwan, R.O.C.

**RESUMEN / SUMMARY:** Curcumin has potential anticancer activity and has been shown to be involved in several signaling pathways including differentiation and apoptosis. Our previous study showed that water-soluble PLGA curcumin nanoparticles (Cur-NPs) triggered apoptotic cell death through regulation of the function of MDR1 and the production of reactive oxygen species (ROS) in cisplatin-resistant human oral cancer CAR cells. In this study, we investigated the anti-proliferative effects of Cur-NPs on human osteosarcoma U2OS cells. The morphology of Cur-NPs showed spherical shape by TEM analysis. The encapsulation efficiency of curcumin in Cur-NPs prepared by single emulsion was 90.5+/−3.0%. Our results demonstrated that the curcumin fragments on the mass spectrum of Cur-NPs and the peaks of curcumin standard could be found on the Cur-NPs spectrum by 1H-NMR spectra analysis. Cur-NPs induced anti-proliferative effects and apoptosis in U2OS cells. Compared to the untreated U2OS cells, more detectable amount of Cur-NPs was found inside the treated U2OS cells. Cur-NPs induced DNA fragmentation and
apoptotic bodies in U2OS cells. Both the activity and the expression levels of caspases-3/-7 and caspase-9 were elevated in the treated U2OS cells. Cur-NPs upregulated the protein expression levels of cleaved caspase-3/caspase-9, cytochrome c, Apaf-1 and Bad and downregulated the protein expression level of p-Akt in U2OS cells. These results suggest Cur-NPs are effective in enhancing apoptosis in human osteosarcoma cells and thus could provide potential for cancer therapeutics.

**AUTORES / Authors:** - Yerushalmi GM; Gilboa Y; Jakobson-Setton A; Tadir Y; Goldchmit C; Katz D; Seidman DS

**INSTITUCIÓN / INSTITUTION:** - Departments of Obstetrics and Gynecology, Chaim Sheba Medical Center, Tel-Hashomer, Israel; Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel.

**RESUMEN / SUMMARY:** - OBJECTIVE: To evaluate the efficacy and safety of 3 months of vaginal mifepristone treatment on leiomyoma volume and related symptoms.

**DESIGN:** Prospective, open-label, two tertiary centers, phase II clinical trial. **SETTING:** Two tertiary medical centers in Israel. **PATIENT(S):** Thirty-three enrolled women, ages 30-53 years, diagnosed with symptomatic uterine fibroids. **INTERVENTION(S):** Patients received 10 mg mifepristone vaginally daily for 3 months. **MAIN OUTCOME MEASURE(S):** Reduction in uterine leiomyoma volume. Improvement in symptoms related to uterine fibroids was assessed with the use of the “Uterine Fibroid Symptoms Quality of Life Questionnaire” (UFS-QoL). The number of bleeding days, safety, and tolerability were secondary measures. **RESULT(S):** Mifepristone treatment significantly reduced leiomyoma volume from 135.3 +/- 22.9 cc at enrollment to 101.2 +/- 22.4 cc after 3 months of treatment. The UFS-QoL Score significantly decreased from 20.7 +/- 0.7 at enrollment to 14.0 +/- 0.8 after 3 months of treatment. The number of bleeding days significantly decreased by 3.5 days. Endometrial biopsies showed no evidence of endometrial hyperplasia or cellular atypia. There were no major side effects during the course of the study, and treatment was well tolerated. **CONCLUSION(S):** Vaginal mifepristone may offer an effective treatment option for women with symptomatic uterine leiomyoma and can improve the patients’ quality of life. **CLINICAL TRIAL REGISTRATION NUMBER:** NCT00881140.

[102]

**TÍTULO / TITLE:** - Estradiol and tamoxifen enhance invasion of endometrial stromal cells in a three-dimensional coculture model of adenomyosis.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / Authors:** - Taylor AH; Kalathy V; Habiba M

**INSTITUCIÓN / INSTITUTION:** - Reproductive Sciences Section, Department of Cancer Studies and Molecular Medicine, Clinical Sciences Building, University of Leicester, Leicester, United Kingdom. Electronic address: aht3@le.ac.uk.

**RESUMEN / SUMMARY:** - OBJECTIVE: To examine the effect of estradiol alone or with progesterone and tamoxifen on the depth of invasion of endometrial stromal cells from women with and without adenomyosis in a three-dimensional (3D) coculture model that includes myocytes. **DESIGN:** Case-controlled, blinded comparison. **SETTING:** Medical school department. **PATIENT(S):** Premenopausal women with and without uterine adenomyosis. **INTERVENTION(S):** Human endometrial stromal and myometrial cells grown in a 3D coculture with crossover between cells from uteri with and without adenomyosis; cocultures treated with tamoxifen, estradiol alone, or estradiol with...
progestrone. MAIN OUTCOME MEASURE(S): Depth of stromal cell invasion into a collagen matrix. RESULT(S): The depth of invasion for adenomyotic stromal cells was statistically significantly higher than for the control stromal cells, whether grown on plain collagen, on collagen containing control or adenomyotic muscle cells. The addition of estradiol or tamoxifen, but not the estradiol and progesterone combination, increased the depth of invasion of both adenomyotic stromal cells and control stromal cells in all cell combinations. When grown on plain collagen, the depth of invasion for control stromal cells and adenomyotic stromal cells increased by 126% and 93% with the use of tamoxifen, and by 71% and 50%, with the use of estradiol. CONCLUSION(S): Both estradiol and tamoxifen enhance stromal cell invasion, but the greater depth of invasion of adenomyotic stromal cells and the enhancing effect of adenomyotic muscle were maintained under all experimental conditions, suggesting an inherent predisposition in affected women.

[103]
TÍTULO / TITLE: - Proton therapy for reirradiation of progressive or recurrent chordoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - McDonald MW; Linton OR; Shah MV
INSTITUCIÓN / INSTITUTION: - Department of Radiation Oncology, Indiana University School of Medicine, Indianapolis, Indiana; Indiana University Health Proton Therapy Center, Bloomington, Indiana. Electronic address: mmcdona2@iuhealth.org.
RESUMEN / SUMMARY: - PURPOSE: To report the results in patients reirradiated with proton therapy for recurrent or progressive chordoma, with or without salvage surgery.
METHODS AND MATERIALS: A retrospective review of 16 consecutive patients treated from 2005 to 2012 was performed. All patients had received at least 1 prior course of radiation therapy to the same area, and all but 1 patient had at least 1 surgical resection for disease before receiving reirradiation. At the time of recurrence or progression, half of the patients underwent additional salvage surgery before receiving reirradiation. The median prior dose of radiation was 75.2 Gy (range, 40-79.2 Gy). Six patients had received prior proton therapy, and the remainder had received photon radiation. The median gross tumor volume at the time of reirradiation was 71 cm(3) (range, 0-701 cm(3)). Reirradiation occurred at a median interval of 37 months after prior radiation (range, 12-129 months), and the median dose of reirradiation was 75.6 Gy (relative biological effectiveness [RBE]) (range. 71.2-79.2 Gy [RBE]), given in standard daily fractionation (n=14) or hyperfractionation (n=2). RESULTS: The median follow-up time was 23 months (range, 6-63 months); it was 26 months in patients alive at the last follow-up visit (range, 12-63 months). The 2-year estimate for local control was 85%, overall survival 80%, chordoma-specific survival 88%, and development of distant metastases 20%. Four patients have had local progression: 3 in-field and 1 marginal. Late toxicity included grade 3 bitemporal lobe radionecrosis in 1 patient that improved with hyperbaric oxygen, a grade 4 cerebrospinal fluid leak with meningitis in 1 patient, and a grade 4 ischemic brainstem stroke (out of radiation field) in 1 patient, with subsequent neurologic recovery. CONCLUSIONS: Full-dose proton reirradiation provided encouraging initial disease control and overall survival for patients with
recurrent or progressive chordoma, although additional toxicities may develop with longer follow-up times.

[104] **TITULO / TITLE:** - Inhibition of focal adhesion kinase induces apoptosis in human osteosarcoma SAOS-2 cells.  
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)  
**AUTORES / AUTHORS:** - Wang J; Zu J; Xu G; Zhao W; Jinglong Y  
**INSTITUCIÓN / INSTITUTION:** - Department of Bone Surgery, First Affiliated Hospital of Harbin Medical University, No. 23 Youzheng street, Harbin, Heilongjiang province, 150001, China.  
**RESUMEN / SUMMARY:** - Focal adhesion kinase (FAK), a non-receptor tyrosine kinase protein, acts as an early modulator of integrin signaling cascade, regulating basic cellular functions. In transformed cells, unopposed FAK signaling has been considered to promote tumor growth, progression, and metastasis. The aim of this study was to assess the role of focal adhesion kinase in human osteosarcoma SAOS-2 cells. SAOS-2 cells were transfected with PGPU6/GFP/shNC, and PGPU6/GFP/FAK-334 (shRNA-334), respectively. Expression of FAK was detected by real-time PCR and western blots. MTT assay was used to examine changes in cell proliferation. Cell apoptosis was analyzed by flow cytometry. The expression of caspase-3,-7,-9 was measured by Western blots. The expression of FAK in SAOS-2 cells significantly decreased in shRNA-334 group contrast to the control group (P < 0.01). Cells proliferation was inhibited by shRNA-334 and shRNA-334 + cisplatin, and the effects were clearly enhanced when cells treated with the anticancer agents. The level of cell apoptosis in shRNA-334 and shRNA-334 + cisplatin group was higher than in the control group (P < 0.01). The current data support evidence that down-regulation of FAK could induce SAOS-2 apoptosis through the caspase-dependent cell death pathway. Inhibition of the kinases may be important for therapies designed to enhance the apoptosis in osteosarcoma.

[105] **TITULO / TITLE:** - Expression of protease-activated receptor-2 in human gastric stromal tumor and its clinicopathological significance.  
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)  
**AUTORES / AUTHORS:** - Wang GJ; Wang YB; Li DN; Deng BB  
**RESUMEN / SUMMARY:** - Aim: To explore the expression of protein-activated receptor-2 (PAR-2) in human gastric stromal tumor and its clinicopathological significance. Methods: The expression of PAR-2 was detected with immunohistochemistry, RT-PCR and Western blot in tumor tissue, peritumoral tissue and gastric normal tissue from 72 patients with gastric stromal tumor. Results: PAR-2 expression was significantly higher in peritumoral tissue (P<0.05) and tumor tissue (P<0.01) than in gastric normal tissue,
and significantly higher in tumor tissue than in peritumoral tissue (P<0.01). With the increase in NIH grade, PAR-2 expression was elevated in tumor tissues. PAR-2 expression was strongly associated with mucosal invasion. Conclusion: PAR-2 expression is significantly higher in gastric stromal tumor tissue than in peritumoral tissue and gastric normal tissue. The high expression of PAR-2 may be associated with the invasion and metastasis of gastric stromal tumor.

[106]
TÍTULO / TITLE: - Epigenetic regulation of the pro-apoptosis gene TSSC3 in human osteosarcoma cells.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1016/j.biopha.2013.10.006
AUTORES / AUTHORS: - Li Y; Huang Y; Lv Y; Meng G; Guo QN
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Xinqiao Hospital, Third Military Medical University, Chongqing 400037, China.
RESUMEN / SUMMARY: - Promoter hypermethylation can lead to a loss of genetic imprinting in carcinogenesis. The mechanism for the loss of expression of the imprinted gene TSSC3 has not been investigated in cases of osteosarcoma. In this study, we treated osteosarcoma cell lines with 5-Aza-CdR, which is a widely-used DNA methyltransferase inhibitor, and found dose-dependent reduction in cell growth, conversion of cell morphology to a non-motile phenotype, and obvious increase in apoptosis. In addition, we also found that 5-Aza-CdR reactivated TSSC3 expression through demethylation of the promoter regions. These findings indicate that the TSSC3 gene is silenced through hypermethylation of the promoter regions, a mechanism commonly associated with gene silencing in cancer. Finally, we examined the role of TSSC3 in human osteosarcoma SaOS2 cells. We showed that TSSC3 overexpression suppressed SaOS2 cell growth and increased apoptosis through caspase-3 upregulation, thereby, suggesting that TSSC3 may play a pro-apoptosis role to maintain the normal balance of growth. Taken together, these observations suggest that the epigenetic regulation of TSSC3, a pro-apoptosis gene, provides valuable insights into possible osteosarcoma therapies.

[107]
TÍTULO / TITLE: - A mechanistic study on the metastasis inducing function of FUS-CHOP fusion protein in liposarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1002/ijc.28638
AUTORES / AUTHORS: - Patil N; Rasheed SA; Abba M; Leupold JH; Schwarzbach M; Allgayer H
INSTITUCIÓN / INSTITUTION: - Department of Experimental Surgery and Molecular Oncology of Solid Tumors, Medical Faculty Mannheim, University of Heidelberg and German Cancer Research Center, Heidelberg, Germany.
The FUS-CHOP fusion protein has been found to be instrumental for specific oncogenic processes in liposarcoma, but its ability to induce metastasis and the underlying mechanisms by which this can be achieved remain unknown. In order to dissect its functional role in this context, we stably overexpressed this protein in SW872 liposarcoma and HT1080 fibrosarcoma cell lines, and were able to demonstrate that forced expression of FUS-CHOP significantly increases migration and invasion, as well as enhances lung and liver metastasis in the in vivo chicken chorioallantoic membrane (CAM) model, that is proliferation independent. Additionally, FUS-CHOP enhances the expression of matrix-metalloproteinases -2 and -9, and transactivates their promoters in vitro. Mutational analysis showed that C/EBP-beta (-769/-755), NF-kappaB (-525/-516) and CREB/AP-1 (-218/-207) sites were important for MMP-2 and NF-kappaB (-604/-598), AP-1 (-539/-532) and AP-1 (-81/-72) for MMP-9 transactivation. Moreover, a direct in vivo interaction of FUS-CHOP was observed in case of the MMP-2 promoter within region (-769/-207). siRNA data revealed that MMP-2 expression is essential in the FUS-CHOP induced metastatic phenotype. MMP-2 mRNA and protein expression correlated significantly with FUS-CHOP positivity in 46 resected patient liposarcoma tissues. We have for the first time provided substantial evidence for the FUS-CHOP oncoprotein as an inducer of metastasis that is due to the transcriptional induction of specific tumor-associated proteases. Insights gained from this study not only support a deeper understanding of the mechanistic properties of FUS-CHOP, but also open up new avenues for targeted therapy.

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various cancers. We show that inhibition of Hh signaling reduces Yap1 expression and knockdown of Yap1 significantly inhibits tumor progression. Moreover, long non-coding RNA H19 is aberrantly expressed and induced by upregulated Hh signaling and Yap1 overexpression. Our results demonstrate that aberrant Hh signaling in mature osteoblasts is responsible for the pathogenesis of osteoblastic osteosarcoma through Yap1 and H19 overexpression. Oncogene advance online publication, 21 October 2013; doi:10.1038/onc.2013.433.

[109] TÍTULO / TITLE: - Gallic acid induces apoptosis and inhibits cell migration by upregulating miR-518b in SW1353 human chondrosarcoma cells.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Liang W; Li X; Li Y; Li C; Gao B; Gan H; Li S; Shen J; Kang J; Ding S; Lin X; Liao L
INSTITUCIÓN / INSTITUTION: - Research Base of Traditional Chinese Medicine Syndrome, Fujian University of Traditional Chinese Medicine, Fuzhou 350122, P.R. China.
RESUMEN / SUMMARY: - Gallic acid (GA), a natural agent, is widely distributed in plants with a range of biological effects and has been of potential interest as anticancer agent. However, its effects on chondrosarcoma cell apoptosis are still undefined. In the present study, the possible mechanisms of GA-induced apoptosis were explored in SW1353 cells, a human chondrosarcoma cell line. Our results showed that GA inhibited cell viability dose- and time-dependently. Morphological examination of GA-treated cells exhibited the typical features of cell death, such as rounding up of the cells and cell shrinkage. Wound-healing assay indicated that GA inhibited the migratory abilities of SW1353 cells. Hoechst 33258 staining assay and Annexin V/PI staining assay exhibited apoptosis induction by GA. To determine the molecular mechanism of GA-induced apoptosis, the expression levels Bcl-2, Bax, caspase-3 and caspase-9 were determined in SW1353 cells treated with GA. We found that GA downregulated the expression of the anti-apoptotic protein Bcl-2, and upregulated the expression of the pro-apoptotic protein Bax, and the activation of caspase-3 and caspase-9. To identify the possible mechanisms, the changes of microRNA expression were tested using the miRCURY LNA expression array. It was observed that the miR-518b gene was upregulated in treated cells. Taken together, these data show that GA induces apoptosis and inhibits cell migration by upregulating miR-518b in SW1353 cells.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Resumen / Summary: Background: Undifferentiated embryonal sarcoma of the liver (UESL) represents less than 5% of all malignant hepatic tumors in childhood. It is considered an aggressive neoplasm with an unfavorable prognosis. The aim of this paper is to present a single center experience in the treatment of children with UESL.

Materials and Methods: Ten children with UESL were treated between 1981 and 2012. Age at diagnosis ranged from 4 months to 17 years (median age, 6 years and 9 months). Surgery after neoadjuvant chemotherapy (CHT) was performed in 7 patients, and in 3 patients primary surgery was done. Adjuvant chemotherapy was administered in all 10 patients (CYVADIC, CAV, CAV/ETIF/IF+ADM, CDDP/PLADO). Right hemihepatectomy was performed in 1 patient, extended right hemihepatectomy in 6, and partial resection of the right lobe (segments V-VI, segment V) in 2 patients. One patient with unresectable tumor affecting both lobes was listed for liver transplantation (LTx). Results: Follow-up from diagnosis ranged from 50 to 222 months (mean 138 months). Among 9 patients treated with partial liver resection, distant metastases/local recurrence was not observed in any, and disease-free survival in this group is 100% (9 patients alive). The patient that underwent liver transplantation died of multiorgan failure 4 months postoperatively. However, this patient was misdiagnosed as having hepatoblastoma (HBL) and received PLADO chemotherapy. The overall survival rate is 90%. Conclusion: Excellent results with long-term survival can be achieved in children with UESL with conventional therapy, including a combination of neoadjuvant and adjuvant chemotherapy and surgery, even in large extensively growing tumors.

[111] Título / Title: Pulmonary artery sarcoma masquerading as pulmonary embolism.

Resumen / Summary: Enlace al Resumen / Link to its Summary

Revista / Journal: Am J Respir Crit Care Med. 2013 Nov 1;188(9):1161. doi: 10.1164/rccm.201304-0715IM.

Autores / Authors: Adekolu O; Lundbye J; Manthous C

Institución / Institution: 1 Department of Medicine, University of Connecticut, Farmington, Connecticut.

[112] Título / Title: Targeted imaging of Ewing sarcoma in preclinical models using a 64Cu-labeled anti-CD99 antibody.

Resumen / Summary: Enlace al Resumen / Link to its Summary


Autores / Authors: - Enlace al texto completo (gratuito o de pago) 1158/1078-0432.CCR-13-1660
AUTORES / AUTHORS: - O’Neill AF; Dearling JL; Wang Y; Tupper T; Sun Y; Aster JC; Calicchio ML; Perez-Atayde AR; Packard AB; Kung AL
INSTITUCIÓN / INSTITUTION: - Pediatric Oncology, Harvard Medical School, Dana-Farber Cancer Institute.

RESUMEN / SUMMARY: - PURPOSE: Ewing sarcoma is a tumor of the bone and soft tissue characterized by diffuse cell membrane expression of CD99 (MIC2). Single-site, surgically resectable disease is associated with an excellent 5-year event-free survival; conversely, patients with distant metastases have a poor prognosis. Non-invasive imaging is the standard approach to identifying sites of metastatic disease. We sought to develop a CD99-targeted imaging agent for staging Ewing sarcoma and other CD99-expressing tumors. EXPERIMENTAL DESIGN: We identified a CD99 antibody with highly specific binding in vitro and labeled this antibody with 64Cu. Mice with either subcutaneous Ewing sarcoma xenograft tumors or micrometastases were imaged with the 64Cu-labeled anti-CD99 antibody and these results were compared to conventional MRI and FDG-PET imaging. RESULTS: 64Cu-labeled anti-CD99 antibody demonstrated high avidity for the CD99 positive subcutaneous tumors, with a high tumor-to-background ratio, greater than that demonstrated with FDG-PET. Micrometastases, measuring 1-2 mm on MRI, were not detected with FDG-PET but readily visualized with the 64Cu-labeled anti-CD99 antibody. Probe biodistribution studies demonstrated high specificity of the probe for CD99 positive tumors. CONCLUSIONS: 64Cu-labeled anti-CD99 antibody can detect subcutaneous Ewing sarcoma tumors and metastatic sites with high sensitivity, outperforming FDG-PET in preclinical studies. This targeted radiotracer may have important implications for the diagnosis, surveillance, and treatment of Ewing sarcoma. Similarly, it may impact the management of other CD99 positive tumors.

[TITULO / TITLE: - Wnt Pathway in Osteosarcoma, from Oncogenic to Therapeutic.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cai Y; Cai T; Chen Y
INSTITUCIÓN / INSTITUTION: - School of Pharmacy, Jinan University, Guangzhou, China.

RESUMEN / SUMMARY: - Osteosarcoma is the most common malignant bone tumor in children and adolescents. Although pathologic characteristics of this disease are clear and well established, much remains to be understood about this tumor, particularly at the molecular signaling level. Secreted signaling molecules of the Wnt family have been widely investigated and found to play a central role in controlling embryonic bone development, bone mass and postnatal bone regeneration. A variety of studies also suggest that Wnt signaling pathway is closely associated with bone malignancies, including breast or prostate cancer induced bone metastasis, multiple myeloma, as well as osteosarcoma. Here, we provide an overview of the role of Wnt signaling pathway in osteosarcoma development and progression, highlighting the aberrant activation of Wnt pathway in this bone malignancy. We also discuss the potential therapeutic applications for the treatment of osteosarcoma targeting Wnt pathway. J. Cell. Biochem. © 2013 Wiley Periodicals, Inc.
TÍTULO / TITLE: - Association of RECQL5 gene polymorphisms and osteosarcoma in a Chinese Han population.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Zhi LQ; Ma W; Zhang H; Zeng SX; Chen B
INSTITUCIÓN / INSTITUTION: - Department of Orthopedics, The First Affiliated Hospital of Xi'an Jiaotong University, No. 227, YanTa West Rd, Xi'an, 710061, China.
RESUMEN / SUMMARY: - Despite the knowledge on many genetic variants present in osteosarcoma, the complexity of this disease precludes placing its biology into a simple conceptual framework. RECQL is a DNA helicase involved in DNA mismatch repair and has been reported to be associated with many human cancers. We aimed to investigate the association of RECQL genetic polymorphism with osteosarcoma in a Chinese population. We selected three polymorphisms of the RECQL5 gene (rs820196, rs820200, and rs4789223) in the present study. TaqMan method was utilized for genotyping these three SNPs in 212 patients with osteosarcoma and 240 age- and sex-matched noncancer controls. In our study, we found that CC genotype in rs820196 (17.5 vs 8.3 %, P = 0.005) and AA genotype in rs4789223 (21.7 vs 14.2, P < 0.001) were more frequent in osteosarcoma group compared to the control group, respectively. We also found that the C allele of rs820196 (OR = 1.492, 95 % CI 1.138 approximately 1.951; P = 0.004) and A allele of rs4789223 (OR = 1.767, 95 % CI: 1.354 approximately 2.301; P < 0.001) were common in the osteosarcoma patients than those in the control subjects, respectively. Haplotype analysis showed that TTA (OR = 3.469, 95 % CI 1.798 approximately 6.695; P < 0.001) was associated with increased risk for osteosarcoma. However, the TTG (OR = 0.578, 95 % CI 0.442 approximately 0.756) was associated with decreased risk for osteosarcoma. Our results suggested that RECQL5 genetic polymorphisms were associated with osteosarcoma in a Chinese population.

TÍTULO / TITLE: - Methylglyoxal induces oxidative stress and mitochondrial dysfunction in osteoblastic MC3T3-E1 cells.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Suh KS; Choi EM; Rhee SY; Kim YS
INSTITUCIÓN / INSTITUTION: - Research Institute of Endocrinology, Kyung Hee University Hospital, Seoul, Republic of Korea.
RESUMEN / SUMMARY: - Methylglyoxal is a reactive dicarbonyl compound produced by glycolytic processing and identified as a precursor of advanced glycation end products. The elevated methylglyoxal levels in patients with diabetes are believed to contribute to diabetic complications, including bone defects. The objective of this study was to evaluate the effect of methylglyoxal on the function of osteoblastic MC3T3-E1 cells. The data indicated that methylglyoxal decreased osteoblast differentiation and
induced osteoblast cytotoxicity. Pretreatment of MC3T3-E1 cells with aminoguanidine (a carbonyl scavenger), Trolox (an antioxidant), and cyclosporin A (a blocker of the mitochondrial permeability transition pore) prevented methylglyoxal-induced cytotoxicity in MC3T3-E1 cells. However, BAPTA/AM (an intracellular Ca2+ chelator) and dantrolene (an inhibitor of endoplasmic reticulum Ca2+ release) did not reverse the cytotoxic effect of methylglyoxal. Methylglyoxal increased the formation of intracellular reactive oxygen species, mitochondrial superoxide, and cardiolipin peroxidation in osteoblastic MC3T3-E1 cells. Methylglyoxal also decreased the mitochondrial membrane potential and intracellular ATP and nitric oxide levels, suggesting that carbonyl stress-induced loss of mitochondrial integrity contributes to the cytotoxicity of methylglyoxal. Furthermore, the results demonstrated that methylglyoxal induced protein adduct formation, inactivation of glyoxalase I, and activation of glyoxalase II. Aminoguanidine reversed all aforementioned effects of methylglyoxal. Taken together, these data support the notion that high methylglyoxal concentrations have detrimental effects on osteoblasts through a mechanism involving oxidative stress and mitochondrial dysfunction.

[116]

**TÍTULO / TITLE:** - Association between CTLA-4 genetic polymorphisms and susceptibility to osteosarcoma in Chinese Han population.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - Endocrine. 2013 Sep 28.

**AUTORES / AUTHORS:** - He J; Wang J; Wang D; Dai S; Yv T; Chen P; Ma R; Diao C; Lv G

**INSTITUCIÓN / INSTITUTION:** - Department of Spine, The Second Xiangya Hospital Affiliated with Central South University, No. 139 Middle Renmin Road, Changsha, 410011, Hunan Province, People’s Republic of China.

**RESUMEN / SUMMARY:** - Osteosarcoma (OS) is the most common malignant primary bone tumor in the world. The cytotoxic T-lymphocyte antigen-4 gene (CTLA-4) is an important candidate gene for influencing the development of OS. This study aimed to investigate the potential association of CTLA-4 genetic polymorphisms with OS risk in Chinese Han population. A total of 415 OS patients and 431 healthy controls were enrolled in this study. The created restriction site-polymerase chain reaction (CRS-PCR) and DNA sequencing methods were used to detect the genotyping of CTLA-4 c.75G>C and c.326G>A genetic polymorphisms. We observed that the genotypes/alleles of c.75G>C and c.326G>A genetic polymorphisms were statistically associated with the increased risk of OS (for c.75G>C, CC versus (vs.) GG: OR 1.72, 95 % CI 1.09-2.74; C vs. G: OR 1.30, 95 % CI 1.06-1.60; for c.326G>A, AA vs. GG: OR 2.12, 95 % CI 1.31-3.42; A vs. G: OR 1.31, 95 % CI 1.07-1.61). The allele-C/genotype-CC of c.75G>C and allele-A/genotype-AA of c.326G>A may contribute to OS susceptibility. These data indicate that CTLA-4 genetic polymorphisms are potentially related to OS risk in Chinese Han population, and might be used as molecular markers for evaluating the risk of OS.

[117]
Calcitonin-induced NF-kappaB Activation Up-regulates Fibronectin Expression in MG63 Osteosarcoma Cells.

RESUMEN / SUMMARY: Salmon calcitonin has been used extensively as a therapeutic tool in the regulation of bone remodeling. However, there is a growing body of evidence indicating that the calcitonin peptides are involved in regulation of cell growth, differentiation, survival and tissue development. In the present study, we investigated the effect of calcitonin in cell matrix interactions in MG63 cell line. Our results demonstrated that calcitonin increases cell growth of MG63 osteosarcoma cells in parallel with serine/threonine protein kinase B (AKT/PKB) activation. Moreover, calcitonin induced up-regulation of fibronectin expression in a nuclear factor-kappa B (NF-kappaB)-dependent manner, accompanied by enhanced enzymatic activity of matrix metalloproteinase-9 (MMP-9) and increased expression of tissue inhibitors of MMP-1 and -2. MMP-9 stimulation with calcitonin was accompanied by an increase in protein expression of the alpha5beta1 integrin receptor. To our knowledge, our results demonstrate, for the first time, that calcitonin is a potent inducer of fibronectin, an extracellular matrix component that is suggested to have a pro-oncogenic and healing effect, in a NF-kappaB-dependent manner.

OBJECTIVE: To elucidate the clinical profile of a concomitant diagnosis of adenomyosis in women with leiomyomas. DESIGN: Retrospective questionnaire-based study. SETTING: Academic medical center. PATIENT(S): The study sample comprised a total of 560 women: 159 women with adenomyosis and leiomyomas and 401 women with leiomyomas alone. INTERVENTION(S): Mailing of a symptom questionnaire. MAIN OUTCOME MEASURE(S): Comparison of women undergoing hysterectomy with adenomyosis and leiomyomas and women with leiomyomas alone. RESULT(S): Women with a concomitant diagnosis of adenomyosis and leiomyomas had significantly higher scores for disease burden during the menstrual period before surgery: heavy bleeding episodes and passing blood clots. Furthermore, women with adenomyosis and leiomyomas reported higher scores of distress regarding pelvic pain occurring during the menstrual period and pelvic pain not associated with the menstrual cycle. Moreover, in multivariate analysis, older age (odds
ratio (OR 1.10, 95% confidence interval [CI] 1.04-1.18), gravidity (OR 1.44, 95% CI 1.12-1.74), and pelvic pain occurring during the menstrual period (OR 1.27, 95% CI 1.06-1.54) increase the odds of having adenomyosis and not only leiomyomas.

CONCLUSION(S): Adenomyosis contributes to symptomatology in women with concomitant adenomyosis and leiomyomas.

[119]
TÍTULO / TITLE: - Inhibition of Six1 promotes apoptosis, suppresses proliferation, and migration of osteosarcoma cells.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Hua L; Fan L; Aichun W; Yongjin Z; Qingqing C; Xiaojian W
INSTITUCIÓN / INSTITUTION: - Department of Orthopedics, Haian Hospital of Traditional Chinese Medicine, 55 Ninghai Middle Road, Haian, 226600, Jiangsu Province, People’s Republic of China.

RESUMEN / SUMMARY: - Sineoculis homeobox homolog 1 (Six1) is one of the transcription factors that act as master regulators of development and is frequently dysregulated in cancers. However, the biological role of Six1 is not clear in osteosarcoma. To address the expression of Six1 in osteosarcoma cells, three osteosarcoma cell lines (U2OS, SaOS-2, and MG63) and a human osteoblastic cell line (hFOB1.19) were used to detect the expression of Six1 by quantitative real-time polymerase chain reaction and western blotting. The results showed that Six1 was upregulated in osteosarcoma cell lines compared to human osteoblastic cell line hFOB1.19. To investigate the role of Six1 in osteosarcoma cells, 3-(4,5-dimethylthiazol-2-yl)-2,5-diphenyltetrazolium bromide assay, flow cytometry analysis, and transwell chamber assays were used to determine the effects of Six1 on the cell viability, cycle, apoptosis, and migration properties in U2OS cells. The results showed that Six1 could promote U2OS cell proliferation and migration, and suppress U2OS cell apoptosis. In addition, we investigated the effects of Six1 on the expression of following proteins (cyclin D1, caspase-3, and vascular endothelial growth factor-C (VEGF-C)). Results showed that Six1 could increase the expression of cyclin D1 and VEGF-C, and decrease the expression of caspase-3. All these data suggested that Six1 might be involved in the promotion of growth, proliferation, and migration of U2OS cells, as well as the inhibition of apoptosis of U2OS cells. These data might provide information for the prediction of osteosarcoma prognosis and potential targets for therapy of osteosarcoma.

[120]
TÍTULO / TITLE: - Differential microRNA expression profiles between malignant rhabdoid tumor and epithelioid sarcoma: miR193a-5p is suggested to downregulate SMARCB1 mRNA expression.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Malignant rhabdoid tumor and epithelioid sarcoma are classified as tumors of uncertain differentiation. However, it is controversial whether these tumors are distinct entities because they share similar histological and immunohistochemical features such as the existence of rhabdoid cells or complete loss of SMARCB1 protein expression. MicroRNAs are small non-coding RNAs, and it is suggested that knowledge of microRNA expression profiles in cancer may have substantial value for diagnostics. We first analyzed microRNA expression profiles in 13 frozen materials (five malignant rhabdoid tumors, two proximal type epithelioid sarcomas, and six conventional type epithelioid sarcomas) and subsequently examined the specific microRNA expressions in 29 paraffin-embedded materials (8 malignant rhabdoid tumors, 13 proximal type epithelioid sarcomas, and 8 conventional type epithelioid sarcomas) and 13 previously described frozen materials by quantitative RT-PCR. According to the unsupervised hierarchical clustering of microRNA, proximal type epithelioid sarcoma and conventional type epithelioid sarcoma were classified into the same category, whereas malignant rhabdoid tumor was a distinct category from both types of epithelioid sarcoma. In addition, when malignant rhabdoid tumor with SMARCB1 gene alterations and proximal type and conventional type epithelioid sarcoma with no SMARCB1 gene alterations were compared, 56 microRNAs were isolated as being significantly different (ANOVA, P<0.05). Among them, quantitative RT-PCR using frozen and paraffin-embedded materials demonstrated that expression levels of miR193a-5p (P=0.002), which has been suggested to downregulate SMARCB1 mRNA expression, showed statistically different expression levels between malignant rhabdoid tumor and epithelioid sarcoma with no SMARCB1 gene alterations. These results suggest that epithelioid sarcoma, especially proximal type epithelioid sarcoma, and malignant rhabdoid tumor are distinct tumors with respect to the microRNA expression profiles and that miR193a-5p may have an important role in the inhibition of SMARCB1 mRNA expression.

Modern Pathology advance online publication, 29 November 2013; doi:10.1038/modpathol.2013.213.

[121] TITULO / TITLE: - A Rare Type of Secondary Cancer in a Child With Acute Lymphoblastic Leukemia: Malignant Fibrous Histiocytoma.

1097/MPH.0b013e318290c65c
For development of secondary cancer after treatment, the latency period varies between 5 and 10 years. In this case, a 13 year-old-boy diagnosed as high-risk ALL was treated with chemotherapy and prophylactic cranial radiotherapy at a dose of 1800 cGy. Six years after the end of treatment he developed a 5x5x4 cm mass at the right temporal region of the cranium. The mass was excised totally with clear surgical margin. Pathology of mass has been diagnosed as malignant fibrous histiocytoma (MFH), recently referred to as an undifferentiated pleomorphic sarcoma (UPS). After treatment of childhood ALL, reported cases of secondary MFH is extremely rare in the literature. Herein we present a case of MFH/UPS that developed as a secondary cancer 6 years after the end of ALL treatment.

[122]  
TÍTULO / TITLE: High ALK mRNA expression has a negative prognostic significance in rhabdomyosarcoma.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Bonvini P; Zin A; Alaggio R; Pawel B; Bisogno G; Rosolen A
INSTITUCIÓN / INSTITUTION: 1] Pediatric Oncology-Hematology Clinic, University-Hospital of Padova, via Giustiniani 3, 35100 Padova, Italy [2] Institute of Pediatric Research Citta della Speranza, 35100 Padova, Italy.
RESUMEN / SUMMARY: Background: Anaplastic lymphoma kinase (ALK) is a receptor tyrosine kinase aberrantly expressed in cancer, but its clinical and functional importance remain controversial. Mutation or amplification of ALK, as well as its expression levels assessed by conventional immunohistochemistry methods, has been linked to prognosis in cancer, although with potential bias because of the semi-quantitative approaches. Herein, we measured ALK mRNA expression in rhabdomyosarcoma (RMS) and determined its clinical impact on patients' stratification and outcome. Methods: Specimens were obtained from RMS patients and cell lines, and ALK expression was analysed by quantitative RT-PCR, western blotting, IHC, and copy number analysis. Results: High ALK mRNA expression was detected in the vast majority of PAX3/7-FOXO1-positive tumours, whereas PAX3/7-FOXO1-negative RMS displayed considerably lower amounts of both mRNA and protein. Notably, ALK mRNA distinguished unfavourable PAX3/7-FOXO1-positive tumours from PAX3/7-FOXO1-negative RMS (P<0.0001), and also correlated with larger tumour size (P<0.05) and advanced clinical stage (P<0.01), independently of fusion gene status. High ALK mRNA levels were of prognostic relevance by Cox univariate regression analysis and correlated with increased risk of relapse (P=0.001) and survival (P=0.01), whereas by multivariate analysis elevated ALK mRNA expression resulted a negative prognostic marker when clinical stage was not included. Conclusion: Quantitative assessment of ALK mRNA expression helps to improve risk stratification of RMS patients and identifies tumours with adverse biological characteristics and aggressive behaviour. British Journal of Cancer advance online publication, 22 October 2013; doi:10.1038/bjc.2013.653 www.bjcancer.com.

[123]
Mode of action of trabectedin in myxoid liposarcomas.

To elucidate the mechanisms behind the high sensitivity of myxoid/round cell liposarcoma (MRCL) to trabectedin and the suggested selectivity for specific subtypes, we have developed and characterized three MRCL xenografts, namely ML017, ML015 and ML004 differing for the breakpoint of the fusion gene FUS-CHOP, respectively of type I, II and III. FUS-CHOP binding to the promoters of some target genes such as Pentraxin 3 or Fibronectin 1, assessed by chromatin immunoprecipitation, was strongly reduced in the tumor 24 h after the first or the third weekly dose of trabectedin, indicating that the drug at therapeutic doses causes a detachment of the FUS-CHOP chimera from its target promoters as previously shown in vitro. Moreover, the higher sensitivity of MRCL types I and II appears to be related to a more prolonged block of the transactivating activity of the fusion protein. Doxorubicin did not affect the binding of FUS-CHOP to target promoters. Histologically, the response to trabectedin in ML017 and ML015 was associated with a marked depletion of non-lipogenic tumoral cells and vascular component, as well as lipidic maturation as confirmed by PPARgamma2 expression in western Blot. By contrast, in ML004 no major changes either in the cellularity or in the amount of mature were found, and consistently PPARgamma2 was null. In conclusion, the data support the view that the selective mechanism of action of trabectedin in MRCL is specific and related to its ability to cause a functional inactivation of the oncogenic chimera with consequent derepression of the adipocytic differentiation.

Advanced chondrosarcomas: role of chemotherapy and survival.

BACKGROUND: There are limited data about the role of chemotherapy in patients with advanced chondrosarcomas. METHODS: The medical charts of 180 patients with advanced chondrosarcomas having received chemotherapy in 15 participating institutions between 1988 and 2011 were reviewed. RESULTS:
Median age was 52 years. Sixty-three percent of patients had conventional chondrosarcoma and 88% had metastatic disease. Combination chemotherapy was delivered in 98 cases (54.5%). One hundred and thirty-one patients (73%) received an anthracycline-containing regimen. Using RECIST, the objective response rate was significantly different according to histological subtype, being 31% for mesenchymal chondrosarcoma, 20.5% for dedifferentiated chondrosarcoma, 11.5% for conventional chondrosarcoma and 0% for clear-cell chondrosarcoma (P = 0.04). Median progression-free survival (PFS) was 4.7 months [95% confidence interval (CI) 3-6.5]. Performance status (PS) >/=2, number of metastatic sites >/=1 and single-agent regimen were independently associated with poor PFS. Median overall survival (OS) was 18 months (95% CI 14.5-21.6). PS, number of metastatic sites and palliative surgery were independently associated with OS. CONCLUSIONS: Conventional chemotherapy have very limited efficacy in patients with advanced chondrosarcoma, the highest benefit being observed in mesenchymal and dedifferentiated chondrosarcoma. These data should be used as a reference for response and outcome in the assessment of investigational drugs in advanced chondrosarcoma.

[125]

**TITULO / TITLE:** - Mid-term outcome after curettage with polymethylmethacrylate for giant cell tumor around the knee: higher risk of radiographic osteoarthritis?

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - van der Heijden L; van de Sande MA; Heineken AC; Fiocco M; Nelissen RG; Dijkstra PD

**INSTITUCIÓN / INSTITUTION:** - Department of Orthopedic Surgery, Leiden University Medical Center, P.O. Box 9600, 2300 RC Leiden, the Netherlands. E-mail address for L. van der Heijden: lvanderheijden@lumc.nl.

**RESUMEN / SUMMARY:** - BACKGROUND: It has been suggested that, when a patient has a giant cell tumor, subchondral bone involvement close to articular cartilage and a hyperthermic reaction from polymethylmethacrylate (PMMA) are risk factors for the development of osteoarthritis. We determined the prevalence, risk factors, and clinical relevance of osteoarthritis on radiographs after curettage and application of PMMA for the treatment of giant cell tumors around the knee. METHODS: This retrospective single-center study included fifty-three patients with giant cell tumor around the knee treated with curettage and PMMA between 1987 and 2007. The median age at the time of follow-up was forty-two years (range, twenty-three to seventy years). There were twenty-nine women. Radiographic evidence of osteoarthritis was defined, preoperatively and postoperatively, as Kellgren and Lawrence grade 3 or 4 (KL3-4). We studied the influence of age, sex, tumor-cartilage distance, subchondral bone involvement (</=3 mm of residual subchondral bone), subchondral bone-grafting, intra-articular fracture, multiple curettage procedures, and complications on progression to KL3-4. Functional outcomes and quality of life were assessed with the Short Form-36 (SF-36), Musculoskeletal Tumor Society (MSTS) score, and Knee injury and Osteoarthritis Outcome Score (KOOS). RESULTS: After a median duration of follow-up of eighty-six months (range, sixty to 285 months), six patients (11%) had progression to
KL3, two (4%) had progression to KL4, and one had preexistent KL4. No patient underwent total knee replacement. The hazard ratio for KL3-4 was 9.0 (95% confidence interval [CI] = 2.0 to 41; p = 0.004) when >70% of the subchondral bone was affected and 4.2 (95% CI = 0.84 to 21; p = 0.081) when the tumor-cartilage distance was <=3 mm. Age, sex, subchondral bone-grafting, intra-articular fracture, multiple curettage procedures, and complications did not affect progression to KL3-4. Patients with KL3-4 reported lower scores on the KOOS symptom subscale (58 versus 82; p = 0.01), but their scores on the other KOOS subscales, the MSTS score (21 versus 24), and the SF-36 (76 versus 81) were similar to those for the patients with KL0, 1, or 2 (KL0-2). CONCLUSIONS: Seventeen percent of patients with giant cell tumor around the knee had radiographic findings of osteoarthritis after treatment with curettage and PMMA. A large amount of subchondral bone involvement close to articular cartilage increased the risk for osteoarthritis. The function and quality of life of the patients with KL3-4 were comparable with those for the patients with KL0-2, suggesting that radiographic findings of osteoarthritis at the time of intermediate follow-up had a modest clinical impact. Treatment with curettage and PMMA is safe for primary and recurrent giant cell tumors, even large tumors close to the joint. LEVEL OF EVIDENCE: Therapeutic Level IV. See Instructions for Authors for a complete description of levels of evidence.

[126]

**TITULO / TITLE:** Oxymatrine induces mitochondria dependent apoptosis in human osteosarcoma MNNG/HOS cells through inhibition of PI3K/Akt pathway.

**RESUMEN / SUMMARY:**

**Enlace al Resumen / Link to its Summary**

**REVISTA / JOURNAL:** Tumour Biol. 2013 Sep 29.

**AUTORES / AUTHORS:** Zhang Y; Sun S; Chen J; Ren P; Hu Y; Cao Z; Sun H; Ding Y

**INSTITUCIÓN / INSTITUTION:** Institute of Osteosarcoma, Tangdu Hospital of the Fourth Military Medical University, 1 Xinsi Road, Xi’an, Shaanxi, 710038, China.

**RESUMEN / SUMMARY:** The cytostatic drug from traditional Chinese medicinal herb has acted as a chemotherapeutic agent used in treatment of a wide variety of cancers. Oxymatrine, classified as a quinolizidine alkaloid, is a phytochemical product derived from Sophora flavescens, and has been reported to possess anticancer activities. However, the cancer growth inhibitory effects and molecular mechanisms in human osteosarcoma MNNG/HOS cell have not been well studied. In the present study, the cytotoxic effects of oxymatrine on MNNG/HOS cells were examined by MTT and bromodeoxyuridine (BrdU) incorporation assays. The percentage of apoptotic cells and the level of mitochondrial membrane potential (Δψm) were assayed by flow cytometry. The levels of apoptosis-related proteins were measured by Western blot analysis or enzyme assay Kit. Our results showed that treatment with oxymatrine resulted in a significant inhibition of cell proliferation and DNA synthesis in a dose-dependent manner, which has been attributed to apoptosis. Furthermore, we found that oxymatrine considerably inhibited the expression of Bcl-2 whilst increasing that of Bax. This promoted mitochondrial dysfunction, leading to the release of cytochrome c from the mitochondria to the cytoplasm, as well as the activation of caspase-9 and -3. Moreover, addition of oxymatrine to MNNG/HOS cells also attenuated phosphatidylinositol 3-kinase (PI3K) Akt signaling pathway cascade, evidenced by the
dephosphorylation of P13K and Akt. Likewise, oxymatrine significantly suppressed tumor growth in female BALB/C nude mice bearing MNNG/HOS xenograft tumors. In addition, no evidence of drug-related toxicity was identified in the treated animals by comparing the body weight increase and mortality. Therefore, these findings should be useful for understanding the apoptotic cellular mechanism mediated by oxymatrine and might offer a therapeutic potential advantage for human osteosarcoma chemoprevention or chemotherapy.

[127] TÍTULO / TITLE: - Dual targeting of EWS-FLI1 activity and the associated DNA damage response with Trabectedin and SN38 synergistically inhibits Ewing sarcoma cell growth.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Grohar PJ; Segars LE; Yeung C; Pommier Y; D'Incalci M; Mendoza A; Helman LJ
INSTITUCIÓN / INSTITUTION: - Division of Hematology-Oncology, Department of Pediatrics, Vanderbilt University and Vanderbilt Ingram Cancer Center.

PURPOSE: The goal of this study is to optimize the activity of trabectedin for Ewing sarcoma by developing a molecularly targeted combination therapy. EXPERIMENTAL DESIGN: We have recently shown that trabectedin interferes with the activity of EWS-FLI1 in Ewing sarcoma cells. In this report, we build on this work to develop a trabectedin based combination therapy with improved EWS-FLI1 suppression that also targets the drug associated DNA damage to ES cells. RESULTS: We demonstrate by siRNA experiments that EWS-FLI1 drives the expression of the Werner Syndrome protein (WRN) in ES cells. Since WRN deficient cells are known to be hypersensitive to camptothecins, we utilize trabectedin to block EWS-FLI1 activity, suppress WRN expression and selectively sensitize ES cells to the DNA damaging effects of SN38. We show that trabectedin and SN38 are synergistic, demonstrate an increase in DNA double strand breaks, an accumulation of cells in S-phase and a low picomolar IC50. In addition, SN38 cooperates with trabectedin to augment the suppression of EWS-FLI1 downstream targets, leading to an improved therapeutic index in vivo. These effects translate into the marked regression of two Ewing sarcoma xenografts at a fraction of the dose of camptothecin used in other xenograft studies. CONCLUSIONS: These results provide the basis and rationale for translating this drug combination to the clinic. In addition, the study highlights an approach that utilizes a targeted agent to interfere with an oncogenic transcription factor and then exploits the resulting changes in gene expression to develop a molecularly targeted combination therapy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

Enlace al texto completo (gratuito o de pago) 1016/j.ejso.2013.09.002

Di Monta G; Caraco C; Benedetto L; La Padula S; Marone U; Tornesello ML; Buonaguro FM; Simeone E; Ascierto PA; Mozzullo N

Department of Surgery “Melanoma, Soft Tissues, Head and Neck, Skin Cancers”, Istituto Nazionale dei Tumori Fondazione Pascale Napoli 80131 Naples, Italy. Electronic address: gidimonta@libero.it.

BACKGROUND: Electrochemotherapy (ECT) is a novel modality for the treatment of skin nodules and cutaneous or subcutaneous tumors that allows delivery of low and non-permeant drug into cells. The aim of this prospective single-center study was to evaluate ECT efficacy in the local treatment of Classic Kaposi’s sarcoma (CKS) skin localization stage I-II sec. Brambilla et al. METHODS: Nineteen consecutive patients affected by classic KS were included in this study. All patients underwent blood sampling and concurrent incisional biopsy for histological diagnosis and Kaposi’s sarcoma related herpes virus 8 (HHV-8) molecular analysis. ECT treatment of KS cutaneous lesions were performed according to the European Standard Operating Procedures of Electrochemotherapy (ESOPE). The primary endpoint of the study was the evaluation of ECT efficacy in the treatment of KS skin nodules and the assessment of HHV-8 viral load in the peripheral blood following the ECT therapy. RESULTS: Complete response (CR) was observed in 14 (73.6%) patients after first ECT session, while 3 (15.7%) and 2 (10.5%) out of 19 patients received a second and a third ECT treatment, respectively. Clinical response dragged out the whole follow-up period that ranged between 6 and 31 months with a median of 16 months. CONCLUSIONS: Clinical management of CKS skin localizations still represents a challenging task for surgeons and oncologists. Therefore, according to this and other author’s recent experiences, ECT is claimed to become the “new standard of care” as first line treatment strategy for stage I-II CKS patients.

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Sarcomas are a heterogeneous family of mesenchymal malignancies that employ an impressive variety of pathogenic mechanisms. The traditional role of the pathologist in this field has been to ensure accurate diagnosis and histologic grading to direct therapy. More recently, with the advent of targeted therapies directed at particular molecular alterations, the role of the pathologist has expanded and increased awareness of the genetic features of sarcomas is needed to deliver optimal multidisciplinary care. This review discusses these trends and briefly enumerates many of the molecular derangements and targeted agents currently used or under investigation in soft tissue sarcoma. A few sarcomas are highlighted in more detail to illustrate how pathologists can exert positive influence on patient care-not just
through diagnosis and grading, but with molecular characterizations as well. Featured sarcomas include alveolar soft part sarcoma, dermatofibrosarcoma protuberans, gastrointestinal stromal tumor, inflammatory myofibroblastic tumor, and PEComas. This article is protected by copyright. All rights reserved.

[130] TÍTULO / TITLE: Osteoblastic differentiation of human stem cells derived from bone marrow and periodontal ligament under the effect of enamel matrix derivative and transforming growth factor-Beta.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Houshmand B; Behnia H; Khoshzaban A; Morad G; Behrouzi G; Dashti SG; Khojasteh A

RESUMEN / SUMMARY: Purpose: To increase the understanding of the applicability of biomaterials and growth factors in enhancing stem cell-based bone regeneration modalities, this study evaluated the effects of enamel matrix derivative (EMD) and recombinant human transforming growth factor-beta (rhTGF-beta) on osteoblastic differentiation of human bone marrow mesenchymal stem cells (hBMSCs) as well as human periodontal ligament stem cells (hPDLSCs). Materials and Methods: hBMSCs and hPDLSCs were obtained, and identification of stem cell surface markers was performed according to the criteria of the International Society for Cellular Therapy. Each group of stem cells was separately treated with a serial dilution of EMD (10, 50, and 100 mug/mL) or rhTGF-beta (10 ng/mL). Osteoblastic differentiation was examined through in vitro matrix mineralization by alizarin red staining, and mRNA expression of osteopontin and osteonectin was determined by quantitative reverse-transcriptase polymerase chain reaction. hPDLSCs were further assessed for osteocalcin mRNA expression. Stem cells cultured in osteogenic medium were employed as a standard positive control group. Results: In none of the experimental groups were bone-related mRNAs detected subsequent to treatment with EMD for 5, 10, and 15 days. Alizarin red staining on day 21 was negative in EMD-treated BMSC and PDLSC cultures. In rhTGF-beta-supplemented BMSC culture, expression of osteonectin mRNA was demonstrated on day 15, which was statistically comparable to the positive control group. Nevertheless, extracellular matrix mineralization was inhibited in both groups of stem cells. Conclusions: Within the limitations of this study, it could be concluded that EMD with a concentration of 10, 50, or 100 mug/mL has no appreciable effect on osteoblastic differentiation of BMSCs and PDLSCs. Application of rhTGF-beta increased osteonectin mRNA expression in BMSCs. This finding corroborates the hypothesis that TGF-beta might be involved in early osteoblastic maturation.


RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Shui W; Zhang W; Yin L; Nan G; Liao Z; Zhang H; Wang N; Wu N; Chen X; Wen S; He Y; Deng F; Zhang J; Luu HH; Shi LL; Hu Z; Haydon RC; Mok J; He TC

**INSTITUCIÓN / INSTITUTION:** - Department of Orthopaedic Surgery, The First Affiliated Hospital of Chongqing Medical University, Chongqing, 400016, China; Institute of Orthopaedic Research and Education, Chongqing Medical University, Chongqing, 400016, China.

**RESUMEN / SUMMARY:** - Successful bone tissue engineering at least requires sufficient osteoblast progenitors, efficient osteoinductive factors, and biocompatible scaffolding materials. We have demonstrated that BMP9 is one of the most potent factors in inducing osteogenic differentiation of mesenchymal progenitors. To facilitate the potential use of cell-based BMP9 gene therapy for bone regeneration, we characterize the in vivo osteoconductive activities and bone regeneration potential of three clinically-used scaffold materials, type I collagen sponge, hydroxyapatite-tricalcium phosphate (HA-TCP) and demineralized bone matrix (DBM), using BMP9-expressing C2C12 osteoblastic progenitor cells. We find that recombinant adenovirus-mediated BMP9 expression effectively induces osteogenic differentiation in C2C12 cells. Although direct subcutaneous injection of BMP9-transduced C2C12 cells forms ectopic bony masses, subcutaneous implantation of BMP9-expressing C2C12 cells with collagen sponge or HA-TCP scaffold yields the most robust and mature cancellous bone formation, whereas the DBM carrier group forms no or minimal bone masses. Our results suggest that collagen sponge and HA-TCP scaffold carriers may provide more cell-friendly environment to support the survival, propagation, and ultimately differentiation of BMP9-expressing progenitor cells. This line of investigation should provide important experimental evidence for further pre-clinical studies in BMP9-mediated cell based approach to bone tissue engineering.

**[132] TÍTULO / TITLE:** - Osthole inhibits proliferation and induces apoptosis in human osteosarcoma cells.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Ding Y; Lu X; Hu X; Ma J; Ding H

**RESUMEN / SUMMARY:** - Objective: The purpose of this study was to investigate the effect of osthole on osteosarcoma cell proliferation and apoptosis. Method: Cell counting Kit-8 assay was performed to establish the effects of osthole on osteosarcoma MG-63 cell proliferation. Annexin V-FITC/PI was performed to analyze the apoptotic rate of the cells. Result: The inhibitory effects of osthole on the expression of BCL-2, BAX, and caspase-3 were detected by Western blotting. Osthole inhibited the growth of human osteosarcoma MG-63 cells by inhibiting cell proliferation and induced cell apoptosis. Western blotting demonstrated that osthole downregulated the expressions of BCL-2 and caspase-3 and upregulated the expression of BAX in human osteosarcoma cells. Conclusion: Osthole can inhibit osteosarcoma cell proliferation and induced apoptosis effectively in a dose-dependent manner through downregulating the expression of BCL-2 and caspase-3 proteins levels and upregulating the expression of BAX proteins levels.
Case 199: aggressive angiomyolipoma with renal vein thrombosis and pulmonary fat embolus.

Enlace al Resumen / Link to its Summary


Enlace al texto completo (gratuito o de pago) 1148/radiol.13121187

Yarmish G; Dipoce J

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Association of D2-40 and MMP-1 expression with cyst formation in lung metastatic lesions of cutaneous angiosarcoma on the scalp: immunohistochemical analysis of 23 autopsy cases.

Enlace al Resumen / Link to its Summary


Enlace al texto completo (gratuito o de pago) 1016/j.humpath.2013.07.022

Masuzawa M; Mikami T; Numata Y; Tokuyama W; Masuzawa M; Murakumo Y; Okayasu I; Katsuoka K

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Cutaneous angiosarcoma of the scalp can rapidly develop into pulmonary metastasis. The pulmonary metastatic lesions display a unique appearance, so-called thin-walled cysts, which cause a fatal relapsed pneumothorax by rupturing. We analyzed 23 autopsy cases of angiosarcoma with pulmonary metastasis to elucidate the mechanism of the thin-walled cyst development. Of the 23 cases of cutaneous angiosarcoma of the scalp with pulmonary metastasis, radiological examination revealed pulmonary metastatic lesions as thin-walled cysts (39%), nodules (39%), mixed cysts and nodules (13%), and ground-glass opacity (9%). All the cases but one with cystic metastases were complicated by pneumothorax. The cystic lesions were accompanied by podoplanin (D2-40)-positive tumor cells in the luminal surface of the cysts. In both primary cutaneous lesions and pulmonary metastatic lesions, the D2-40 expression was positive for angiosarcoma cells in 100% and 92% of the cases, respectively. While the estrogen-regulated gene (ERG) expression was also positive for most of the primary and metastatic pulmonary angiosarcomas, D2-40 was a more useful marker to differentiate tumor cells from the background than was the ERG expression of the vascular endothelium. Matrix metalloproteinase-1 (MMP-1) expression was also predominant in primary lesions (95%) and pulmonary metastatic lesions (82.6%). Proteinases, like MMP-1, might be associated with a developing thin-walled cyst, although there were no differences in the MMP-1 expression in either the cystic or nodular metastasis. Two extremely aggressive cases showed cystic
metastasis with central necrosis that was not observed in other cases. These results suggest a pathogenesis of thin-walled cysts in some progressive cases.

[135]

TÍTULO / TITLE: - Comparative pathophysiology, toxicology, and human cancer risk assessment of pharmaceutical-induced hibernoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Radi Z; Bartholomew P; Elwell M; Vogel WM
INSTITUCIÓN / INSTITUTION: - Pfizer Worldwide Research and Development, Drug Safety R&D, 1 Burtt Rd., Andover, MA 01810, USA. Electronic address: zaher.radi@pfizer.com.
RESUMEN / SUMMARY: - In humans, hibernoma is a very rare, benign neoplasm of brown adipose tissue (BAT) that typically occurs at subcutaneous locations and is successfully treated by surgical excision. No single cause has been accepted to explain these very rare human tumors. In contrast, spontaneous hibernoma in rats is rare, often malignant, usually occurs in the thoracic or abdominal cavity, and metastases are common. In recent years, there has been an increased incidence of spontaneous hibernomas in rat carcinogenicity studies, but overall the occurrence remains relatively low and highly variable across studies. There have only been four reported examples of pharmaceutical-induced hibernoma in rat carcinogenicity studies. These include phentolamine, an alpha-adrenergic antagonist; varenicline, a nicotine partial agonist; tofacitinib, a Janus kinase (JAK) inhibitor; and hydromorphone, an opioid analgesic. Potential non-genotoxic mechanisms that may contribute to the pathogenesis of BAT activation/proliferation and/or subsequent hibernoma development in rats include: (1) physiological stimuli, (2) sympathetic stimulation, (3) peroxisome proliferator-activated receptor (PPAR) agonism, and/or (4) interference or inhibition of JAK/Signal Transducer and Activator of Transcription (JAK/STAT) signaling. The evaluation of an apparent increase of hibernoma in rats from 2-year carcinogenicity studies of novel pharmaceutical therapeutics and its relevance to human safety risk assessment is complex. One should consider: the genotoxicity of the test article, dose/exposure and safety margins, and pathophysiologic and morphologic differences and similarities of hibernoma between rats and humans. Hibernomas observed to date in carcinogenicity studies of pharmaceutical agents do not appear to be relevant for human risk at therapeutic dosages.

[136]

TÍTULO / TITLE: - MicroRNA-128 promotes proliferation in osteosarcoma cells by downregulating PTEN.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Shen L; Chen XD; Zhang YH
INSTITUCIÓN / INSTITUTION: - Department of Orthopaedic Surgery, Xinhua Hospital, Shanghai JiaoTong University School of Medicine, No.1665 kongjiang Rd., Shanghai, 200092, People’s Republic of China.

RESUMEN / SUMMARY: - MicroRNAs are a class of small noncoding RNAs that function as critical gene regulators through targeting mRNAs for translational repression or degradation. Several studies have indicated that abnormal expression of miRNAs occurs frequently in human osteosarcoma. In the present study, we found that miR-128 expression was significantly increased in osteosarcoma tissues compared to adjacent normal tissues. Ectopic overexpression of miR-128 significantly promoted while suppression of miR-128 by its antisense inhibited the proliferation of MG63 and U2OS cells. At the molecular level, our results demonstrated that miR-128 overexpression could repress expression of PTEN by directly targeting PTEN 3’-untranslated region. Consistently, downstream AKT signaling was altered by miR-128 overexpression or knockdown. Therefore, our results suggest that miR-128 plays an important role in the proliferation of human osteosarcoma cells by directly regulation of PTEN/AKT signaling.

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[137]

TÍTULO / TITLE: - Induction of G2/M phase cell cycle arrest and apoptosis by ginsenoside Rf in human osteosarcoma MG63 cells through the mitochondrial pathway.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Shangguan WJ; Li H; Zhang YH

INSTITUCIÓN / INSTITUTION: - Department of Traditional Chinese Medicine, Renji Hospital, Shanghai Jiaotong University School of Medicine, Shanghai 200127, P.R. China.

RESUMEN / SUMMARY: - Ginsenosides, extracted from the traditional Chinese herb ginseng, are a series of novel natural anticancer products known for their favorable safety and efficacy profiles. The present study aimed to investigate the cytotoxicity of ginsenoside Rf to human osteosarcoma cells and to explore the anticancer molecular mechanisms of ginsenoside Rf. Five human osteosarcoma cell lines (MG-63, OS732, U-2OS, HOS and SAOS-2) were employed to investigate the cytotoxicity of ginsenoside Rf by MTT and colony forming assays. After treatment with ginsenoside Rf, MG-63 cells which were the most sensitive to ginsenoside Rf, were subjected to flow cytometry to detect cell cycle distribution and apoptosis, and nuclear morphological changes were visualized by Hoechst 33258 staining. Caspase-3, -8 and -9 activities were also evaluated. The expression of cell cycle markers including cyclin B1 and Cdk1 was detected by RT-PCR and western blotting. The expression of apoptotic genes Bcl-2 and Bax and the release of cytochrome c were also examined by western blotting. Change in the mitochondrial membrane potential was observed by JC-1 staining in situ. Our results demonstrated that the cytotoxicity of ginsenoside Rf to these human osteosarcoma cell lines was dose-dependent, and the MG-63 cells were the most sensitive to exposure to ginsenoside Rf. Additionally, ginsenoside Rf induced G2/M phase cell cycle arrest and apoptosis in MG-63 cells. Furthermore, we observed
upregulation of Bax and downregulation of Bcl-2, Cdk1 and cyclin B1, the activation of caspase-3 and -9 and the release of cytochrome c in MG-63 cells following treatment with ginsenoside Rf. Our findings demonstrated that ginsenoside Rf induces G2/M phase cell cycle arrest and apoptosis in human osteosarcoma MG-63 cells through the mitochondrial pathway, suggesting that ginsenoside Rf, as an effective natural product, may have a therapeutic effect on human osteosarcoma.
OBJECTIVE: Angiosarcomas are aggressive, malignant soft tissue neoplasms of endothelial origin and occur rarely in the female genital tract. There is lack of consensus on risk factors for poor outcome and optimal treatment. To this end, we performed a clinicopathologic review and survival analysis. METHODS: We report a case of a woman with an angiosarcoma of the vagina. Published English literature was reviewed for angiosarcomas of the vulva, vagina, uterus, and ovary. Survival was evaluated by using Kaplan-Meier analysis and the effect of clinical and demographic variables on survival by using Cox regression analysis. RESULTS: A total of 51 patients were identified with a median age of 47 years (range, 17-87 years). Two of the patients had an angiosarcoma of the vulva; 2 had an angiosarcoma of the vagina; 18 had an angiosarcoma of the uterus, and 29 had an angiosarcoma of the ovary. Five-year overall survival was 27% (SE, 8%). Most patients presented with locoregional disease, having surgery as their primary intervention. Overall, adjuvant therapy significantly improved survival (hazards ratio, 0.17; 95% confidence interval, 0.05-0.59; adjusted for age and tumor size). Adjuvant treatment consisted of radiotherapy for angiosarcomas of the vulva, vagina, and uterus and chemotherapy for ovarian angiosarcomas. Subgroup analysis of the female genital tract site was hampered by the small number of cases. CONCLUSIONS: This review supports the use of surgical and adjuvant radiotherapy for angiosarcomas of the vulva, vagina, and uterus. Cytoreductive surgery and adjuvant chemotherapy remain the primary treatment of angiosarcomas of the ovary.

[140]
TÍTULO / TITLE: CD8 tumor-infiltrating lymphocytes at primary sites as a possible prognostic factor of cutaneous angiosarcoma.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: Fujii H; Arakawa A; Utsumi D; Sumiyoshi S; Yamamoto Y; Kitoh A; Ono M; Matsumura Y; Kato M; Konishi K; Shiga T; Sano S; Sakaguchi S; Miyagawa-Hayashino A; Takahashi K; Uezato H; Miyachi Y; Tanioka M
INSTITUCIÓN / INSTITUTION: Department of Dermatology, Kyoto University Graduate School of Medicine, Kyoto, Japan.

RESUMEN / SUMMARY: Tumor-infiltrating lymphocytes (TILs) have been reported as a prognostic factor in various cancers and are a promising target for immunotherapy. To investigate whether TILs have any impact on the prognosis of angiosarcoma patients, 55 non-treated patients (40 patients at stage 1 with cutaneous localized tumors, 4 patients at stage 2 with lymph node metastases and 11 patients at stage 3 with distant metastases) with angiosarcoma were evaluated retrospectively by
immunohistochemistry stained CD4, CD8, FOXP3 and Ki67. The Kaplan-Meier method was used to estimate overall survival with patients at stage 1. Survival differences were analyzed by the log-rank test. Patients with higher numbers of CD8+ TILs in their primary tumors survived significantly longer compared with patients with lower values. Moreover, the number of CD8 in TILs was positively correlated with a distant metastasis-free period. The total number of primary TILs (CD4 plus CD8) and CD8+ primary TILs of stage 3 patients with distant metastases was positively correlated with their overall survival. To evaluate whether CD8+ effector T cells are activated or differentiated, flow cytometric analysis of peripheral blood mononuclear cells (PBMC) was performed. The percentages of CD8+ T cells producing IFN-gamma in PBMC were significantly higher in patients with angiosarcoma (n = 10) compared not only with that of healthy controls (n = 20) but also patients with advanced melanoma (n = 11). These results suggest that anti-tumor immunity is clinically relevant in angiosarcoma.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Hatakeyama W; Taira M; Chosa N; Kihara H; Ishisaki A; Kondo H
INSTITUCIÓN / INSTITUTION: Department of Prosthodontics and Oral Implantology, Iwate Medical University School of Dentistry, Morioka, Iwate 020-8505, Japan.
RESUMEN / SUMMARY: The development of new osteoconductive bone substitute materials is expected in medicine. In this study, we attempted to produce new hydroxylapatite (HAP)/collagen (Col) composites using two HAP particles of different sizes and porcine type I collagen. The two HAP particles were either nano-sized (40 nm in average diameter; n-HAP) or had macro-pore sizes of 0.5-1.0 mm in length with fully interconnected pores (m-HAP). The aim of this study was to investigate the effects of apatite particle size in two HAP/Col composites on the osteogenic differentiation profile in osteoblast-like cells (SaOS-2). We created a collagen control sponge (Col) and two HAP/Col composite sponges (n-HAP/Col and m-HAP/Col) using freeze-drying and dehydrothermal cross-linking techniques, and then punched out samples of 6 mm in diameter and 1 mm in height. The SaOS-2 cells were cultured on three test materials for 1, 2, 3 and 4 weeks. Total RNA was extracted from the cultured cells and the expression of osteogenic differentiation-related genes was evaluated by reverse transcription PCR (RT-PCR) using primer sets of alkaline phosphatase (ALP), type 1 collagen (COL1), bone sialoprotein (BSP) and osteocalcin precursor [bone gamma-carboxyglutamate (gla) protein (BGLAP)] genes, as well as the beta-actin gene. The cells were also cultured on Col, n-HAP/Col and m-HAP/Col specimens for 1 and 4 weeks, and were then observed under a scanning electron microscope (SEM). The experimental results were as follows: RT-PCR indicated that osteogenic differentiation, particularly the gene expression of BSP, was most accelerated when the cells were cultured on n-HAP/Col specimens, followed by m-HAP/Col, whilst the weakest acceleration was observed when the cells were cultured on Col specimens.
As shown by the SEM images, the SaOS-2 cells were fibroblastic when cultured on Col specimens for up to 4 weeks; they were fibroblastic when cultured on n-HAP/Col specimens for 1 week, but appeared as spheroids, while actively phagocytizing n-HAP particles at 4 weeks; however, they appeared as deformed fibroblasts when cultured on m-HAP/Col specimens, detached from the particles. Despite limited experimental results, our study suggests that n-HAP/Col may be employed as a new osteoconductive bone substitute material.

[142]
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Tivoli YA; Thomas JA; Chen AF; Weiss ET
INSTITUCIÓN / INSTITUTION: Nova Southeastern University/Broward General Medical Center, Fort Lauderdale, Florida.

[143]
TÍTULO / TITLE: Role of radiation therapy in the conservative management of sarcoma within an irradiated field.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Modesto A; Filleron T; Chevreau C; Le Pechoux C; Rochaix P; Le Guelllec S; Ducassou A; Gangloff D; Ferron G; Delannes M
RESUMEN / SUMMARY: PURPOSE: To report on clinical outcome and toxicity profile after combined treatment that included radiation therapy (RT) in patients with localized sarcoma within an irradiated field. PATIENTS AND METHODS: Individual clinical data from all consecutive patients diagnosed and treated for a localized SIF between January 2000 and October 2011 at the Institut Claudius Regaud, Toulouse, France, were retrospectively reviewed. Outcomes of patients with SIF who underwent adjuvant or definitive radiotherapy were compared with patients who did not receive further RT. RESULTS: Of the 27 patients eligible for this study: surgery alone (S), surgery followed by RT (S + RT) or definitive RT (RT) was performed in 16, 8 and 2 cases respectively. The rate of unresectable, gross or microscopically positive margin disease among the 10 re-irradiated patients was significantly higher than the non re-irradiated group (90% vs. 12% p < 0.001). After a median follow-up of 3.8 years, there was a trend toward longer survival and better local control in the subgroup of patients who received adjuvant or definitive RT compared to the rest of the cohort with an acceptable toxicity profile. The 4-year relapse free survival rates of patients treated with and without RT were 53% and 27% respectively (p = 0.09). CONCLUSION: SIF
complete surgical resection is often difficult to achieve, enhancing the risk of relapse. RT should be discussed in case of unresectable tumor or after suboptimal surgery as part of intensified local management that has a curative intent.

[144]
TÍTULO / TITLE: Angiosarcoma of the scalp successfully treated with pazopanib.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Tomita H; Koike Y; Asai M; Ogawa F; Abe K; Tanioka M; Utani A
INSTITUCIÓN / INSTITUTION: Department of Dermatology, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan.

[145]
TÍTULO / TITLE: Deferoxamine promotes osteoblastic differentiation in human periodontal ligament cells via the nuclear factor erythroid 2-related factor-mediated antioxidant signaling pathway.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Chung JH; Kim YS; Noh K; Lee YM; Chang SW; Kim EC
INSTITUCIÓN / INSTITUTION: Department of Periodontology, School of Dentistry, Kyung Hee University, Seoul, Korea.
RESUMEN / SUMMARY: BACKGROUND AND OBJECTIVE: Recently it was reported that deferoxamine (DFO), an iron chelator, stimulates bone formation from MG63 and mesenchymal stem cells, but inhibits differentiation in rat calvarial cells; however, the effect of DFO on osteoblastic differentiation in human periodontal ligament cells (hPDLCs) has not been reported. The aim of this study was to investigate the effects and the possible underlying mechanism of DFO on osteoblastic differentiation of hPDLCs. MATERIAL AND METHODS: The effect of DFO on osteoblast differentiation was determined by the staining intensity of calcium deposits with Alizarin red and by RT-PCR analysis of the expression of osteoblastic markers. Signal transduction pathways were analyzed by western blotting. RESULTS: DFO increased osteogenic differentiation in a concentration-dependent manner by expression of the mRNA for differentiation markers and calcium nodule formation. Exposure of hPDLCs to DFO resulted in increases in the production of reactive oxygen species and in the levels of nuclear factor erythroid 2-related factor (Nrf2) protein in nuclear extractions, as well as a dose-dependent increase in the expression of Nrf2 target genes, including glutathione (GSH), glutathione S-transferase, gamma-glutamylcysteine lygase, glutathione reductase and glutathione peroxidase. Pretreatment with Nrf2 small interfering RNA, GSH depletion by buthionine sulfoximine and diethyl maleate, and with antioxidants by N-acetylcysteine and vitamin E, blocked DFO-stimulated osteoblastic differentiation. Furthermore, pretreatment with GSH depletion and antioxidants blocked DFO-induced p38 MAPK, ERK, JNK and nuclear factor-kappaB pathways.
CONCLUSION: These data indicate, for the first time, that nontoxic DFO promotes osteoblastic differentiation of hPDLCs via modulation of the Nrf2-mediated antioxidant pathway.

[146]
**TÍTULO / TITLE:** - Extraluminal GI stromal tumor of the jejunum diagnosed by EUS at double-balloon endoscopy.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](https://doi.org/10.1016/j.gie.2013.09.016)
**AUTORES / AUTHORS:** - Nakamura M; Ohmiya N; Hirooka Y; Kawashima H; Yamamura T; Ishihara M; Yamada K; Nagura A; Yoshimura T; Miyahara R; Funasaka K; Itoh A; Ohno E; Ando T; Watanabe O; Uehara K; Yoshioka Y; Nagino M; Goto H
**INSTITUCIÓN / INSTITUTION:** - Department of Gastroenterology and Hepatology, Nagoya University Graduate School of Medicine, Nagoya, Japan.

[147]
**TÍTULO / TITLE:** - Expression of insulin-like growth factor II mRNA-binding protein 3 (IMP3) in sacral chordoma.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](https://doi.org/10.1007/s11060-013-1274-4)
**AUTORES / AUTHORS:** - Zhou M; Chen K; Yang H; Wang G; Lu J; Ji Y; Wu C; Chen C
**INSTITUCIÓN / INSTITUTION:** - Department of Orthopedic Surgery, The First Affiliated Hospital of Soochow University, 188 Shizi St, Suzhou, 215006, Jiangsu, China.
**RESUMEN / SUMMARY:** - Sacral chordoma is a rare and aggressive tumor, with a high rate of local recurrence even when the tumor is radically resected. The fundamental knowledge of its biological behavior remains unknown. Insulin-like growth factor II mRNA-binding protein 3 (IMP3) is one of the RNA binding proteins and is expressed during embryogenesis and in various malignant tumors. This study evaluated expression of IMP3 in sacral chordoma for association with patient’s clinicopathological factors. A total of 32 patients with sacral chordoma (17 male and 15 female) and 10 samples of distant normal tissues were collected for analysis of IMP3 expression using immunohistochemistry. Association between IMP3 expression and clinicopathological factors (such as patient’s age, gender, tumor location, tumor size, surrounding muscle invasion, Ki-67 expression, and tumor recurrence) were statistically analyzed. IMP3 was expressed in 20 (62.5 %) patients, whereas there was no expression in the 10 distant normal tissues. IMP3 expression was associated with tumor invasion into the surrounding muscle (P = 0.028), high levels of Ki-67 expression (P = 0.009), and tumor recurrence (P = 0.012). The log-rank test revealed that patients with positive IMP3 expression had a shorter continuous disease-free survival time than those with negative IMP3 expression (P = 0.016). IMP3 expression was independent of age, gender, tumor location and tumor size. These results indicate that IMP3 was overexpressed in sacral chordoma and this expression was associated with tumor invasion and recurrence; thus, IMP3 may play an important role in tumor progression.
and could serve as a prognostic biomarker for sacral chordoma and IMP3 could be used as a potential therapeutic target for the treatment of sacral chordoma.

[148]

TÍTULO / TITLE:  - The innocent bystander: papillary fibroelastoma.
RESUMEN / SUMMARY:  - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS:  - Li A; Azarisman SM; Teo KS; Worthley MI; Sidharta S; Glenie T; Samaraie L; Chua SK; Bailie TJ; Stuklis R; Worthley SG
INSTITUCIÓN / INSTITUTION:  - Cardiovascular Research Centre, Royal Adelaide Hospital, University of Adelaide, Adelaide, South Australia, Australia; Department of Medicine, Ruttonjee Hospital, Wan Chai, Hong Kong. Electronic address: liyw@graduate.hku.hk.

[149]

TÍTULO / TITLE:  - Pleomorphic liposarcoma: A clinicopathological, immunohistochemical and molecular cytogenetic study of 32 additional cases.
RESUMEN / SUMMARY:  - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS:  - Wang L; Ren W; Zhou X; Sheng W; Wang J
INSTITUCIÓN / INSTITUTION:  - Department of Pathology, Cancer Hospital, Fudan University, Shanghai, China; Department of Oncology, Shanghai Medical College, Fudan University, Shanghai, China.
RESUMEN / SUMMARY:  - The purpose of this study is to report the author’s experience with 32 cases of pleomorphic liposarcoma to further broaden the clinicopathological spectrum. The tumours occurred equally in males and females with ages ranging from 11 to 83 years (median, 56 years). Tumour site included the extremities (17 cases), abdomen/retroperitoneum (4 cases), internal organs (5 cases), thoracic cavity/mediastinum (2 cases), orbit, neck, groin and scrotum (1 case each). The diagnostic pleomorphic lipoblasts were identified in 31 primary tumours and one recurrent tumor but varied widely in proportion between cases or different areas of the same tumor. Four tumors contained sheets or focal aggregates of lipoblasts with epithelioid morphology. The nonlipogenic component in 26 cases had an appearance of undifferentiated pleomorphic sarcoma, whereas in six cases it was consistent with intermediate to high grade myxofibrosarcoma. The pleomorphic and epithelioid lipoblasts displayed variable expression of S100 protein. There was no signal of amplified MDM2 gene in 10 cases tested by fluorescence in situ hybridization. This study further illustrates that pleomorphic liposarcoma is a distinctive entity with no relationship to either well differentiated liposarcoma or dedifferentiated liposarcoma. Albeit very rare, pleomorphic liposarcoma can occur in teenaged patients and internal organs.
Exomic analysis of myxoid liposarcomas, synovial sarcomas, and osteosarcomas.

Bone and soft tissue sarcomas are a group of histologically heterogeneous and relatively uncommon tumors. To explore their genetic origins, we sequenced the exomes of 13 osteosarcomas, eight myxoid liposarcomas (MLPS), and seven synovial sarcomas (SYN). These tumors had few genetic alterations (median of 10.8). Nevertheless, clear examples of driver gene mutations were observed, including canonical mutations in TP53, PIK3CA, SETD2, AKT1, and subclonal mutation in FBXW7. Of particular interest were mutations in H3F3A, encoding the variant histone H3.3. Mutations in this gene have only been previously observed in gliomas. Loss of heterozygosity of exomic regions was extensive in osteosarcomas but rare in SYN and MLPS. These results provide intriguing nucleotide-level information on these relatively uncommon neoplasms and highlight pathways that help explain their pathogenesis.

Lapatinib alters the malignant phenotype of osteosarcoma cells via downregulation of the activity of the HER2-PI3K/AKT-FASN axis in vitro.

Lapatinib, an inhibitor of human epidermal growth factor receptor 2 (HER2) phosphorylation, has been reported to inhibit several types of tumors such as HER2-overexpressing breast cancer. However, the effect of lapatinib on the malignant phenotype of human osteosarcoma (OS) cells and the potential molecular mechanisms remain unclear. To elucidate the effect of lapatinib on OS, two OS cell lines, U2-OS and MG-63, were utilized in the present study. Various concentrations of lapatinib were used to treat OS cells for different time durations. Cell proliferation was evaluated by MTT and colony formation assays. Flow cytometry (FCM) was used to evaluate cell apoptosis. Wound healing and Transwell invasion assays were performed to examine the migratory and invasive abilities of the cells.
investigate the possible molecular mechanisms involved, the expression of p-HER2, phosphatidylinositol 3-kinase (PI3K), p-AKT, AKT and fatty acid synthase (FASN) protein was detected by western blotting. MTT assays showed that lapatinib inhibited the proliferation of U2-OS and MG-63 cells in a dose- and time-dependent manner, and the rate of colony formation of the lapatinib-treated cells was significantly lower when compared to those cells not treated with lapatinib in both cell lines. FCM assay revealed a significantly higher apoptotic rate in the lapatinib-treated OS cells. Wound healing and Transwell invasion assays revealed that the migratory and invasive abilities of OS cells were significantly inhibited by lapatinib (P<0.05). Western blotting showed that lapatinib suppressed the activity of HER2-PI3K/AKT-FASN in U2-OS and MG-63 cells in vitro. These results suggest that lapatinib may alter the malignant phenotype of OS cells via downregulation of the activity of the HER2-PI3K/AKT-FASN signaling pathway in vitro. Thus, lapatinib may be an effective chemotherapeutic agent for the treatment of osteosarcoma.

[152]

Título / Title: Smooth muscle differentiation identifies two classes of poorly differentiated pleomorphic sarcomas with distinct outcome.

Resumen / Summary: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Perot G; Mendiboure J; Brouste V; Velasco V; Terrier P; Bonvalot S; Guillou L; Ranchere-Vince D; Aurias A; Coindre JM; Chibon F

INSTITUCIÓN / INSTITUTION: 1] Institut Bergonie, Department of Pathology, Bordeaux, France [2] INSERM U916, Bordeaux, France.

Resumen / Summary: The clinical relevance of accurately diagnosing pleomorphic sarcomas has been shown, especially in cases of undifferentiated pleomorphic sarcomas with myogenic differentiation, which appear significantly more aggressive. To establish a new smooth muscle differentiation classification and to test its prognostic value, 412 sarcomas with complex genetics were examined by immunohistochemistry using four smooth muscle markers (calponin, h-caldesmon, transgelin and smooth muscle actin). Two tumor categories were first defined: tumors with positivity for all four markers and tumors with no or incomplete phenotypes. Multivariate analysis demonstrated that this classification method exhibited the strongest prognostic value compared with other prognostic factors, including histological classification. Secondly, incomplete or absent smooth muscle phenotype tumor group was then divided into subgroups by summing for each tumor the labeling intensities of all four markers for each tumors. A subgroup of tumors with an incomplete but strong smooth muscle differentiation phenotype presenting an intermediate metastatic risk was thus identified. Collectively, our results show that the smooth muscle differentiation classification method may be a useful diagnostic tool as well as a relevant prognostic tool for undifferentiated pleomorphic sarcomas. Modern Pathology advance online publication, 29 November 2013; doi:10.1038/modpathol.2013.205.
It Takes Two to Tango: Dual Inhibition of PI3K and MAPK in Rhabdomyosarcoma.

Enlace al Resumen / Link to its Summary


Enlace al texto completo (gratuito o de pago) 1158/1078-0432.CCR-13-2177

Authors

Jahangiri A; Weiss WA

The PI3K/AKT/mTOR and RAS/RAF/MAPK pathways play essential roles in rhabdomyosarcoma. Singular targeting of each pathway is ineffective due to extensive cross-talk and compensatory feedback between these two pathways. Dual blockade with inhibitors of PI3K and MAPK in combination synergistically inhibits growth of rhabdomyosarcoma both in vitro and in vivo. Clin Cancer Res; 19(21); 5811-3. ©2013 AACR.

Vanadium and cancer treatment: Antitumoral mechanisms of three oxidovanadium(IV) complexes on a human osteosarcoma cell line.

Enlace al Resumen / Link to its Summary


Enlace al texto completo (gratuito o de pago) 1016/j.jinorgbio.2013.10.009

Authors

Leon IE; Butenko N; Di Virgilio AL; Muglia CI; Baran EJ; Cavaco I; Etcheverry SB

We report herein the antitumor actions of three oxidovanadium(IV) complexes on MG-63 human osteosarcoma cell line. The three complexes: VO(oda), VO(oda)bipy and VO(oda)phen (oda=oxodiacetate), caused a concentration dependent inhibition of cell viability. The antiproliferative action of VO(oda)phen could be observed in the whole range of concentrations (at 2.5μM), while VO(oda)bipy and VO(oda) showed a decrease of cell viability only at higher concentrations (at 50 and 75μM, respectively) (p<0.01). Moreover, VO(oda)phen caused a decrease of lysosomal and mitochondrial activities at 2.5μM, while VO(oda) and VO(oda)bipy affected neutral red uptake and mitochondrial metabolism at 50μM (p<0.01). On the other hand, no DNA damage studied by the Comet assay could be observed in MG-63 cells treated with VO(oda) at 2.5-10μM. Nevertheless, VO(oda)phen and VO(oda)bipy induced DNA damage at 2.5 and 10μM, respectively (p<0.01). The generation of reactive oxygen species increased at 10μM of VO(oda)phen and only at 100μM of VO(oda) and VO(oda)bipy (p<0.01). Besides, VO(oda)phen and VO(oda)bipy triggered apoptosis as determined by externalization of the phosphatidylserine. The determination of DNA cleavage by agarose gel electrophoresis showed that the ability of VO(oda)(bipy) is similar to that of VO(oda),
while VO(oda)(phen) showed the highest nuclease activity in this series. Overall, our results showed a good relationship between the bioactivity of the complexes and their structures since VO(oda)phen presented the most potent antitumor action in human osteosarcoma cells followed by VO(oda)bipy and then by VO(oda) according to the number of intercalating heterocyclic moieties.

[155]
TÍTULO / TITLE: - Clinical outcomes of Kyocera Modular Limb Salvage system after resection of bone sarcoma of the distal part of the femur: the Japanese Musculoskeletal Oncology Group study.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Nakamura T; Matsumine A; Uchida A; Kawai A; Nishida Y; Kunisada T; Araki N; Sugiura H; Tomita M; Yokouchi M; Ueda T; Sudo A
INSTITUCIÓN / INSTITUTION: - Department of Orthopaedic Surgery, Mie University Graduate School of Medicine, Tsu, Japan.
RESUMEN / SUMMARY: - PURPOSE: The Japanese Musculoskeletal Oncology Group have developed an original prosthesis called the Kyocera Modular Limb Salvage system (KMLS system). This prosthesis has a semi-rotating hinge joint and is particularly designed for people with an Asian body type. The metallic parts of the prosthesis are made entirely of titanium alloy. The purpose of this study is to evaluate the clinical outcomes of treatment using this system following tumour resection of primary bone sarcoma of the distal femur. METHODS: Between 2002 and 2010, 82 patients with primary bone sarcomas of the distal femur were treated. Seventeen patients underwent stem cementation, while 65 patients were treated with cementless prostheses. The mean follow-up period after surgery was 61 months. RESULTS: Complications were observed in 28 of the 82 patients. Forty-one complications occurred in these 28 patients. Thirteen prostheses (16 %) required revision surgery due to complications, including five cases of stem breakage, three deep infections, three cases of aseptic loosening, one case of displacement of the shaft cap and one case of breakage of the tibial tray. The five-year overall prosthetic survival rate was 80.0 %. Four of the 82 patients underwent subsequent amputation due to local recurrence. The five-year limb salvage rate was 94.5 %. The mean function score according to the scoring system of the Musculoskeletal Tumour Society was 21.8 points (72.5 %). CONCLUSIONS: Although further follow-up is required to determine the performance, this prosthesis is considered to be satisfactory for reconstruction of the distal femur after resection of bone sarcoma.

[156]
TÍTULO / TITLE: - Preclinical validation of Aurora kinases-targeting drugs in osteosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Nakamura T; Matsumine A; Uchida A; Kawai A; Nishida Y; Kunisada T; Araki N; Sugiura H; Tomita M; Yokouchi M; Ueda T; Sudo A
INSTITUCIÓN / INSTITUTION: - Department of Orthopaedic Surgery, Mie University Graduate School of Medicine, Tsu, Japan.
RESUMEN / SUMMARY: - PURPOSE: The Japanese Musculoskeletal Oncology Group have developed an original prosthesis called the Kyocera Modular Limb Salvage system (KMLS system). This prosthesis has a semi-rotating hinge joint and is particularly designed for people with an Asian body type. The metallic parts of the prosthesis are made entirely of titanium alloy. The purpose of this study is to evaluate the clinical outcomes of treatment using this system following tumour resection of primary bone sarcoma of the distal femur. METHODS: Between 2002 and 2010, 82 patients with primary bone sarcomas of the distal femur were treated. Seventeen patients underwent stem cementation, while 65 patients were treated with cementless prostheses. The mean follow-up period after surgery was 61 months. RESULTS: Complications were observed in 28 of the 82 patients. Forty-one complications occurred in these 28 patients. Thirteen prostheses (16 %) required revision surgery due to complications, including five cases of stem breakage, three deep infections, three cases of aseptic loosening, one case of displacement of the shaft cap and one case of breakage of the tibial tray. The five-year overall prosthetic survival rate was 80.0 %. Four of the 82 patients underwent subsequent amputation due to local recurrence. The five-year limb salvage rate was 94.5 %. The mean function score according to the scoring system of the Musculoskeletal Tumour Society was 21.8 points (72.5 %). CONCLUSIONS: Although further follow-up is required to determine the performance, this prosthesis is considered to be satisfactory for reconstruction of the distal femur after resection of bone sarcoma.
Background: Aurora kinases are key regulators of cell cycle and represent new promising therapeutic targets in several human tumours. Methods: Biological relevance of Aurora kinase-A and -B was assessed on osteosarcoma clinical samples and by silencing these genes with specific siRNA in three human osteosarcoma cell lines. In vitro efficacy of two Aurora kinases-targeting drugs (VX-680 and ZM447439) was evaluated on a panel of four drug-sensitive and six drug-resistant human osteosarcoma cell lines. Results: Human osteosarcoma cell lines proved to be highly sensitive to both drugs. A decreased drug sensitivity was observed in doxorubicin-resistant cell lines, most probably related to ABCB1/MDR1 overexpression. Both drugs variably induced hyperploidy and apoptosis in the majority of cell lines. VX-680 also reduced in vitro cell motility and soft-agar clonning efficiency. Drug association experiments showed that VX-680 positively interacts with all conventional drugs used in osteosarcoma chemotherapy, overcoming the cross-resistance observed in the single-drug treatments. Conclusion: Aurora kinase-A and -B represent new candidate therapeutic targets for osteosarcoma. In vitro analysis of the Aurora kinases inhibitors VX-680 and ZM447439 indicated in VX-680 a new promising drug of potential clinical usefulness in association with conventional osteosarcoma chemotherapeutic agents.

[157] TITULO / TITLE: - Pulmonary embolization as the primary clinical manifestation of giant renal angiomyolipoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1016/j.athoracsur.2013.01.060
AUTORES / AUTHORS: - Yu L; Gu T; Xiu Z
INSTITUCIÓN / INSTITUTION: - Department of Cardiac Surgery, The First Affiliated Hospital of China Medical University, Shenyang, P. R. China.

[158] TITULO / TITLE: - DNA damage and cell cycle arrest induced by protoporphyrin IX in sarcoma 180 cells.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1159/000354479
AUTORES / AUTHORS: - Li Q; Wang X; Zhang K; Li X; Liu Q; Wang P
INSTITUCIÓN / INSTITUTION: - Key Laboratory of Medicinal Resources and Natural Pharmaceutical Chemistry, Ministry of Education, National Engineering Laboratory for
RESUMEN / SUMMARY: - BACKGROUND: Porphyrin derivatives have been widely used in photodynamic therapy as effective sensitizers. Protoporphyrin IX (PpIX), a well-known hematoporphyrin derivative component, shows great potential to enhance light induced tumor cell damage. However, PpIX alone could also exert anti-tumor effects. The mechanisms underlying those direct effects are incompletely understood. This study thus investigated the putative mechanisms underlying the anti-tumor effects of PpIX on sarcoma 180 (S180) cells. METHODS: S180 cells were treated with different concentrations of PpIX. Following the treatment, cell viability was evaluated by the 3-(4, 5-dimethylthiazol-2-yl)-2, 5-diphenyltetrazoliumbromide (MTT) assay; Disruption of mitochondrial membrane potential was measured by flow cytometry; The trans-location of apoptosis inducer factor (AIF) from mitochondria to nucleus was visualized by confocal laser scanning microscopy; DNA damage was detected by single cell gel electrophoresis; Cell cycle distribution was analyzed by DNA content with flow cytometry; Cell cycle associated proteins were detected by western blotting.

RESULTS: PpIX (>\= 1 microg/ml) significantly inhibited proliferation and reduced viability of S180 cells in a dose-dependent manner. PpIX rapidly and significantly triggered mitochondrial membrane depolarization, AIF (apoptosis inducer factor) translocation from mitochondria to nucleus and DNA damage, effects partially relieved by the specific inhibitor of MPTP (mitochondrial permeability transition pore). Furthermore, S phase arrest and upregulation of the related proteins of P53 and P21 were observed following 12 and 24 h PpIX exposure. CONCLUSION: PpIX could inhibit tumor cell proliferation by induction of DNA damage and cell cycle arrest in the S phase.

[159]

TITULO / TITLE: Matrine promotes G0/G1 arrest and down-regulates cyclin D1 expression in human rhabdomyosarcoma cells.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Guo L; Xue TY; Xu W; Gao JZ
INSTITUCIÓN / INSTITUTION: Department of Pediatrics Hospital Affiliated to Xuzhou Medical College Xuzhou, Jiangsu, China - xuety2006@163.com

RESUMEN / SUMMARY: Matrine has a broad-spectrum of anti-cancer effects and is efficient in the inhibition of proliferation of hepatoma cells, leukemia cells and neuroblastoma cell. However, its efficacy and tentative mechanisms in rhabdomyosarcoma have not been addressed before. This study aimed to investigate the effects of Matrine on cell cycle and expression of cyclin D1 in human rhabdomyosarcoma cells (RD cell line). RD cell line was treated with different concentrations (0, 0.5, 1.0, and 1.5 mg/mL) of Matrine, and cell proliferation and cell cycle were evaluated using, respectively, MTT assay and flow cytometry. The effect of Matrine on cyclin D1 mRNA levels was measured by RT-PCR. There was a dose-dependent inhibition of proliferation in the matrine-treated group (inhibition of proliferation rate in control cells 12.70 +/- 0.35%; Matrine-treated cells [0.5, 1.0, and 1.5 mg/mL]: 31.16 +/- 0.11%, 42.96 +/- 0.9%, and 57.26 +/- 0.8%). The G0 / G1 ratio in study groups were, respectively, 58.44 +/- 3.57%, 64.79 +/- 2.03%, 69.97 +/- 2.89%
and 75.03 +/- 1.23%. Cyclin D1 mRNA levels progressively diminished (control group ratio of cyclin D1 / beta-actin: 0.59 +/- 0.06; Matrine: 0.35 +/- 0.05, 0.27 +/- 0.02 and 0.04 +/- 0.03). All aforementioned changes were significant (P<0.05). In conclusion, Matrine markedly suppresses cell proliferation in RD cells by decreasing expression of cyclin D1 mRNA and blocking the cell cycle at the G0 / G1 stage.

[160]
TÍTULO / TITLE: - Use of tissue expander in pelvic Ewing’s sarcoma treated with radiotherapy.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Perez-Munoz I; Grimer RJ; Spooner D; Carter S; Tillman R; Abudu A; Jeys L
INSTITUCIÓN / INSTITUTION: - Hospital Infanta Cristina, Avenida Nueve de Junio, 2, 28981 Parla, Madrid, España. Electronic address: israel.perezmun@salud.madrid.org.
RESUMEN / SUMMARY: - INTRODUCTION: The local treatment option for pelvic Ewing sarcoma (ES) remains uncertain and challenging as surgery is often disabling while radiotherapy alone has a higher risk of local recurrence but not necessarily a worse survival. The aim is to analyse the outcome of patients with pelvic ES after radiotherapy as the primary local treatment in combination with a temporary intrapelvic surgically placed tissue expander (TE) to reduce bowel complications. MATERIALS AND METHODS: 20 patients were retrospectively analysed. All patients had neoadjuvant and adjuvant chemotherapy. We identified survival, time to develop local recurrence and metastasis, dose of radiotherapy administered, local complications related to the use of the tissue expander and bowel effects of radiotherapy. RESULTS: The median follow-up was 41 months. 14 patients were stage IIb and six stage III. There were no problems after insertion of the TE and only one patient who developed mild diarrhoea. Local recurrence occurred in six patients. At the last follow-up 12 patients have died from sarcoma, five are disease free and three have had recurrent disease. CONCLUSIONS: In this paper we reviewed pelvic Ewing sarcoma with all the special considerations that this entails. We think that tissue expander can be safely used when radiotherapy is chosen to treat pelvic ES. It does appear to prevent bowel problems and is a low morbidity procedure. New treatment approaches should be considered to give a chance of cure to those patients with “bad prognostic” pelvic ES.

[161]
TÍTULO / TITLE: - Comparative expression of thioredoxin-1 in uterine leiomyomas and myometrium.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Hou P; Zhao L; Li Y; Luo F; Wang S; Song J; Bai J
INSTITUCIÓN / INSTITUTION: - Faculty of Environmental Science and Engineering, Kunming University of Science and Technology, Kunming 650500, China.
RESUMEN / SUMMARY: Uterine leiomyomas are benign tumors that develop from smooth muscle cells (SMCs). The reactive oxygen species (ROS) have been shown to be involved in the signaling pathways that stimulate proliferation of a variety of cell types. Thioredoxin-1 (TRX-1) is a redox-regulating protein, which is overexpressed in various tumors. In the present study, we investigated the expressions of TRX-1 and its related molecules in uterine leiomyomas and matched adjacent myometrium. Our results showed the expression of TRX-1 was increased in leiomyomas compared with the matched adjacent myometrium by quantitative RT-PCR and western blotting. FOXO3A expression was increased in leiomyomas compared with myometrium by western blotting. The mRNA levels of hypoxia-inducible factor-1alpha, cyclooxygenase-2 and cyclin D1 were increased in leiomyomas compared with the adjacent myometrium. The mRNA level of (thioredoxin-1-binding protein) TBP-2 in leiomyomas was not altered when compared with the matched adjacent myometrium. These results suggest that TRX-1 and some of its related molecules are associated with the pathogenesis of uterine leiomyomas. The identification of TRX-1 signaling pathways leading to cell proliferation points to another potential therapeutic target for treatment and/or prevention of uterine leiomyomas.

TÍTULO / TITLE: Involvement and targeted intervention of dysregulated hedgehog signaling in osteosarcoma.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Lo WW; Wunder JS; Dickson BC; Campbell V; McGovern K; Alman BA; Andrulis IL

INSTITUCIÓN / INSTITUTION: Department of Molecular Genetics, University of Toronto, Ontario, Canada; Lunenfeld-Tanenbaum Research Institute, Mount Sinai Hospital, Toronto, Ontario, Canada.

RESUMEN / SUMMARY: BACKGROUND: During development, the Hedgehog pathway plays important roles regulating the proliferation and differentiation of chondrocytes, providing a template for growing bone. In this study, the authors investigated the components of dysregulated Hedgehog signaling as potential therapeutic targets for osteosarcoma. METHODS: Small-molecule agonists and antagonists that modulate the Hedgehog pathway at different levels were used to investigate the mechanisms of dysregulation and the efficacy of Hedgehog blockade in osteosarcoma cell lines. The inhibitory effect of a small-molecule Smoothened (SMO) antagonist, IPI-926 (saridegib), also was examined in patient-derived xenograft models. RESULTS: An inverse correlation was identified in osteosarcoma cell lines between endogenous glioma-associated oncogene 2 (GLI2) levels and Hedgehog pathway induction levels. Cells with high levels of GLI2 were sensitive to GLI inhibition, but not SMO inhibition, suggesting that GLI2 overexpression may be a mechanism of ligand-independent activation. In contrast, cells that expressed high levels of the Hedgehog ligand gene Indian hedgehog (IHH) and the target genes patched 1 (PTCH1) and GLI1 were sensitive to modulation of both SMO and GLI, suggesting ligand-dependent activation. In 2 xenograft models, active autocrine and paracrine, ligand-dependent Hedgehog signaling was identified. IPI-926 inhibited the Hedgehog signaling.
interactions between the tumor and the stroma and demonstrated antitumor efficacy in 1 of 2 ligand-dependent models. CONCLUSIONS: The current results indicate that both ligand-dependent and ligand-independent Hedgehog dysregulation may be involved in osteosarcoma. It is the first report to demonstrate Hedgehog signaling crosstalk between the tumor and the stroma in osteosarcoma. The inhibitory effect of IPI-926 warrants additional research and raises the possibility of using Hedgehog pathway inhibitors as targeted therapeutics to improve treatment for osteosarcoma. Cancer 2013. Esta es una cita bibliográfica que va por delante de la publicación en papel. La fecha indicada en la cita provista, NO corresponde con la fecha o la cita bibliográfica de la publicación en papel. La cita bibliográfica definitiva (con el volumen y su paginación) saldrá en 1 ó 2 meses a partir de la fecha de la emisión electrónica-online. *** This is a bibliographic record ahead of the paper publication. The given date in the bibliographic record does not correspond to the date or the bibliographic citation on the paper publication. The publisher will provide the final bibliographic citation (with the volume, and pagination) within 1 or 2 months from the date the record was published online. © 2013 American Cancer Society.

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[163]

**TÍTULO / TITLE:** - Four cases of solitary fibrous tumour of the eye and orbit: one with sarcomatous transformation after radiotherapy and one in a 5-year-old child's eyelid.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Blandamura S; Alaggio R; Bettini G; Guzzardo V; Valentini E; Bedogni A

**INSTITUCIÓN / INSTITUTION:** - Pathological Anatomy, DIMED-Department of Medicine, University of Padova, Padova, Italy.

**RESUMEN / SUMMARY:** - BACKGROUND: Solitary fibrous tumour (SFT) is quite a rare neoplasm involving the eye and the orbit. It is described as showing benign behaviour in adults, but malignant cases are exceptionally reported in this location. This report describes four new cases of SFT/giant cell angiofibroma (GCA) of the eyelid and orbit, one in a 5-year-old child, and one with sarcomatous dedifferentiated transformation occurring 9 years after radiotherapy. METHODS: Four cases of ocular SFT/GCA were retrieved from the database of the Pathological Anatomy Unit, University of Padova; immunohistochemistry and RT-PCR were used to identify COL1A1-PDGFB fusion gene transcripts in all cases. RESULTS: In case 1, late relapse 9 years later was characterised by abrupt transition into a high-grade component, associated with a non-distinctive high-grade sarcomatous area. The latter component was CD34, CD99 and Bcl2 negative and smooth muscle actin positive. Molecular characterisation showed the absence of COL1A1-PDGFB fusion transcripts in cases 1, 3 and 4, excluded diagnosis of giant cell fibroblastoma in all cases. Analysis could not be performed in case 2. CONCLUSIONS: An eyelid SFT/GCA in a 5-year-old child is the youngest case reported in the literature, indicating that the tumour is not exclusive to adults. The case with sarcomatous transformation, with dedifferentiated features occurring 9 years after radiotherapy, raises some questions about the choice of treatment for ocular SFT, in which excision is sometimes difficult without devastating surgery.
RESUMEN / SUMMARY: BACKGROUND AND OBJECTIVES: Prior studies have demonstrated postoperative infection may confer a survival benefit after osteosarcoma resection. Our aim was to determine whether infection after soft tissue sarcoma resection has similar effects on metastasis, recurrence and survival. METHODS: A retrospective review was conducted; 396 patients treated surgically for a soft tissue sarcoma between 2000 and 2008 were identified. Relevant oncologic data were collected. Fifty-six patients with a postoperative infection were compared with 340 patients without infection. Hazard ratios and overall cumulative risk were evaluated. RESULTS: There was no difference in survival, local recurrence or metastasis between patients with or without a postoperative infection. Patients were evenly matched for age at diagnosis, gender, smoking status, and diabetes status. Tumor characteristics did not differ between groups in tumor size, location, depth, grade, margin status, stage, and histologic subtype. There was no difference in utilization of chemotherapy or radiation therapy between groups. From our competing risk model, only positive margin status significantly impacted the risk of local recurrence. An increase in tumor size corresponded to an increased risk of metastasis and death. CONCLUSIONS: Postoperative infection neither conferred a protective effect, nor increased the risk of adverse oncologic outcomes after soft tissue sarcoma resection. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.
in tumor spindle cells and in intratumor stromal vascular endothelial cells was analyzed with regard to clinicomorphological features. Tumor AKT related to lack of marked extravasated erythrocytes, tumor PTEN to presence of intratumor hemosiderin (p = 0.04 for both comparisons). Presence of both extravasated erythrocytes and hemosiderin related directly to endothelial stromal vascular nuclear PTEN and to low endothelial mTOR (p = 0.4 and 0.03, respectively). High tumor 4EBP1 related to a high slit-type abnormal vascular component (p = 0.04). The results of our study suggest pro-permeability or pro-angiogenic roles for 4EBP1 and PTEN and, opposite roles for AKT and mTOR in KS. Our hypotheses warrant further studies to obtain more generally applicable results.

[166]
**TITULO / TITLE:** - Marginal miss or radioresistance? The pattern of local recurrence after operation and 3D planned radiation treatment in soft tissue sarcoma of the extremities and the limb girdles; an analysis based on image fusion.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**REVISTA / JOURNAL:** - Acta Oncol. 2013 Nov 8.

**AUTORES / AUTHORS:** - Sampo MM; Tuomikoski L; Tarkkanen M; Jaaskelainen AS; Tukiainen EJ; Beule A; Tenhunen M; Bohling TO; Blomqvist CP
**INSTITUCIÓN / INSTITUTION:** - Department of Oncology, Helsinki University Central Hospital (HUCH), Finland.
**RESUMEN / SUMMARY:** - Background. Most local recurrences have developed in the clinical target volume in previously published series after combined modality treatment for soft tissue sarcoma. However, marginal misses were seen in almost 20% of the patients. The aim of the present study was to determine the location of the recurrence and the total dose at the centre point of the local recurrence for future radiation therapy planning. Material and methods. We included only patients with images in digital form, during 1999-2006 (n = 17), treated for soft tissue sarcoma with combined surgical therapy and radiotherapy at Helsinki University Central Hospital. Image fusion was used to determine the location of the recurrence in relation to radiation therapy target. Results. In the present study utilising digital image fusion, in patients with 3D CT-based radiation treatment planning the risk of marginal miss was low as only one patient of 17 relapsed outside the target. Estimated mean radiation dose at the site of local recurrence was 49.1 Gy in patients with positive margins and 48.1 Gy in patients with negative margins. Conclusion. The risk of marginal miss in soft tissue sarcoma is low after modern 3D planned radiation treatment combined with surgery. More generous use of boost might improve in-target local control.

[167]
**TITULO / TITLE:** - Long-term survival after excision of a giant esophageal gastrointestinal stromal tumor with imatinib mesylate resistance: report of a case.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

- Enlace al texto completo (gratuito o de pago) 1007/s00595-013-0770-7
A 69-year-old woman underwent 4 months of imatinib mesylate chemotherapy for a diagnosed gastrointestinal stromal tumor of the esophagus. This treatment was suspended because of its side effects and because radiological examinations showed that the tumor had not changed or had even increased slightly in size. Thus, we performed esophagectomy via left thoracotomy and removed a tumor that measured 18 x 17 x 10 cm. Immunohistochemical examination revealed positive reactions for c-kit and CD34, suggestive of a high-risk malignancy. The patient was discharged from hospital on postoperative day 30, and has remained well with no sign of tumor recurrence for more than 5 years, without adjuvant chemotherapy.
single cell, and for the first time demonstrates that these clonal cells differentiate into fibroblast and SMC subpopulation as the fibroid grows.

[169]
**TÍTULO / TITLE:** - Gastrointestinal stromal tumors: molecular markers and genetic subtypes.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Barnett CM; Corless CL; Heinrich MC

**INSTITUCIÓN / INSTITUTION:** - Hematology and Medical Oncology, Division of Hematology/Oncology, Portland VA Medical Center, OHSU Knight Cancer Institute, Oregon Health & Science University, Mail Code L586, 3181 Southwest Sam Jackson Park Road, Portland, OR 97239, USA.

**RESUMEN / SUMMARY:** - Mutation-activated signaling from the KIT and PDGFRA kinases has been successfully targeted in gastrointestinal stromal tumors (GISTs), with subtle differences between the mutations serving to refine prognosis and more precisely tailor therapy. There is a growing understanding of the molecular drivers of GISTs lacking mutations in KIT or PDGFRA, so called wild-type GISTs, further aiding in management decisions. This article provides an overview of all the known molecular subtypes of GIST and provides information about clinical correlates, treatment, and prognosis depending on the subtype.

[170]
**TÍTULO / TITLE:** - Update in treatment and targets in Ewing sarcoma.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Cote GM; Choy E

**INSTITUCIÓN / INSTITUTION:** - Division of Hematology Oncology, Yawkey Center for Outpatient Care, Massachusetts General Hospital, Harvard Medical School, 32 Fruit Street, Boston, MA 02114, USA.

**RESUMEN / SUMMARY:** - The improvement in outcome for patients with localized and metastatic Ewing sarcoma since the development of cytotoxic chemotherapy remains one of the most profound advances in oncology and one of the proudest achievements of sarcoma researchers. Identification of molecular targets for new treatments has become an intense area within Ewing sarcoma research. The development of improved preclinical Ewing sarcoma models and advanced molecular techniques will build on knowledge of EWS/FLI1 function, EWS/FLI1 transcription targets, and the other critical driver events in these tumors.

[171]
**TÍTULO / TITLE:** - Treatment of localized sarcomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1016/j.hoc.2013.07.006
AUTORES / AUTHORS: - Gronchi A; Raut CP
INSTITUCIÓN / INSTITUTION: - Department of Surgery, Sarcoma Service, Fondazione IRCCS Istituto Nazionale dei Tumori, Via Venezian 1, Milan 20133, Italy.
RESUMEN / SUMMARY: - Surgery remains the only potentially curative therapy in the management of localized adult soft tissue sarcomas and gastrointestinal stromal tumors. There are over 50 different unique histologic types of soft tissue sarcomas, with different patterns of recurrence and prognosis. Surgical principles and sensitivity to locoregional and systemic treatments vary considerably based on the histologic type and anatomic location, as discussed in detail in this review.

[172]
TÍTULO / TITLE: - Update on targets and novel treatment options for high-grade osteosarcoma and chondrosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1016/j.hoc.2013.07.012
AUTORES / AUTHORS: - van Oosterwijk JG; Anninga JK; Gelderblom H; Cleton-Jansen AM; Bovee JV
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Leiden University Medical Center, PO Box 9600, L1-Q, Leiden RC 2300, The Netherlands.
RESUMEN / SUMMARY: - Osteosarcoma and chondrosarcoma are the 2 most common malignant bone tumors. This review discusses the clinicopathologic features, recent preclinical developments, and targets currently being or to be validated in the clinic.

[173]
TÍTULO / TITLE: - Cervical neoplasia-related factors and decreased prevalence of uterine fibroids among a cohort of African American women.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1016/j.fertnstert.2013.09.021
AUTORES / AUTHORS: - Moore KR; Smith JS; Laughlin-Tommaso SK; Baird DD
INSTITUCIÓN / INSTITUTION: - Department of Epidemiology, Gillings School of Global Public Health, University of North Carolina at Chapel Hill, Chapel Hill, North Carolina; Epidemiology Branch, National Institute of Environmental Health Sciences, National Institutes of Health, Research Triangle Park, North Carolina.
RESUMEN / SUMMARY: - OBJETIVO: To investigate whether the previously reported inverse association between cervical neoplasia and uterine fibroids is corroborated. DESIGN: Cross-sectional analysis of enrollment data from an ongoing prospective study of fibroid development. SETTING: Not applicable. PATIENT(S): Self-reported data on abnormal Pap smear, colposcopy, and cervical treatment were obtained from
1,008 African American women ages 23-34 with no previous fibroid diagnosis and no reported history of human papillomavirus vaccination. Presence of fibroids was assessed at a standardized ultrasound examination. INTERVENTION(S): None. MAIN OUTCOME MEASURE(S): The association between the three cervical neoplasia-related variables and the presence of fibroids was evaluated with logistic regression to estimate age-adjusted and multivariable-adjusted odds ratios (aORs). RESULT(S): Of the analysis sample, 46%, 29%, and 14% reported a prior abnormal Pap smear, colposcopy, and cervical treatment, respectively. Twenty-five percent had fibroids at ultrasound. Those reporting cervical treatment had a 39% (aOR, 0.61; 95% confidence interval [CI] [0.38-0.96]) reduction in fibroid risk. Weak nonsignificant associations were found for abnormal Pap smear and colposcopy. CONCLUSION(S): Although a protective-type association of cervical neoplasia with uterine fibroids seems counterintuitive, a causal pathway is possible, and the findings are consistent with two prior studies. Further investigation is needed on the relationship between fibroids and cervical neoplasia and human papillomavirus-related mechanisms.

[174]

**TÍTULO / TITLE:** - Intravascular adenomyomatosis: expanding the morphologic spectrum of intravascular leiomyomatosis.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Hirschowitz L; Mayall FG; Ganesan R; McCluggage WG

**INSTITUCIÓN / INSTITUTION:** - *Department of Cellular Pathology, Birmingham Women’s NHS Foundation Trust, Birmingham daggerDepartment of Histopathology, Musgrove Park Hospital, Taunton double daggerDepartment of Pathology, Royal Group of Hospitals Trust, Belfast, Northern Ireland, UK.

**RESUMEN / SUMMARY:** - Intravascular leiomyomatosis (IVL) is characterized by the presence of smooth muscle in venous and lymphatic spaces within the myometrium. Although the intravascular component usually consists solely of typical smooth muscle or variants of smooth muscle differentiation, we report 5 cases in which the intravascular component also included endometrioid glandular and stromal elements. We propose the term “intravenous adenomyomatosis” to describe this unusual variant of IVL. The mean age of the patients in this series was 50.2 years, slightly older than that of patients with conventional IVL. In addition to intravenous adenomyomatosis, both adenomyosis and leiomyomas were identified in all of our cases, supporting the hypothesis that the intravascular smooth muscle component in IVL is derived from associated myometrial pathology rather than from vessel walls. In our series, intravenous adenomyomatosis had a similar benign clinical behavior to most cases of IVL with no metastatic or recurrent disease identified at follow-up in 4 cases for which follow-up information was available. The main differential diagnoses are adenomyosis with vascular involvement, low-grade endometrial stromal sarcoma (ESS), including ESS with smooth muscle and glandular differentiation, and adenosarcoma with lymphovascular invasion. The possibility of intravenous adenomyomatosis should be
borne in mind when considering these diagnoses, particularly ESS and adenosarcoma, which have different implications for patient management and prognosis.

[175]
TÍTULO / TITLE: - Kaposis sarcoma in the early post-transplant period in a kidney transplant recipient.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Ercan Z; Demir ME; Merhametsiz O; Yayar O; Ulas T; Ayli MD

[176]
TÍTULO / TITLE: - Musculoskeletal sarcomas in the forearm and hand: standard treatment and microsurgical reconstruction for limb salvage.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Muramatsu K; Ihara K; Yoshida K; Tominaga Y; Hashimoto T; Taguchi T
INSTITUCIÓN / INSTITUTION: - Department of Orthopedic Surgery, Yamaguchi University School of Medicine, 1-1-1 Minami-Kogushi, Ube, Yamaguchi 755-8505, Japan. muramatu@yamaguchi-u.ac.jp.
RESUMEN / SUMMARY: - Sarcomas in the forearm and hand are very rare, accounting for less than 1% of all upper-limb tumors and clinical outcomes after surgery and adjuvant therapies are uncertain. The forearm and hand present specific challenges due to their unique anatomical structures. There is little soft tissue and each compartment is narrow, such important structures exist in close proximity. Anatomic constraints make it difficult to achieve wide surgical margins. Although sarcomas often metastasize to the lung, the overall survival rate is excellent. Wide marginal resection during initial surgery is the most predictive factor for tumor control. The role of reconstructive surgery following wide excision for sarcoma of the forearm and hand is even more important than elsewhere in the body because excision is likely to cause bone, tendon and nerve defects, leading to severe functional deficits. Multiple options exist for bony and soft tissue reconstruction of the upper limb, with the choice dependent upon tumor type, wound characteristics, surgeon preference and the patients’ functional requirements. Success should be measured not just by stable wound coverage but also by preservation of patient’s health, limb cosmesis, sensation and function. Careful preoperative planning with consideration of all the possible resected structures should improve patient outcomes.

[177]
TÍTULO / TITLE: - Targeted therapies in rare sarcomas: IMT, ASPS, SFT, PEComa, and CCS.
Resumen: Este artículo destaca los datos actualmente disponibles sobre la actividad de tratamiento médico en un subgrupo de entidades raras dentro de los sarcomas de tejido blandos, incluyendo el tumor miofibroblástico inflamatorio, sarcoma de partes blandas alveolar, tumor fibroso solitario, tumor perivascular de células epitelioides malignas (PEComa) y sarcoma de células selectras clara.

Título: Therapies for soft-tissue sarcomas.

Resumen: Los sarcomas son un grupo heterogéneo de tumores que se originan en tejidos de origen mesenquimatoso. Las opciones actuales para pacientes con enfermedad avanzada son limitadas, y solo 2 drogas han sido aprobadas para estos tipos de enfermedades en el último década. Aunque varias drogas se están desarrollando actualmente para sarcoma de tejido blando como un todo, el mejor entendimiento de la biología del sarcoma ha llevado a la emergencia de terapias específicas de subtipo. Este artículo revisa los datos clínicos recientes sobre terapias emergentes para sarcoma de tejido blando.


Resumen: El sarcoma asociado a radiación (RT-AS) es una enfermedad infrecuente con una incidencia inferior a 1% de todos los sarcomas blandos. El pronóstico global es bastante desalentador con altas tasas de recurrencias y...
poor overall survival. There is an obvious paucity of data regarding clinical outcomes of patients with breast RT-AS.

Methods: We identified all patients with RT-AS treated at the Memorial Sloan-Kettering Cancer Center between 1982-2011 and collected their correlative clinical information. Results: We identified 79 women with RT-AS with a median age of 68 (range 36-87). The median interval between radiation and development of RT-AS was 7 years (range 3-19). The median time to local and distant recurrence was 1.29 years (95% CI 0.72-NA) and 2.48 years (95% CI 1.29-NA), respectively. The median disease-specific survival was 2.97 years (95% CI 2.21-NA). Independent predictors of worse disease-specific survival included age \( \geq 68 \) years (HR 3.11, 95% CI 1.20-8.08, \( P=0.020 \)) and deep tumors (HR 3.23, 95% CI 1.02-10.21, \( P=0.046 \)).

Conclusion: RT-AS has high local/distant recurrence rates, limited duration on standard chemotherapy and poor disease-specific survival.

[180]
**TÍTULO / TITLE:** Invasive growth patterns of juvenile nasopharyngeal angiofibroma: radiological imaging and clinical implications.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - Acta Radiol. 2013 Oct 16.

**AUTORES / AUTHORS:** Szymanska A; Szymanski M; Czekajska-Chehab E; Szczeroło-Trojanowska M

**INSTITUCIÓN / INSTITUTION:** Department of Interventional Radiology and Neuroradiology, Medical University of Lublin, Lublin, Poland.

**RESUMEN / SUMMARY:** Juvenile nasopharyngeal angiofibroma is a benign lesion with locally aggressive nature. Knowledge of its typical growth patterns is crucial for precise preoperative staging and adequate preoperative patient counseling. This pictorial essay focuses on characteristic radiological features and paths of invasive growth of this rare tumor. Also, the impact of accurate preoperative evaluation of tumor extensions on surgical planning and results of treatment are discussed.

[181]
**TÍTULO / TITLE:** Unroofing and grasp-and-snare techniques in the management of a large, duodenal lipoma by duodenoscope combined with a double-channel endoscope.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Yong P; Bing-Yin Z; Tao W; Li-Jun T; Fu-Zhou T

**INSTITUCIÓN / INSTITUTION:** Center of General Surgery, General Hospital of Chengdu Military Region of PLA, Chengdu City, Sichuan Province, China.

[182]
**TÍTULO / TITLE:** Luminal lipoma: the “pot-of-gold” sign.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
[183] **TÍTULO / TITLE:** MR imaging in a case of osteosarcoma in the temporomandibular joint.

**REVISTA / JOURNAL:** Dentomaxillofac Radiol. 2013 Nov 18.

**AUTORES / AUTHORS:** Uchiyama Y; Matsumoto K; Murakami S; Kanesaki T; Matsumoto A; Kishino M; Furukawa S

**INSTITUCIÓN / INSTITUTION:** Osaka University Graduate School of Dentistry Oral and Maxillofacial Radiology 1-8 Yamadaoka JAPAN Suita Osaka 565-0871 +81-6-6879-2970 +81-6-6879-2967 Osaka University Graduate School of Dentistry.

**RESUMEN / SUMMARY:** Osteosarcoma of temporomandibular joint (TMJ) is rare. We report a case of osteosarcoma in TMJ in a 62-year-old female preoperatively diagnosed to have a benign tumor. and discuss the usefulness and limits of MR imaging using a TMJ coil as a diagnosis.

[184] **TÍTULO / TITLE:** The role of angiogenic factors in fibroid pathogenesis: potential implications for future therapy.

**REVISTA / JOURNAL:** Hum Reprod Update. 2013 Sep 29.

**AUTORES / AUTHORS:** Tal R; Segars JH

**INSTITUCIÓN / INSTITUTION:** Department of Obstetrics and Gynecology, Maimonides Medical Center, Brooklyn, NY 11219, USA.

**RESUMEN / SUMMARY:** Background It is well established that tumors are dependent on angiogenesis for their growth and survival. Although uterine fibroids are known to be benign tumors with reduced vascularization, recent work demonstrates that the vasculature of fibroids is grossly and microscopically abnormal. Accumulating evidence suggests that angiogenic growth factor dysregulation may be implicated in these vascular and other features of fibroid pathophysiology. Methods Literature searches were performed in PubMed and Google Scholar for articles with content related to angiogenic growth factors and myometrium/leiomyoma. The findings are hereby reviewed and discussed. Results Multiple growth factors involved in angiogenesis are differentially expressed in leiomyoma compared with myometrium. These include epidermal growth factor (EGF), heparin-binding-EGF, vascular endothelial growth factor, basic fibroblast growth factor, platelet-derived growth factor, transforming growth factor-beta and adrenomedullin. An important paradox is that although leiomyoma tissues are hypoxic, leiomyoma feature down-regulation of key molecular regulators of
the hypoxia response. Furthermore, the hypoxic milieu of leiomyoma may contribute to fibroid development and growth. Notably, common treatments for fibroids such as GnRH agonists and uterine artery embolization (UAE) are shown to work at least partly via anti-angiogenic mechanisms. Conclusions: Angiogenic growth factors play an important role in mechanisms of fibroid pathophysiology, including abnormal vasculature and fibroid growth and survival. Moreover, the fibroid’s abnormal vasculature together with its aberrant hypoxic and angiogenic response may make it especially vulnerable to disruption of its vascular supply, a feature which could be exploited for treatment. Further experimental studies are required in order to gain a better understanding of the growth factors that are involved in normal and pathological myometrial angiogenesis, and to assess the potential of anti-angiogenic treatment strategies for uterine fibroids.

[185]
TÍTULO / TITLE: - Effect of neoadjuvant treatment in the management of osteosarcomas of the head and neck.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Mucke T; Mitchell DA; Tannapfel A; Wolff KD; Loeffelbein DJ; Kanatas A
INSTITUCIÓN / INSTITUTION: - Department of Oral and Maxillofacial Surgery, Technische Universität München, Klinikum Rechts der Isar, Ismaninger Str. 22, 81675, Munich, Germany, th.mucke@gmx.de.
RESUMEN / SUMMARY: - PURPOSE: Osteosarcomas of the craniomaxillofacial region in adults are rare malignant tumors with many sites of origin. The purpose of this study was to analyze the outcome of adult patients suffering from osteosarcomas and investigate whether neoadjuvant chemotherapy would be beneficial to overall outcome. PATIENTS AND METHODS: The medical records of 36 patients treated during 2002-2012 were reviewed. All patients suffered from primary osteosarcomas of the craniomaxillofacial region. RESULTS: The mean survival of patients was 64.49 +/- 23.52 months. The 2- and 5-year overall survival rates in the neoadjuvant treatment group were 100 and 66.7 %; in the surgery only group, the overall survival rates were 66.7 and 41.7 %, respectively. The neoadjuvant treatment (p = 0.017), tumor size (p = 0.004), tumor location (p = 0.02), and age (p < 0.0001) were significant parameters influencing survival, whereas other tumor-related or demographic factors had no significant influence on survival. CONCLUSIONS: Early identification of osteosarcoma of the craniomaxillofacial region and combined treatment by neoadjuvant chemotherapy with radical surgery are the most important strategies in dealing with these sarcomas. If possible, this treatment option should be followed unless contraindicated by other factors.

[186]
TÍTULO / TITLE: - Molecular studies and therapeutic targeting of Kaposi’s sarcoma herpesvirus (KSHV/HHV-8) oncogenesis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Kaposi’s sarcoma herpesvirus or human herpesvirus-8 (KSHV/HHV-8) is the etiological agent of Kaposi’s sarcoma (KS), an AIDS-defining angioproliferative neoplasm that continues to be a major global health problem and, of primary effusion lymphoma (PEL), a rare incurable B-cell lymphoma. This review describes the research from our laboratory and its collaborators to uncover molecular mechanisms of viral oncogenesis in order to develop new pathogenesis-based therapies to the KSHV-induced AIDS malignancies KS and PEL. They include the discovery of the viral angiogenic oncogene G protein-coupled receptor (vGPCR), the development of mouse models of KSHV and oxidative stress-induced KS, the identification of the role of Rac1-induced ROS in viral oncogenesis of KS and the development of novel therapeutic approaches able to target both latent and lytic oncogenic KSHV infection.

Technique to Displace Bowel Loops in MRI-Guided High-Intensity Focused Ultrasound Ablation of Fibroids in the Anteverted or Anteflexed Uterus.

OBJECTIVE. In MRI-guided high-intensity focused ultrasound ablation of uterine fibroids, bowel interposition in the sonication path is often problematic. The purpose of this article is to discuss a bowel-manipulation technique to displace the bowel loop, which consisted of sequential applications of urinary bladder filling, rectal filling, and urinary bladder emptying. CONCLUSION. This technique contributed to a decreased screening failure rate and succeeded in consistently displacing the bowel loop, thus allowing safe treatment of fibroids in the anteverted or anteflexed uterus.

Cytoreductive surgery and hyperthermic intraperitoneal chemotherapy in the management of recurrent high-grade uterine sarcoma with peritoneal dissemination.
OBJETIVO: Peritoneal sarcomatosis from primary uterine sarcoma (US) is a rare condition. Conventional therapeutic modalities have failed to improve survival and outcomes among patients with high-grade US with extrapelvic spread. Cytoreductive surgery (CRS) with hyperthermic intraperitoneal chemotherapy (HIPEC) has shown improved outcomes for peritoneal carcinomatosis from other epithelial primaries with similar clinical presentation. We report our experience applying this treatment in 3 patients with recurrent high-grade US with peritoneal dissemination.

STUDY DESIGN: This retrospective review of a prospective database of 378 patients with peritoneal dissemination of cancer treated with CRS/HIPEC identified 3 patients with recurrent high-grade US. Follow-up for disease progression was carried out by physical examination and computed tomography scan of the chest, abdomen, and pelvis. RESULTS: Two leiomyosarcomas and 1 adenosarcoma with sarcomatous overgrowth were identified. Two of the 3 had failed standard treatment with surgery and systemic chemotherapy before CRS/HIPEC was performed. Follow-up ranged from 34 to 140 months. All 3 patients are alive, 2 with no evidence of disease (NED), and 1 alive with disease. Adramycin/cisplatin was used for HIPEC in 1 case (140 months with NED), whereas melphalan was used in the other 2 cases (53 months alive with disease, 34 months with NED). Two patients underwent 1 CRS/HIPEC, whereas 1 required 3 CRS/HIPEC due to disease recurrence. CONCLUSION: CRS/HIPEC shows promise as a treatment modality for the management of selected patients with recurrent high-grade US with peritoneal dissemination. Further studies are warranted.

[189]

TÍTULO: Spontaneous expulsion of a large left-colon lipoma.
RESUMEN: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Chahri N; Querol V; Ballesta E; Marti M; Garrigo J

[190]

TÍTULO: Are meningeal hemangiopericytoma and mesenchymal chondrosarcoma the same?: a study of HEY1-NCOA2 fusion.
RESUMEN: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Fritchie KJ; Jin L; Ruano A; Oliveira AM; Rubin BP
INSTITUCIÓN / INSTITUTION: Mayo Clinic, Dept of Anatomic Pathology, Hilton 11, 200 First St, SW, Rochester, MN 55905; fritchie.karen@mayo.edu.
RESUMEN / SUMMARY: OBJECTIVES: Meningeal hemangiopericytoma (HPC) and mesenchymal chondrosarcoma are aggressive neoplasms with a propensity to involve
the meninges and dura. In addition to similar clinical presentations, both meningeal HPC and mesenchymal chondrosarcoma share overlapping morphologic features, including ovoid cells, variable collagen deposition, and a branching vascular pattern. Recently, a novel HEY1-NCOA2 fusion was reported as a recurrent event in mesenchymal chondrosarcomas. METHODS: Thirteen mesenchymal chondrosarcomas and 18 meningeal HPCs were identified from surgical pathology archives, and the tumors were evaluated for HEY1-NCOA2 fusion with reverse transcriptase-polymerase chain reaction (RT-PCR). RESULTS: HEY1-NCOA2 fusion transcript was detected in all six cases of mesenchymal chondrosarcoma but in none of the meningeal HPC cases (0/11) that were evaluable with RT-PCR. CONCLUSIONS: These results show that (1) meningeal HPC and mesenchymal chondrosarcoma are distinct at the molecular level, and (2) the identification of HEY1-NCOA2 can be used as an auxiliary diagnostic tool to differentiate these entities.

[191]
TITULO / TITLE: - The protective effects of Achyranthes bidentata root extract on the antimycin A induced damage of osteoblastic MC3T3-E1 cells.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Suh KS; Lee YS; Choi EM
INSTITUCIÓN / INSTITUTION: - Research Institute of Endocrinology, Kyung Hee University Hospital, 1, Hoegi-dong, Dongdaemun-gu, Seoul, 130-702, Republic of Korea.
RESUMEN / SUMMARY: - Achyranthes bidentata (A. bidentata) Blume is a medicinal herb with the property of strengthening bones and muscles and ensuring proper downward flow of blood in terms of the therapeutic theory of traditional medicine. In the present study, the effect of A. bidentata root extract (AE) on osteoblast function was investigated in osteoblastic MC3T3-E1 cells. AE caused a significant elevation of alkaline phosphatase activity, collagen synthesis, osteocalcin production, and mineralization in the cells (P < 0.05). AE also decreased the production of TNF-alpha, IL-6, and RANKL induced by antimycin A, mitochondrial electron transport inhibitor. Exposure of MC3T3-E1 cells to antimycin A caused significant reduction of cell viability and mineralization. However, pretreatment with AE prior to antimycin A exposure significantly reduced antimycin A-induced cell damage by preventing mitochondrial membrane potential dissipation, ATP loss, ROS release, and nitrotyrosine increase, suggesting that AE may be useful for protecting mitochondria against a burst of oxidative stress. Moreover, AE increased the phosphorylation of cAMP-response element-binding protein inhibited by antimycin A. Our study demonstrates that A. bidentata could significantly prevent osteoblast damage in aged patients.

[192]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
**OBJECTIVE:** To describe the clinical and imaging characteristics of a new lymphatic disorder with a unique histological pattern and poor prognosis.

**STUDY DESIGN:** An observational, retrospective study identified and characterized 20 patients with distinct lymphatic histopathology referred to the Vascular Anomalies Center at Boston Children’s Hospital between 1995 and 2011.

**RESULTS:**

- The median age at onset was 6.5 years (range, birth to 44 years).
- Clinical and radiologic findings suggested a generalized process. The most common presentations were respiratory symptoms (50%), hemostatic abnormalities (50%), and an enlarging, palpable mass (35%).
- All patients had mediastinal involvement; 19 patients developed pericardial (70%) and/or pleural effusions (85%). Extrathoracic disease manifested in bone and spleen and less frequently in abdominal viscera, peritoneum, integument, and extremities.
- Despite aggressive procedural and medical therapies, the 5-year survival was 51% and the overall survival was 34%. Mean interval between diagnosis and death was 2.75 years (range, 1-6.5 years).

**CONCLUSIONS:** We describe a clinicopathologically distinct lymphatic anomaly. We propose the term kaposiform lymphangiomatosis (KLA) because of characteristic clusters or sheets of spindled lymphatic endothelial cells accompanying malformed lymphatic channels. The intrathoracic component is most commonly implicated in morbidity and mortality; however, extrathoracic disease is frequent, indicating that KLA is not restricted to pulmonary lymphatics. The mortality rate of KLA is high despite aggressive multimodal therapy.
MED subunits in a specific tumor. Thus, the aim of this study was to investigate for the first time the gene expression profile of the whole MED complex in human osteosarcoma (OS). To this purpose, we have examined all the MED subunit genes in three OS cell lines compared to normal osteoblasts by real-time RT-PCR. Interestingly, our findings indicate that the expression of most of the MED genes is altered in OS. Moreover, a very high overexpression of MED20 and MED31 can be observed in all the analyzed OS cells, thus suggesting for the first time a potential role of these subunits in human malignancies. Overall, this study may open the way to other functional studies exploring the role of the whole complex in cancer development and progression. These findings may lead to the identification of novel biomarkers, which can be used also in combination with imaging techniques for early detection, and/or to develop novel targets for innovative therapeutic approaches.

[194]
TÍTULO / TITLE: - Risk of Kaposi Sarcoma Among Immigrants to Sweden.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Mousavi SM; Sundquist J; Hemminki K
INSTITUCIÓN / INSTITUTION: - Division of Molecular Genetic Epidemiology, C050, German Cancer Research Center (DKFZ), Im Neuenheimer Feld 580, DE-69120 Heidelberg, Germany. smmousavi@yahoo.com.
RESUMEN / SUMMARY: - Abstract is missing (Short).

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[195]
TÍTULO / TITLE: - Right atrial and ventricular angiosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Segers D; Galuzina J; Verdijk RM; Manintveld OC
INSTITUCIÓN / INSTITUTION: - Department of Cardiology, Erasmus MC, Rotterdam, The Netherlands.

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[196]
TÍTULO / TITLE: - Homozygous missense and nonsense mutations in BMPR1B cause acromesomelic chondrodysplasia-type Grebe.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Graul-Neumann LM; Deichsel A; Wille U; Kakar N; Koll R; Bassir C; Ahmad J; Cormier-Daire V; Mundlos S; Kubisch C; Borck G; Klopopcki E; Mueller TD; Doelken SC; Seemann P
Acromesomelic chondrodysplasias (ACDs) are characterized by disproportionate shortening of the appendicular skeleton, predominantly affecting the middle (forearms and forelegs) and distal segments (hands and feet). Here, we present two consanguineous families with missense (c.157T>C, p.(C53R)) or nonsense (c.657G>A, p.(W219*)) mutations in BMPR1B. Homozygous affected individuals show clinical and radiographic findings consistent with ACD-type Grebe. Functional analysis of the missense mutation C53R revealed that the mutated receptor was partially located at the cell membrane. In contrast to the wild-type receptor, C53R mutation hindered the activation of the receptor by its ligand GDF5, as shown by reporter gene assay. Further, overexpression of the C53R mutation in an in vitro chondrogenesis assay showed no effect on cell differentiation, indicating a loss of function. The nonsense mutation (c.657G>A, p.(W219*)) introduces a premature stop codon, which is predicted to be subject to nonsense-mediated mRNA decay, causing reduced protein translation of the mutant allele. A loss-of-function effect of both mutations causing recessive ACD-type Grebe is further supported by the mild brachydactyly or even non-penetration of these mutations observed in the heterozygous parents. In contrast, dominant-negative BMPR1B mutations described previously are associated with autosomal-dominant brachydactyly-type A2.

European Journal of Human Genetics advance online publication, 16 October 2013; doi:10.1038/ejhg.2013.222.

[197] TÍTULO / TITLE: - In vitro modulation of MMP-2 and MMP-9 in adult human sarcoma cell lines by cytokines, inducers and inhibitors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Roomi MW; Kalinovsky T; Monterrey J; Rath M; Niedzwiecki A

INSTITUCIÓN / INSTITUTION: - Dr. Rath Research Institute, Santa Clara, CA 95050, USA.

RESUMEN / SUMMARY: - The highly aggressive adult sarcomas are characterized by high levels of matrix metalloproteinase (MMP)-2 and -9, which play crucial roles in tumor invasion and metastasis by degradation of the extracellular membrane leading to cancer cell spread to distal organs. We examined the effect of cytokines, mitogens, inducers and inhibitors on MMP-2 and MMP-9 secretion in chondrosarcoma (SW-1353), fibrosarcoma (HT-1080), liposarcoma (SW-872) and synovial sarcoma (SW-982) cell lines. The selected compounds included natural cytokines and growth factors, as well as chemical compounds applied in therapy of sarcoma and natural compounds that have demonstrated anticancer therapeutic potential. MMP-2 and MMP-9 secretions were analyzed by gelatinase zymography following 24-h exposure to the tested agents and quantitated by densitometry. Fibrosarcoma, chondrosarcoma, liposarcoma and synovial sarcoma showed bands corresponding to MMP-2 and MMP-9 with dose-dependent enhancement of MMP-9 with phorbol 12-myristate 13-acetate.
(PMA) treatment. In chondrosarcoma cells, tumor necrosis factor (TNF)-alpha had a stimulatory effect on MMP-9 and insignificant effect on MMP-2 and interleukin (IL)-1beta stimulated MMP-9 and MMP-2. In fibrosarcoma and liposarcoma cells, TNF-alpha had a profound stimulatory effect on MMP-9, but no effect on MMP-2 and in synovial sarcoma an inhibitory effect on MMP-2 and no effect on MMP-9. IL-1beta had a slight inhibitory effect on fibrosarcoma, liposarcoma and synovial sarcoma MMP-2 and MMP-9 except for MMP-9 in synovial sarcoma which showed slight stimulation. Lipopolysaccharide (LPS) stimulated expression of MMP-2 in fibrosarcoma and chondrosarcoma while inhibited it in liposarcoma. Doxycycline, epigallocatechin gallate and the nutrient mixture inhibited MMP-2 and MMP-9 in all cell lines. Actinomycin-D, cyclohexamide, retinoic acid, and dexamethasone inhibited MMP-2 and -9 in chondrosarcoma and fibrosarcoma cells. Our results show that cytokines, mitogens, inducers and inhibitors have an up or down regulatory effect on MMP-2 and MMP-9 expression in adult sarcoma cell lines, suggesting these agents may be effective strategies to treat these cancers.
TÍTULO / TITLE: Clinical Neuropathology practice guide 6-2013: morphology and an appropriate immunohistochemical screening panel aid in the identification of synovial sarcoma by neuropathologists.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Keith JL; Bilbao J; Croul S; Ang LC; Guiot MC; Rossiter J; Ghorab Z; Hawkins C; Karamchandani J

RESUMEN / SUMMARY: AIMS: Pathologists are under increasing pressure to accurately subclassify sarcomas, yet neuropathologists have limited collective experience with rare sarcoma types such as synovial sarcoma. We reviewed 9 synovial sarcomas affecting peripheral nerve diagnosed by neuropathologists and explored the morphologic and immunohistochemical differences between these and MPNST. Our goal was to make practical recommendations for neuropathologists regarding which spindle cell tumors affecting nerve should be sent for SYT-SSX testing. METHODS: Clinical records and genetics were reviewed retrospectively and central pathology review of 9 synovial sarcomas and 6 MPNST included immunohistochemistry for SOX10, S100, BAF47, CK (lmw, pan, CK7, CK19), EMA, CD34, bcl2, CD99, and neurofilament. RESULTS: Common synovial sarcoma sites were brachial plexus, spinal and femoral nerve, none were “intra-neural”, all had the SYTSSX1 translocation, and 6/9 were monophasic with myxoid stroma and distinct collagen. Half of the monophasic synovial sarcomas expressed CK7, CK19 or panCK in a “rare positive cells pattern”, 8/9 (89%) expressed EMA, and all were SOX10 immunonegative with reduced but variable BAF47 expression. CONCLUSIONS: We recommend that upon encountering a cellular spindle cell tumor affecting nerve neuropathologists consider the following: 1) SYT-SSX testing should be performed on any case with morphology suspicious for monophasic synovial sarcoma including wiry or thick bands of collagen and relatively monomorphous nuclei; 2) neuropathologists should employ a screening immunohistochemical panel including one of CK7, panCK or CK19, plus EMA, S100 and SOX10, and 3) SYT-SSX testing should be performed on any spindle cell tumor with CK and/or EMA immunopositivity if SOX10 immunostaining is negative or only labels entrapped nerve elements.


RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Maillet D; Goulvent T; Rimokh R; Vacher-Lavenu MC; Pautier P; Alexandre J; Pujade-Laurraine E; Devouassoux-Shisheboran M; Treilleux I; Ray-Coquard I

INSTITUCIÓN / INSTITUTION: Centre Leon Berard, Lyon, France; Universite Claude Bernard Lyon1 - Universite de Lyon, Lyon, France.
OBJECTIVE: Ovarian sex cord-stromal tumors (SCSTs) are rare and their diagnosis is often difficult to establish. Recently, immunostaining and molecular analysis for Forkhead box L2 (FOXL2) have been developed in this pathology. This study aims to assess the benefit of an algorithm incorporating these new tools for a better diagnosis and classification of SCSTs.

METHODS: Seventy-two tumors with a potential diagnosis of SCSTs were addressed by 37 different pathologists to one French rare ovarian tumor expert center, member of the Rare Malignant Ovarian Tumor network (TMRO). Then a “second opinion” (SO) through an algorithm incorporating immunostaining (IHC) and molecular analysis of FOXL2 was performed for all these cases. This algorithm was then validated by all pathologists of the TMRO network.

RESULTS: After a second opinion including molecular analysis and immunostaining for FOXL2 the initial diagnosis was changed in 15 of 72 samples (21%). FOXL2 mutation was present in 44 out of 47 adult granulosa cell tumors (94%), in 3 out of 8 thecomas (37%), in 1 out of 10 Sertoli-Leydig cell tumors (SLSTs) (10%) and in 3 out of 5 undifferentiated-SCSTs (Und-SCSTs) (60%). Immunoexpression of FOXL2 was available in 45 cases of SCSTs: FOXL2 was expressed in 44 of them (98%).

CONCLUSIONS: A second opinion in an expert center for all cases of SCSTs is fundamental to get an optimal classification of these rare tumors. This second opinion could be performed with an algorithm which integrates FOXL2 mutation and expression status of FOXL2 in order to standardize the practice.

TÍTULO / TITLE: Submucosal fibroids and the relation to heavy menstrual bleeding and anemia.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Puri K; Famuyide AO; Erwin PJ; Stewart EA; Laughlin-Tommaso SK

INSTITUCIÓN / INSTITUTION: Division of Gynecology, Department of Obstetrics and Gynecology, Mayo Clinic, Rochester, MN.

RESUMEN / SUMMARY: OBJECTIVE: The objective of the study was to determine the contribution of submucosal fibroids (SMs) to heavy menstrual bleeding (HMB) and anemia among women with HMB. STUDY DESIGN: Our retrospective study included premenopausal women who presented to a tertiary care center for HMB between January 2007 and October 2011. All women in this cohort underwent flexible office hysteroscopy (n = 1665) and 259 (15.6%) had SMs. We also reviewed the clinical ultrasounds (n = 914) from these women to determine whether SMs (n = 148) or any fibroids (n = 434) were present in the uterus. Clinical evaluation of bleeding included hemoglobin and pictorial blood loss assessment charts. RESULTS: In our cohort, hysteroscopically diagnosed SMs were associated with significantly lower hemoglobin (adjusted difference -0.35 g/dL; 95% confidence interval [CI], -0.56 g/dL to -0.13 g/dL) and higher risk of anemia (odds ratio [OR], 1.46; 95% CI, 1.04-2.03). Women with ultrasound-diagnosed SMs had lower hemoglobin and anemia, but results were not significant once adjusted for confounders (hemoglobin: adjusted difference -0.21 g/dL; 95% CI, -0.47 g/dL to 0.06 g/dL; and anemia: OR, 1.28; 95% CI, 0.82-1.97). Ultrasound-
diagnosed fibroids anywhere in the uterus were not associated with hemoglobin (P = .7) or anemia (P = .8). Self-reported pictorial blood loss assessment charts scores did not differ between women with and without fibroids diagnosed by either hysteroscopy or ultrasound (P = .4 and P = .9, respectively). CONCLUSION: SMs were related to lower hemoglobin and higher risk of anemia but not self-reported bleeding scores. Diagnostic modality was important: hysteroscopically diagnosed SMs had lower hemoglobin and more anemia than ultrasound-diagnosed SMs. This may explain the inconsistent results in the literature.

[202]

TÍTULO / TITLE: - Incidence and survival patterns of cranial chordoma in the United States.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Chambers KJ; Lin DT; Meier J; Remenschneider A; Herr M; Gray ST
INSTITUCIÓN / INSTITUTION: - Department of Otolaryngology-Head and Neck Surgery, Massachusetts Eye and Ear Infirmary, and Department of Otology and Laryngology, Harvard Medical School, Boston, MA.
RESUMEN / SUMMARY: - Objective: To determine trends in survival patterns for cranial chordoma in the United States. Study Design: A cross-sectional analysis of a national healthcare database. Methods: From the Surveillance, Epidemiology, and End Results (SEER) program of the National Cancer Institute, all cases of microscopically confirmed cranial chordoma, between 1973 and 2009, were examined. Age-adjusted incidence and survival rates were calculated and stratified by treatment. Additionally, in order to assess trends over time, comparisons in survival were conducted for three calendar year cohorts: 1975-1984, 1985-1994, and 1995-2004. Results: A total of 594 cases of microscopically confirmed chordoma involving cranial sites were identified, which accounted for 42% of all chordomas. Age-adjusted incidence rate (IR) of all chordomas was 0.089 per 100,000. Overall median survival time with surgery plus radiation was 9.2 years. Age and treatment modality were found to influence patient survival. Specifically, age >50 years was associated with a significant increase in mortality rate (P <.05). Five-year survival for the 1975-1984, 1985-1994, and 1995-2004 cohort was 48.5%, 73.0%, and 80.7%, respectively, with improved survival in the more recent cohorts (P<0.01). Conclusion: This study provides new data regarding survival patterns of cranial chordoma in the United States, with a trend toward improvement in survival in recent decades.

[203]

TÍTULO / TITLE: - Fibromyxoid pseudotumor of the ligamentum teres treated with fresh osteochondral allograft.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

●● Enlace al texto completo (gratuito o de pago) 1007/s00256-013-1752-y
Intra-articular tumors and tumor-like conditions of the hip are rare. When they occur, they can interrupt normal articular congruency, leading to pain and joint dysfunction. If these conditions result in large osteochondral defects, they pose challenging reconstructive problems in young patients. We describe a case of a 29-year-old man who presented with a 2-year history of right hip pain. Advanced imaging demonstrated an expansile lesion in the region of his ligamentum teres (LT), eroding a significant portion of his femoral head and expanding the cotyloid fossa. He was treated with surgical hip dislocation, excision of the lesion, and femoral head reconstruction with fresh osteochondral (OC) allograft transplantation via press-fit technique. Histologic examination of the mass showed a benign fibromyxoid pseudotumor. Although non-neoplastic masses have been described in almost all organ systems, to our knowledge this is the first description of this entity affecting the native hip joint. It is only the second description of using press-fit OC allografting in the femoral head. This case adds to the body of literature defining symptomatic LT pathology that may benefit from surgical management. It underscores the need to study the ligament further, as the ability to diagnose and treat intra-articular hip pathology has improved with modern imaging and methods of open and arthroscopic hip surgery.

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Steroid receptor co-activator-3 promotes osteosarcoma progression through up-regulation of FoxM1.

Increasing evidence suggests that the three homologous members of steroid receptor co-activator (SRC) family (SRC-1, SRC-2, and SRC-3) play key roles in enhancing cell proliferation in various human cancers, such as breast, prostate, and hepatocellular carcinoma. However, the function of SRC-3 in osteosarcoma remains largely unexplored. In the current study, we found that SRC-3, but not SRC-1 and SRC-2, was dramatically up-regulated in human osteosarcoma tissues, compared with adjacent normal tissues. To explore the functions of SRC-3 in osteosarcoma, in vitro studies were performed in MG63 and U2OS cells. SRC-3 overexpression promoted osteosarcoma cell proliferation, whereas knockdown of SRC-3 inhibits its proliferation. In support of these findings, we further demonstrated that SRC-3 up-regulated FoxM1 expression through co-activation of C/EBPgamma. Together our results show that SRC-3 drives osteosarcoma progression and imply it as a therapeutic target to abrogate osteosarcoma.
MED12 exon 2 mutations in uterine and extrauterine smooth muscle tumors.

Mutations in exon 2 of the MED12 gene have been reported in 50% to 70% of uterine leiomyomas. To determine the frequency of MED12 mutations in various types of smooth muscle tumors as well as normal uterine myometrium adjacent to a leiomyoma, we selected a total of 143 cases for analysis of MED12 exon 2 mutations by polymerase chain reaction and Sanger sequencing. MED12 mutations were detected in 54% of classical uterine leiomyomas (15/28) and in 15% of cases in myometrium adjacent to leiomyomas (2/13); 34% of leiomyoma/leiomyomatosis in pelvic/retroperitoneal sites (10/29); 0% of extrauterine leiomyomas (0/29); 8% of smooth muscle tumor of uncertain malignant potential (1/12); 30% of uterine leiomyosarcomas (6/20); and 4% of extrauterine leiomyosarcomas (1/25). Mutations were clustered around codons 44, 40, 41, and 36, and consisted primarily of single nucleotide substitutions and small in-frame deletions. Our results confirm the findings of similar recent studies and further show that pelvic and retroperitoneal leiomyomas harbor an increased frequency of MED12 mutations (34%) as compared with other extrauterine sites (0%; P = 0.0006), and that histologically unremarkable adjacent myometrium can harbor similar MED12 mutations. These findings suggest that smooth muscle tumors in pelvic/retroperitoneal sites are subject to the same mutational changes as those of uterine myometrium, and that these mutations may precede the gross or histological development of a leiomyoma.

Variability in functional p53 reactivation by PRIMA-1(Met)/APR-246 in Ewing sarcoma.

Background: Though p53 mutations are rare in ES, there is a strong indication that p53 mutant tumours form a particularly bad prognostic group. As such, novel treatment strategies are warranted that would specifically target and
eradicate tumour cells containing mutant p53 in this subset of ES patients. Methods: PRIMA-1(Met), also known as APR-246, is a small organic molecule that has been shown to restore tumour-suppressor function primarily to mutant p53 and also to induce cell death in various cancer types. In this study, we interrogated the ability of APR-246 to induce apoptosis and inhibit tumour growth in ES cells with different p53 mutations. Results: APR-246 variably induced apoptosis, associated with Noxa, Puma or p21(WAF1) upregulation, in both mutant and wild-type p53 harbouring cells. The apoptosis-inducing capability of APR-246 was markedly reduced in ES cell lines transfected with p53 siRNA. Three ES cell lines established from the same patient at different stages of the disease and two cell lines of different patients with identical p53 mutations all exhibited different sensitivities to APR-246, indicating cellular context dependency. Comparative transcriptome analysis on the three cell lines established from the same patient identified differential expression levels of several TP53 and apoptosis-associated genes such as APOL6, PENK, PCDH7 and MST4 in the APR-246-sensitive cell line relative to the less APR-246-sensitive cell lines. Conclusion: This is the first study reporting the biological response of Ewing sarcoma cells to APR-246 exposure and shows gross variability in responses. Our study also proposes candidate genes whose expression might be associated with ES cells’ sensitivity to APR-246. With APR-246 currently in early-phase clinical trials, our findings call for caution in considering it as a potential adjuvant to conventional ES-specific chemotherapeutics.

[207]

TÍTULO / TITLE: Antitumor activity of neurokinin-1 receptor antagonists in MG-63 human osteosarcoma xenografts.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Munoz M; Berger M; Rosso M; Gonzalez-Ortega A; Carranza A; Covenas R

INSTITUCIÓN / INSTITUTION: Virgen del Rocio University Hospital, Research Laboratory on Neuropeptides, Seville, España.

RESUMEN / SUMMARY: Osteosarcoma is a highly malignant bone tumor in children and adolescents. Aprepitant is a selective highaffinity antagonist of the human neurokinin1 (NK1) receptor (NK1R) with robust antitumor activity. No data exist on the presence of NK1R in osteosarcoma and whether this tumor responds to NK1R antagonists. Here, we analyzed the expression of NK1R in the human osteosarcoma cell line MG-63 with western blot analysis and PCR and found significant expression both at the protein and mRNA levels. We further studied the growth inhibitory capacity of aprepitant and other NK1R antagonists on MG-63 in vitro using an MTS cytotoxicity assay and DAPI staining. All antagonists induced tumor growth inhibition and apoptosis. Synergism was observed for the combination of L-733,060 with common cytostatic drugs in MG-63, but not in non-malignant HEK293 cells. Pretreatment of HEK293 with L-733,060 prior to exposure to cytostatic drugs partially protected HEK293 cells from inhibition by these drugs. Furthermore, nanomolar concentrations of substance P (SP), the natural ligand of the NK1R, increased the growth rate of MG63 cells and micromolar concentrations of aprepitant inhibited SP-induced growth in a
dosedependent manner. In vivo, a xenograft for MG-63 was created in nude mice and treated with peritumoral s.c. injections of fosaprepitant, which resulted in a significant reduction of tumor volume. Collectively, we demonstrated for the first time that the NK1R is expressed in human osteosarcoma cell line MG63 and that this receptor can be targeted with NK1R antagonists both in vitro as well as in vivo.
Most ovarian sex cord-stromal tumors (SCSTs) can be categorized on the basis of conventional histology, but approximately 10% of cases are unclassified because they present indeterminate or overlapping morphologic features. Immunohistochemical and molecular studies of unclassified ovarian SCST are very limited, but recently, it has been demonstrated that 2 major subgroups of SCST, adult-type granulosa cell tumor and Sertoli-Leydig cell tumor, are characterized by somatic mutations in FOXL2 and DICER1, respectively. In this study, 12 diagnostically problematic ovarian SCST, including 9 unclassified tumors, were investigated for FOXL2 and DICER1 mutations and for immunohistochemical expression of calretinin, CD56, CD99, estrogen receptor alpha, estrogen receptor beta, FOXL2, inhibin, progesterone receptor, and steroidogenic factor-1. Four of 11 tumors with satisfactory analysis showed a FOXL2 mutation; 3 of these cases were reported initially as unclassified SCST and 1 as Sertoli-Leydig cell tumor. Conversely, 3 cases with an original diagnosis of granulosa cell tumor were FOXL2 mutation-negative, and none of 7 tumors with satisfactory analysis demonstrated a DICER1 mutation. All tumors expressed at least 4 of the immunomarkers examined, although staining was often focal and there was no consistent correlation with tumor morphology. In conclusion, molecular analysis is useful in the assessment of diagnostically challenging ovarian SCST. The absence of FOXL2 and DICER1 mutations in most unclassified SCST suggests that these could represent a distinct tumor subgroup with different molecular pathogenesis. Immunohistochemical profiles overlap with those of better categorized SCST, but staining may be focal or negative emphasizing the requirement for antibody panels in diagnostic assessment.
RESUMEN / SUMMARY: - Infantile fibrosarcoma (IFS) is a malignant neoplasm, arising in children younger than 2 years of age and with a hallmark chromosomal translocation t(12;15)(p13;q26) encoding an ETV6-NTRK3 fusion oncoprotein. A review of the world literature found no reported cases of molecularly proven IFS with distant metastatic spread at presentation. We report the case of a 2-month-old infant girl presenting with a chest wall primary IFS bearing and expressing the ETV6-NTRK3 fusion, who had several pulmonary metastatic deposits at diagnosis. She achieved complete remission with chemotherapy and surgery. To our knowledge, this is the first reported case of molecularly proven IFS with distant metastatic spread.

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TÍTULO / TITLE: - Expansible kaposiform hemangioendothelioma deformed thoracic cage in an adult.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Wu CH; Tsai TM; Liau JY; Chang YL; Lai HS; Lee JM

INSTITUCIÓN / INSTITUTION: - Department of Surgery, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan.

RESUMEN / SUMMARY: - Kaposiform hemangioendothelioma is a vascular tumor that commonly presents as a cutaneous mass, is observed in children, and is associated with Kasabach-Merritt phenomenon. Herein we report a case of kaposiform hemangioendothelioma with chest wall deformity in an adult who did not show the Kasabach-Merritt phenomenon or cutaneous lesions. To our knowledge, this is the first case of asymptomatic kaposiform hemangioendothelioma arising from the pleura and deforming the chest wall. The patient was treated with tumor excision and chest wall reconstruction.

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TÍTULO / TITLE: - Chromosomal aberrations in primary PDGFRA-mutated gastrointestinal stromal tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Schaefer IM; Delfs C; Cameron S; Gunawan B; Agaimy A; Ghadimi BM; Haller F

INSTITUCIÓN / INSTITUTION: - Department of Pathology, University Medical Center Gottingen, Robert-Koch-Strasse 40, Gottingen D-37075, Germany. Electronic address: inga-marie.schaefer@med.uni-goettingen.de.

RESUMEN / SUMMARY: - Approximately 15% of gastrointestinal stromal tumors (GISTs) harbor mutations in the platelet-derived growth factor receptor alpha
(PDGFRA) gene. Chromosomal aberrations play a crucial role in tumor progression and correlate with clinical behavior. Imbalances, particularly in PDGFRA-mutated GISTs, have not yet been evaluated in larger series. We analyzed 53 PDGFRA-mutated GISTs (including 2 with corresponding metastases) for chromosomal imbalances by conventional comparative genomic hybridization and compared them with a historical collective of 122 KIT-mutated GISTs. PDGFRA exon 18 mutations (91% of cases) and exon 12 mutations (9% of cases) correlated significantly with gastric and intestinal sites, respectively. The most common aberrations were identical to those found in KIT-mutated GISTs, with -14q in 70%, -1p in 28%, and -22q in 17% of cases. Overall, there were significantly fewer chromosomal aberrations compared with KIT-mutated GISTs, with a mean of 2.8 (0.6 gains, 2.1 losses) aberrations per tumor. There was a statistically significant association of more than 5 chromosomal imbalances with intermediate/high-risk categories. Regarding specific chromosomal aberrations, -9p, -13q, and -22q correlated with intermediate/high risk, and -1p and +8q with poorer survival, although progression occurred in only 2 cases. Altogether, PDGFRA-mutated GISTs display the same chromosomal aberrations as KIT-mutated GISTs, although they have a lower degree of chromosomal instability in line with their generally favorable outcome.

[214]

**TITULO / TITLE:** - MYC amplification and overexpression in primary cutaneous angiosarcoma: a fluorescence in-situ hybridization and immunohistochemical study.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Shon W; Sukov WR; Jenkins SM; Folpe AL

**INSTITUCIÓN / INSTITUTION:** - Department of Laboratory Medicine and Pathology, Mayo Clinic, Rochester, MN, USA.

**RESUMEN / SUMMARY:** - MYC, a proto-oncogene located on chromosome 8q24, is involved in the control of cell proliferation and differentiation. Previous studies have documented high-level MYC gene amplification and MYC overexpression by immunohistochemistry (IHC) in post-irradiation angiosarcomas, but not in primary cutaneous angiosarcoma (AS-C) or in other radiation-associated vascular proliferations, such as atypical vascular lesions. Prompted by our recent finding of MYC amplification in a primary hepatic AS, we analyzed a large number of well-characterized AS-C for MYC amplification and protein overexpression. Formalin-fixed, paraffin-embedded blocks from 38 AS-C were retrieved from our archives and were examined by IHC analysis and fluorescence in-situ hybridization (FISH), using a commercially available antibody and probe. For FISH analysis, the number of copies of MYC was compared with the control gene, CEN8 (MYC/CEN8 ratio). All cases occurred on sun-exposed skin; no patient was known to have a history of therapeutic irradiation. Possible associations between survival and a wide variety of clinicopathological variables were evaluated using the log-rank test. By IHC analysis, MYC overexpression was present in 9/38 (24%) AS-C (2-3+: 6 cases, 16%; 1+: 3 cases, 8%). By FISH analysis, 2/5 (40%) informative cases with 2-3+ immunostaining showed high-level gene amplification. One additional case with 3+ immunostaining showed higher level aneusomy of chromosome 8 (5-8 MYC and CEN8). Two out of
fourteen (14%) IHC-negative cases also carried MYC amplification (one high level and one lower level). Low copy number gain of chromosome 8 (3-5 MYC and CEN8) was observed in AS-C with or without MYC expression. MYC amplification and MYC protein overexpression were not correlated with clinical outcome. We have shown, for the first time, MYC gene amplification and protein overexpression in primary (non-radiation-associated) AS of the skin. MYC protein overexpression in cases lacking gene amplification likely reflects other mechanisms of MYC activation. The study of a larger number of AS-C showing MYC amplification may be necessary to determine whether the behavior of such cases differs from their more common non-amplified counterparts.

Modern Pathology advance online publication, 4 October 2013; doi:10.1038/modpathol.2013.163.

[215]

**TÍTULO / TITLE:** Surgical outcomes for 131 cases of carcinosarcoma of the hepatobiliary tract.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Okabayashi T; Shima Y; Iwata J; Iiyama T; Sumiyoshi T; Kozuki A; Tokumaru T; Hata Y; Noda Y; Morita M

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**RESUMEN / SUMMARY:** Carcinosarcoma of the hepatobiliary tract is highly aggressive and has a poor prognosis even after curative resection. The purpose of this study was to collate and analyze published data to clarify the surgical outcome of carcinosarcoma of the hepatobiliary tract and the relationships between potential prognostic factors and survival after surgery. We surveyed worldwide literature from 1970 to 2012 and obtained clinicopathological data for 131 patients who had undergone surgical resection for carcinosarcoma of the hepatobiliary tract, including one patient from our clinic. The relationships between potential prognostic factors and survival rates were examined using the Kaplan-Meier method and the log-rank test. The overall 1-, 3-, and 5-year survival rates for patients with carcinosarcoma of the hepatobiliary tract after surgery were 44.0, 29.3, and 27.0 %, respectively. In univariate analyses, age and gender were not significant prognostic factors; however, advanced stage according to the classification of the Union for International Cancer Control in resected specimens was significantly associated with a shorter survival time after surgery. Although carcinosarcoma of the hepatobiliary tract remains a rare disease worldwide, its poor prognosis, even after curative resection, demands further epidemiological and pathological study that could lead to the development of new management strategies.

[216]

**TÍTULO / TITLE:** Gastrointestinal stromal tumors of the stomach: the role of laparoscopic resection. Single-centre experience of 38 cases.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary

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RESUMEN / SUMMARY: - INTRODUCTION: Laparoscopic resection is considered the gold standard of treatment only for small gastric gastrointestinal stromal tumor (GIST). MATERIALS AND METHODS: Between January 2004 and September 2012, 38 consecutive gastric GISTs were operated on by laparoscopic approach, without conversions. Thirty-five cases were primary GISTs and three were bleeding GISTs with hepatic metastases non-responding to conservative therapy treated by emergency surgery. RESULTS: Median tumor size was 3.63 cm (1.8-17 cm). In two cases tumor size was <2 cm, between 2 and 5 cm in 26 cases, between 5 and 10 cm in eight cases, and >10 cm in two cases. In two cases, localization was in the cardia, fundus in ten cases, lesser curve in 11 cases, greater curve in 12 cases, and antrum in three cases. We performed 24 wedge resections, eight transgastric resections and six antrectomies. An Endo-GIA was used in 25 cases, and a manual laparoscopic reconstruction with extramucosal suture was performed in 13 cases. No postoperative mortality and morbidity was observed. The routine use of laparoscopy allowed us to perform resections in 100 % of cases, even in those where preoperative imaging suggested an open approach according to the current guidelines. CONCLUSIONS: The use of a pre-resection endobag avoids spillage and seeding, thus increasing the possibility of resection. In conclusion, we consider the laparoscopic approach as mandatory in all cases, always considering the possibility of converting to the open technique when necessary.

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AUTORES / AUTHORS: - Yan L; Ding L; Tang R; Chen ZJ

INSTITUCIÓN / INSTITUTION: - Center for Reproductive Medicine, Department of Obstetrics and Gynecology, Provincial Hospital Affiliated to Shandong University, Jinan, PR China.

RESUMEN / SUMMARY: - Aims: To study the effect of adenomyosis on in vitro fertilization/intracytoplasmic sperm injection (IVF/ICSI) outcomes in infertile patients. Methods: We performed a retrospective, database-searched cohort study based on 10,268 patients undergoing controlled ovarian hyperstimulation and IVF/ICSI between 2009 and 2011 in our unit. Adenomyosis was diagnosed by transvaginal ultrasound. A high-quality matched cohort study with strict inclusion criteria was conducted. We compared the basic characteristics and main IVF/ICSI outcomes between the two groups. Results: We identified 83 patients with adenomyosis, of whom we included 77, and strictly matched them to 77 patients without adenomyosis. Higher day 3 estrogen levels and a longer duration of gonadotropin stimulation days were found in women with adenomyosis.
with adenomyosis compared to control subjects. Patients with adenomyosis had a nonsignificant trend toward a lower clinical pregnancy rate and a higher miscarriage rate (p = 0.103 and 0.09, respectively). The delivery rate was significantly lower in the adenomyosis group in comparison to the matched controls (p = 0.022). Conclusions: Within the limitations of a retrospective study (albeit with a remarkably large number of observations), our results suggest that transvaginal ultrasound-diagnosed adenomyosis could have a negative impact on the main IVF/ICSI outcomes. Improving the diagnostic validity and scoring of disease severity in patients with adenomyosis is suggested. © 2013 S. Karger AG, Basel.

[218]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Kurucu N; Sari N; Celasun B; Sarihan H; Ahmetoglu A; Ilhan IE
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RESUMEN / SUMMARY: - Gastrointestinal stromal tumor (GIST) is the most common mesenchymal neoplasm of the gastrointestinal tract. Only 1.5% to 2% of all GISTs are observed in children and adolescents. Most of the pediatric cases are between 10 and 18 years of age, with a median age of 13 years. GIST is extremely rare in the newborn period. We could find only 5 reports on the neonatal cases. Herein, we have reported a case with abdominal tumor that was identified by prenatal ultrasonography and magnetic resonance imaging, and diagnosed as GIST on the seventh day of life. We have also reviewed the neonatal GIST cases reported in the English literature.

[219]
TÍTULO / TITLE: - Lipoma of the middle ear: An unusual presentation in a 6 year old child.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Aldosari B
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local recurrence for extremity soft-tissue sarcomas for survival. METHODS: Three hundred and sixty-three patients who underwent resection of localized primary extremity soft-tissue sarcomas with curative intent were selected from the United States Department of Defense Automated Central Tumor Registry. Outcomes for local recurrence, distant recurrence, disease-specific survival, and overall survival were analyzed according to clinical, pathological, and treatment variables with use of the Kaplan-Meier method (log-rank test) and the multivariate Cox regression model.

RESULTS: Positive margins (hazard ratio, 1.99 [95% confidence interval, 1.15 to 3.45]), local recurrence (hazard ratio, 2.93 [95% confidence interval, 1.38 to 6.23]), and distant recurrence (hazard ratio, 12.13 [95% confidence interval, 5.97 to 24.65]) were significantly associated with overall survival on multivariate Cox regression analysis. However, for disease-specific survival, local recurrence was not significant and tumor size of >10 cm (hazard ratio, 2.83 [95% confidence interval, 1.15 to 6.95]), positive margins (hazard ratio, 1.95 [95% confidence interval, 1.05 to 3.63]), and distant recurrence (hazard ratio, 9.46 [95% confidence interval, 4.37 to 20.47]) were independent adverse prognostic factors. The disease-specific survival rate for patients with localized soft-tissue sarcomas was 89% (95% confidence interval, 85% to 92%) for five years and 75% (95% confidence interval, 70% to 81%) for ten years.

CONCLUSIONS: Positive surgical margins are consistently associated with adverse survival-related outcomes in localized soft-tissue sarcomas of the extremity. Local recurrence had a significant impact on overall survival, but not on disease-specific survival.

[222]
TITULO / TITLE: - Disseminated Peritoneal Leiomyomatosis Clinically and Radiologically Mimicking Malignancy.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - K R A; Rema P; Jayasree K

[223]
TITULO / TITLE: - MR imaging features of spindle cell lipoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Kirwadi A; Abdul-Halim R; Fernando M; Highland A; Kotnis N
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content with grade 0 representing 0 % fat signal and grade 4 100 % fat signal. The degree of fat suppression and contrast-enhancement pattern were also recorded. The excision specimens were independently reviewed by a consultant histopathologist. The histology specimens were assessed for the amount of internal fat and non-adipose tissue and graded using the same scale applied for the imaging. Where core needle biopsy (CNB) was performed, the CNB specimens were also examined for positive features of SCL. RESULTS: In our study, 93 % (13/14) of our patients were male and the average age was 58 years. 65 % (9/14) of the lesions presented in the upper back, shoulder, or neck. All lesions were subcutaneous. 35 % (5/14) of the SCLs demonstrated grade 3 (>75 %) or grade 4 (100 %) fat signal on MR examination. 35 % (5/14) of the lesions had grade 2 (25-75 %) fat signal and 29 % (4/14) of the lesions demonstrated grade 0 (0 %) or grade 1 (<25 %) fat signal. 43 % (6/14) of lesions demonstrated homogenous fat suppression, 28 % (4/14) showed focal areas of high internal signal, and 28 % (4/14) had diffuse internal high signal on fluid-sensitive fat-saturated sequences. 86 % (6/7) of the cases demonstrated septal/nodular enhancement. The diagnosis was evident on the CNB specimen in 100 % (9/9) cases. The histopathology fat content grade was in agreement with the imaging grade in 86 % (12/14) cases. CONCLUSIONS: The internal signal pattern of SCL can range broadly, with low fat content lesions seen almost as commonly as intermediate and high fat content lesions. We also found that the fat:non-fat internal MR signal pattern of these lesions is accurately reflected in their composition at histology.


RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Prasad SC; Piccirillo E; Nuseir A; Sequino G; De Donato G; Paties CT; Sanna M

INSTITUCION / INSTITUTION: Department of Otology and Skull Base Surgery, Gruppo Otologico, Piacenza Civil Hospital, Piacenza, Italy.

RESUMEN / SUMMARY: Objective: To study the clinical features, tumor characteristics and outcomes of giant cell tumors (GCTs) in the skull base based on long-term follow-up. We also report the largest series of GCTs in the temporal bone and the lateral skull base. Materials and Methods: A retrospective study was conducted of all GCTs managed at the Gruppo Otologico, a quaternary referral skull base institute, in Italy from 1993 to 2013. The clinical features, investigations, surgical management and follow-up were recorded. The surgical approaches used were infratemporal fossa approach (ITFA) type B and D and middle cranial fossa (MCF) approaches. Results and Observations: A total of 7 patients with GCTs of the skull base were treated at our institution. The principal complaints were hearing loss reported in 6 (85.71%) patients, tinnitus in 5 (71.43%) and swelling in 3 (42.9%). Pure-tone audiometry showed conductive hearing loss in 5 (71.43%) patients. High-resolution CT scan and MRI with gadolinium enhancement were done in all patients. Radiology showed involvement of the ITF and middle ear in 6 (85.71%) patients each, temporomandibular joint in 4 (57.14%) patients, invasions of the squamous part of the temporal bone, mastoid, MCF
and greater wing of sphenoid in 3 (42.9%) patients each and the petrous bone in 2 (28.6%) patients. ITFA type B was applied as an approach for tumor removal in 5 (71.43%) patients, including a case where an additional MCF approach was employed, and ITFA type D and the transmastoid approach were applied in 1 (14.3%) patient each. Total tumor removal and successful cure was achieved in 6 (85.71%) patients. Subtotal removal leading to recurrence and eventual mortality was the result in 1 (14.3%) patient. Conclusions: A thorough knowledge of the anatomy of the skull base and the various skull base approaches is necessary to tackle GCTs. ITFA type B and D combined with MCF approaches provide good exposure of the tumor with minimal postoperative sequelae and good locoregional control. Recurrence due to either subtotal removal or suboptimal treatment may have disastrous consequences for the patient. © 2013 S. Karger AG, Basel.

[225]

**TÍTULO / TITLE:** - Organ preservation surgery for low- and intermediate-grade laryngeal chondrosarcomas: Analysis of 16 cases.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Piazza C; Del Bon F; Grazioli P; Mangili S; Barbieri D; Nicolai P; Peretti G

**INSTITUCIÓN / INSTITUTION:** - Department of Otorhinolaryngology-Head and Neck Surgery, University of Brescia, Brescia, Italy.

**RESUMEN / SUMMARY:** - OBJECTIVES/HYPOTHESIS: To demonstrate that endoscopic resection (ER), open partial laryngectomies, and cricotracheal resection and anastomosis (CTRA) achieve a good balance between oncologic radicality and organ preservation for laryngeal low-grade chondrosarcoma (LCS) and intermediate-grade chondrosarcoma (ICS). STUDY DESIGN: Retrospective series in an academic institution. METHODS: Between 2001 and 2013, we treated 13 cricoid, two thyroid, and one arytenoid LCS and ICS. Two cricoid and the only arytenoid LCS were managed by ER. Two thyroid ala LCS were submitted to laminectomy. Five ICS and six LCS of the cricoid received CTRA. RESULTS: Nine patients only required tracheotomy, removed after a maximum of 14 days. Three patients required a nasogastric feeding tube, removed after a maximum of 8 days. Immediate complications included one bleeding, one cervical emphysema, and one partial anastomotic dehiscence. The only late complication was anastomotic stenosis that was resolved by laser resection. All patients regained regular oral feeding and a voice ranging from normal to moderate dysphonia. At the last follow-up, two patients died of unrelated causes, seven are alive with asymptomatic and radiologically stable residual disease, and seven are alive without evidence of persistent disease. One patient received total laryngectomy 11 years after CTRA for recurrent symptomatic disease. CONCLUSIONS: Organ preservation surgery for laryngeal LCS and ICS represents a treatment option with low morbidity, good quality of life, and fair possibility to obtain oncologic radicality. LEVEL OF EVIDENCE: 4 Laryngoscope, 2013.

[226]
**TÍTULO / TITLE:** - HDAC5 promotes osteosarcoma progression by upregulation of Twist 1 expression.

**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)

**REVISTA / JOURNAL:** - Tumour Biol. 2013 Oct 5.

**AUTORES / AUTHORS:** - Chen J; Xia J; Yu YL; Wang SQ; Wei YB; Chen FY; Huang GY; Shi JS

**INSTITUCIÓN / INSTITUTION:** - Department of Orthopedic, Huashan Hospital, Fudan University, Shanghai, 200040, China.

Histone deacetylases (HDACs) form a family of enzymes, which have fundamental roles in the epigenetic regulation of gene expression and contribute to the growth, differentiation, and apoptosis of cancer cells. In this study, we firstly investigated the biological function of HDAC5 in osteosarcoma cells. We found that mRNA and protein levels of HDAC5 were upregulated in osteosarcoma tissues and cell lines. Furthermore, overexpression of HDAC5 could promote cell proliferation in osteosarcoma cell lines. In contrast, HDAC5 knockdown using small interfering RNA inhibited cell proliferation. At the molecular level, we demonstrated that HDAC5 promoted mRNA expression of twist 1, which has been reported as an oncogene. Together, these results highlighted for the first time an unrecognized link between HDAC5 and osteosarcoma progression and demonstrated that its specific inhibition might contribute to the treatment of tumorigenesis.

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**TÍTULO / TITLE:** - Liposarcomas: diagnostic pitfalls and new insights.

**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)


**AUTORES / AUTHORS:** - Dei Tos AP

**INSTITUCIÓN / INSTITUTION:** - Departments of Pathology and Oncology, General Hospital of Treviso, Treviso, ITALY.

Liposarcomas represents the commonest histotype among soft tissue sarcomas. They actually represents a heterogeneous group of distinctive lesions that poses several diagnostic difficulties. The current WHO classification of soft tissue tumors and bone recognizes four major liposarcoma subtypes: 1. atypical lipomatous tumor/well-differentiated (WD) liposarcoma (which includes the adipocytic, sclerosing, inflammatory, and spindle cell variants); 2. dedifferentiated liposarcoma; 3. myxoid liposarcoma, 4. pleomorphic liposarcoma. These four main subgroups are characterized by distinctive morphologies, unique genetic findings as well as distinct clinical behavior. Accurate classification requires the integration of morphologic, immunohistochemical, and (in selected situations) genetic findings, and represents the essential step in order to address patients to the best available treatment. This review will focus on main diagnostic pitfalls encountered in routine diagnosis of liposarcoma, underlining the diagnostic value of combining morphology with cytogenticities and molecular genetics. This article is protected by copyright. All rights reserved.

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[227]

[228]
Molecular pathological analysis of sarcomas using paraffin-embedded tissue: current limitations and future possibilities.

Sarcomas of soft tissue and bone are rare neoplasms that can be separated into a large number of different diagnostic entities. Over the years, a number of diagnostic markers have been developed that aid pathologists in reaching the appropriate diagnoses. Many of these markers are sarcoma-specific proteins that can be detected by immunohistochemistry in formalin-fixed, paraffin-embedded (FFPE) sections. In addition, a wide range of molecular studies have been developed that can detect gene mutations, gene amplifications or chromosomal translocations in FFPE material. Until recently, most sequencing-based approaches relied on the availability of fresh frozen tissue. However, with the advent of next-generation sequencing technologies, FFPE material is increasingly being used as a tool to identify novel immunohistochemistry markers, gene mutations, and chromosomal translocations, and to develop diagnostic tests.

Rhabdoid morphology in gastrointestinal stromal tumours (GISTs) is associated with PDGFRA mutations but does not imply aggressive behaviour.

AIMS: Rhabdoid morphology resembling that of the aggressive paediatric rhabdoid tumours occurs in various malignancies usually lacking characteristic SMARCB1 (INI1) loss. Little is known about the clinicopathological and molecular characteristics of the rhabdoid phenotype in gastrointestinal stromal tumours (GISTs). METHODS AND RESULTS: Six gastric rhabdoid GISTs were examined by immunohistochemistry, KIT and platelet-derived growth factor receptor-alpha gene (PDGFRA) mutation analysis, and comparative genomic hybridization (CGH). All tumours expressed KIT, PDGFRA, DOG-1, and SMARCB1 (two of six with a mosaic pattern). Five of six tumours harboured PDGFRA mutations (D842V in four; N659K in one), and one case was wild type for KIT/PDGFRA and succinate dehydrogenase (SDH) A-negative and SDHB-negative by immunohistochemistry. CGH revealed aberrations typical of GISTs (-1p, -14, and -22q in three, five, and three cases, respectively), with a mean of 1.7 aberrations in the epithelioid component and 2.7 in the rhabdoid component. None showed progression (mean follow-up of 25 months).

CONCLUSIONS: Rhabdoid gastric GISTs are associated with epithelioid morphology.
and PDGFRA mutations. They harbour CGH aberrations that are typical of ordinary GISTs in both tumour components. The presence of additional genetic alterations in the rhabdoid areas indicates evolution from the epithelioid components, and possible genetic and biological progression. On the basis of our series and previous reports, rhabdoid morphology in GISTs presumably does not imply aggressiveness.

[230]
**TÍTULO / TITLE:** - BRAF Mutation in “Sarcomas”: A Possible Method to Detect Dedifferentiated Melanomas.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Cipriani NA; Letovanec I; Hornicek FJ; Mullen JT; Duan Z; Borger DR; Petur Nielsen G

**INSTITUCIÓN / INSTITUTION:** - Department of Pathology, The University of Chicago, Chicago, IL, USA.

**RESUMEN / SUMMARY:** - AIMS: BRAF is mutated in 50-60% of melanomas, but BRAF mutation in sarcomas has not been systematically evaluated. Some melanomas are spindled and may show no immunohistochemical evidence of melanocytic differentiation. Similarly, many sarcomas are undifferentiated, i.e. undifferentiated pleomorphic sarcomas (UPS). Diagnosing melanoma versus sarcoma in an undifferentiated spindle cell malignancy can be challenging. Our aim was to evaluate the prevalence of BRAF mutation in sarcomas and the use of BRAF mutational status in the diagnosis of spindle cell malignancies. METHODS & RESULTS: BRAF mutational analysis was performed on tissue from 104 patients: 90 with sarcoma only (50 UPS) and 14 with sarcoma and melanoma (7 UPS). In the sarcoma-only group, BRAF mutation was absent. In the sarcoma-melanoma group, three sarcomas showed BRAF mutation; all were UPS, occurred after the melanomas and did not stain for melanocytic markers. One melanoma-sarcoma pair showed identical BRAF V600E mutations. CONCLUSIONS: The presence of BRAF mutation in these tumors raises the possibility that poorly-differentiated spindle cell malignancies with BRAF mutation may represent melanomas, and BRAF mutational analysis should be considered in a patient with a spindle cell malignancy and a history of melanoma, as a positive result may indicate dedifferentiated melanoma. This article is protected by copyright. All rights reserved.

[231]
**TÍTULO / TITLE:** - Regorafenib: A Novel Multitargeted Tyrosine Kinase Inhibitor for Colorectal Cancer and Gastrointestinal Stromal Tumors.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Crona DJ; Keisler MD; Walko CM

**INSTITUCIÓN / INSTITUTION:** - University of North Carolina (UNC) Eshelman School of Pharmacy, NC, USA.
RESUMEN / SUMMARY: - OBJECTIVE: To review currently available literature on the oral multikinase inhibitor regorafenib and its role in the treatment of metastatic colorectal cancer (mCRC), and imatinib- and sunitinib-resistant gastrointestinal stromal tumors (GISTs). DATA SOURCES: A comprehensive literature search was performed of PubMed/MEDLINE and American Society of Clinical Oncology (ASCO) abstracts (through August 2013). STUDY SELECTION/DATA EXTRACTION: Preclinical pharmacological and phase I to III trials data analyzing regorafenib efficacy and safety in mCRC or imatinib- and sunitinib-resistant GIST patients were evaluated. All available English-language, peer-reviewed articles and ASCO abstracts with relevant information were reviewed. DATA SYNTHESIS: Regorafenib was approved for mCRC in September 2012 and for imatinib- and sunitinib-resistant GISTs in February 2013. Regorafenib is an inhibitor of stromal, angiogenic, and oncogenic receptor tyrosine kinases, as well as the RAF/MEK/ERK signaling pathway. Phase III CORRECT (Regorafenib Monotherapy for Previously Treated Metastatic Colorectal Cancer) trial data demonstrated an overall survival benefit for mCRC patients treated with regorafenib (6.4 vs 5.0 months; P = .0052). Phase III GRID (Gastrointestinal Stromal Tumors After Failure of Imatinib and Sunitinib) trial data revealed a progression-free survival benefit in imatinib- and sunitinib-resistant GIST patients (4.8 vs 0.9 months; P < .0001). Its adverse event (AE) profile is comparable to that of other multikinase inhibitors. The most commonly observed grade >/=3 AEs included hypertension, hand-foot skin reaction, rash, diarrhea, and fatigue. CONCLUSIONS: Regorafenib is a novel oral multikinase inhibitor that has shown promising results for patients with advanced, unresectable or metastatic treatment-refractory CRCs or imatinib- and sunitinib-resistant GISTs.

TÍTULO / TITLE: - MACC1 is involved in the regulation of proliferation, colony formation, invasion ability, cell cycle distribution, apoptosis and tumorigenicity by altering Akt signaling pathway in human osteosarcoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Zhang K; Tian F; Zhang Y; Zhu Q; Xue N; Zhu H; Wang H; Guo X

INSTITUCIÓN / INSTITUTION: - Department of Orthopaedics, Chinese PLA General Hospital & Chinese PLA Medical School, No. 28 Fuxing Road, Haidian District, Beijing, 100853, China.

RESUMEN / SUMMARY: - There is mounting evidence that metastasis-associated in colon cancer-1 (MACC1) plays pivotal roles in development and progression of many tumors, particularly in osteosarcoma (OS). However, its precise roles and molecular mechanisms remain to be delineated in OS. In the current study, we found that the levels of MACC1 mRNA and protein in four OS cell lines (MG-63, HOS, SaOS-2 and U2OS) were significantly higher than that in hFOB1.19 osteoblast (P < 0.05). The vector pcDNA-MACC1 contributed to the increase of MACC1 level in MG-63 cells, whereas MACC1 siRNA evoked the decrease of MACC1 level in U2OS cells. In addition, MACC1 downregualtion caused the inhibition of cell proliferation in vitro, colony formation, invasion and tumor growth in vivo, arrested cell cycle in G0/G1 phase
and induced cell apoptosis in U2OS cells, and reversed effects were observed in MG-63 cells by MACC1 upregulation. Most notably, MACC1 depletion markedly inactivated Akt signaling pathway in U2OS cells, conversely, MACC1 upregulation evidently activated Akt signaling pathway in MG-63 cells. Collectively, our data presented herein suggest that biological implications triggered by MACC1 may be tightly associated with the status of Akt signaling pathway in OS.
PD for operable DGISTs if clear surgical margins are achieved. Comprehensive treatment is necessary.

[234]

**TÍTULO / TITLE:** - Immunohistochemistry of soft tissue tumours - review with emphasis on 10 markers.

**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](https://doi.org/10.1111/his.12298)


**AUTORES / AUTHORS:** - Miettinen M

**INSTITUCIÓN / INSTITUTION:** - Laboratory of Pathology, National Cancer Institute, Bethesda, MD, USA.

**RESUMEN / SUMMARY:** - Immunohistochemistry is an integral component in the proper analysis of soft tissue tumours, and a simple panel of six markers is useful in practical triage: CD34, desmin, epithelial membrane antigen (EMA), keratin cocktail AE1/AE3, S100 protein and alpha smooth muscle actin (SMA). These markers frequently assist in the differential diagnosis of fibroblastic, myoid, nerve sheath and perineurial cell tumours, synovial and epithelioid sarcoma and others. However, they all are multispecific, so that one has to be cognizant of their distribution in normal and neoplastic tissues. Four additional useful markers for specific tumour types are discussed here: CD31 and ERG for vascular endothelial tumours, and KIT and DOG1/Ano-1 for gastrointestinal stromal tumours (GISTs). However, hardly any marker is totally monospecific for any one type of tumour. Furthermore, variably lineage-specific markers do not usually distinguish between benign and malignant proliferations, so that this distinction has to be made on histological grounds. Immunohistochemical evaluation is most useful, efficient and cost-effective when used in the context of careful histological evaluation by an experienced pathologist, aware of all diagnostic entities and their histological spectra. Additional diagnostic steps that must be considered in difficult cases include clinicoradiological correlation and additional sampling of remaining wet tissue, if possible.

[235]

**TÍTULO / TITLE:** - Ewing Sarcoma/Peripheral Primitive Neuroectodermal Tumor in the Adrenal Gland of an Adolescent: A Case Report and Review of the Literature.

**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](https://doi.org/10.1097/MPH.0000000000000058)


**AUTORES / AUTHORS:** - Yoon JH; Kim H; Lee JW; Kang HJ; Park HJ; Park KD; Park BK; Shin HY; Park JD; Park SH; Ahn HS

**INSTITUCIÓN / INSTITUTION:** - *Department of Pediatrics, Division of Hematology/Oncology, Cancer Research Institute double daggerDepartment of Pathology, Seoul National University College of Medicine, Seoul daggerCenter for Pediatric Oncology, National Cancer Center, Goyang, Korea.

**RESUMEN / SUMMARY:** - Ewing sarcoma/peripheral primitive neuroectodermal tumors (ES/pPNETs) typically occur in the long or flat bones, the chest wall, extraskeletal soft
tissue, or less frequently, in solid organs. They can arise from anywhere in the body; however, ES/pPNETs arising from the adrenal gland are very rare, especially in children and adolescents. Herein, the authors report a case of an ES/pPNET in the adrenal gland of a 17-year-old girl, who was successfully treated with a multimodal treatment, with a brief review of the pertinent literature.

[236]

TÍTULO / TITLE: - Sampling modality influences the predictive value of grading in adult soft tissue extremity sarcomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Khoja H; Griffin A; Dickson B; Wunder J; Ferguson P; Howarth D; Kandel R
INSTITUCIÓN / INSTITUTION: - From the Departments of Pathology and Laboratory Medicine (Drs Khoja, Dickson, Howarth, and Kandel) and Orthopaedic Surgery (Mr Griffin and Drs Wunder and Ferguson), Mount Sinai Hospital, Toronto, Ontario, Canada; and the Departments of Laboratory Medicine and Pathobiology (Drs Dickson, Howarth, and Kandel) and Surgery (Drs Wunder and Ferguson), University of Toronto, Toronto, Ontario, Canada.

RESUMEN / SUMMARY: - Context.-Histologic grade is one of the best predictors of outcome in adult soft tissue sarcomas. Current grading systems were validated on resection specimens; however, there has been a trend toward the use of biopsies to diagnosis these tumors. Objectives.-To determine whether the grade of an extremity soft tissue sarcoma determined on tissue obtained by either core needle biopsy or incisional biopsy is predictive of metastasis- or disease-free survival, and whether either sampling modality is superior. Design.-One hundred three core needle biopsies and 107 incisional biopsies of nonmetastatic spindle cell sarcomas of the extremities were retrieved from the archives. All cases had a minimum 2-year follow-up. Patient data and outcome and tumor characteristics were recorded. Tumors were reviewed and evaluated using the French Federation of Cancer Centers Sarcoma Group grading system. Kaplan-Meier survival curves were generated to correlate tumor grade with metastasis- and disease-free survival for both groups. Results.-Patient and tumor characteristics were similar between groups except that more tumors were grade 3 and superficial in the incisional biopsy group. Grade determined on core needle biopsy was not predictive of either metastasis-free survival (P = .59) or disease-free survival (P = .50). In contrast, grade determined on incisional biopsy was predictive of both metastasis-free survival (P < .001) and disease-free survival (P = .001). Conclusions.-Biopsy, particularly core needle biopsy, represents a convenient diagnostic tool, particularly in the context of neoadjuvant therapy. However, based on these results incisional biopsy is recommended if grading is to be used to predict prognosis in spindle cell soft tissue sarcomas of the extremities.

[237]
**TÍTULO / TITLE:** - In vitro modulation of MMP-2 and MMP-9 in pediatric human sarcoma cell lines by cytokines, inducers and inhibitors.

**RESUMEN / SUMMARY:** - The highly aggressive pediatric sarcomas are characterized by high levels of matrix metalloproteinase (MMP)-2 and MMP-9, which play crucial roles in tumor invasion and metastasis by degradation of the extracellular membrane leading to cancer cell spread to distal organs. We examined the effects of cytokines, mitogens, inducers and inhibitors on MMP-2 and -9 expression in osteosarcoma (U2OS) and rhabdomyosarcoma (RD). The selected compounds included natural cytokines and growth factors, as well as chemical compounds applied in therapy of sarcoma and natural compounds that have demonstrated anticancer therapeutic potential. These cell lines were cultured in their respective media to near confluence and the cells were washed with PBS and incubated in serum-free medium with various concentrations of several cytokines, mitogens and inhibitors. After 24 h the media were removed and analyzed for MMP-2 and -9 by gelatinase zymography and quantitated by densitometry. Osteosarcoma and rhabdomyosarcoma showed bands corresponding to MMP-2 and -9 with dose-dependent enhancement of MMP-9 with phorbol 12-myristate 13-acetate (PMA) treatment. Tumor necrosis factor-alpha, interleukin-1beta and LPS enhanced osteosarcoma U2OS MMP-9 secretion but had no effect on MMP-2 secretion. Tumor necrosis factor-alpha stimulated rhabdomyosarcoma MMP-2 expression, but had no effect on MMP-9 secretion. Doxycycline, epigallocatechin gallate, nutrient mixture (NM), actinomycin-D, cyclohexamide, retinoic acid and dexamethasone inhibited MMP-2 and -9 in U2OS osteosarcoma cells. PMA-treated RD cells showed dose-response inhibition of MMP-9 by doxycycline and epigallocatechin gallate and both MMPs by NM. Dexamethasone and actinomycin-D showed inhibition of MMP-2 secretion of RD cells. Our results show that cytokines, mitogens and inducers show variable upregulation of U2OS osteosarcoma and RD rhabdomyosarcoma MMP-2 and -9 secretion, and inhibitors demonstrate downregulation under stimulatory conditions, suggesting the application of these agents for the development of effective therapies in pediatric sarcomas.

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**TÍTULO / TITLE:** - Surgical management of juvenile nasopharyngeal angiofibroma: Analysis of 162 cases 1995-2012.

**RESUMEN / SUMMARY:** - The highly aggressive pediatric sarcomas are characterized by high levels of matrix metalloproteinase (MMP)-2 and MMP-9, which play crucial roles in tumor invasion and metastasis by degradation of the extracellular membrane leading to cancer cell spread to distal organs. We examined the effects of cytokines, mitogens, inducers and inhibitors on MMP-2 and -9 expression in osteosarcoma (U2OS) and rhabdomyosarcoma (RD). The selected compounds included natural cytokines and growth factors, as well as chemical compounds applied in therapy of sarcoma and natural compounds that have demonstrated anticancer therapeutic potential. These cell lines were cultured in their respective media to near confluence and the cells were washed with PBS and incubated in serum-free medium with various concentrations of several cytokines, mitogens and inhibitors. After 24 h the media were removed and analyzed for MMP-2 and -9 by gelatinase zymography and quantitated by densitometry. Osteosarcoma and rhabdomyosarcoma showed bands corresponding to MMP-2 and -9 with dose-dependent enhancement of MMP-9 with phorbol 12-myristate 13-acetate (PMA) treatment. Tumor necrosis factor-alpha, interleukin-1beta and LPS enhanced osteosarcoma U2OS MMP-9 secretion but had no effect on MMP-2 secretion. Tumor necrosis factor-alpha stimulated rhabdomyosarcoma MMP-2 expression, but had no effect on MMP-9 secretion. Doxycycline, epigallocatechin gallate, nutrient mixture (NM), actinomycin-D, cyclohexamide, retinoic acid and dexamethasone inhibited MMP-2 and -9 in U2OS osteosarcoma cells. PMA-treated RD cells showed dose-response inhibition of MMP-9 by doxycycline and epigallocatechin gallate and both MMPs by NM. Dexamethasone and actinomycin-D showed inhibition of MMP-2 secretion of RD cells. Our results show that cytokines, mitogens and inducers show variable upregulation of U2OS osteosarcoma and RD rhabdomyosarcoma MMP-2 and -9 secretion, and inhibitors demonstrate downregulation under stimulatory conditions, suggesting the application of these agents for the development of effective therapies in pediatric sarcomas.

[238]
Objective: The purpose of this study was to report on a series of 162 patients presenting with juvenile nasopharyngeal angiofibroma in a single academic hospital during the past 17 years in an effort to compare outcomes between open and transnasal endoscopic approach, and to define an ideal treatment strategy. Study Design: Patients who received either open or endoscopic surgery with minimum follow-up of 6 months were selected. Local control and complications were compared between groups. Methods: Retrospectively, clinical data, surgical reports, pre- and postoperative images and follow-up information were reviewed and analyzed. Results: All patients were male subjects from 8 to 41 years old. Ninety-six patients were treated by transpalatal or transmaxillary approach and the remaining sixty-six patients were treated using transnasal endoscopic approach with without labiogingival incision. When compared to the open surgery group, the endoscopic surgery group showed a lower median intraoperative blood loss (800 vs. 1100 ml, \(P=0.017\)), number of postoperative complications (1 vs. 10). In addition, recurrence statistically correlated with Radkowski’s classification and patient age. Conclusion: Transnasal endoscopic approach can be successfully used for Radkowski’s stages I-IIb tumors and selective IIc-IIIb lesions, allowing for less blood loss, postoperative complications and a lower percentage of recurrence in comparison to open surgery. The management of recurrent tumor is complex, should be individually tailored and should take into account tumor location, patient age, complications of treatment and the possibility of spontaneous involution, to better define treatment strategy.
INTRODUCTION: Primary cardiac sarcomas (PCS) are rare tumours of dismal prognosis. METHODS: Data of 124 patients with PCS referred to institutions of the French Sarcoma Group (FSG) from 1977 and 2010 were reviewed. RESULTS: Median age was 48.8 years. PCS were poorly-differentiated sarcomas (N=45, 36.3%), angiosarcomas (N=40, 32.3%), leiomyosarcomas (N=16, 12.9%) and others (N=23, 18.6%). At diagnosis, 100 patients (80.6%) were localised and 24 (19.4%) metastatic. Tumours were located in the right (N=47, 38.8%), left atrial cavities (N=45, 37.2%) or encompassed several locations in nine cases (7.4%). Surgery was performed in 81 cases (65.3%). Heart transplant was performed in five patients. Radiotherapy adjuvant (N=18, 14.5%) or alone (N=6, 4.8%) was performed in non-metastatic patients only (N=24, 19.4%). With a median follow-up of 51.2 months, median overall survival (OS) was 17.2 months for the entire cohort, 38.8 months after complete resection versus 18.2 after incomplete resection and 11.2 months in non-resected patients. Radiotherapy was associated with improved progression-free survival (PFS) on multivariate analysis. Chemotherapy was significantly associated with better OS only in non-operated patients but not in operated patients. In non-metastatic patients, surgery (hazard ratio [HR]=0.42, p<0.001), male gender (HR=0.56, p=0.032) was associated with better OS and surgery (HR=0.61; p=0.076), radiotherapy (HR=0.43; p=0.004) and chemotherapy (HR=0.30, p=0.003) improved PFS. CONCLUSION: Only surgical resection is associated with a perspective of prolonged survival. Chemotherapy is associated with a better outcome in non-resected patients.

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TÍTULO / TITLE: - Extranasopharyngeal angiofibroma of the left lower turbinate: A case report.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Nazar R; Naser A; Rubio F; Ortega G

INSTITUCIÓN / INSTITUTION: - Departamento de Otorrinolaringología, Hospital Clínico de la Universidad de Chile, Santiago de Chile, Chile. Electronic address: rnazars@gmail.com.

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TÍTULO / TITLE: - Are There Useful CT Features to Differentiate Renal Cell Carcinoma From Lipid-Poor Renal Angiomyolipoma?

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Yang CW; Shen SH; Chang YH; Chung HJ; Wang JH; Lin AT; Chen KK

INSTITUCIÓN / INSTITUTION: - 1 Division of Urology, Department of Surgery, Taipei Veterans General Hospital, Taipei, Taiwan.
OBJECTIVE. This study was an attempt to identify key CT features that can potentially be used to differentiate between lipid-poor renal angiomyolipoma and renal cell carcinoma (RCC). MATERIALS AND METHODS. We conducted an analysis of patients who received nephrectomy or renal biopsy from 2002 to 2011 with suspected RCC. We included tumors smaller than 7 cm with a completed three-phase CT examination. A radiologist and a urology fellow, blinded to histopathologic diagnosis, recorded the imaging findings by consensus and compared the values for each parameter between lipid-poor angiomyolipoma, RCC subtypes, and RCC as a group. Multivariate logistic regression analysis was performed for each univariate significant feature. RESULTS. The sample in our study consisted of 132 patients with 135 renal tumors, including 51 men (age range, 26-84 years; mean age, 57 years) and 81 women (age range, 29-91 years; mean age, 57 years). These tumors included 33 lipid-poor angiomyolipomas, 54 clear-cell RCC, 31 chromophobe RCC, and 17 papillary RCC. Multivariate analysis revealed four significant parameters for differentiating RCC as a group from lipid-poor angiomyolipoma (angular interface, p = 0.023; hypodense rim, p = 0.045; homogeneity, p = 0.005; unenhanced attenuation > 38.5 HU, p < 0.001), five for clear-cell RCC, two for chromophobe RCC, and one for papillary RCC. Lipid-poor angiomyolipoma and clear-cell RCC showed early strong enhancement and a washout pattern, whereas chromophobe RCC and papillary RCC showed gradual enhancement over time. CONCLUSION. Specific CT features can potentially be used to differentiate lipid-poor renal angiomyolipoma from renal cell carcinoma.

[243]

TITULO / TITLE: Low-grade fibromyxoid sarcoma with nuclear pleomorphism arising in the subcutis of a child.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Sedrak MP; Parker DC; Gardner JM

INSTITUCIÓN / INSTITUTION: Department of Pathology, University of Texas Medical Branch, Galveston, TX, USA.

RESUMEN / SUMMARY: Low-grade fibromyxoid sarcoma (LGFS) represents a rare soft tissue tumor that was first characterized in 1987. LGFS usually presents as a large, deeply situated mass in adults and is characterized by deceptively bland histopathologic features. LGFS is less common in superficial soft tissue and in children. It is distinctly uncommon for LGFS to exhibit nuclear pleomorphism. Herein, we present a case of a 10-year-old male who presented with a subcutaneous back mass that displayed features typical for LGFS as well as scattered large, hyperchromatic and pleomorphic nuclei. The constellation of clinicopathologic features, including the young age of the patient, the small size and superficial location of the tumor and the presence of scattered nuclear pleomorphism are all unusual features for LGFS. Fluorescent in situ hybridization (FISH) with a break-apart probe for FUS revealed the presence of a FUS gene rearrangement confirming the diagnosis of LGFS. This case highlights the importance of maintaining a high index of suspicion for LGFS even in the context of small, superficially-located tumors, pediatric patients or tumors with scattered nuclear pleomorphism.
**TÍTULO / TITLE:** Recurrent adult-type rhabdomyoma: a rare differential diagnosis of "swellings in the masticatory muscle".

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Schlittenbauer T; Rieker R; Amann K; Schmitt C; Wehrhan F; Mitsimponas K; Schlegel KA; Agaimy A

**RESUMEN / SUMMARY:** Rhabdomyomas are rare benign mesenchymal tumors with skeletal muscle differentiation and a predilection for the head and neck area. A 38-year-old man presented with persistent, slowly growing, painless swelling in the left inner cheek for 2(1/2) years. The lesion was detected during routine dental examination and was considered to represent a mucocele. The mass was removed via a transoral surgical approach, followed by a local recurrence 6 months later that was again surgically removed. The patient is alive and well 2 months after last surgery. Adult-type rhabdomyoma is a rare, occasionally recurring, benign mesenchymal tumor that should be included in the differential diagnosis of submucosal swellings in the oral cavity including the masticatory musculature. Adult-type rhabdomyoma of the cheek and masticatory area are exceptionally rare with no more than 3 cases reported to date.

**TÍTULO / TITLE:** Examination of the cutoff value of postchemotherapy increase in tumor volume as a predictor of subsequent oncologic events in stage IIB osteosarcoma.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Jeon DG; Kong CB; Cho WH; Song WS; Cho SH; Choi SW; Lee SY

**INSTITUCIÓN / INSTITUTION:** Department of Orthopedic Surgery, Korea Cancer Center Hospital, Seoul, Korea.

**RESUMEN / SUMMARY:** BACKGROUND: Tumor enlargement after chemotherapy is a predictor of a poor histological response, poor survival, and local recurrence. However, the cutoff point of tumor enlargement for predicting subsequent oncologic events has not been determined. METHODS: We retrospectively reviewed 567 patients who were treated at our institute for stage IIB osteosarcoma. We used receiver operating characteristic (ROC) curve analysis of tumor volume increase for the prediction of subsequent metastasis or local recurrence, and calculated diagnostic indices for different cutoff values. RESULTS: A tumor volume increase of >15% predicted subsequent metastasis or local recurrence with a sensitivity of 64.7%, a specificity of 81.5%, a positive predictive value of 71.6%, and a negative predictive value of 76.1%. Increases in tumor volumes based on this cutoff value were able to predict subsequent oncologic events in all clinical subgroups, except in cases of rare pathologic subtypes. However, for tumors in the proximal humerus, a cutoff value of 25% had optimal predictive value. CONCLUSIONS: This study shows that a cutoff value of 15% for tumor volume increase is useful for predicting subsequent metastasis or local...
recurrence. Our results suggest that tumor enlargement after chemotherapy serves as an easily assessable clinical parameter for risk-adapted therapy. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.

[246]

**TÍTULO / TITLE:** - Ewing Sarcoma of the Acetabulum in Children: A “Growth Plate-based” Surgical Strategy.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1097/BPO.0000000000000112

**AUTORES / AUTHORS:** - Sales de Gauzy J; Lafontan V; Ursei M; Accadbled F

**INSTITUCIÓN / INSTITUTION:** - Service de Chirurgie Orthopedique et Traumatologique, Hopital des Enfants, Toulouse, France.

**RESUMEN / SUMMARY:** - BACKGROUND:: Surgical strategy for Ewing sarcoma (ES) of the pelvis relies on Enneking classification. In adults, in case the acetabulum is involved, excision of the entire acetabulum is needed and often leads to loss of function. In children, the surgeon may adopt a strategy, such as in metaphyseal tumors of long bones where an unaffected growth plate allows a transepiphyseal resection, therefore sparing the joint. METHODS:: We present a “triradiate cartilage strategy” for the excision of ES of the pubic component of the acetabulum, which allows a wide resection while preserving most of the socket. The approach is ilioinguinal. Osteotomies of the iliopectineal and ischio pubisrami are performed, followed by hip arthrotomy and anterior dislocation. Transacetabular resection is achieved using osteotomes, under image intensifier guidance, with no further reconstruction. Cases are presented for 2 boys aged 6.5 and 9.5 years, treated with chemotherapy and transacetabular resection. RESULTS:: Resections were all rated R0. Patient #1 remained asymptomatic, including during sport activities, and had normal hip range of motion at 12-year follow-up. Radiograph demonstrated mild protrusio acetabuli. Patient #2 had no complain at 3-year follow-up with normal hip range of motion, although he presented with an equinus gait. Radiograph demonstrated a well-covered femoral head without medial shift. CONCLUSIONS:: A “growth plate-based” surgical strategy can be adapted to malignant pelvic tumors in skeletally immature children. This technique leaves intact the ilio ischiatic component of the triradiate cartilage, which, according to Ponseti, contributes the most to the growth of the acetabulum. Joint sparing improves the functional result and decreases the risk of complication. LEVEL OF EVIDENCE:: IV.

[247]

**TÍTULO / TITLE:** - A case of lung metastasis in myxoinflammatory fibroblastic sarcoma: analytical review of one hundred and thirty eight cases.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1007/s00264-013-2048-5
RESUMEN / SUMMARY: - PURPOSE: Myxoinflammatory fibroblastic sarcoma (MIFS) is a rare soft tissue tumour first identified at the end of the 1990s. This study presents our experience and literature reviews focusing on risk of recurrence. METHODS: Rizzoli Orthopaedic Institute database and literature were searched for patients with MIFS observed from 1997 to 2012. Data were analysed in a new database. RESULTS: Five patients underwent surgery at our institute, and 133 cases were retrieved from the literature. Not all clinicopathological data were available: 76/138 were men (55 %), median age was 45 [interquartile range (IQR) 34-56] years, median tumour size was three (IQR two to five) centimetres. Common sites of occurrence were hand (24 %), fingers (23 %) and foot (20 %). Pain was present at diagnosis in 14/82 patients (17 %), with a median duration of seven (IQR three to 12) months. Surgery was performed for a suspected benign tumour in 88 patients (74 %). Resection was incomplete in 45/71 cases (63 %); re-excision was performed in 32/45 (71 %). At a median follow-up of 26 months, 26/118 patients (22 %) developed recurrent disease; median time to recurrence was 15 months (IQR seven to 26). Actuarial relapse-free survival (RFS) at one, three and five years was 93 %, 72 % and 67 %, respectively. At univariate analysis, only symptom duration of six months or less was significantly associated with a worse RFS (p = 0.046). Metastatic disease to lymph nodes and/or lungs was observed in four patients (3 %). CONCLUSIONS: Clinicopathological findings confirm the low-grade nature of MIFS. However, local recurrence occurs, and patients may be affected by aggressive forms with a potential for distant metastases. Follow-up is strongly advised.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Chen L; Ding XY; Wang CS; Si MJ; Du LJ; Lu Y
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Ruijin Hospital, Shanghai Jiao Tong University School of Medicine, No.197, Ruijin 2nd Road, Shanghai 200025, China; Department of Radiology, Yueyang Hospital, Shanghai University of Traditional Chinese Medicine, No.110, Ganhe Road, Shanghai 200437, China. Electronic address: chenliang0106@yahoo.com.cn.
RESUMEN / SUMMARY: - OBJECTIVE: Our purpose was, through the comparison of the characteristics of time-intensity curve on triple-phase dynamic contrast-enhanced MRI among groups of giant cell tumor of bone (GCTB), recurrent benign giant cell tumor of bone (RBGCTB), and secondary malignant giant cell tumor of bone (SMGCTB), to find clues to predict the malignant transformation of GCTB. SUBJECTS AND METHODS: 21 patients diagnosed as GCTB were included in this study. All cases took recurrence after intralesional curettage. 9 cases were confirmed as
SMGCTB and 12 cases were confirmed as RBGCTB. Cases were divided into four groups: group A, GCTB (n=9); group B, SMGCTB (n=9); group C, GCTB (n=12); group D, RBGCTB (n=12). Enhancement index (EI) of lesions on DCEMRI was calculated using formula: EI(t)=\[S(t)-S(0)]/S(0), where S(0) was signal intensity of lesion on non-contrast-enhanced T1-weighted images and S(t) was signal intensity of lesion on DCEMRI (t=30, 60, 180s). Enhancement index of each group in each phase was compared using One-Way ANOVA analysis. Slope values of time-intensity curve were compared by the same way. RESULTS: Time-intensity curve of SMGCTB was characterized by a steep upward slope followed by an early and rapid washout phase. Time-intensity curve of GCTB and RBGCTB was characterized by a steep slope followed by a relatively slow washout phase. No significant difference in enhancement index was found in the first phase (p>0.05). There was significant difference in the second and the third phase (p<0.05). Enhancement index of group B (SMGCTB) was smaller. There was no difference in rising slope value (p>0.05). CONCLUSIONS: Dynamic contrast-enhanced MRI appears a helpful method to find new clues to predict malignant transformation of GCTB.

[249]

**TÍTULO / TITLE:** Imaging appearance of renal epithelioid angiomyolipomas.

**RESUMEN / SUMMARY:**


**AUTORES / AUTHORS:** Ryan MJ; Francis IR; Cohan RH; Davenport MS; Weizer A; Hafez K; Kunju LP

**INSTITUCIÓN / INSTITUTION:** From the *Departments of Radiology, daggerUrology, and double daggerPathology, University of Michigan Hospitals, Ann Arbor, MI.

**RESUMEN / SUMMARY:** OBJECTIVE: This study aimed to describe the computed tomographic (CT) imaging appearance of renal epithelioid angiomyolipomas (eAMLs). METHODS: The CT scans and electronic medical records of 8 patients with histologically confirmed eAMLs identified by biopsy and/or surgical excision who had available imaging performed between 1995 and 2012 were reviewed. Preoperative CT imaging appearance, histologic features, and clinical follow-up were recorded for each patient. RESULTS: Macroscopic fat was identified in 3 (38%) of 8 eAMLs on preoperative CT imaging. Seven of the eAMLs demonstrated postcontrast enhancement of greater than 20 Hounsfield units. None of the eAMLs showed evidence of local invasion, vascular involvement, or distant metastases on the initial preoperative CT; however, 1 patient developed local recurrence and another developed distant metastatic disease on follow-up imaging. CONCLUSIONS: Epithelioid angiomyolipomas may or may not demonstrate macroscopic fat. Those with macroscopic fat do not possess any CT imaging characteristics that allow them to be distinguished from typical angiomyolipomas. Epithelioid angiomyolipomas without macroscopic fat are indistinguishable from renal cancers.

[250]
**TÍTULO / TITLE:** - Malignant soft tissue tumors in children.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1016/j.ocln.2013.07.002

**AUTORES / AUTHORS:** - Thacker MM

**INSTITUCIÓN / INSTITUTION:** - Department of Orthopedic Surgery, Nemours - Alfred I duPont Hospital for Children, 1600 Rockland Road, Wilmington, DE 19803, USA.

Electronic address: mihir.thacker@nemours.org.

**RESUMEN / SUMMARY:** - Soft tissue masses are frequently seen in children. Although most are benign or reactive, soft tissue sarcomas (STS), both rhabdomyosarcoma (most common) and non-rhabdo STS, do occur in the extremities. Appropriate evaluation of extremity soft tissue tumors often includes a biopsy as the clinical and imaging features may not be enough to establish a definitive diagnosis. Much needs to be done for improving the treatment of these rare but often devastating sarcomas. Given the small numbers of these cases seen at various centers, collaborative efforts should be made to further our understanding and improve the management of these challenging cases.

*[251]*

**TÍTULO / TITLE:** - Pediatric fronto-orbital cemento-ossifying fibroma: a report of two cases.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - Clin Neuropathol. 2013 Nov 11.

- Enlace al texto completo (gratuito o de pago) 5414/NP300674

**AUTORES / AUTHORS:** - Pant I; Chaturvedi S; Dua RK; Singh G; Kumari R

*[252]*

**TÍTULO / TITLE:** - Differentiation of myxoid liposarcoma by magnetic resonance imaging: a histopathologic correlation.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1177/0284185113508114

**AUTORES / AUTHORS:** - Lowenthal D; Zeile M; Niederhagen M; Fehlberg S; Schnapauff D; Pink D; Tunn P; Reichardt P; Hamm B; Dudeck O

**INSTITUCIÓN / INSTITUTION:** - Department of Radiology and Nuclear Medicine, Otto-von-Guericke University, Magdeburg, Germany.

**RESUMEN / SUMMARY:** - BACKGROUND: Myxoid liposarcomas represent a heterogeneous group of soft tissue tumors in which prognosis is dependent on differentiation. PURPOSE: To identify magnetic resonance imaging (MRI) criteria to distinguish low-grade from high-grade myxoid liposarcomas. MATERIAL AND METHODS: MR images of 30 histologically proven myxoid liposarcomas were retrospectively reviewed. Tumors were evaluated according to size, localization, tumor border, and structure as well as tumor composition. These imaging criteria were correlated with histopathological findings. RESULTS: Nineteen myxoid liposarcomas were histologically classified as low-grade myxoid liposarcomas, whereas 11 were
considered high-grade myxoid liposarcomas. Mean tumor volume of low-grade myxoid liposarcomas (710.1 +/- 960.1 ccm) was significantly smaller as compared to high-grade myxoid liposarcomas (2737.0 +/- 3423.7 ccm; P = 0.04). In addition to necrotic areas, three tumor components - fatty, myxoid, as well as contrast-enhancing non-fatty, non-myxoid - could be identified. The mean fraction of fatty tumor areas in low-grade myxoid liposarcomas was 10 +/- 11% as compared to 6 +/- 4% for high-grade myxoid liposarcomas (P = 0.66). Myxoid components accounted for 88 +/- 16% in low-grade myxoid liposarcomas, but only for 45 +/- 25% in high-grade myxoid liposarcomas (P < 0.0001). The non-fatty, non-myxoid tumor fraction was significantly higher in high-grade myxoid liposarcomas (50 +/- 25%) as compared to low-grade myxoid liposarcomas (2 +/- 9%; P < 0.0001). A proportion of > 5% of this tumor fraction was found to be a precise unique predictor for high-grade myxoid liposarcomas with a sensitivity of 100% and a specificity of 95%. CONCLUSION: Tumor components with contrast-enhancing non-fatty, non-myxoid imaging features were predominantly found in high-grade myxoid liposarcomas, which may histologically resemble round cell clusters.

[253]
TITULO / TITLE: - Dedifferentiated laryngeal chondrosarcoma: combined morphologic and functional imaging with PET/MRI.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Purohit BS; Dulguerov P; Burkhardt K; Becker M
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Geneva University Hospital, Rue Gabrielle PerretGentil 4, CH-1211, Geneva 14, Switzerland.
RESUMEN / SUMMARY: - Chondrosarcoma of the larynx is a rare, low-grade malignancy in terms of histology and clinical behavior. We present an unusual case of laryngeal chondrosarcoma, which developed a large dedifferentiated component on recurrence after primary surgery. The diagnosis of dedifferentiation was suggested in view of the morphological and metabolic findings on hybrid positron emission tomography magnetic resonance imaging (PET/MRI) and was subsequently confirmed surgically. Whole-organ, slice-by slice radiologic-histologic correlation revealed excellent delineation of the well differentiated and dedifferentiated tumor components with PET/MRI. PET/MRI can provide additional functional information to supplement the morphological mapping and histopathology of these tumors.

[254]
TITULO / TITLE: - The paternally imprinted DLK1-GTL2 locus is differentially methylated in embryonal and alveolar rhabdomyosarcomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Schneider G; Bowser MJ; Shin DM; Barr FG; Ratajczak MZ
INSTITUCIÓN / INSTITUTION: - Stem Cell Institute at the James Graham Brown Cancer Center, University of Louisville, Louisville, KY, USA.
RESUMEN / SUMMARY: Parental imprinting of differentially methylated regions (DMRs) contributes to appropriate expression of several developmentally important genes from paternally or maternally derived chromosomes. Rhabdomyosarcoma (RMS) is the most common soft-tissue sarcoma in children and is associated with altered expression of certain parentally imprinted genes. As previously reported, RMS cells display loss of imprinting (LOI) of the DMR at the IGF2-H19 locus, resulting in insulin-like growth factor 2 (IGF2) transcription from both paternally and maternally inherited chromosomes, and overall IGF2 overexpression. As the DLK1-GTL2 locus is structurally similar to the IGF2-H19 locus, the status of parental imprinting of the DLK1-GTL2 locus was studied in RMS. We observed that while both embryonal and alveolar rhabdomyosarcomas (ERMS and ARMS, respectively) show LOI of the DMR at the IGF2-H19 locus, imprinting of the DMR at the DLK1-GTL2 locus varies in association with the histological subtype of RMS. We found that, while ERMS tumors consistently show LOI of the DMR at the DLK1-GTL2 locus, ARMS tumors have erasure of imprinting (EOI) at this locus. These changes in imprinting status of the DLK1-GTL2 locus result in a higher GTL2/DLK1 mRNA ratio in ARMS as compared to ERMS. This difference in imprinting elucidates a novel genetic difference between these two RMS subtypes and may provide a potential diagnostic tool to distinguish between these subtypes.

[255]

TÍTULO / TITLE: - Endobronchial Lipoma: An Unusual Cause of Bronchial Obstruction.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
 AUTORES / AUTHORS: - Sacristan Bou L; Fernandez Robledo E; Pena Blas F
INSTITUCIÓN / INSTITUTION: - Servicio de Neumología, Hospital General de Tomelloso, Tomelloso, Ciudad Real, España. Electronic address: lirios_sacristan@yahoo.es.

[256]

TÍTULO / TITLE: - The Spatiotemporal Dynamics of Scene Gist Recognition.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
 AUTORES / AUTHORS: - Larson AM; Freeman TE; Ringer RV; Loschky LC
RESUMEN / SUMMARY: - Viewers can rapidly extract a holistic semantic representation of a real-world scene within a single eye fixation, an ability called recognizing the gist of a scene, and operationally defined here as recognizing an image’s basic-level scene category. However, it is unknown how scene gist recognition unfolds over both time and space-within a fixation and across the visual field. Thus, in 3 experiments, the current study investigated the spatiotemporal dynamics of basic-level scene categorization from central vision to peripheral vision over the time course of the critical first fixation on a novel scene. The method used a window/scotoma paradigm in which images were briefly presented and processing times were varied using visual masking.
The results of Experiments 1 and 2 showed that during the first 100 ms of processing, there was an advantage for processing the scene category from central vision, with the relative contributions of peripheral vision increasing thereafter. Experiment 3 tested whether this pattern could be explained by spatiotemporal changes in selective attention. The results showed that manipulating the probability of information being presented centrally or peripherally selectively maintained or eliminated the early central vision advantage. Across the 3 experiments, the results are consistent with a zoom-out hypothesis, in which, during the first fixation on a scene, gist extraction extends from central vision to peripheral vision as covert attention expands outward. (PsycINFO Database Record © 2013 APA, all rights reserved).
[258]

**TÍTULO / TITLE:** Peritoneal debulking/intraperitoneal chemotherapy-non-sarcoma.

**RESUMEN / SUMMARY:** [Enlace al Resumen / Link to its Summary](#)


**AUTORES / AUTHORS:** Kelly KJ; Nash GM

**INSTITUCIÓN / INSTITUTION:** Department of Surgery, Memorial Sloan-Kettering Cancer Center, New York, New York.

**RESUMEN / SUMMARY:** The combination of cytoreductive surgery (CRS) and intraperitoneal chemotherapy (IPC) is widely practiced for appendiceal, colorectal, gastric, and ovarian cancers with isolated peritoneal metastasis as well as for primary peritoneal cancer. The aim of this report is to explain the rationale and available techniques for CRS and IPC, and to highlight disease-specific considerations that should be taken into account when evaluating potential candidates for CRS and IPC. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.

[259]

**TÍTULO / TITLE:** Pulmonary inflammatory myofibroblastic tumor and IgG4-related inflammatory pseudotumor: a diagnostic dilemma.

**RESUMEN / SUMMARY:** [Enlace al Resumen / Link to its Summary](#)


**AUTORES / AUTHORS:** Bhagat P; Bal A; Das A; Singh N; Singh H

**INSTITUCIÓN / INSTITUTION:** Department of Histopathology, Post Graduate Institute of Medical Sciences & Research (PGIMER), Sector -12, Chandigarh, 160012, India.

**RESUMEN / SUMMARY:** IgG4-related inflammatory pseudotumor (IPT) and inflammatory myofibroblastic tumor (IMT) share morphological features like a prominent fibroblastic/myofibroblastic proliferation and the presence of inflammatory cells. Since IPT is managed conservatively and IMT is treated by surgical excision, it is important to differentiate these two lesions. The aim of this study is to highlight morphological and immunohistochemical features that distinguish IPT and IMT. Clinicopathological characteristics of cases diagnosed as pulmonary IPT or IMT from 1997 to 2013 were reviewed. The histological features were studied on hematoxylin and eosin-stained sections. Immunohistochemistry was done for IgG, IgG4, ALK-1, SMA, desmin, and CD34 for classification into IPT and IMT. Of the ten patients, seven were male and the age ranged from 4 to 58 years. The tumor size ranged from 1.5 to 4.0 cm in diameter. Histologically, proliferation of bland-looking spindle cells along with fibrosis and an inflammatory infiltrate comprising of lymphocytes and plasma cells were the common morphological features of both lesions. The spindle cell proliferation was more marked in IMT whereas lymphoplasmacytoid infiltrate was more prominent in IPT. Obstructive phlebitis was observed only in cases of IPT. IgG4 expression was noted in IPT, and the number of IgG4-positive plasma cells and the ratio of IgG4+/IgG+ plasma cells were significantly lower in IMT than in IgG4-related IPT. Expression of anaplastic lymphoma kinase (ALK) was observed only in IMT, but not in IgG4-related
IPT. The proportion of proliferating spindle cells, lymphoplasmacytic infiltrate, obstructive phlebitis, IgG4+ plasma cells and the ratio of IgG4+/IgG+ plasma cells, and ALK expression are helpful in differentiating these morphologically similar but biologically different lesions, which require different treatment modalities.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Mangham DC; Kindblom LG
INSTITUCIÓN / INSTITUTION: - Department of Musculoskeletal Pathology, Royal Orthopaedic Hospital NHS Trust, Robert Aitken Institute of Clinical Research and School of Cancer Sciences, Medical School, Birmingham University, Birmingham, UK; Department of Musculoskeletal Pathology, Robert Jones and Agnes Hunt Orthopaedic Hospital NHS Trust, Oswestry, UK.
RESUMEN / SUMMARY: - Soft tissue tumours that rarely metastasize have been afforded their own subcategory in recent WHO classifications. This review discusses the nature of these tumours and the difficulty in constructing useful simple classifications for heterogeneous and complex groups of tumours. We also highlight the specific rarely metastasizing soft tissue tumours that have been recently added to the WHO classification (phosphaturic mesenchymal tumour, pseudomyogenic haemangioendothelioma) and those entities where there have been recent important defining genetic discoveries (myxoinflammatory fibroblastic sarcoma, solitary fibrous tumour, myoepitheliomas).

[261] TÍTULO / TITLE: - Round cell sarcomas beyond Ewing: emerging entities.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Antonescu C
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Memorial Sloan-Kettering Cancer Center, New York, NY, USA.
RESUMEN / SUMMARY: - Primitive small blue round cell tumours (SBRCT) of childhood and young adults have been problematic to diagnose and classify. Diagnosis is also complicated in cases with atypical morphology, aberrant immunoprofiles and unusual clinical presentations. Even with the increased use of ancillary techniques in archival material, such as immunohistochemistry and molecular/genetic methods, a proportion of these tumours cannot be subclassified into specific histological types. A subset of tumours resembling microscopically the Ewing sarcoma family of tumours (EFT), being composed of primitive small round cells and occurring in paediatric or young adult age groups, remain unclassified, being negative for EWSR1, SS18(SYT), DDIT3(CHOP) and FOXO1(FKHR) gene rearrangements by FISH/RT-PCR. A small number of cases sharing the undifferentiated EFT appearance have been characterized recently carrying BCOR-CCNB3 or CIC- DUX4 fusions. However, based on the somewhat
limited number of cases, it remains unclear if these newly defined genetic entities belong to any of the pre-existing clinicopathological disorders or represent altogether novel conditions. This review presents the latest molecular findings related to these SBRCTs, beyond the common EWSR1-ETS fusions. Specific attention has been paid to morphological features not associated typically with classic EFT, and the value of ancillary tests that can be applied when dealing with EWSR1-negative SBRCTs is discussed.

[262]
TÍTULO / TITLE: - Gastrointestinal stromal tumours: from KIT to succinate dehydrogenase.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Doyle LA; Hornick JL
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Brigham and Women’s Hospital, Harvard Medical School, Boston, MA, USA.
RESUMEN / SUMMARY: - The discovery of activating mutations in the tyrosine kinase receptor genes KIT and PDGFRA has led to the development of effective targeted therapies for gastrointestinal stromal tumours (GISTs). Specific genotypes, in part, predict the response to treatment with tyrosine kinase inhibitors. However, ~10% of GISTs lack such mutations (often referred to as ‘wild-type’ GISTs). Recent insights into the biology of ‘wild-type’ GISTs have resulted in clinically significant subclassification of this heterogeneous group of tumours, a large subset of which are now known to represent succinate dehydrogenase-deficient GISTs. Recognition of this distinctive class of tumours has critical implications for prognosis, therapy, clinical follow-up, and genetic counselling. Other uncommon genetic groups include neurofibromatosis type I-associated and BRAF-mutant GISTs. This review provides an update on the diagnosis and pathogenesis of these less common classes of GISTs, summarizes the clinical and pathological features associated with particular genotypes, and discusses mechanisms of resistance to targeted therapies.

[263]
TÍTULO / TITLE: - Syndrome-associated soft tissue tumours.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Coffin CM; Davis JL; Borinstein SC
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Microbiology, and Immunology, Vanderbilt University School of Medicine, Nashville, TN, USA.
RESUMEN / SUMMARY: - Soft tissue neoplasms may be associated with a variety of genetic disorders and malformation syndromes, especially when they arise in children, adolescents and early adulthood. This review summarizes the principal histopathological types of soft tissue tumours which occur in various syndromes, with an emphasis on pathological features, genetic aspects and considerations for the diagnostic pathologist.
The evolving classification of soft tissue tumours - an update based on the new 2013 WHO classification.

The new World Health Organization (WHO) classification of soft tissue tumours was published in early 2013, almost 11 years after the previous edition. While the number of newly recognized entities included for the first time is fewer than that in 2002, there have instead been substantial steps forward in molecular genetic and cytogenetic characterization of this family of tumours, leading to more reproducible diagnosis, a more meaningful classification scheme and providing new insights regarding pathogenesis, which previously has been obscure in most of these lesions. This brief overview summarizes changes in the classification in each of the broad categories of soft tissue tumour (adipocytic, fibroblastic, etc.) and also provides a short summary of newer genetic data which have been incorporated in the WHO classification.

Failure of echocardiography to detect a large left atrial myxoma.


Cardiac angiosarcoma: A paradigmatical case?

[267]
**TÍTULO / TITLE:** - Plexiform Fibromyxoma: Report of Two Pediatric Cases, Including the First Example in the Esophagus.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**REVISTA / JOURNAL:** - Pediatr Dev Pathol. 2013 Oct 25.
   - Enlace al texto completo (gratuito o de pago) 2350/13-09-1373-OA.1
**AUTORES / AUTHORS:** - Duckworth LV; Gonzalez RS; Martelli M; Liu C; Coffin CM; Reith J
**INSTITUCIÓN / INSTITUTION:** - a University of Florida, Pathology, Immunology, and Laboratory Medicine.
**RESUMEN / SUMMARY:** - Abstract Plexiform fibromyxoma is a distinctive mesenchymal neoplasm usually arising in the gastric antrum. We report two cases of this entity in pediatric patients, including the first case arising in the esophagus. The patients were a 16-year-old female who presented with chest pain and was found on CT scan to have a mid-esophageal mass at the level of the carina, and an 11-year-old female with a gastric mass. Both patients underwent surgical resection of their tumors, which histologically exhibited a plexiform growth pattern with multiple nodules in the muscularis propria and infiltrative borders. The nodules were composed of a rich myxoid stroma with bland uniform spindle cells, no mitoses or necrosis, and delicate blood vessels in the background. Immunohistochemical studies demonstrated the tumor cells were immunoreactive with smooth muscle actin, and negative with S-100, CD34, desmin, and c-kit. We report the first case of plexiform fibromyxoma originating in the esophagus, emphasize its occurrence in pediatric patients, and review the related literature.

[268]
**TÍTULO / TITLE:** - Pancreas-sparing duodenectomy for gastrointestinal stromal tumor.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
   - Enlace al texto completo (gratuito o de pago) 1016/j.amjsurg.2013.05.009
**AUTORES / AUTHORS:** - Yamashita S; Sakamoto Y; Saiura A; Yamamoto J; Kosuge T; Aoki T; Sugawara Y; Hasegawa K; Kokudo N
**INSTITUCIÓN / INSTITUTION:** - Hepato-Biliary-Pancreatic Surgery Division, Department of Surgery, Graduate School of Medicine, University of Tokyo, 7-3-1 Hongo, Bunkyo-ku, Tokyo 113-8655, Japan.
**RESUMEN / SUMMARY:** - BACKGROUND: Pancreas-sparing duodenectomy (PSD) is a promising alternative procedure to pancreaticoduodenectomy for the treatment of duodenal tumors with low-grade malignant behavior. METHODS: Between March 2003 and September 2012, PSD was performed in 7 patients with a gastrointestinal stromal tumor (GIST) in the second (n = 5) or third (n = 2) portions of the duodenum. The short- and long-term outcomes of treatment were analyzed in all patients. RESULTS: The median blood loss was 160 mL, and the median operative time was 315 minutes. No pancreatic leakage or perioperative mortality occurred. Surgical margins were negative in all cases. All patients were alive at the median follow-up time of 42 months after PSD. The recurrence-free 5-year survival rate was 53% in all patients. Hepatic
metastases developed in 2 of the 5 patients with high- or intermediate-grade risks at the time of diagnosis. Hepatic resection was performed, and imatinib mesylate was administered in the 2 cases. CONCLUSIONS: Good short- and long-term outcomes and surgical curability were observed in patients treated with PSD for duodenal GIST.
[272]
**Título / Title:** Leiomyoma Presenting as a Massive Calcified Circumferential Esophageal Mass.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1016/j.athoracsur.2013.01.097

**Autores / Authors:** Walters DM; Vaughn NH; Isbell JM; Jeffus SK; Atkins KA; Sauer BG; Jones DR

**Institución / Institution:** Department of Surgery, University of Virginia, Charlottesville, Virginia.

**Resumen / Summary:** Esophageal leiomyoma is the most common benign esophageal neoplasm and often presents as an incidental finding or with nonspecific symptoms such as dysphagia or chest pain. Surgical enucleation is the mainstay of treatment and may be accomplished using both open and thoracoscopic approaches. We present a case of a 57-year-old man who presented with a massive circumferential calcified leiomyoma.

[273]

**Título / Title:** Huge inflammatory myofibroblastic tumor of pleura with concomitant nuchal fibroma.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1016/j.athoracsur.2013.01.082

**Autores / Authors:** Dongel I; Yazkan R; Duman L; Ozturk O; Kapucuoglu FN

**Institución / Institution:** Department of Thoracic Surgery, Suleyman Demirel University, Medical Faculty, Isparta, Turkey. Electronic address: drdongel@hotmail.com.

**Resumen / Summary:** Inflammatory myofibroblastic tumor (IMT) is a rare benign neoplasm. It is a challenging disease because the symptoms and radiologic findings are diverse and nonspecific. Although pulmonary IMT is the most common form, pleural origin is an extremely rare clinical entity. Nuchal fibroma (NF) is another rare benign neoplasm. We report herein a case of pleural IMT with concomitant NF in a 15-year-old girl. To the best of our knowledge, this is the first report suggesting an association between IMT and NF, and our case had the largest reported intrathoracic
IMT. Moreover, we found a possible association between IMT and increased CA-125 levels.

[274]
TÍTULO / TITLE: - Sarcomatous degeneration in fibrous dysplasia of the rib cage.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
● Enlace al texto completo (gratuito o de pago) 1016/j.athoracsur.2013.04.131
AUTORES / AUTHORS: - Van Rossem C; Pauwels P; Somville J; Camerlinck M; Bogaerts P; Van Schil PE
INSTITUCIÓN / INSTITUTION: - Department of Thoracic and Vascular Surgery, Antwerp University Hospital, Edegem, Belgium.
RESUMEN / SUMMARY: - Malignant degeneration in fibrous dysplasia is a rare occurrence. Most cases are reported in polyostotic fibrous dysplasia with predisposition of the femur, tibia, maxilla, and mandible. The most commonly observed malignant tumors are osteosarcoma, fibrosarcoma, and chondrosarcoma. We describe a case of a low-grade osteosarcoma occurring in polyostotic fibrous dysplasia of the rib cage in a 50-year-old man.

[275]
TÍTULO / TITLE: - Clear cell chondrosarcoma: Cytologic findings in six cases.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
● Enlace al texto completo (gratuito o de pago) 1002/dc.23043
AUTORES / AUTHORS: - Jiang XS; Pantanowitz L; Bui MM; Esther R; Budwit D; Dodd LG
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Duke University Medical Center, Durham, North Carolina.
RESUMEN / SUMMARY: - Clear cell chondrosarcoma (CCCS) is a rare variant of chondrosarcoma characterized, in most instances, by indolent behavior and a long interval to progression of disease. CCCS commonly occurs in adult individuals and has a proclivity for the epiphysis of long bones, although it has been reported in other sites. This lesion is difficult to diagnose preoperatively. Factors contributing to difficulty in recognizing this lesion include its relative scarcity as well as its tendency to be confused with other lesions on imaging studies. In the following, we report six cases of CCCS initially diagnosed by fine needle aspiration and/or touch preparations of needle biopsy samples. The cytologic features identified include large, plasmacytoid cells with foamy cytoplasm as well as extracellular chondroid type matrix material. Definitive diagnosis was made in each case by recognizing the "clear cell" nature of the tumor on cell block material. Diagn. Cytopathol 2013. © 2013 Wiley Periodicals, Inc.
TÍTULO / TÍTULO: - Fine-Needle aspiration cytology of primary renal angiosarcoma with histopathologic and immunocytochemical correlation: A Case Report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Grapsa D; Sakellariou S; Politi E
INSTITUCIÓN / INSTITUTION: - Cytopathology Department, Areteion University Hospital, Athens, Greece.
RESUMEN / SUMMARY: - Primary renal angiosarcoma is an extremely rare neoplasm, with fewer than 28 cases reported thus far in the English literature. We report for the first time the cytomorphology and immunocytochemistry of this tumor in liquid-based (ThinPrep) fine-needle aspiration (FNA) samples in correlation with the conventional cytologic and histopathologic findings. Conventional smears showed pleomorphic tumor cells focally arranged in structures suggesting anastomosing vascular channels, while ThinPrep smears were less cellular with fewer and smaller tumor cells arranged in clusters or rosette-like formations. Immunocytochemical staining demonstrated positive results for vimentin, CD31, and CD34 and negative staining for epithelial markers, thus supporting the diagnosis of a mesenchymal tumor of vascular origin. The diagnosis of primary renal angiosarcoma was established after histopathologic evaluation of a metastatic liver nodule. The cytological differential diagnosis of this neoplasm and the utility of the ThinPrep method as a diagnostic adjunct to conventional FNA cytology are further discussed. Diagn. Cytopathol. 2013. © 2013 Wiley Periodicals, Inc.

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[277]
TÍTULO / TITLE: - Future directions for pediatric and young adult bone sarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Finney J; Kent PM; Batus M

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[278]
TÍTULO / TITLE: - 18F-FDG PET/CT Imaging in Primary Cardiac Angiosarcoma: Diagnosis and Follow-Up.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Tan H; Jiang L; Gao Y; Zeng Z; Shi H
INSTITUCIÓN / INSTITUTION: - From the Departments of *Nuclear Medicine, and daggerRadiation Oncology, Zhongshan Hospital, Fudan University, Shanghai, China.
RESUMEN / SUMMARY: - A 40-year-old woman presented with recurrent chest tightness and shortness of breath for 10 days. The echocardiogram and MRI revealed
a mass in the right atrium. FDG PET/CT was performed for further evaluation. The images demonstrated abnormally increased activity in the right atrial mass. The result of pathological examination after the surgical removal of the mass was consistent with angiosarcoma. Despite of receiving aggressive therapy including radiation, the patient had recurrent and metastatic malignancy 8 months later.

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**TÍTULO / TITLE:** - Beyond the GIST: mesenchymal tumors of the stomach.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Kang HC; Menias CO; Gaballah AH; Shroff S; Taggart MW; Garg N; Elsayes KM

**INSTITUCIÓN / INSTITUTION:** - Departments of Diagnostic Radiology and Pathology, University of Texas M.D. Anderson Cancer Center, 1515 Holcombe Blvd, Unit 1473, Houston, TX 77030; Department of Radiology, Mayo Clinic, Tucson, Ariz.

**RESUMEN / SUMMARY:** - Intramural gastric masses arise in the wall of the stomach (generally within the submucosa or muscularis propria), often with intact overlying mucosa. These tumors are typically mesenchymal in origin and have overlapping radiologic appearances. A combination of features such as location, attenuation, enhancement, and growth pattern may suggest one diagnosis over another. Gastrointestinal stromal tumors (GISTs) account for the majority of intramural tumors and can vary widely in appearance, from small intraluminal lesions to exophytic masses that protrude into the peritoneal cavity, commonly with areas of hemorrhage or necrosis. A well-circumscribed mass measuring -70 to -120 HU is a lipoma. Leiomyomas usually manifest as low-attenuation masses at the gastric cardia. Homogeneous attenuation is a noteworthy characteristic of schwannomas, particularly for larger lesions that might otherwise be mistaken for GISTs. A hypervascular mass in the antrum is a common manifestation of glomus tumors. Hemangiomas are also hypervascular but often manifest in childhood. Inflammatory fibroid polyps usually arise as a polypoid mass in the antrum. Inflammatory myofibroblastic tumors are infiltrative neoplasms with a propensity for local recurrence. Plexiform fibromyxomas are rare, usually antral tumors. Carcinoid tumors are epithelial in origin, but often submucosal in location, and therefore should be distinguished from other intramural lesions. Multiple carcinoid tumors are associated with hypergastrinemia, either in the setting of chronic atrophic gastritis or Zollinger-Ellison syndrome. Sporadic solitary carcinoid tumors not associated with hypergastrinemia have a higher rate of metastasis. Histopathologic analysis, including immunohistochemistry, is usually required for diagnosis of intramural masses.

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**TÍTULO / TITLE:** - Development of enterohepatic fistula after embolization in ileal gastrointestinal stromal tumor: A case report.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
Enlace al texto completo (gratuito o de pago) 3748/wjg.v19.i43.7816

AUTORES / AUTHORS: - Lee YH; Koo JS; Jung CH; Chung SY; Lee JJ; Kim SY; Hyun JJ; Jung SW; Choung RS; Lee SW; Choi JH

INSTITUCION / INSTITUTION: - Yun Ho Lee, Ja Seol Koo, Chang Ho Jung, Sang Yoon Chung, Jae Joong Lee, Seung Young Kim, Jong Jin Hyun, Sung Woo Jung, Rok Seon Choung, Sang Woo Lee, Jai Hyun Choi, Division of Gastroenterology, Department of Internal Medicine, Korea University Ansan Hospital, 516 Gojan-dong, Danwon-gu, Ansan-si, Gyeonggi-do 425-707, South Korea.

RESUMEN / SUMMARY: - Gastrointestinal stromal tumor (GIST) is a rare mesenchymal tumor of the gastrointestinal tract that has been associated with the formation of fistulas to adjacent organs in few case reports. However, GIST with enterohepatic fistula has not been reported. Here we report the case of an enterohepatic fistula that occurred after embolization of a liver mass originating in the distal ileum. An 87-year-old woman was hospitalized for melena. On initial conventional endoscopy, a bleeding focus in the gastrointestinal tract was not found. Because of massive hematochezia, enteroscopy was performed through the anus. A protruding, ulcerative mass was found in the distal ileum that was suspected to be the source of the bleeding; a biopsy sample was taken. Electrocoagulation was not successful in controlling the bleeding; therefore, embolization was performed. After embolization, the patient developed a high fever and severe abdominal tenderness with rebound tenderness. Follow-up abdominopelvic computed tomography revealed an enterohepatic fistula between the liver and distal ileum. The fistula was treated surgically by segmental resection of the distal ileum and unlooping of the liver mass.
Enlace al texto completo (gratuito o de pago) 1007/s00247-013-2817-8
AUTORES / AUTHORS: - Calvo-García MA; Lim FY; Stanek J; Bitters C; Kline-Fath BM
INSTITUCIÓN / INSTITUTION: - Department of Radiology, MLC 5031, Cincinnati Children’s Hospital Medical Center, 3333 Burnet Ave., Cincinnati, OH, 45229-3026, USA, maria.calvo@cchmc.org.
RESUMEN / SUMMARY: - We present a prenatal case of congenital peribronchial myofibroblastic tumor referred as a congenital pulmonary airway malformation (CPAM) with hydrops and polyhydramnios at 30 weeks’ gestational age. US and fetal MRI findings did not fit with the referral diagnosis, raising the possibility of intrinsic lung tumor. Fetal hydrops worsened and the baby was successfully delivered by ex utero intrapartum treatment (EXIT) to resection at 31 weeks’ gestational age. To the best of our knowledge, this is a unique case of congenital peribronchial myofibroblastic tumor that underwent comprehensive prenatal evaluation and EXIT procedure with good outcome.

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[TÍTULO / TITLE]: - Left atrial metastasis of Hurthle-cell thyroid carcinoma mimicking myxoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Giovanella L; Treglia G; Ceriani L; Weidner S; Perriard U; Bongiovanni M
INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine and PET/CT Centre, Oncology Institute of Southern Switzerland, Via Ospedale, 12, 6500, Bellinzona, Switzerland.

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[TÍTULO / TITLE]: - Structural and molecular features of the endomyometrium in endometriosis and adenomyosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Benagiano G; Brosens I; Habiba M
INSTITUCIÓN / INSTITUTION: - Department of Gynaecology, Obstetrics and Urology, Sapienza University, 00161 Rome, Italy.
RESUMEN / SUMMARY: - BACKGROUNDAdenomyosis and endometriosis were initially described as ‘adenomyoma’. When the retrograde menstruation theory became widely accepted to explain the pathogenesis of endometriosis, since it does not explain adenomyosis, the two conditions came to be seen as distinct entities. However, emerging evidence suggests that both diseases may be linked to changes in the inner portion of the myometrium. In addition, similar anomalies were found in the eutopic endometrium of the two conditions and the debate has re-opened. A common origin for both adenomyosis and endometriosis would have relevance not only for understanding uterine function and pathophysiology, but also for clinical management and treatment.METHODSThe Scopus and Medline databases were searched for all
original articles published in English up to the end of 2012. Search terms included ‘adenomyosis'; ‘endometriosis'; ‘endometrium'; ‘eutopic endometrium'; ‘inner myometrium'; ‘junctional zone'. Special attention was paid to articles comparing features of eutopic endometrium in the two conditions.

**RESULTS**

A number of similarities exist between adenomyosis and endometriosis and, by using magnetic resonance and laparoscopy, it was found that, at least in some subgroups, the two conditions often coexist. In both situations the inner myometrium (or junctional zone) is altered, although alterations are much more marked in adenomyosis where a thickness >12 mm is today considered sufficient for diagnosis. Research has shown differences between the eutopic endometrium of women with both diseases when compared with controls. There is an immune dysfunction and there are alterations of adhesion molecules, cell proliferation and apoptosis. An increase in cytokines and inflammatory mediators has also been observed. Finally, the presence of oxidative stress and anomalies in free-radical metabolism may alter uterine receptivity. When the two conditions were compared, dissimilarities were also observed in the extent of apoptosis inhibition and in the expression of some inflammatory mediators. It is not clear if observed differences are primarily related to presenting symptoms. Finally, both conditions are steroid dependent and research suggests a role for epigenetic mechanisms. The analysis indicates that much of the published research may have been influenced by the method of diagnosis and/or has not been controlled for the presenting symptoms, the concomitant presence of both diseases or full consideration of fluctuations within cycle phase.

**CONCLUSIONS**

It is difficult to draw firm conclusions from existing evidence since major diagnostic limitations still exist and there is a systematic bias in clinical presentation. In addition, scanty information is available on the natural history of endometriosis and no studies exist on the natural history of adenomyosis. Notwithstanding these limitations, a number of similarities, but also some differences have been found between the eutopic endometrium in the two diseases. These findings need to be taken with considerable caution as the few instances where the research was repeated yielded conflicting results.

[285]

**TÍTULO / TITLE:** - Thrombotic thrombocytopenic purpura and cardiac papillary fibroelastoma: a ‘unique coexistence’.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Ramakrishnan Geethakumari P; Rubin A; Varadi G

**INSTITUCIÓN / INSTITUTION:** - aDepartment of Internal Medicine bDivision of Cardiology cDivision of Hematology-Oncology, Albert Einstein Medical Center, Philadelphia, Pennsylvania, USA.

**RESUMEN / SUMMARY:** - Thrombotic thrombocytopenic purpura (TTP), a complex thrombotic microangiopathy, remains an evolving enigma. A 49-year-old African-American woman presented with acute left hemiplegia, an ischemic cerebrovascular accident involving the right middle cerebral artery. Sequential appearance of thrombocytopenia and evidence of microangiopathic haemolysis led to the diagnosis of
acquired idiopathic autoimmune TTP. This was managed with plasma exchange (PEX) and steroids. Early haematologic relapse within a month was managed with the addition of rituximab attaining sustained remission. The patient presented 3 years later with acute confusion and expressive aphasia due to multiple infarcts involving the left parieto-occipital cortex. Transoesophageal echocardiography demonstrated a pedunculated 6 mm mitral valvular mass consistent with a papillary fibroelastoma. Anticoagulation was instituted and the patient was continued on therapeutic oral warfarin. A haematologic relapse of TTP eventually emerged and was managed with PEX, steroids and rituximab. This vignette demonstrates several dilemmas in the clinical presentation, diagnosis and management of TTP in current day practice. Rituximab has adjuvant benefits to PEX and is being investigated as potential first-line therapy. Monitoring ADAMTS13 activity and inhibitor titre, as in our case, prove to have prognostic significance. Cardiac fibroelastomas are rare benign cardiac tumours usually arising from valvular endocardium with thromboembolic potential. One of the proposed mechanisms of origin of these masses is organizing thrombi in the setting of endocardial injury and inflammation questioning a possible link to thrombotic microangiopathy. To the best of our knowledge, this is the first report of this unique coexistence.

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TÍTULO / TITLE: Laryngeal Chondrosarcoma of the Arytenoid Cartilage Presenting as Bilateral Vocal Fold Immobility: A Case Report and Literature Review.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Hu R; Xu W; Liu H; Chen X
INSTITUCIÓN / INSTITUTION: Department of Otorhinolaryngology-Head and Neck Surgery, Beijing Tongren Hospital, Capital Medical University, Beijing, China.
RESUMEN / SUMMARY: OBJECTIVES: To describe an atypical case of laryngeal chondrosarcoma of arytenoid cartilage presenting as bilateral vocal fold immobility and to avoid potential missed diagnosis. METHODS: Our case study included a detail history, physical and radiological examination, laryngeal electromyography (LEMG), and surgical treatment and pathology analysis. We compared it with the previously discussed cases of chondrosarcoma of arytenoid cartilage in the literature. RESULTS: Chondrosarcomas of the arytenoid cartilage is rare, and to date only approximately 10 cases have been reported. We reported a case of a 51-year-old man with 1 month of persistent dyspnea presenting with bilateral vocal fold immobility without neoplasms in larynx. The LEMG showed no obvious abnormality. The cervical-enhanced computed tomography (CT) found no significant signs of a mass except for localized high-density areas in arytenoid cartilage. Right arytenoidectomy and biopsy were performed under general anesthesia with CO2 laser with the pathological diagnosis of chondroma. A total laryngectomy was performed 2 years later, and low-grade chondrosarcoma was the final diagnosis. CONCLUSIONS: Laryngeal chondrosarcomas of the arytenoid cartilage are rare. It is easily neglected, especially in those cases presenting with idiopathic vocal fold immobility without any obvious signs of neoplasms. The LEMG
TÍTULO / TITLE: - Hyperexpression of HOXC13, located in the 12q13 chromosomal region, in welldifferentiated and dedifferentiated human liposarcomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Cantile M; Galletta F; Franco R; Aquino G; Scognamiglio G; Marra L; Cerrone M; Malzone G; Manna A; Apice G; Fazioli F; Botti G; De Chiara A
INSTITUCIÓN / INSTITUTION: - Division of Pathology, Istituto Nazionale Tumori ‘Fondazione G. Pascale’-IRCCS, 80131 Naples, Italy.

RESUMEN / SUMMARY: - Liposarcoma (LPS) is the most common soft tissue neoplasm in adults and is characterized by neoplastic adipocyte proliferation. Some subtypes of LPSs show aberrations involving the chromosome 12. The most frequent are t(12;16) (q13;p11) present in more than 90% of myxoid LPSs and 12q13-15 amplification in well-differentiated and dedifferentiated LPSs. In this region, there are important oncogenes such as CHOP (DDIT3), GLI, MDM2, CDK4, SAS, HMGA2, but also the HOXC locus, involved in development and tumor progression. In this study, we evaluated the expression of HOXC13, included in this chromosomal region, in a series of adipocytic tumors. We included 18 well-differentiated, 4 dedifferentiated, 11 myxoid and 6 pleomorphic LPSs as well as 13 lipomas in a tissue microarray. We evaluated the HOXC13 protein and gene expression by immunohistochemistry and quantitative PCR. Amplification/translocation of the 12q13-15 region was verified by FISH. Immunohistochemical HOXC13 overexpression was observed in all well-differentiated and dedifferentiated LPSs, all characterized by the chromosome 12q13-15 amplification, and confirmed by quantitative PCR analysis. In conclusion, our data show a deregulation of the HOXC13 marker in welldifferentiated and dedifferentiated LPSs, possibly related to 12q13-15 chromosomal amplification.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Baffert S; Italiano A; Pierron G; Traore MA; Rapp J; Escande F; Ghnassia JP; Terrier P; Voegeli AC; Ranchere-Vince D; Coindre JM; Pedeutour F
INSTITUCIÓN / INSTITUTION: - Institut Curie, departement de sante publique, 26, rue d'Ulm, 75005 Paris, France.

RESUMEN / SUMMARY: - Sarcomas represent a complex and heterogeneous group of rare malignant tumors and their correct diagnosis is often difficult. Recent molecular biological techniques have been of great diagnostic use and there is a need to assess the cost of these procedures in routine clinical practice. Using prospective and
observational data from eight molecular biology laboratories in France, we used "microcosting" method to assess the cost of molecular biological techniques in the diagnosis of five types of sarcoma. The mean cost of fluorescence in situ hybridization (FISH) was 318 euro (273-393) per sample; mean reverse transcription polymerase chain reaction (RT-PCR) cost ranged from 300 euro (229-481) per formalin-fixed, paraffin-embedded specimen to 258 euro (213-393) per frozen specimen; mean quantitative polymerase chain reaction (Q-PCR) cost was 184 euro (112-229) and mean CGH-array cost was 332 euro (329-335). The cost of these recently implemented techniques varied according to the type of sarcoma; the method of tissue collection and local organizational factors including the level of local expertise and investment. The cost of molecular diagnostic techniques needs to be balanced against their respective performance.

[289]
**TÍTULO / TITLE:** - Long-term conservative management of a giant cardiac fibroma.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Massin M; Ould AF; Jacquemart C; Damry N
**INSTITUCIÓN / INSTITUTION:** - Division of Pediatric Cardiology, Queen Fabiola Children’s University Hospital, Free University of Brussels (ULB), Belgium.

**RESUMEN / SUMMARY:** - A giant cardiac fibroma was discovered during evaluation for a soft systolic murmur in an asymptomatic 2-week-old girl. Echocardiography and magnetic resonance imaging showed a large intraventricular solid mass developed at the expense of the left ventricular lateral wall. Tumour progression resulted in failure to thrive and ventricular arrhythmia between 2 and 18 months of age. At that time, complete resection seemed unfeasible and conservative management with heart failure and antiarrhythmic medications was chosen. All drugs were discontinued when the patient was 5 years old. Since that time, the mass is stable and the patient is strictly asymptomatic. Conservative strategy seems to be acceptable in selected cases but close follow-up is mandatory.

[290]
**TÍTULO / TITLE:** - Regulation of onco and tumor suppressor MiRNAs by mTORC1 inhibitor PRP-1 in human chondrosarcoma.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Galoian KA; Guettouche T; Issac B; Qureshi A; Temple HT
**INSTITUCIÓN / INSTITUTION:** - Department of Orthopaedics, Miller School of Medicine, University of Miami, 1600 NW 10th Avenue, Suite 8012 (D27), Miami, FL, 33136, USA, kgaloian@med.miami.edu.

**RESUMEN / SUMMARY:** - Metastatic chondrosarcoma of mesenchymal origin is the second most common bone malignancy and does not respond either to chemotherapy or radiation; therefore, the search for new therapies is relevant and urgent. This study aimed to reveal the comparative analysis of miRNAs and their targets in human JJ012
chondrosarcoma cell line between control and experimental samples, treated with mTORC1 inhibitor, cytostatic antiproliferative proline-rich polypeptide (PRP-1). Examination of tumor-specific microRNA expression profiles has revealed widespread deregulation of these molecules in diverse cancers. It was reported that microRNAs can function as novel biomarkers for disease diagnostics and therapy, as well as a novel class of oncogenes and tumor suppressor genes. mTORC1 inhibitor PRP-1 caused significant upregulation of tumor suppressors, such as miR20a, miR125b, and miR192; and downregulation of onco miRNAs, miR509-3p, miR589, miR490-3p, miR550 in human chondrosarcoma JJ012 cell line.

[291]

**TÍTULO / TITLE:** - Prostate cancer derived prostatic acid phosphatase promotes an osteoblastic response in the bone microenvironment.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


[●● Enlace al texto completo (gratuito o de pago)](1007/s10585-013-9625-2)

**AUTORES / AUTHORS:** - Larson SR; Chin J; Zhang X; Brown LG; Coleman IM; Lakely B; Tenniswood M; Corey E; Nelson PS; Vessella RL; Morrissey C

**INSTITUCIÓN / INSTITUTION:** - Genitourinary Cancer Research Laboratory, Department of Urology, University of Washington, Box 356510, Seattle, WA, 98195, USA.

**RESUMEN / SUMMARY:** - Approximately 90 % of patients who die of prostate cancer (PCa) have bone metastases, often promoting osteoblastic lesions. We observed that 88 % of castration-resistant PCa (CRPC) bone metastases express prostatic acid phosphatase (PAP), a soluble secreted protein expressed by prostate epithelial cells in predominate osteoblastic (n = 18) or osteolytic (n = 15) lesions. Additionally, conditioned media (CM) of an osteoblastic PCa xenograft LuCaP 23.1 contained significant levels of PAP and promoted mineralization in mouse and human calvaria-derived cells (MC3T3-E1 and HCO). To demonstrate that PAP promotes mineralization, we stimulated MC3T3-E1 cells with PAP and observed increased mineralization, which could be blocked with the specific PAP inhibitor, phosphonic acid. Furthermore, the mineralization promoted by LuCaP 23.1 CM was also blocked by phosphonic acid, suggesting PAP is responsible for the mineralization promoting activity of LuCaP 23.1. In addition, gene expression arrays comparing osteoblastic to osteolytic CRPC (n = 14) identified betacellulin (BTC) as a gene upregulated during the osteoblastic response in osteoblasts during new bone formation. Moreover, BTC levels were increased in bone marrow stromal cells in response to LuCaP 23.1 CM in vitro. Because new bone formation does occur in osteoblastic and can occur in osteolytic CRPC bone metastases, we confirmed by immunohistochemistry (n = 36) that BTC was highly expressed in osteoblasts involved in new bone formation occurring in both osteoblastic and osteolytic sites. These studies suggest a role for PAP in promoting the osteoblastic reaction in CRPC bone metastases and identify BTC as a novel downstream protein expressed in osteoblasts during new bone formation.

[292]
**Título / Title:** Radiologic assessment of earliest, best, and plateau response of gastrointestinal stromal tumors to neoadjuvant imatinib prior to successful surgical resection.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


**Autores / Authors:** Tirumani SH; Shinagare AB; Jagannathan JP; Krajewski KM; Ramaiya NH; Raut CP

**Institución / Institution:** Department of Imaging, Dana-Farber Cancer Institute, Harvard Medical School, 450 Brookline Ave, Boston, MA 02215, USA; Department of Radiology, Brigham and Women's Hospital, Harvard Medical School, 75 Francis Street, Boston, MA 02115, USA. Electronic address: stirumani@partners.org.

**Resumen / Summary:** BACKGROUND: To determine the timing of earliest, best and plateau response to neoadjuvant imatinib in patients with GIST. MATERIALS AND METHODS: In this IRB-approved retrospective study, we included all 20 patients (10 women; mean age 61 years, range 30-83 years) with KIT-positive primary GIST who received neoadjuvant imatinib and underwent surgery between January 2001 and December 2012. Earliest (earliest time to partial response), best (percentage reduction in longest axial diameter [LAD] and volume correlated with RECIST 1.1 and volumetric criteria) and plateau (time point when there was <10% change in treatment response between two consecutive scans beyond best response) responses were analyzed on review of imaging. RESULTS: Median tumor size at baseline was 7.2 cm (range, 3.0-31.4 cm). Median duration of neoadjuvant imatinib was 32 weeks (IQR, 16-36 weeks). Partial response was noted in 16/20 patients (median interval = 16 weeks; IQR, 7-26 weeks); 4/20 had stable disease. Median time to earliest PR was 16 weeks (IQR, 7-26 weeks). At best response, median decrease in LAD and volume were 43% (IQR, 31-48%) and 83% (IQR, 63-87%), (median interval = 28 weeks; IQR, 18-37 weeks), at which point 10 tumors were resected. Plateau response (45% [IQR, 35-45%] LAD reduction) was noted in the remaining 10 patients (median interval = 34 weeks; IQR, 26-41 weeks) before resection. Tumor size, location or risk category did not correlate with best response or time to best response. CONCLUSION: Best response to neoadjuvant imatinib was seen at 28 weeks irrespective of tumor size and location. Plateau response was seen at 34 weeks, beyond which further treatment may not be beneficial.

[293]

**Título / Title:** Endoscopic ultrasound-guided angiotherapy of a large bleeding gastrointestinal stromal tumor.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


**Autores / Authors:** Kumbhari V; Gondal B; Okolo Iii PI; Lennon AM; Law JK; Singh VK; Saxena P; Shin EJ; Canto MI; Kalloo AN; Khashab MA
**TÍTULO / TITLE:** Immunohistochemical Markers in Fibrohistiocytic Lesions: Factor XIIIa, CD34, S-100 and p75.

**RESUMEN / SUMMARY:** BACKGROUND:: The distinction between dermatofibroma (DF), dermatofibrosarcoma protuberans (DFSP), and other benign and malignant cutaneous spindle cell lesions frequently requires immunohistochemical staining. CD34 and factor XIIIa are the most commonly used immunostains; however, they may exhibit aberrant expression and introduce the potential for misdiagnosis. There is some data supporting that p75 and S100A6 may be additional helpful immunohistochemical markers. METHODS:: We undertook a large case series examining the use of CD34 and factor XIIIa as well as p75 and S100A6 in DF, cellular DF, DFSP, indeterminate fibrohistiocytic lesion, and scar. RESULTS:: As expected, CD34 stained DFSP, although it was usually negative in DF. Factor XIIIa was generally positive in DF and negative in DFSP. There were exceptions in both cases of DF and DFSP. S100A6 was routinely negative in all entities studied. P75 was negative in all cases except DFSP, approximately half of which showed weak and/or patchy positivity. CONCLUSIONS:: We conclude that to date, CD34 and factor XIIIa remain the most reliable immunohistochemical markers for DF and DFSP.

**TÍTULO / TITLE:** Soft tissue angiofibroma: a case report.

**RESUMEN / SUMMARY:** Soft tissue angiofibroma is a recently described neoplasm that typically presents as a slowly growing, painless mass in the soft tissues of the lower extremities. Cytogenetic and molecular studies have identified a recurrent t(5;8) translocation. Treatment is simple excision. Existing data suggest that this tumor is benign and has a low rate of local recurrence. The radiologic and pathologic differential diagnoses for this lesion include both benign and malignant lesions, including plantar fibromatosis, tenosynovial giant cell tumor, fibroma of tendon sheath, epithelioid
sarcoma, and low-grade myxofibrosarcoma. Proper identification of this benign lesion through radiologic and pathologic correlation is important to prevent misdiagnosis of a low-grade sarcoma.

[296]
**TÍTULO / TITLE:** Computed tomography manifestations of a malignant solitary fibrous tumour of the pleura with distinct blood supply from celiac trunk.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** Eur J Cardiothorac Surg. 2013 Sep 27.

**AUTORES / AUTHORS:** Fan F; Zhou H; Zeng Q; Liu Y

**INSTITUCIÓN / INSTITUTION:** Department of Neurosurgery, Xiang Ya Hospital, Central South University, Changsha, Hunan, PR China.

**RESUMEN / SUMMARY:** Solitary fibrous tumours of the pleura (SFTPs) are rare and roughly 80% of them have a benign course. Malignant SFTPs are seldom reported, as are their computed tomography (CT) manifestations. We report the case of a 45-year-old male patient who presented with coughing for over 2 months. A large lesion in his right hemithorax was found by chest X-ray. CT scan showed patchy areas and a sharp time-attenuation curve. CT angiography reconstruction revealed a distinct feeding vessel from celiac trunk. After surgery, the tumour was confirmed to be a malignant SFTP by immunochemistry.

[297]
**TÍTULO / TITLE:** Paraurethral leiomyoma.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Pastor Navarro H; Martinez Ruiz J; Martinez Sanchiz C; Peran Teruel M; Segura Martin M; Pastor Guzman JM; Virseda Rodriguez J

**INSTITUCIÓN / INSTITUTION:** Servicio Urologia. Hospital General de Albacete. Albacete. Spain.

**RESUMEN / SUMMARY:** OBJECTIVE: To describe a case of para-urethral leiomyoma and to review the literature. METHODS: The usual preoperative diagnostic procedures and clinical manifestations are discussed. RESULTS: The mass was resected and, 6 years later, the patient remains asymptomatic and with no recurrence. CONCLUSIONS: Urethral or paraurethral leiomyomas are benign tumors that arise from the urethral or vaginal smooth muscle. Radiological findings (particularly magnetic resonance imaging) may suggest the origin of the tumor before surgery; however, the final diagnosis is determined by histology.

[298]
**TÍTULO / TITLE:** Giant angiomyolipoma in the upper pole of the right kidney.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Peran Teruel M; Fernandez Anguita PJ; Martínez Ruiz J; Núñez Sarrion MA; Gimenez Bachs JM; Virseda Rodriguez J

[299]
TÍTULO / TITLE: - Juvenile nasopharyngeal angiofibroma resection- novel technique to improve posterior/inferior margin control.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Kamat A; Goldstein GH; Kennedy DW

[300]
TÍTULO / TITLE: - Imaging findings of the spinal peripheral Ewing’s sarcoma family of tumours.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Huang WY; Tan WL; Geng DY; Zhang J; Wu G; Zhang BY; Li YX; Yin B
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Huashan Hospital, Fudan University, Shanghai 200041, China.
RESUMEN / SUMMARY: - AIM: To present the neuroradiological and clinical characteristics of Ewing’s sarcoma family of tumours (ESFTs) and to increase awareness of this neoplasm. MATERIALS AND METHODS: The magnetic resonance imaging (MRI) features and clinical presentations of seven patients with pathologically documented ESFTs were retrospectively analysed. The tumour location, morphological features, signal intensity, contrast enhancement characteristics, involvement of the paraspinal soft tissues, and adjacent bony structures were assessed. RESULTS: Most of the ESFTs in young adults were well-circumscribed. The present study demonstrated that ESFTs often have a hypo- or iso-intense signal on T1-weighted imaging and an iso-intense signal on T2-weighted imaging. Spinal ESFTs tended to present homogeneous signal intensity and diffuse enhancement. ESFTs are more likely to occur in the thoracic spine and later to infiltrate into the paraspinal area or vertebral bone. A broad dural attachment is another common feature in the cases presented here. CONCLUSIONS: ESFT is a rare neoplasm that can have significant overlap in imaging appearance compared with other spinal neoplasms. A well-demarcated extradural mass invading the paraspinal soft or vertebral bones, with iso-intense on T2 weighted imaging and homogeneous enhancement could facilitate the diagnosis of spinal ESFT.

[301]
Chemosaturation with percutaneous hepatic perfusion for unresectable metastatic melanoma or sarcoma to the liver: A single institution experience.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Forster MR; Rashid OM; Perez MC; Choi J; Chaudhry T; Zager JS

**INSTITUCIÓN / INSTITUTION:** Department of Surgical Oncology, Levine Cancer Institute, Carolinas Medical Center, Charlotte, North Carolina.

**RESUMEN / SUMMARY:** BACKGROUND: Patients with unresectable melanoma or sarcoma hepatic metastasis have a poor prognosis with few therapeutic options. Percutaneous hepatic perfusion (PHP), isolating and perfusing the liver with chemotherapy, provides a promising minimally invasive management option. We reviewed our institutional experience with PHP. METHODS: We retrospectively reviewed patients with unresectable melanoma or sarcoma hepatic metastasis treated with PHP from 2008 to 2013 and evaluated therapeutic response, morbidity, hepatic progression free survival (hPFS), and overall survival (OS). RESULTS: Ten patients were treated with 27 PHPs (median 3). Diagnoses were ocular melanoma (n = 5), cutaneous melanoma (n = 3), unknown primary melanoma (n = 1), and sarcoma (n = 1). Median hPFS was 240 days, 9 of 10 patients (90%) demonstrated stable disease or partial response to treatment. At a median follow up of 11.5 months, 4 of 10 (40%) remain alive. There were no perioperative mortalities. Myelosuppression was the most common morbidity, managed on an outpatient basis with growth factors. The median hospital stay was 3 days. CONCLUSIONS: Patients with metastatic melanoma and sarcoma to the liver have limited treatment options. Our experience with PHP demonstrates promising results with minimal morbidity and should be considered (pending FDA approval) as a management option for unresectable melanoma or sarcoma hepatic metastasis. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.

The role of the CXCL12-CXCR4/CXCR7 axis in the progression and metastasis of bone sarcomas (Review).

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Liao YX; Zhou CH; Zeng H; Zuo DQ; Wang ZY; Yin F; Hua YQ; Cai ZD

**INSTITUCIÓN / INSTITUTION:** Shanghai Bone Cancer Institute, Shanghai Tenth People’s Hospital, Tongji University School of Medicine, Shanghai 200072, P.R. China.

**RESUMEN / SUMMARY:** Bone sarcomas, which comprise less than 1% of all human malignancies, are a group of relatively rare mesenchymal-derived tumors. They are mainly composed of osteosarcoma, chondrosarcoma and Ewing’s sarcoma. In spite of advances in adjuvant chemotherapy and wide surgical resection, prognosis remains poor due to the high propensity for lung metastasis, which is the leading cause of mortality in patients with bone sarcomas. Chemokines are a superfamily of small pro-
inflammatory chemoattractant cytokines which can bind to specific G protein-coupled seven-span transmembrane receptors. Chemokine 12 (CXCL12), also designated as stromal cell-derived factor-1 (SDF-1), is able to bind to its cognate receptors, chemokine receptor 4 (CXCR4) and chemokine receptor 7 (CXCR7), with high affinity. The binding of CXCL12 to CXCR4/CXCR7 stimulates the activation of several downstream signaling pathways that regulate tumor progression and metastasis. In this review, the structure and function of CXCL12 and its receptors, CXCR4 and CXCR7, as well as many factors affecting their expression are discussed. Phosphoinositide 3-kinase (PI3K) and mitogen-activated protein kinase (MAPK) pathways are the two most important downstream pathways regulated by the CXCL12-CXCR4/CXCR7 interaction. CXCR4 expression in bone sarcomas, including tumor cells and samples and the correlation between CXCR4/CXCR7 expression and the survival of patients with bone sarcomas are also discussed. In addition, we review the involvement of the CXCL12-CXCR4/CXCR7 axis in the growth and metastasis of bone sarcomas and the targeting of this axis in preclinical studies.

[303]
TITULO / TITLE: - A case of prolactinoma with chordoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Hattori Y; Tahara S; Ishii Y; Kitamura T; Inomoto C; Osamura RY; Teramoto A; Morita A
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Nippon Medical School, Tokyo, Japan. Electronic address: yujiro@nms.ac.jp.

[304]
TITULO / TITLE: - Germline SDHC mutation presenting as recurrent SDH deficient GIST and renal carcinoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Gill AJ; Lipton L; Taylor J; Benn DE; Richardson AL; Frydenberg M; Shapiro J; Clifton-Bligh RJ; Chow CW; Bogwitz M
INSTITUCIÓN / INSTITUTION: - *Cancer Diagnosis and Pathology Group, Kolling Institute of Medical Research, Royal North Shore Hospital, St Leonards daggerDepartment of Anatomical Pathology, Royal North Shore Hospital, St Leonards double daggerSydney Medical School, University of Sydney, Sydney section signCancer Genetics, Hormones and Cancer Group, Kolling Institute of Medical Research, Royal North Shore Hospital, St Leonards ||Genetic Medicine and Familial Cancer Centre, Royal Melbourne Hospital, Parkville paragraph signDepartment of Surgery, Monash Medical School, Melbourne **Department of Medical Oncology, Cabrini Hospital, Melbourne daggerdaggerDepartment of Medicine,
TÍTULO / TITLE: - Isolated myxoma of the external auditory canal.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Shadfar S; Scanga L; Dodd L; Buchman CA
INSTITUCIÓN / INSTITUTION: - University of North Carolina Hospitals, Department of Otolaryngology Head and Neck Surgery.
RESUMEN / SUMMARY: - The majority of neoplasms within the external auditory canal are benign. Management of these primary tumors and their local recurrences are discussed herein. We present a case of an isolated myxoma of the external auditory canal with review of the common histopathological and radiographic features. While rare, this highlights the possibility of encountering benign tumor types that carry associated morbidity or mortality due to manifestations outside of the head neck.

TÍTULO / TITLE: - Role of the aromatase inhibitor letrozole in the management of uterine leiomyomas in premenopausal women.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Duhan N; Madaan S; Sen J
INSTITUCIÓN / INSTITUTION: - Department of Obstetrics & Gynecology, Pt. B.D. Sharma Postgraduate Institute of Medical Sciences, Rohtak, Haryana, India. Electronic address: nkadian@gmail.com.
RESUMEN / SUMMARY: - BACKGROUND: Uterine myomas are benign tumours affecting 20-40% women. Various medical and surgical therapeutic options are available but the search for an ideal medical option continues. Aromatase inhibitors have recently been reported to have a potential role in the management of oestrogen-dependent conditions like endometriosis and leiomyoma. OBJECTIVE: To evaluate the effect of letrozole on uterine myoma size and symptomatology in perimenopausal women. STUDY DESIGN: Prospective interventional study conducted on 30 premenopausal women aged between 30 and 55 years with menstrual or pressure symptoms and having a single intrauterine myoma of size 4cm or more with or without one or more additional myomata each of size 2cm or less. They received tablet letrozole 2.5mg a day for 12 weeks, and the effect of the drug on myoma size and volume and symptomatology was studied along with the adverse effect profile and patient satisfaction. RESULTS: The mean myoma size reduced from 5.4+/-1.3cm to 4.3+/-0.9cm (p<0.05) and the myoma volume exhibited a reduction of 52.45% (p=0.00) at the end of 3 months. The symptomatology score showed a significant improvement that persisted up to 3 months after cessation of therapy. No significant effect was
observed on lipid profile, serum estradiol, progesterone, testosterone and FSH and LH levels during the therapy. Nausea and hot flushes were the main adverse effects observed and were self-limiting. CONCLUSION: Letrozole significantly reduces myoma size and volume and also improves the associated symptoms. It has a good adverse effect profile and appears to be a promising medical option for management of uterine myomas.

[307]
TÍTULO / TITLE: - Immunocytochemical analysis of proliferative activity of endometrial and myometrial cell populations in focal and stromal adenomyosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Nepomnyashchikh LM; Lushnikova EL; Molodykh OP; Pichigina AK
INSTITUCIÓN / INSTITUTION: - Institute of Regional Pathology and Pathomorphology, Siberian Division of the Russian Academy of Medical Sciences, Novosibirsk, Russia. pathol@soramn.ru.
RESUMEN / SUMMARY: - Immunocytochemical study has shown that Ki-67 antigen is detected in adenomyosis in both endometrial and myometrial cell populations (in the eutopic and ectopic endometrial glandular epithelium, stromal cells, smooth muscle cells, and vascular endotheliocytes of the endometrium and myometrium), the label index differing significantly in different cell populations. The highest labeled cell index is found in the endometrial gland epitheliocytes in focal adenomyosis (23.2 +/- 2.9%); in the stromal variant this index is by 2.8 times lower despite the fact that this variant is associated with endometrial glandular hyperplasia in the majority of cases. Proliferative activity of secretory epitheliocytes is significantly lower in both adenomyosis variants than in the normal eutopic endometrium. Stromal adenomyosis is characterized by 2-fold higher proliferative activity of the cytogenic stroma than that in focal adenomyosis.

[308]
TÍTULO / TITLE: - Angiosarcomas and other sarcomas of endothelial origin.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cioffi A; Reichert S; Antonescu CR; Maki RG
INSTITUCIÓN / INSTITUTION: - Department of Medicine, Mount Sinai School of Medicine, 1 Gustave L. Levy Place, Box 1128, New York, NY 10029-6574, USA; Department of Pediatrics, Mount Sinai School of Medicine, 1 Gustave L. Levy Place, Box 1128, New York, NY 10029-6574, USA.
RESUMEN / SUMMARY: - Although benign hemangiomas are among the most common diagnoses among connective tissue tumors, angiosarcomas and other sarcomas arising from blood vessels are rare, even among sarcomas. Because endothelial tumors have unique embryonal derivation compared with other sarcomas, it is not surprising they have unique characteristics. Herein are reviewed some of these unique characteristics and therapeutic options for patients with some of these diagnoses,
highlighting the potential of new agents for these tumors, which will in all likelihood also impact treatment on more common cancers.

[309]
TITULO / TITLE: - Leiomyosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Serrano C; George S
INSTITUCIÓN / INSTITUTION: - Fletcher Laboratory, Department of Pathology, Brigham and Women’s Hospital, 75 Francis Street, Thorn 528, Boston, MA 02115, USA; Center for Sarcoma and Bone Oncology, Dana-Farber Cancer Institute, 450 Brookline Avenue, Boston, MA 02215, USA.

RESUMEN / SUMMARY: - This article presents an overview of the current literature about the biology, pathology, and the clinical management of leiomyosarcoma. In addition, the article emphasizes and discusses the current systemic treatment options available for patients with leiomyosarcoma, which range from cytotoxic chemotherapy to target therapies. Particular leiomyosarcoma subtypes, such as uterine leiomyosarcoma and inferior vena cava leiomyosarcoma, are discussed separately.

[310]
TITULO / TITLE: - Liposarcomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Henze J; Bauer S
INSTITUCIÓN / INSTITUTION: - Department of Medical Oncology, Sarcoma Center, West German Cancer Center, University Hospital Essen, University of Duisburg-Essen, Hufelandstrasse 55, Essen 45239, Germany.

RESUMEN / SUMMARY: - Liposarcoma is one of the most common sarcoma subtypes with a heterogeneous biology and clinical behavior. This article gives a comprehensive overview on clinically relevant aspects of pathology and imaging. Prognostic factors and treatment strategies are discussed for different clinical situations and histologic subtypes. This information will be of value to clinicians and interdisciplinary sarcoma teams.

[311]
TITULO / TITLE: - Sarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Wagner AJ
**TÍTULO / TITLE:** - Malignant Potential of an Endometrial Stromal Tumor With Limited Infiltration: A Case Report.

**RESUMEN / SUMMARY:** - Endometrial stromal tumors (ESTs) with limited infiltration were first proposed by Dionigi et al. However, the prognostic significance of these tumors is unclear. We report a case of a 60-year-old woman who presented with a prolapsed uterine corpus and then underwent laparoscopic-assisted vaginal hysterectomy. A very small EST was incidentally found. The tumor manifested focal irregularity and finger-like permeation into the adjacent myometrium not exceeding 3 mm but exceeding 3 in number, features intermediate between a low-grade endometrial stromal sarcoma and an endometrial stromal nodule. By definition, we rendered a descriptive diagnosis of “endometrial stromal tumor with limited infiltration.” A subsequent staging operation confirmed metastasis and, hence, a malignant potential.


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TÍTULO / TITLE: - Unilateral segmentally arranged basaloid follicular hamartomas with osteoma cutis and hypodontia: a case of Happle-Tinschert syndrome.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Lo CS; Wu YF; Hsiao YW; Chung WH; Yang CH
INSTITUCIÓN / INSTITUTION: - Department of Dermatology, Chang Gung Memorial Hospital, Keelung, Taiwan; Chang Gung University College of Medicine, Taoyuan, Taiwan.
RESUMEN / SUMMARY: - Happle-Tinschert syndrome (HTS) is a rare syndrome characterized by segmentally arranged basaloid follicular hamartomas (BFH) associated with ipsilateral osseous, dental and cerebral abnormalities. Happle and Tinschert first reported this disorder in 2008, and three cases with similar presentations have since been reported. We report another case, that of a 40-year-old man, presenting with the characteristic clinical features of HTS.

TÍTULO / TITLE: - Impact of heparin-binding domain of recombinant human osteocalcin-fibronectinIII9-14 on the osteoblastic cell response.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Yun YR; Kim HW; Jang JH
INSTITUCIÓN / INSTITUTION: - Institute of Tissue Regeneration Engineering (ITREN), Dankook University, Cheonan, 330-714, South Korea.
RESUMEN / SUMMARY: - Fibronectin (FN) containing a heparin-binding domain (HBD) and an Arg-Gly-Asp (RGD) domain can promote cell adhesion and proliferation compared to FN that contained only RGD. Here, we have engineered recombinant human osteocalcin (rhOC) with FN type III9-14 (rhOC-FNIIII9-14) containing RGD and HBD to promote the cellular activity of MC3T3-E1 cells, including adhesion, proliferation, and differentiation. RhOC-FNIIII9-14 significantly increased cell adhesion and proliferation of MC3T3-E1 cells compared to rhOC-FNIIII9-10 (P < 0.05). Moreover, rhOC-FNIIII9-14 showed osteogenic differentiation of MC3T3-E1 cells in mineralization activity and osteogenic gene expression.

TÍTULO / TITLE: - Primary clear cell sarcoma of the tongue.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Kraft S; Antonescu CR; Rosenberg AE; Deschler DG; Nielsen GP
INSTITUCIÓN / INSTITUTION: - From the Department of Pathology, Massachusetts General Hospital and Harvard Medical School, Boston (Drs Kraft, Rosenberg, and Nielsen); the Department of Pathology, Memorial Sloan-Kettering Cancer Center, New York, New York (Dr Antonescu); and the Division of Head and Neck Surgery, Massachusetts Eye and Ear Infirmary and Harvard Medical School, Boston (Dr Deschler).

RESUMEN / SUMMARY: - Clear cell sarcoma shares features with melanoma, but frequently shows EWSR1 rearrangements. It is an aggressive tumor typically occurring in the soft tissues of the extremities, with a gastrointestinal variant with less consistent melanocytic differentiation. It is extremely rare in the head and neck region, with no reported cases in the oral cavity. We report a case of an 82-year-old woman with a clear cell sarcoma arising in the tongue, with cervical lymph node metastases. Histologically, the tumor showed some features of gastrointestinal clear cell sarcoma. No osteoclast-type giant cells were present. The tumor cells were positive for S100 protein and negative for other melanocytic markers. Fluorescence in situ hybridization showed rearrangements of EWSR1 and ATF1. This case expands the spectrum of clear cell sarcoma with a gastrointestinal-like variant in a novel site, emphasizing the need to consider it as a differential diagnosis to melanoma in mucosal sites.

[317]


RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Norimatsu Y; Ohsaki H; Masuno H; Kagawa A; Teramoto N; Kobayashi TK

INSTITUCIÓN / INSTITUTION: - Department of Medical Technology, Ehime Prefectural University of Health Sciences, Ehime, Japan.

RESUMEN / SUMMARY: - Objective: The removal of blood components is necessary to improve the quality of the liquid-based cytology (LBC) preparations. In ThinPrep® (TP) samples a cell suspension in a methanol-based fixative undergoes a vacuum filtration method, whereas in SurePath (SP) samples a cell suspension in an ethanol-based fixative is processed through a density gradient centrifugation system prior to gravity deposition of the specimen onto a glass slide. We compared the cyto-architectural features for the cytologic diagnosis of endometrial adenocarcinoma using parallel TP and SP preparations in a previous publication. Study Design: We performed our study on LM8 cells (a cultured osteosarcoma cell line). LM8 cells at a concentration of 1.25 x 103 cell/cm2 were seeded on a 35-mm plate in culture medium, which contained 10% fetal bovine serum (FBS), 100 units/ml penicillin, and 100 mu/ml streptomycin in Dulbecco’s modified Eagle’s medium (DMEM), and aliquots of the cell suspension obtained in this way were compared after the addition of a hemolytic agent, i.e. CytoLyt® (CyL). LBC preparations were then obtained on cell suspensions treated with CyL after different time intervals of hemolysis. Results: Treatment with CyL did not alter the cellularity of the preparation, but reduction of the nuclear area and a tendency towards nuclear chromatin condensation with a subsequent higher brightness were found. Because CyL is a 25% methanol-buffered solution, its alcoholic concentration is
low; it was our impression that, while its fixative effect was weak, its hemolytic effect was high. Water influx or efflux through the cell membrane is controlled by osmotic pressure changes induced by the buffer solution in the CyL solution. While CyL was not shown to alter the cell shape, nuclear shrinkage was thought to be probably due to the increasing cell dehydration caused by longer exposure intervals to methanol.

Conclusion: This study has allowed us to make significant observations on the hemolytic properties of CyL, and on its combined effects with PreservCyt on the cytomyomorphology of cells suspensions. © 2013 S. Karger AG, Basel.

[318]

TÍTULO / TITLE: - Differentiation of aggressive and indolent subtypes of uterine sarcoma using maximum standardized uptake value.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Lee EY; Khong PL; Tse KY; Chan KK; Chu MM; Ngan HY
INSTITUCIÓN / INSTITUTION: - Departments of aDiagnostic Radiology bObstetrics and Gynaecology, Queen Mary Hospital, University of Hong Kong, Pokfulam Road, Hong Kong.

RESUMEN / SUMMARY: - OBJECTIVE: The aim of the study was to elucidate the differential metabolic activities in aggressive and indolent subtypes of uterine sarcomas, which may aid in managing these heterogeneous tumours. METHODS: We retrospectively analysed the PET/computed tomography scans of consecutive patients (N=18) diagnosed with uterine sarcoma at our unit. The patients were divided into indolent (N=4) and aggressive (N=14) tumour groups, and the maximum standardized uptake values (SUVmax) of all lesions (n=134) were measured. The SUVmax of the lesions were compared between the two tumour groups using the Mann-Whitney U-test. We calculated the optimal cutoff value as determined by receiver operating characteristic analysis. A P-value less than 0.05 was considered statistically significant. RESULTS: The mean SUVmax of aggressive (n=104) and indolent tumours (n=30) were significantly different (8.0+/-7.3 vs. 1.9+/-0.9 respectively; P<0.001). A cutoff of SUVmax greater than 4.0 was able to exclude indolent tumours, with 100% specificity and positive predictive value (sensitivity 72%, negative predictive value 50% and accuracy 78%; area under the curve 97%). By applying this same cutoff value on the most metabolic active lesion in each patient, we were able to correctly classify all but one patient into either the aggressive or indolent tumour group with 100% specificity and positive predictive value (sensitivity 93%, negative predictive value 80% and accuracy 94%). CONCLUSION: Aggressive and indolent uterine sarcoma subtypes have differential metabolic activities that can be used to classify them and this can aid in patient management for preoperative surgical planning and treatment stratification.

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[319]

TÍTULO / TITLE: - Metformin inhibits growth and sensitzes osteosarcoma cell lines to cisplatin through cell cycle modulation.
RESUMEN / SUMMARY: Osteosarcoma (OS) is the most common cancer that affects the bone and appears to be resistant to several chemotherapeutic drugs. The aim of the present study was to verify whether the combination of metformin and cisplatin has an effect on OS cell lines. OS cell lines U2OS, 143B and MG63 were treated with metformin, cisplatin or a combination of both drugs. Viability, apoptosis and cell cycle were evaluated to characterize the effects of the treatments. Western blot analyses were used to evaluate protein expression. All OS cell lines were found to be sensitive to metformin with different values of IC50, showing a slowdown of cell cycle associated or not with apoptosis. In particular, metformin was able to sensitize cells to cisplatin, to which all OS cell lines were resistant, demonstrating a synergistic effect in the combined treatment of the two drugs. The data obtained may have clinical relevance for novel therapeutic strategies for the treatment of OS; metformin inhibits tumor cell growth and amplifies the effect of cisplatin.

TÍTULO / TITLE: Mast cell sarcoma mimicking metastatic colon carcinoma.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Schwaab J; Horny HP; Jonescheit J; Metzgeroth G; Schafhausen P; Gaiser T; Marx A; Kienle P; Hofmann WK; Reiter A

INSTITUCIÓN / INSTITUTION: III. Medizinische Klinik, Universitätsmedizin Mannheim, Theodor-Kutzer-Ufer 1-3, 68167, Mannheim, Germany.

RESUMEN / SUMMARY: OBJECTIVE: Equivalent cross-relaxation rate (ECR) imaging (ECRI), which allows quantitation of macromolecular tissue components, is a potentially useful nuclear magnetic resonance (NMR) technique for histopathological diagnosis. The purpose of this study was to compare ECR values among various soft tissue tumors.

TÍTULO / TITLE: Correlation between equivalent cross-relaxation rate and cellular density in soft tissue tumors.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Hamada S; Matsushima S; Sugiura H; Yamada K; Nishida Y; Ishiguro N

INSTITUCIÓN / INSTITUTION: Department of Orthopaedic Surgery, Aichi Cancer Center, Nagoya, Japan, shamada@med.nagoya-u.ac.jp.
histological types and assess the correlation between ECR and tumor cellular image in soft tissue tumors. **MATERIALS AND METHODS:** We performed ECRI to evaluate cellular images of soft tissue tumors and tumorous lesions. Thirty-three patients who underwent evaluation with MRI and ECRI at the first visit were enrolled. Resection or biopsy was performed to obtain a histopathological diagnosis, followed by cell density measurement. ECR values of the histological subgroups were compared, and the correlation between ECR and cell density was analyzed to assess whether ECR can be used as an indicator of histological cell density. **RESULTS:** ECR values for benign tumors varied widely and were not significantly different from those for malignant tumors. However, the mean ECR value was significantly higher for high-grade malignant tumors than for low-grade tumors (p < 0.01). Moreover, a positive correlation was found between ECR and cell density (r = 0.72; p < 0.01). **CONCLUSIONS:** ECR reflects the cell density and malignancy grade of a soft tissue tumor. ECRI could provide cellular imaging and useful clinical information to aid the pre-operative diagnosis of soft tissue tumors.

[322]
**TÍTULO / TITLE:** Giant cell tumor of the distal common bile duct: report of a rare, benign entity that may mimic malignant biliary obstruction.
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary
**REVISTA / JOURNAL:** Int J Colorectal Dis. 2013 Nov 28.
  ● Enlace al texto completo (gratuito o de pago) [1007/s00384-013-1798-2](1007/s00384-013-1798-2)
**AUTORES / AUTHORS:** Kolokotronis T; Glanemann M; Wagner M; Bohle RM; Grunhage F
**INSTITUCIÓN / INSTITUTION:** Department of General, Visceral, Vascular and Pediatric Surgery, University of Saarland Medical School, Homburg Campus, 66421, Homburg, Saar, Germany, theodoros.kolokotronis@uniklinikum-saarland.de.

[323]
**TÍTULO / TITLE:** Synovial osteochondromatosis in the subacromial bursa mimicking calcific tendinitis: Sonographic diagnosis.
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary
  ● Enlace al texto completo (gratuito o de pago) [1002/jcu.22097](1002/jcu.22097)
**AUTORES / AUTHORS:** Kim TK; Lee DH; Park JH; Kim CH; Jeong WK
**INSTITUCIÓN / INSTITUTION:** Department of Orthopaedic Surgery, Teunteun Hospital, Ansan, Korea.
**RESUMEN / SUMMARY:** Synovial osteochondromatosis is an idiopathic benign metaplasia of the synovial membrane rarely found in an extra-articular bursa. We describe the case of a 55-year-old woman with synovial osteochondromatosis in the subacromial bursa mimicking calcific tendinitis. Plain radiographs showed a radiopaque mass over the middle facet of the greater tuberosity, suggesting calcific tendinitis. Sonography, however, showed a loose body in the subacromial bursa, and no evidence of calcification inside the rotator cuff. © 2013 Wiley Periodicals, Inc. J Clin Ultrasound, 2013.
[324]
**TÍTULO / TITLE:** - Metastasectomy for gastrointestinal stromal tumors.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Bamboat ZM; Dematteo RP
**INSTITUCIÓN / INSTITUTION:** - Department of Surgery, Memorial Sloan-Kettering Cancer Center, New York, New York.
**RESUMEN / SUMMARY:** - Gastrointestinal stromal tumor (GIST) is the most common sarcoma of the intestinal tract. Improvements in understanding the molecular pathogenesis of GIST have resulted in novel treatment strategies combining surgery with tyrosine kinase inhibitors (TKIs). Metastasectomy in carefully selected patients who have stable or responsive disease on imatinib should be considered in the multidisciplinary setting. We review existing data on surgical cytoreduction in metastatic GIST while on targeted therapy and compare outcomes with either treatment alone. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.

[325]
**TÍTULO / TITLE:** - Management of recurrent retroperitoneal sarcoma.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Gyorki DE; Brennan MF
**INSTITUCIÓN / INSTITUTION:** - Department of Surgery, Memorial Sloan-Kettering Cancer Center, New York, New York.
**RESUMEN / SUMMARY:** - Approximately 15% of soft tissue sarcomas are retroperitoneal. The occult location and anatomic complexity results in local recurrences in the majority of patients. Predictors of recurrence include histological subtype, completeness of resection, and the hospital case volume. The most significant predictor of outcome following local recurrence is the resectability of the recurrent disease. An understanding of the implication of tumor biology on outcomes is essential in determining optimal management for patients with recurrent retroperitoneal sarcoma. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.

[326]
**TÍTULO / TITLE:** - Characterization of translocations in mesenchymal hamartoma and undifferentiated embryonal sarcoma of the liver.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Mathews J; Duncavage EJ; Pfeifer JD
**INSTITUCIÓN / INSTITUTION:** - Lauren V. Ackerman Laboratory of Surgical Pathology, Department of Pathology, Washington University School of Medicine, St. Louis, MO, USA. Electronic address: jmathews@path.wustl.edu.
RESUMEN / SUMMARY: - BACKGROUND: Mesenchymal hamartoma of the liver (MHL) is an uncommon benign primary liver tumor that typically occurs in the pediatric population, although cases have been described in adults. MHL is sometimes associated with the highly malignant undifferentiated embryonal sarcoma (UES), and the synchronous or metachronous occurrence of MHL and UES suggests they share a common genetic link. Although the exact mechanism of tumorigenesis has not been identified, MHL cases harbor recurring chromosomal rearrangements involving 19q13.

DESIGN: In order to provide more details on the genetic events of MHL tumorigenesis, capture-based next generation sequencing (NGS) targeted to loci recently shown to be involved in a translocation in a case of UES arising in MHL (specifically, the MALAT1 gene on chromosome 11 and a gene poor region termed MHLB1 on chromosome 19) was performed on formalin fixed paraffin embedded tissue from seven cases of MHL.

RESULTS: Chromosome rearrangements involving the MHLB1 locus were identified in three of the seven cases, including the translocation t(11,19)(q13.1;q13.42) involving the MALAT1 gene; the translocation t(2,19)(q31.1;q13.42) involving AK023515, an uncharacterized noncoding gene; and the inversion inv(19,19)(q13.42;q13.43) involving the PEG3 gene encoding a Kruppel-type zinc-finger protein. Rearrangements were exclusively identified in pediatric tumors. In each case, the presence of the rearrangement was confirmed by PCR and interphase FISH. Interphase FISH also demonstrated that the arrangements occur within the spindle cell component but not within the epithelial components of the tumor. CONCLUSIONS: Since the MHLB1 locus contains a CpG-rich region whose methylation regulates C19MC miRNA genes, rearrangements that disrupt this region may contribute to MHL development through alteration of miRNA expression. The demonstration that the loose stromal cells harbor the rearrangements indicates that (some cases of) MHL are a neoplastic process due to a somatic genetic change and not a germline abnormality.

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TÍTULO / TITLE: - Co-existence of Epithelioid and Fibroblastoid Subsets in a Sarcomatoid Renal Carcinoma Cell Line Revealed by Clonal Studies.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Hsieh CH; Chen HC; Chang YH; Pang ST; Kuo ML; Chuang CK; Liao SK

INSTITUCIÓN / INSTITUTION: - Taipei Medical University, Taipei, Taiwan, R.O.C. E-mail: liaosk@h.tmu.edu.tw and Dr. Ming-Ling Kuo, Chang Gung University, Taoyuan, Taiwan, R.O.C. mingling@mail.cgu.edu.tw.

RESUMEN / SUMMARY: - BACKGROUND: The biology of sarcomatoid renal cell carcinoma (RCC) and its conversion from and to the clear cell RCC are not fully understood. We aimed to analyze the sarcomatoid RCC cell line, RCC52, derived from a lymph node metastatic lesion consisting mostly of sarcomatoid RCC cells with occasional clear cell areas. MATERIALS AND METHODS: Representative clonal epithelioid and fibroblastoid sublines isolated from the RCC52 cell line were analyzed alongside the parental line. Cytofluorometric and western blot analyses were used for phenotypic study. Xenotransplantation and in vitro invasive assays were used to determine tumorigenicity and invasiveness. Immunohistology in conjunction with antibodies to paired box gene-2 (PAX2) were used to determine if xenografts or tumor
biopsies had the clear cell component. RESULTS: RCC52 cells grown as monolayers in vitro were all PAX2-negative, and consisted mostly of epithelioid cells and partly of fibroblastoid cells as noted in a previous study, confirming the co-existence of these two cell types in the in vitro growth of exclusive sarcomatoid RCC cells. Immunohistology revealed that the parental line and all epithelioid sublines tested were able to develop into solid tumors consisting mostly of sarcomatoid cells with PAX2-positive clear cells in some areas. The RCC stem cell marker CD105 was selectively expressed by a small proportion of the epithelioid, but not fibroblastoid, sublines, which was in line with the tumorigenic property of the epithelioid sublines containing cancer stem cells (CSCs). In contrast, only fibroblastoid sublines exhibited migratory/invasive properties, as determined by in vitro assays. CONCLUSION: Our findings confirm the presence of two distinct subsets in the RCC52 line, and suggest the epithelioid subset being able to de-differentiate to clear cells, albeit partially, and harboring CSCs as an emerging therapeutic target in order to achieve effective treatment of this malignancy.
Computed tomography demonstrated a large, heterogeneously enhancing, soft tissue mass with no macroscopic fat above the right kidney with tumor thrombus extending into the inferior vena cava and right atrium. Positron Emission Tomography scanning demonstrated intense Fluorodeoxyglucose avidity in the primary tumor and tumor thrombus. The presumptive radiological diagnosis was adrenocortical carcinoma, but surgical pathology revealed a dedifferentiated liposarcoma. We conclude that suprarenal retroperitoneal liposarcoma should be included in the differential diagnosis for an apparent adrenal mass with venous invasion.

[330]
TÍTULO / TITLE: - Emergent transcutaneous embolization in an advanced carcinosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Davis F; Schaiberger GG; Rodriguez Y; Odden A
INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine (FD, GGS, YR), University of Michigan, Ann Arbor, Michigan; and University of Miami Miller School of Medicine (AO, GGS), Coral Gables, Florida.
RESUMEN / SUMMARY: - Active hemorrhage is a life-threatening complication of advanced tumors. It often signifies the terminal stage of the disease and therefore is usually treated with palliative care. Transcutaneous arterial embolization (TAE) is a safe, noninvasive procedure that halts acute tumor-related bleeding thereby providing effective life-saving treatment for patients with non-operable tumors. Carcinosarcoma is an uncommon tumor that generally affects the head and neck, respiratory tract, colon, uterus, ovaries, and fallopian tubes. The authors present an interesting case of a rare manifestation in an unusual location. A 60-year-old Caucasian male, who presented with abdominal and groin pain, was found to have a large carcinosarcoma in the retroperitoneal space. The tumor was complicated with an active bleed. Since he was not a candidate for surgical intervention, a TAE was performed. Two days later, the patient was discharged to hospice where he was able to live out the rest of his life.

[331]
TÍTULO / TITLE: - Hepatobiliary and Pancreatic: Large pancreatic liposarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Kuramoto K; Hashimoto D; Abe S; Chikamoto A; Beppu T; Iyama K; Baba H
INSTITUCIÓN / INSTITUTION: - Department of Gastroenterological Surgery, Kumamoto University Graduate School of Medical Sciences, Kumamoto, Japan.

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Enlace al Resumen / Link to its Summary

Infantile congenital fibrosarcomas are rare neoplasms that usually present on the extremities, and although they are locally invasive, they rarely metastasize. They are commonly misdiagnosed as hemangiomas or other vascular tumors, so further evaluation by pathology is required for proper diagnosis. We describe a newborn with a neoplastic growth of the lower lip that was thought to be an infantile hemangioma that did not respond to therapy. When the child was 2 months old, an incisional biopsy demonstrated a fibrosarcoma. This case highlights congenital infantile fibrosarcoma as a mimic of infantile hemangioma. To our knowledge this is first case report of congenital infantile fibrosarcoma involving the lip.

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Enlace al texto completo (gratuito o de pago)

INTRODUCTION: Pulmonary sarcomas overall are very uncommon and comprise only 0.5 % of all primary lung malignancies. The diagnosis is established only after sarcoma-like primary lung malignancies and a metastatic extrathoracic sarcoma have been excluded. Synovial sarcoma accounts for ~8 % of soft-tissue sarcomas. Synovial sarcoma arising from the pleura has rarely been reported. METHODS: We report a case of a 58-year-old woman who complained of right-sided chest pain and shortness of breath. Chest CT scan revealed a large heterogeneous mass, occupying most of the right hemithorax. Histologic diagnosis was supplemented by interphase cytogenetic (FISH) analysis. RESULTS: Computed tomography guided Tru-cut biopsy was suspicious for a sarcomatous or fibrous malignancy. However, intraoperative frozen-section diagnostics confirmed the diagnosis of a sarcoma. Immunohistochemistry showed that tumor cells expressed epithelial membrane antigen, CD99 and BCL2. Based on immunohistochemistry, the diagnosis of synovial sarcoma was suspected and was confirmed by FISH analysis. The patient was treated with right upper bilobectomy. Due to R1-resection status, postsurgical systemic chemotherapy was administered. CONCLUSIONS: Primary pulmonary synovial sarcoma is a rare primary lung tumor. Due to extensive size of the
tumor with pleural and mediastinal invasion only a R1-resection status could be achieved by thoracic surgery.

[334]
TÍTULO / TITLE: - Stereotactic radiosurgery of intracranial chordomas, chondrosarcomas, and glomus tumors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Kano H; Lunsford LD
INSTITUCIÓN / INSTITUTION: - Department of Neurological Surgery, The Center for Image-Guided Neurosurgery, UPMC Presbyterian, University of Pittsburgh School of Medicine, University of Pittsburgh, Suite B-400, 200 Lothrop Street, Pittsburgh, PA 15213, USA. Electronic address: kanoh@upmc.edu.
RESUMEN / SUMMARY: - Chordomas and chondrosarcomas are rare, slow-glowing, locally aggressive tumors with high recurrence rates. Stereotactic radiosurgery (SRS) is an important management option for patients with recurrent or residual chordomas and chondrosarcomas. Glomus jugulare tumor are rare highly vascularized tumors that arise from the paraganglionic structures of the glossopharyngeal and vagal nerves. Because of their highly vascular nature and surgically formidable anatomic location, curative resection often proves challenging. SRS can be used as an up-front treatment or as an additional treatment for patients with recurrent or residual glomus jugulare tumor after surgical resection.

[335]
TÍTULO / TITLE: - Angiosarcoma Involving Native Abdominal Aortic Aneurysm Sac after Endograft Repair.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Fenton J; Veenstra M; Bove P
INSTITUCIÓN / INSTITUTION: - Department of Vascular Surgery William Beaumont Hospital, Royal Oak, MI.
RESUMEN / SUMMARY: - Primary angiosarcoma of the aorta is a rare malignancy that is characterized by rapid proliferation and propensity for metastasis. It has been reported only 35 times in the surgical literature. This case report presents a 66-year-old man diagnosed with angiosarcoma of his native aorta 7 years after endograft repair of an abdominal aortic aneurysm. We then reviewed the world surgical literature for occurrence, tumorigenic studies, prognosis, and management of aortic angiosarcoma. Because native aortic tissue is retained after endovascular repair of an abdominal aortic aneurysm, the treating physician should have an awareness of this pathology and entertain the diagnosis as appropriate.
Giant cell angiofibroma or localized periorbital lymphedema?

Giant cell angiofibroma represents a rare soft tissue neoplasm with a predilection for the orbit. We recently encountered a mass removed from the lower eyelid of a 56-year-old female that histopathologically resembled giant cell angiofibroma. The process consisted of haphazardly arranged CD34-positive spindled and multinucleated cells within an edematous, densely vascular stroma. However, the patient had recently undergone laryngectomy and radiotherapy for a laryngeal squamous cell carcinoma. A similar mass had arisen on the contralateral eyelid, and both had developed several months post-therapy. Lymphedema of the orbit can present as tumor-like nodules and in some cases may share histopathologic features purported to be characteristic of giant cell angiofibroma. A relationship between giant cell angiofibroma and lymphedema has not been established, but our case suggests there may be one. The potential overlap of these two conditions should be recognized, as should other entities that may enter the differential diagnosis.

Nevoid basal cell carcinoma syndrome with a unilateral giant ovarian fibroma in a Japanese 6-year-old girl.

Nevoid basal cell carcinoma syndrome (NBCCS) is characterized by basal cell carcinoma, skeletal abnormalities, benign tumors including ovarian fibroma, and various other phenotypic expressions. Ovarian fibromas in NBCCS before puberty are very rare. We report a 6-year-old prepubescent girl with NBCCS showing skeletal abnormalities, medulloblastoma, and ovarian fibromas. The patient was referred to our hospital owing to abdominal distension. On admission, a huge elastic hard tumor was palpable and computed tomography showed a huge tumor of the left ovary. We performed a left salpingo-oophorectomy and diagnosed the tumor as a benign fibroma. Further examination of the computed tomography images showed skeletal abnormalities. In addition, the patient had a history of medulloblastoma at the age of 4 years. Therefore, we diagnosed NBCCS. A genetic examination indicated a novel 1 bp deletion in exon 18 (c.3055delG). Sequence analysis of exon 18 using DNA from the ovarian tumor revealed a mutant allele (c.3055delG) dominant to the wild-type allele, thus suggesting loss of heterozygosity in the PTCH1 gene, which is known to be associated with NBCCS.
associated with NBCCS. Conclusion On the basis of our experience, physicians treating pediatric ovarian tumors should be aware that such huge benign ovarian tumors may be a phenotype of NBCCS, as shown in our patient. In addition, genetic examination focusing on the PTCH1 gene might be important for diagnosis of NBCCS in pediatric patients.

[338]
TITULO / TITLE: - Dermatofibroma-associated dystrophic calcification.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
- Enlace al texto completo (gratuito o de pago) 1111/cup.12255
AUTORES / AUTHORS: - Famenini S; Cassarino DS
INSTITUCIÓN / INSTITUTION: - Kaiser Permanente Los Angeles Medical Center, Departments of Dermatology and Pathology, Los Angeles, CA, USA.

[339]
TITULO / TITLE: - Congenital dermatofibrosarcoma with associated hypertrichosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
- Enlace al texto completo (gratuito o de pago) 1111/cup.12259
AUTORES / AUTHORS: - Berry RS; Berry TM; Haney M; Shetty A; Yu L; Smidt AC
INSTITUCIÓN / INSTITUTION: - Department of Pathology, University of New Mexico School of Medicine, Albuquerque, NM.

[340]
TITULO / TITLE: - Aleukemic cutaneous myeloid sarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
- Enlace al texto completo (gratuito o de pago) 1111/cup.12231
AUTORES / AUTHORS: - Aboutalebi A; Korman JB; Sohani AR; Hasserjian RP; Louissaint A Jr; Le L; Kraft S; Duncan LM; Nazarian RM
INSTITUCIÓN / INSTITUTION: - Department of Dermatology, Harvard Medical School, Boston, MA, USA.
RESUMEN / SUMMARY: - BACKGROUND: Aleukemic cutaneous myeloid sarcoma (CMS) represents an important yet rare entity denoting the presence of a cutaneous myeloid leukemic infiltrate without concurrent peripheral blood or bone marrow disease. The clinicopathologic diagnosis remains elusive due to isolated skin findings and variable immunostaining. Cytogenetic and molecular findings have infrequently been reported. METHODS: Twenty-five patients with CMS were identified in the Massachusetts General Hospital pathology database between 2004 and 2012. Patients were excluded if concurrent blood or marrow acute myeloid leukemia (AML), myelodysplastic syndrome or lymphoproliferative disorder were diagnosed. RESULTS: Three patients were identified: a neonate with recurrent CMS and marrow disease that
never met diagnostic criteria for AML and two patients relapsing as CMS without concurrent blood or marrow disease following chemotherapy-induced complete remission. Histology showed atypical mononuclear cell interstitial dermal infiltrates. All cases were CD68+, lysozyme+ and CD117-; one of two were CD34+; two of three were myeloperoxidase negative. 11q23 rearrangement, t(1;14), NPM1 (nucleophosmin I), FLT3-ITD (Fms-like tyrosine kinase 3-internal tandem duplication), and novel FLT3-D835 mutations were identified. CONCLUSION: An isolated atypical cutaneous infiltrate may represent aleukemic CMS and should prompt a search for other extramedullary sites of involvement. Immunohistochemistry, molecular and cytogenetic studies can help differentiate aleukemic CMS from benign and malignant, monocytic and histiocytic mimickers, and may potentially indicate therapy and prognosis.

[341]

TÍTULO / TITLE: - Angiomyolipoma with minimal fat and non-clear cell renal cell carcinoma: differentiation on MDCT using classification and regression tree analysis-based algorithm.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Woo S; Cho JY; Kim SH; Kim SY

INSTITUCIÓN / INSTITUTION: - Department of Radiology, Seoul National University College of Medicine, Seoul, Republic of Korea.

RESUMEN / SUMMARY: - BACKGROUND: Differentiation between angiomyolipoma with minimal fat (AMLmf) and non-clear cell renal cell carcinoma (ncRCC) may be difficult owing to lack of macroscopic fat in AMLmf. However, the differential points between AMLmf and ncRCC has not been well established in the literature.

PURPOSE: To evaluate quantitative triphasic multidetector computed tomography (MDCT) features that differentiate between small AMLmf and ncRCC, and to integrate them to develop a simple and easy diagnostic algorithm. MATERIAL AND METHODS: This study was approved by the Institutional Review Board; informed consent was waived. Triphasic MDCT images of pathologically-proven AMLmfs (n = 24) and ncRCCs (n = 55) of 79 patients were retrospectively evaluated. Age, sex, size, long-to-short axis ratio (LSR), attenuation and enhancement degree in all phases, unenhanced tumor-kidney attenuation difference (UTKAD) in Hounsfield units (HU) were compared with Chi-square analysis, independent-samples t-test, and receiver-operating characteristic (ROC) curves. A criterion was formulated with classification and regression tree analysis (CART). Thereafter, CART-based algorithm was tested with additional interpretations from two radiologists. Intra- and inter-observer variability was analyzed with Bland-Altman analysis. RESULTS: LSR was greater in AMLmf than ncRCC (P < 0.001). AMLmf showed higher attenuation (all phases), CMP enhancement, and wash-out than ncRCC (P < 0.001). UTKAD was greater in AMLmf than ncRCC (P < 0.001). ROC curve analysis yielded area under the curves of 0.936, 0.888, and 0.853 using UTKAD, unenhanced attenuation, and LSR. CART-based algorithm (UTKAD >7.5 HU, LSR > 1.23) predicted AMLmf with sensitivity, specificity, PPV, and NPV of 87.5%, 96.4%, 91.3%, and 94.6%. Mean intra- and inter-observer difference was -0.1/0.03 HU and -1.0/0.09 HU for UTKAD/LSR, respectively. These interpretations changed the final diagnosis in 1.3% (1/79) and 5.1% (4/79).
patients for radiologists 1 and 2. CONCLUSION: Triphasic MDCT was useful for不同iating AMLmf and nccRCC. CART-based algorithm using UTKAD > 7.5 andLSR > 1.23 was simple and accurate in predicting AMLmf.

[342]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AutORES / AUTHORS: - Rau CL; Yen TH; Wu LC; Huang YY; Jaw FS; Liou TH
INSTITUCIÓN / INSTITUTION: - From the Institute of Biomedical Engineering, National Taiwan University, Taipei, Taiwan (C-LR, L-CW, Y-YH, F-SJ); and Department of Physical Medicine and Rehabilitation (C-LR, T-HY, T-HL) and Department of Orthopedics (L-CW), Shang-Ho Hospital, Taipei Medical University, Taipei, Taiwan.
RESUMEN / SUMMARY: - A wrist mass is rarely caused by a ruptured tendon in the forearm. The common pathologies are ganglia, tendon tenosynovitis, and giant cell tumors of tendon sheaths. Less common causes are nerve sheath tumors, vascular lesions, or an accessory muscle belly. The authors investigated a case of neglected ruptured flexor carpi ulnaris tendon that mimics a mass in the wrist. To the authors’ knowledge, this is the first case report in relevant literature. During investigation, the high-resolution musculoskeletal ultrasound suggested a soft tissue tumor or a ruptured flexor carpi ulnaris tendon. The magnetic resonance imaging scan indicated an accessory flexor carpi ulnaris muscle belly. The diagnosis of ruptured flexor carpi ulnaris tendon was confirmed by surgical exploration. This case indicates that ultrasound may be better suited than magnetic resonance imaging in evaluating a wrist mass for its accuracy, availability, and portability.

[343]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Baba Y; Hayashi S; Ikeda S; Jinguji M; Nakajo M; Nakajo M
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Graduate School of Medical and Dental Sciences, Kagoshima University, 8-35-1 Sakuragaoka, Kagoshima-shi, Kagoshima, 890-8520, Japan, yasutaka@m3.kufm.kagoshima-u.ac.jp.
RESUMEN / SUMMARY: - PURPOSE: Transcatheter arterial embolization (TAE) with absolute ethanol is widely accepted as a therapeutic procedure for renal angiomyolipoma (AML). We aim to evaluate the split renal function before and after AE for renal AML by using 99m-technetium (99mTc)-mercaptoacetyltriglycine 3 (MAG3) renography. METHODS: This study was approved by the Institutional Review Board. The study population comprised 11 renal AML patients (three males, eight females, age 55.1 +/- 13.8 years, AML in eight right and three left kidneys) who received
unilateral renal TAE with absolute ethanol from April 2002 to January 2013. Blood renal function (i.e. serum creatinine and estimated glomerular filtration rate [eGFR] and split effective renal plasma flow [ERPF]) calculated on 99mTc-MAG3 renography was compared before and within 1 week after renal AE. Statistical analysis was calculated using Wilcoxon signed-ranked test. RESULTS: TAE for renal AML was technically successful in all patients. Serum creatinine and eGFR did not change before and after TAE. ERPF on the embolized kidney did not change before (127.3 +/- 60.8 ml/min) and after (127.6 +/- 47.4 ml/min) TAE (p = 0.9726). ERPF on the nonembolized kidney showed a statistically significant increase before (152.5 +/- 46.8 ml/min) and within 1 week after (169.1 +/- 41.5 ml/min) TAE (p = 0.0093 and p < 0.05, respectively).

CONCLUSION: TAE for renal AML may not induce renal dysfunction on the embolized kidney and may immediately increase the renal blood flow of the nonembolized kidney.

[344]

TÍTULO / TITLE: - Kaposiform hemangioendothelioma complicated by kasabach-merritt phenomenon: ultrastructural observation and immunohistochemistry staining reveal the trapping of blood components.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Yuan SM; Hong ZJ; Chen HN; Shen WM; Zhou XJ
INSTITUCIÓN / INSTITUTION: - Department of Plastic Surgery, Jinling Hospital, School of Medicine, Nanjing University, Nanjing, Jiangsu, China.
RESUMEN / SUMMARY: - Abstract Kaposiform hemangioendothelioma (KHE), a borderline tumor of endothelial origin, is associated with Kasabach-Merritt phenomenon, characterized by profound thrombocytopenia and consumptive coagulopathy resulting from the localized intravascular coagulation (LIC) in the tumor. Previous studies have suggested that the trapping of blood components, including platelets, may underlie the LIC in KHE. However, more evidence is needed to support this hypothesis. In this study, one case of a Chinese infant with a KHE in the left arm was complicated by Kasabach-Merritt phenomenon. The tumor was partially resected and the sample was used for ultrastructural observation and immunohistochemistry staining of Glut-1. Ultrastructural observation found the trapping of erythrocytes, platelets, macrophages, and lymphocytes in the slit-like channels of the tumor nodules, and phagocytic vesicles in the cytoplasm of neoplastic cells. Immunohistochemistry staining further showed numerous Glut-1(+) erythrocytes in the channels. In conclusion, our results provided compelling morphological evidence of the trapping of blood components in KHE, which may interpret the LIC in the tumor and subsequent consumptive coagulopathy.

[345]

TÍTULO / TITLE: - Psammomatoid Juvenile Ossifying Fibroma of the Maxilla: Radical Surgery with Maxillary Resection in a 7-year-old Girl.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Halama D; Hierl T; Wickenhauser C; Pausch NC
INSTITUCIÓN / INSTITUTION: Maxillofacial Surgery, University Hospital Leipzig, Leipzig, Germany.


AUTORES / AUTHORS: Mashhour MA; Abdel Rahman M
INSTITUCIÓN / INSTITUTION: Department of Orthopaedic Surgery, Orthopaedic Oncology Unit, Faculty of Medicine, Ain Shams University, Abbasseia Square, Cairo, Egypt, mohamed.mashhour@gmail.com.

RESUMEN / SUMMARY: PURPOSE: Various methods for the treatment of chondroblastoma of bone have been used including simple curettage, or combined with bone grafting, in addition to the use of adjuvant therapy. However, local recurrence still represents a challenge in the management of this aggressive tumour. This study focuses on evaluating the role of intraregional extended curettage together with the use of adjuvant cryotherapy and autogenous bone grafting in the treatment of benign chondroblastoma of bone aiming to decrease the recurrence rate. METHODS: All patients with chondroblastoma included in this study underwent intralesional extended curettage, adjuvant cryotherapy using liquid nitrogen, and autogenous iliac crest bone grafting. Follow up for healing of chondroblastoma lesions and detection of any local recurrence was assessed on clinical and radiological bases. The functional outcome was assessed by the Musculoskeletal Tumour Society scoring system. RESULTS: The mean follow-up period was 49 months. The average time for bone healing was 7.4 months. Our rate of local recurrence is 7.1 %. Two patients (14.3 %) developed physeal growth arrest. One patient had superficial skin sloughing (7.1 %). None of the cases had pathological fracture. The mean Musculoskeletal Tumour Society functional score was 92.7 %. CONCLUSION: Chondroblastoma is an aggressive benign bone tumour with a high rate of recurrence. The use of high-speed burr combined with adjuvant intralesional cryotherapy and iliac crest autogenous bone grafting is a reliable method of treatment with a low rate of recurrence.

TÍTULO / TITLE: - Oral Fibroma With HPV-Associated Epithelial Dysplasia: Even in Fibromas You Should Expect the Unexpected.

RESUMEN / SUMMARY: - Fibromas are the most common soft tissue lesions of the oral cavity and are generally attributed to trauma. Koilocytic dysplasia refers to human papillomavirus (HPV)-related epithelial cytopathic effect. Koilocytic dysplasia is
considered neoplastic. Herein, we report a case of oral fibroma with HPV-induced dysplastic changes of the surface epithelium confirmed by immunohistochemical stains for p16 and p53 as well as HPV in situ hybridization.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
•● Enlace al texto completo (gratuito o de pago)
1097/COC.0b013e31829b5667
AUTORES / AUTHORS: - Vijay A; Ram L
INSTITUCIÓN / INSTITUTION: - Departments of *General Surgery daggerAnaesthesiology, Hamad Medical Corporation, Doha, Qatar.
RESUMEN / SUMMARY: - Retroperitoneal liposarcomas are rare mesenchymal tumors of the retroperitoneum that typically present with advanced disease and often carry a poor prognosis. Because of their rarity and anatomic location, these malignant tumors can cause a diagnostic dilemma and present several therapeutic challenges. They are usually associated with a high rate of recurrence despite grossly complete resection, thus requiring long-term and often indefinite follow-up. Relevant data on this topic was procured and synthesized with the aid of a comprehensive Medline search in addition to oncologic, pathologic, urologic, radiologic, and surgical literature review on retroperitoneal sarcomas. This article provides an in-depth review into the natural history, pathology, clinical manifestations, and prognostic features of retroperitoneal liposarcomas. It also discusses the reliability of diagnostic procedures and novel curative approaches that are currently being evaluated for the disease.

TÍTULO / TITLE: - Role of matrix metalloproteinase-10 in the BMP-2 inducing osteoblastic differentiation.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Mao L; Yano M; Kawao N; Tamura Y; Okada K; Kaji H
INSTITUCIÓN / INSTITUTION: - Department of Physiology and Regenerative Medicine, Kinki University Faculty of Medicine, Osaka-Sayama 589-8511, Japan.
RESUMEN / SUMMARY: - Fibrodysplasia ossificans progressiva (FOP) is a skeletal disorder with progressive heterotopic ossification in skeletal muscle. A mutation causing constitutive activation in a bone morphogenetic protein (BMP) type 1 receptor [ALK2(R206H)] is found in most patients with FOP. However, the details in the heterotopic ossification of muscle in FOP and the role of matrix metalloproteinase-10 (MMP-10) in bone remain to be fully elucidated. In the present study, we investigated the role of MMP-10 in the differentiation of mouse myoblastic C2C12 cells into osteoblasts. MMP-10 was extracted as a factor, whose expression was most extensively enhanced by ALK2 (R206H) transfection in C2C12 cells. MMP-10 significantly augmented the levels of Osterix, type 1 collagen, alkaline phosphatase (ALP) and osteocalcin mRNA as well as ALP activity enhanced by BMP-2 in C2C12 cells. Moreover, a reduction in endogenous MMP-10 levels by siRNA significantly
decreased the levels of Runx2, Osterix, type 1 collagen, ALP and osteocalcin mRNA enhanced by BMP-2 in these cells. In addition, MMP-10 increased the phosphorylation of Smad1/5/8 as well as enhanced the levels of Smad6 and Smad7 mRNA induced by BMP-2. In conclusion, the present study first demonstrated that MMP-10 promotes the differentiation of myoblasts into osteoblasts by interacting with the BMP signaling pathway. MMP-10 may play some important role in the heterotopic ossification of muscle in FOP.

[350]
TÍTULO / TITLE: - Low grade B cell lymphoma arising in a background of multifocal extra-adrenal myelolipoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Noll A; Boone J; Cunningham M; Mammen J; Tawfik O
INSTITUCIÓN / INSTITUTION: - Department of Pathology and Laboratory Medicine, The University of Kansas Medical Center, Kansas City, KS 66160, USA; phone: 913 588 1180, fax: 913 588 1185, e mail: otawfik@kumc.edu.
RESUMEN / SUMMARY: - Myelolipomas are rare, benign, non-functioning tumors composed of an admixture of mature adipose tissue and hematopoietic elements. Extra-adrenal myelolipomas are extremely rare, but have been reported in multiple sites including the omentum, presacral, and retroperitoneal areas, along with the thorax, kidneys, liver and stomach. We report a case of a 68-year-old man with low-grade B-cell lymphoma arising in a background of recurrent multifocal extra-adrenal myelolipoma. Pathological evaluation of the lesion and bone marrow showed foci of lymphoid aggregate that were confirmed to be monoclonal B lymphoma by flow cytometry. To our knowledge, this is only the third reported case to feature such a rare combination of diseases. The clinical, radiological, and pathological differential diagnostic findings are discussed.

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[351]
TÍTULO / TITLE: - Leiomyoma arising within a mature cystic teratoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Pathology. 2013 Dec;45(7):691-3. doi: 10.1097/PAT.0b013e3182a9a375
AUTORES / AUTHORS: - Jakate K; Srigley J; Plotkin A
INSTITUCIÓN / INSTITUTION: - *University of Toronto daggerTrillium Health Partners, McMaster University double daggerTrillium Health Partners, University of Toronto, Ontario, Canada.

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[352]
TÍTULO / TITLE: - Rapidly progressing primary splenic angiosarcoma with fatal hemorrhagic event.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cho EA; Choi WY; Kim SH; Hong JY; Jung SH; Kim MJ; Hwang JE; Bae WK; Shim HJ; Lee KH; Cho SH; Chung IJ
RESUMEN / SUMMARY: - Primary angiosarcoma of the spleen is an extremely rare malignancy. Splenic angiosarcoma is difficult to diagnose, and treatment is often challenging. The prognosis is dismal, with a mean survival of less than 1 year. Currently, no standard treatment methods have been established. Anecdotal reports suggest that early diagnosis and prompt splenectomy followed by cytotoxic chemotherapy can be effective treatment modalities. We report a case of rapidly progressing primary splenic angiosarcoma with multiple liver and bone metastases and a fatal hemorrhagic outcome.

[353]
TÍTULO / TITLE: - A very rare cause of neck pain: primary ewing sarcoma of the axis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Aydin R; Bilgici MC; Dagcinar A
INSTITUCIÓN / INSTITUTION: - From the *Department of Radiology, University of Ondokuz Mayis, Faculty of Medicine, Samsun; and daggerDepartment of Neurosurgery, University of Marmara, Faculty of Medicine, Istanbul, Turkey.
RESUMEN / SUMMARY: - We report the case of a 7-year-old boy who presented with a 1-month history of neck pain, left-sided torticollis, and no neurological deficit. Computed tomography and magnetic resonance imaging revealed an expansile lesion in the axis, with epidural and prevertebral soft tissue components. Histopathologic examination of the biopsy specimen revealed primary vertebral Ewing sarcoma. This is the first case of primary vertebral Ewing sarcoma that has presented with torticollis. It is essential for physicians to be familiar with this condition and the associated imaging findings because early diagnosis of such cases is the key to better prognosis.

[354]
TÍTULO / TITLE: - Endobronchial Ultrasound-Guided Transbronchial Needle Aspiration of an Intravascular Sarcoma Metastasis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Dusemund F; Schneider T; Zeisel C; Rothermundt C; Kluckert T; Schmid S; Brutsche MH
INSTITUCIÓN / INSTITUTION: - Division of Pneumology, Department of Internal Medicine, Kantonspital St. Gallen, St. Gallen, Switzerland.
RESUMEN / SUMMARY: - The role of endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) in the diagnosis of endovascular lesions has rarely been described. We report a case of EBUS-TBNA of a solid mass in the left pulmonary
artery in a patient with synovial sarcoma of the kidney, which was performed without complications and led to the diagnosis of metastatic disease. EBUS-TBNA seems to be a rapid, minimally invasive, safe and effective diagnostic procedure in selected cases of endovascular lesions.

[355]
**TÍTULO / TITLE:** Rapid growth of left atrial myxoma after radiofrequency ablation.
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** Rubio Alvarez J; Martinez de Alegria A; Sierra Quiroga J; Adrio Nazar B; Rubio Taboada C; Martinez Comendador JM
**INSTITUCIÓN / INSTITUTION:** Departments of Cardiac Surgery (Drs. Adrio Nazar, Martinez Comendador, Rubio Alvarez, Rubio Taboada, and Sierra Quiroga) and Radiology (Dr. Martinez de Alegria), University Hospital, 15706 Santiago de Compostela, España.

**RESUMEN / SUMMARY:** Atrial myxoma is the most common benign tumor of the heart, but its appearance after radiofrequency ablation is very rare. We report a case in which an asymptomatic, rapidly growing cardiac myxoma arose in the left atrium after radiofrequency ablation. Two months after the procedure, cardiovascular magnetic resonance, performed to evaluate the right ventricular anatomy, revealed a 10 x 10-mm mass (assumed to be a thrombus) attached to the patient’s left atrial septum. Three months later, transthoracic echocardiography revealed a larger mass, and the patient was diagnosed with myxoma. Two days later, a 20 x 20-mm myxoma weighing 37 g was excised. To our knowledge, the appearance of an atrial myxoma after radiofrequency ablation has been reported only once before. Whether tumor development is related to such ablation or is merely a coincidence is uncertain, but myxomas have developed after other instances of cardiac trauma.

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[356]
**TÍTULO / TITLE:** Pediatric lipoblastoma of the neck.
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** Choi HJ; Lee YM; Lee JH; Kim JW; Tark MS
**RESUMEN / SUMMARY:** BACKGROUND: Lipoblastoma is a rare, benign, and encapsulated tumor arising from embryonic white fat. Most of the cases occur in the extremities and the trunk; only a few cases in the head and the neck are reported. Thus, we present a case of lipoblastoma of the neck with a review of the literature.
**PATIENT AND METHOD:** A 1-year-old male infant presented to our hospital, with a history of painless swelling in the left side of the neck for 3 months that was rapidly enlarged. His birth history and medical history were unremarkable. A physical examination demonstrated a soft and compressible mass in the left side of the neck. The mass was nontender to palpation and mobile without cellulitic changes in the overlying skin. A computed tomographic scan showed that the mass is heterogenous, has low attenuation in nature, and is 3.8 x 2.8 x 9 cm in size. **RESULT:** Under general anesthesia, transverse cervical incision was made through the neck wrinkle, and there was no invasion of any of the neck structures. Complete surgical excision
demonstrated yellowish-white, irregular lobules of immature fat cells separated by a loose and myxoid connective tissue. Grossly, the mass was a homogeneous tan-pink gelatinous mass. A microscopic examination demonstrated a small number of capillaries and mature fat cells, and differentiating immature lipoblastoma cells were detected in the myxoid stroma. A pathologic finding confirmed the diagnosis of lipoblastoma. The postoperative course was uneventful. The patient underwent follow-up for 1 year after the operation, and there was no evidence of recurrence.

CONCLUSIONS: The most common presentation of lipoblastoma is a painless, rapidly enlarging neck mass. Published reports showed that most of them occur before the age of 3 years. Complete surgical excision is the treatment of choice. Although lipoblastoma is an extremely rare benign tumor, it should be considered in the diagnosis of neck mass in children younger than 3 years.

[357]

TÍTULO / TITLE: A Murmur-free Giant Myxoma Discovered Incidentally on Abdominal Ultrasonography.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Nishizaki Y; Yamagami S; Myojin M; Sesoko M; Yamashita H; Suzuki R; Daida H
INSTITUCIÓN / INSTITUTION: Department of Cardiology, Juntendo University School of Medicine, Juntendo Tokyo Koto Geriatric Medical Center, Japan.

RESUMEN / SUMMARY: Patients with myxoma normally present with cardiovascular symptoms due to mitral valve obstruction caused by the tumor. However, some cases are difficult to diagnose because the findings of auscultation are normal and there are no cardiovascular symptoms. A 62-year-old man presented at a nearby clinic with a fever. No cardiac murmurs were heard on a physical examination. Abdominal ultrasonography was conducted to evaluate the origin of the fever, and a giant left atrial myxoma was discovered incidentally. Although many myxoma cases are found on transthoracic echocardiography, we herein describe a case of a giant left atrial myxoma incidentally discovered on abdominal ultrasonography.

[358]

TÍTULO / TITLE: ENHANCED DEPTH IMAGING OPTICAL COHERENCE TOMOGRAPHY FEATURES OF CHOROIDAL OSTEOMA.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Pellegrini M; Invernizzi A; Giani A; Staurenghi G
INSTITUCIÓN / INSTITUTION: Eye Clinic, Department of Biomedical and Clinical Sciences, Luigi Sacco Hospital, University of Milan, Milan, Italy.
RESUMEN / SUMMARY: PURPOSE:: To describe the choroidal findings in eyes affected by choroidal osteoma imaged by enhanced depth imaging optical coherence tomography. METHODS:: Retrospective case series. RESULTS:: Seven eyes from five patients with choroidal osteoma were included in the study. Patients mean age of presentation was 26 years (median, 34; range, 6-37 years) and mean best-corrected
visual acuity was 20/32 (median, 20/20; range, 20/20-20/200). Enhanced depth imaging optical coherence tomography examination revealed normal inner retina in all the cases and normal outer retina in three eyes. Abnormalities included irregularities in external limiting membrane (n = 2), myoid zone (n = 1), ellipsoid junction (n = 4), cone outer segments of photoreceptors (n = 5), and retinal pigment epithelium (n = 3). Choroidal analysis revealed thinned (n = 4) or non visible (n = 2) choriocapillaris, thinned (n = 3) or non visible (n = 4) medium vessels, and thinned large vessels layer (n = 4). The osteoma showed multiple intralesional layers (n = 5), a sponge-like appearance (n = 7), and intralesional vessels (n = 7). The sclero-choroidal junction was visible in all cases. Choroidal neovascularization was found in four eyes. CONCLUSION: Analysis of eyes affected by choroidal osteoma revealed a characteristic sponge-like tumor appearance with the presence of multiple intralesional layers. The lesion showed a typical transparency with visibility of sclero-choroidal junction in all cases.

[359]
**Título / Title:** Enhanced CT and FDG PET/CT in Malignant Solitary Fibrous Tumor of the Lung.

**Resumen / Summary:**

Enlace al Resumen / Link to its Summary


Enlace al texto completo (gratuito o de pago)

1097/RLU.0000000000000281

**Autores / Authors:** Dong A; Zuo C; Wang Y; Cui Y

**Institución / Institution:** From the Departments of *Nuclear Medicine, Pathology, and Cardiac and Thoracic Surgery, Changhai Hospital, Second Military Medical University, Shanghai, China.

**Resumen / Summary:** Although some histological features of solitary fibrous tumor are related to its aggressive behavior, tumors with benign features may also follow an aggressive behavior. An 18-year-old male patient was referred after the detection of multiple bilateral pulmonary lesions on a chest x-ray in a health examination 2 months ago. These lesions showed inhomogeneous enhancement on enhanced CT and intense FDG uptake (SUVmax, 21.8) on PET/CT. CT-guided biopsy revealed histologically benign solitary fibrous tumor.

[360]
**Título / Title:** Surgical management of synchronous central giant cell granuloma and ossifying fibroma of the mandible.

**Resumen / Summary:**

Enlace al Resumen / Link to its Summary


Enlace al texto completo (gratuito o de pago)

1097/SCS.0b013e3182a2ddc4

**Autores / Authors:** Fasolis M; Zavattero E; Garzino-Demo P; Ramieri G; Berrone S

**Institución / Institution:** From the Division of Maxilofacial Surgery, Head and Neck Department, San Giovanni Battista Hospital, University of Turin, Turin, Italy.
RESUMEN / SUMMARY: - We describe the surgical management of an uncommon case of synchronous presentation of central giant cell granuloma and ossifying fibroma in the mandible. A mandibular resection was performed and a fibula-free flap was harvested to reconstruct the defect.

TÍTULO / TITLE: - Unusual early-stage pancreatic sarcomatoid carcinoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Ren CL; Jin P; Han CX; Xiao Q; Wang DR; Shi L; Wang DX; Chen H
INSTITUCIÓN / INSTITUTION: - Chuan-Li Ren, Ping Jin, Chong-Xu Han, Laboratory Medicine and Pathology Department, Northern Jiangsu People’s Hospital and Clinical Medical College of Yangzhou University, Yangzhou 225001, Jiangsu Province, China.
RESUMEN / SUMMARY: - Sarcomatoid carcinoma of the pancreas (SCP) is a very rare pathological type of carcinoma that usually has a poor prognosis. Its pathogenesis has not been elucidated. We herein report a case of an early-stage SCP involving successful treatment and a good prognosis. The patient was a 48-year-old Chinese man with a 5-mo history of vague abdominal pain. Ultrasonography revealed a 93 mm x 94 mm x 75 mm mass of mixed echogenicity in the tail of the pancreas. Laboratory test results were within the normal range, with the exception of an obviously increased pretreatment neuron-specific enolase level. The plasma transforming growth factor (TGF)beta1 and interleukin-11 levels were obviously increased according to enzyme-linked immunosorbent assay. Microscopically, the excised tumor tissue comprised cancer cells and mesenchymal cells. Immunohistochemical analysis was positive for alpha-1-antichymotrypsin, pan-cytokeratin, cytokeratin 19, cytokeratin 8/18, and vimentin and negative for CD68 and lysozyme. The pathogenetic mechanism of this case shows that TGFbeta1 may regulate the epithelial-to-mesenchymal transition in SCP. With early eradication of the tumor and systemic therapy, this patient has been alive for more than 3 years without tumor recurrence or distant metastasis. This case is also the first to show that TGFbeta1 may regulate the epithelial-to-mesenchymal transition in early-stage SCP.

TÍTULO / TITLE: - Wide excision and anterolateral thigh perforator flap reconstruction for dermatofibrosarcoma protuberans of the face.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Longo B; Paolini G; Belli E; Costantino B; Pagnoni M; Santanelli F
INSTITUCIÓN / INSTITUTION: - From the *Plastic Surgery Unit, and daggerMaxillofacial Surgery Unit, Department of Neuroscienze, Salute Mentale e Organi di Senso, Sant’Andrea Hospital, School of Medicine and Psychology, Sapienza University of Rome, Rome, Italy.

RESUMEN / SUMMARY: - Dermatofibrosarcoma protuberans is a rare cutaneous malignant tumor associated with a high cure rate but with a high incidence of local recurrence. Because of its tentacle-like subcutaneous infiltrating pattern that extends far beyond the clinically visible skin lesion, a wide resection margin is recommended. Hence, its localization to the head-and-neck regions, although rare, represents a real challenge for both the oncologic surgeon and the reconstructive surgeon, who aim to achieve a radical resection of the tumor with the best possible aesthetic outcome. A case of a 21-year-old Mediterranean man who presented with a 7-month history of a slowly growing subcutaneous lesion of the left preauricular region is reported. A diagnosis of dermatofibrosarcoma protuberans CD34+ was confirmed through surgical biopsy, and the patient subsequently underwent a wide en bloc local surgical resection, followed by anterolateral thigh perforator free flap reconstruction. Healing was uneventful. Initially, there was some facial nerve neurapraxia; however, this completely subsided within 3 months after the surgery. At the 13-month follow-up, the patient was completely well and free from the disease.

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[363]

TÍTULO / TITLE: - Plexiform fibrohistiocytic tumor of bone.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Yalcinkaya U; Uz Unlu M; Bilgen MS; Yazici Z

INSTITUCIÓN / INSTITUTION: - Department of Surgical Pathology, Uludag University Medical School, Bursa, Turkey.

RESUMEN / SUMMARY: - Plexiform fibrohistiocytic tumor is an extremely rare soft tissue tumor with a low malignancy potential. The patient is usually a child or a young adolescent and the tumor is usually localized in the upper extremities. We report on a case of a 21-year-old male with a plexiform fibrohistiocytic tumor in the left fibula admitted to our hospital due to a swelling and pain in the left lower extremity. Radiologically a lytic lesion in the distal end of left fibula consistent with a non-aggressive lesion with low biological activity was found. Treated with curettage, the specimen revealedplexiform proliferation of mononuclear histiocyte-like cells, multinucleated osteoclast-like cells, and spindle fibroblast-like cells in variable proportions histopathologically. Immunohistochemical stains were positive for CD68 in scattered fashion in histiocytes and giant cells, and spindle like cells showed positivity for smooth muscle actin. Under electron microscopy, rough endoplasmic reticulum and collagen bundles in the spindle cells suggested fibroblastic differentiation. Also multiple large electron-dense lysosomal granules in histiocytoid cells were found. Multinucleated giant cells exhibited osteoclast-like appearance. All these findings suggested plexiform fibrohistiocytic tumor. Interestingly, the tumor was localized in bone. During the follow up for 27 months after the resection, there was no recurrence or metastasis.
- Multiple linear leiomyomas of the forehead as the presenting sign of Reed syndrome.

- A 62-year-old female presented with a linear arrangement of multiple asymptomatic, discrete, dome-shaped, smooth, skin-colored papules and nodules involving the left forehead. Histopathology showed a poorly circumscribed nodule of haphazardly arranged fascicles of smooth muscle cells involving the papillary and superficial reticular dermis. Genetic testing revealed the patient to be heterozygous for the R233H mutation in the fumarate hydratase gene. Clinical, microscopic, and genetic findings were consistent with a diagnosis of Reed syndrome. Reed syndrome is a rare disorder defined by cutaneous and uterine leiomyomas and, uncommonly, renal cell carcinoma.

- AIDS-related Kaposi's sarcoma in Brazil: trends and geopolitical distribution.

- BACKGROUND: AIDS-related Kaposi's sarcoma (KS) is a unique model of the relationship between viral infection, immunity, environmental, and genetic factors in viral cancers. The goal was to determine the distribution of KS cases among Brazilian geopolitical regions, looking at the ecological relationship with median CD4 cell count. METHODS: Ecological study using Brazilian National Diseases Reporting Databases: 1982-2009. Subjects >/=13 years of age who have KS cited in their AIDS reporting form were selected, and demographic and HIV exposure data were collected. RESULTS: We found 11,731 KS cases in the period, with a prevalence of 2.4% among AIDS cases; 88% were male, and 68% lived in the Southeast region, which accounted for 59% of AIDS cases. The regional and national prevalence trends were similar, although the highest proportion among women was found in the North region, which has the lowest number of both AIDS and KS cases. Heterosexual transmission accounted for 87% of HIV among women compared to 18% among men. Fifty-seven percent of all KS cases were diagnosed before antiretroviral therapy (ART). Injection drug use accounted for 11% of KS cases. Median survival was 472 days before the ART era and 1482 after it (P < 0.001). Median CD4 counts increased in all
regions in the period as ART coverage expanded, and a resulting correlating decline in KS cases was observed. CONCLUSIONS: Prevalence of KS declined after the introduction of ART in all regions of Brazil, suggesting individual protection conveyed by ART.

[366]
TÍTULO / TITLE: - Multiple dermatofibromas subsequent to folliculitis.  
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary  
 ●● Enlace al texto completo (gratuito o de pago) 1684/ejd.2013.2164
AUTORES / AUTHORS: - Watanabe K; Fukuda H; Niiyama S; Oharasaki T; Mukai H
INSTITUCIÓN / INSTITUTION: - Department of Dermatology.

[367]
TÍTULO / TITLE: - Chondrosarcomas of the head and neck.  
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary  
 ●● Enlace al texto completo (gratuito o de pago) 1007/s00405-013-2807-3
AUTORES / AUTHORS: - Coca-Pelaz A; Rodrigo JP; Triantafyllou A; Hunt JL; Fernandez-Miranda JC; Strojan P; de Bree R; Rinaldo A; Takes RP; Ferlito A
INSTITUCIÓN / INSTITUTION: - Department of Otolaryngology, Hospital Universitario Central de Asturias, Oviedo, España.
RESUMEN / SUMMARY: - Chondrosarcoma represents approximately 11% of all primary malignant bone tumors. It is the second most common sarcoma arising in bone after osteosarcoma. Chondrosarcomas of the head and neck are rare and may involve the sinonasal tract, jaws, larynx or skull base. Depending on the anatomical location, the tumor can produce a variety of symptoms. Computed tomography and magnetic resonance imaging are the preferred imaging modalities. The histology of conventional chondrosarcoma is relatively straightforward; major challenges are the distinction between grade I chondrosarcomas and chondromas, and the differential diagnosis with chondroblastic osteosarcoma and chondroid chordoma. Surgery alone or followed by adjuvant radiotherapy is the treatment of choice. Radiotherapy alone has also been reported to be effective and can be considered if mutilating radical surgery is the only curative alternative. The 5-year survival for chondrosarcoma reaches 80%; distant metastases and/or local recurrences significantly worsen prognosis. The present review aims to summarize the current state of information about the biology, diagnosis and management of these rare tumors.

[368]
TÍTULO / TITLE: - Laparoscopy-assisted resection of ileocecal intussusception caused by ileal pedunculated lipoma.  
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary  
 ●● Enlace al texto completo (gratuito o de pago) 9738/INTSURG-D-13-00011.1
AUTORES / AUTHORS: - Saito K; Osawa H; Morohara K; Nakamura K; Kimura S; Okada A; Sakai M; Wada W; Yasuda N; Suzuki Y; Kuwano H
INSTITUCIÓN / INSTITUTION: - 1 Department of Surgery, Isesaki Municipal Hospital, Isesaki, Japan.
RESUMEN / SUMMARY: - Abstract We report on a case of ileal lipoma that prolapsed into the ascending colon and was resected by laparoscopy-assisted surgery. A 31-year-old male Japanese patient was admitted to our hospital because of hematochezia and anemia. Colonoscopy revealed a pedunculated polyp arising from the ileum. Colonicoscopy showed a low-density mass in the ascending colon. A diagnosis of pedunculated ileal lipoma with intussusception was made, and laparoscopy-assisted surgery was performed. The intussusception was reduced by resection of the lipoma. The surgical specimen was a 40 x 30 x 25 mm round tumor with a long stalk 11 cm in length. Microscopic examination of the specimen revealed ileal lipoma. Laparoscopic surgery is recommended for benign tumors of the small intestine because it is minimally invasive.
A case of melanoma with rhabdomyoblastic differentiation is presented in the context of the previously reported cases. The emerging literature seeking to identify the molecular basis of rhabdoid and rhabdomyoblastic differentiation, as well as their poor prognosis, is reviewed. The combination of a diverse range of morphology and the potential for spontaneous primary tumor regression, despite metastasis, makes the accurate diagnosis of melanoma challenging. Histopathology review is often recommended in these cases, as is referral to a specialized cancer center for discussion in a multidisciplinary meeting. Improved recognition of this rare pattern of melanoma morphology may provide the means for omics-based techniques to identify novel therapeutic targets to improve the prognostic outlook for these patients.

[371] Fibroma of the tendon sheath of the long head of the biceps tendon.

A fibroma of the tendon sheath of the long head of the biceps tendon is a benign tumor that is less common than giant cell tumor of the tendon sheath. Both tumors may present as a painless, slowly enlarging mass. Radiological findings may be similar for both tumors. Histologically, fibroma of the tendon sheath lacks the hemosiderin-laden macrophages that are typical for giant cell tumor of the tendon sheath. We report on a 49-year-old woman with fibroma of the tendon sheath of the long head of the biceps tendon. In our case, on MR images, we observed band-like hypointense areas centrally in the tumor, mild patchy contrast enhancement, and most importantly, no decrease of signal intensity on gradient echo images. These characteristics reflected histological findings.

[372] Intra-articular fibroma of tendon sheath arising in the acromioclavicular joint.

Fibromas of the tendon sheath arising in the acromioclavicular joint are rare. They may present as a painless, slowly enlarging mass. Radiologically, they may be similar to other lesions such as ganglion cysts and bursitis. We report on a 32-year-old woman with a fibroma of the tendon sheath arising in the acromioclavicular joint. In our case, on MR images, we observed a well-defined, hypointense mass centrally in the tumor, mild patchy contrast enhancement, and no decrease of signal intensity on gradient echo images. These characteristics reflected histological findings.
**RESUMEN / SUMMARY:** Fibroma of the tendon sheath, a rare benign soft tissue tumor that most often occurs in the distal upper extremities (hands and wrist), is exceedingly rare to present as an intraarticular mass. Presented here is the first case in the English literature, to our knowledge, of a fibroma of the tendon sheath arising in the acromioclavicular joint. The patient presented with recurrent shoulder pain with activity without antecedent trauma. Radiographs were essentially normal. MR images demonstrated a lobulated, heterogeneous mass with contrast enhancement arising from the acromioclavicular joint. Following surgical resection, histopathology revealed hypocellular collagen matrix with spindle-shaped fibroblasts, confirming the diagnosis of fibroma of tendon sheath. The imaging features of the fibroma of the tendon sheath and a brief review of the literature are presented.

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**TÍTULO / TITLE:** Nodular fasciitis in the buccal region with rapid growth after incisional biopsy mimicking sarcoma.

**RESUMEN / SUMMARY:** Nodular fasciitis (NF) is a reactive and proliferative fibroblastic lesion that occurs predominantly in the upper limbs but rarely develops in the oral cavity. This lesion can be misdiagnosed as malignant owing to its frequent display of rapid growth, rich cellularity, and high mitotic activity. Unlike a sarcoma, NF can resolve spontaneously or after an incisional biopsy. We describe a challenging case involving a lesion in the buccal region that rapidly enlarged after incisional biopsy. This variation of clinical behavior illustrates the difficulty in predicting whether NF will continue to grow or regress. Clinicians dealing with cases of an enlarging fibrous lesion of short duration should remain aware of this disease entity and its potential diagnostic dilemma.

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**TÍTULO / TITLE:** Carcinosarcoma of the Uterine Corpus on 18F-FDG PET/CT in a Postmenopausal Woman With Elevated AFP.

**RESUMEN / SUMMARY:** Carcinosarcoma of the Uterine Corpus on 18F-FDG PET/CT in a Postmenopausal Woman With Elevated AFP.
RESUMEN / SUMMARY: - Uterine carcinosarcoma (termed malignant mixed mullerian tumor) is a rare neoplasm of the uterus with a poor prognosis. There have been very few cases in the literature describing the PET/CT findings of uterine carcinosarcoma. We report a case of tissue-proven carcinosarcoma of the uterine corpus in a 65-year-old woman with elevated serum alpha-fetoprotein (AFP), whose F-FDG PET/CT showed a 10.3-cm mass in the uterus with uneven high FDG uptake. The SUVmax was 12.8. After surgery, the patient received 6 courses of chemotherapy, and the serum levels of AFP decreased to reference range.

[375]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Lu D; Yang H; Liu SX; Zhao Y; Chen F
INSTITUCIÓN / INSTITUTION: - From the Department of Otorhinolaryngology, Head and Neck Surgery, West China Hospital, Sichuan University, Sichuan, China.
RESUMEN / SUMMARY: - Angiomyolipomas are benign neoplasms composed of smooth muscle, vasculature, and mature adipose tissue, which most commonly occur in the kidney and located in the head and neck region. A very rare neoplasm, there are only 3 cases of angiomyolipoma in the parotid gland that have been reported to date. Here, we report a case of a 38-year-old man who had a slow-growing mass in the parotid gland for the past 7 years. The results of a physical examination revealed a rubbery mass that was 2.5 cm in diameter in the below superficial lobe of the left parotid gland. A computed tomographic scan showed a heterogeneous and lobulated nodule with a well-defined margin, which was resected through partial parotidectomy with preservation of the facial nerve. A histologic finding revealed an angiomyolipoma of the parotid gland. In conclusion, angiomyolipoma should be considered in the differential diagnosis of rubbery parotid gland masses.

[376]
TÍTULO / TITLE: - Unusual presentation of monostatic fibrous dysplasia in zygoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Verma A; Jindal N; Singh V; Sethi P
INSTITUCIÓN / INSTITUTION: - From the *Department of Oral & Maxillofacial Surgery, PDM Dental College, Bahadurgarh; daggerDepartment of Oral & Maxillofacial Surgery, Government Dental College, PGIMS, Rohtak; and double daggerDepartment of Oral Pathology, Swami Devi Dayal Hospital & Dental College, Panchkula, India.
RESUMEN / SUMMARY: - Fibrous dysplasia is a nonneoplastic, developmental disease of the bone that begins in childhood with obscure etiology. Clinically, it is presented as
a continuously growing, painless mass at late childhood. Maxilla and mandible are mostly involved in facial skeleton. Involvement of the zygomatic bone is rare. Fibrous dysplasia of the zygomatic bone may cause orbital dystopia, diplopia, proptosis, loss of visual acuity, swelling, mass formation, or facial asymmetry. We present 1 case of fibrous dysplasia with isolated zygomatic bone involvement.
LEIOMYOSARCOMAS OF THE HEAD AND NECK

Leiomyosarcoma of the head and neck is an extremely rare entity that because of its infrequency has been associated with both delayed diagnosis and misdiagnosis. Sinonasal tract is the most common site in this region. The overall prognosis is poor. It is necessary for appropriate immunohistochemical investigation for accurate diagnosis. Tonsillar leiomyosarcoma presented only 1 case in the English-language literature (PubMed, Ovid, and Proquest databases). We report a second case of leiomyosarcoma arising in the tonsil in a 38-year-old woman. Level of Evidence: Level IV therapeutic study.

TÍTULO / TITLE: - Solitary osteoma in the zygomatic arch.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

TÍTULO / TITLE: - Postradiation sarcoma from a free flap.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Postradiation sarcomas constitute approximately 0.5% to 5.5% of all sarcomas. They develop locally approximately 3 to 20 years after the administration of radiotherapy (RT). They are generally high-grade tumors. Osteosarcomas, fibrosarcomas, malignant fibrous histiocytoma, angiosarcomas, and leiomyosarcomas are the most frequently observed. It is rare for these tumors to originate from free flaps, and this patient report is one of the first in the literature. **PATIENT:** A 59-year-old man was operated on because of ethmoid sinus cancer in 2004, and the reconstruction was performed with a rectus abdominis free muscle flap. He received postoperative RT and subsequently presented to our clinic with a medially protruding mass on his upper jaw. A biopsy was performed. Its pathologic diagnosis was reported as malignant mesenchymal tumor. Computed tomography and magnetic resonance imaging were performed, demonstrating that the mass originated from the free muscle flap (m. rectus abdominis) at the front wall of the sphenoid sinus. A total excision of the free muscle flap and near-total maxillectomy were performed. The pathologic finding was reported as leiomyosarcoma with bone invasion. **DISCUSSION:** With the advancement of medical and pharmaceutical technologies, our patient’s life expectancy is increasing. In long-living patients who have received RT, tumors can develop 20 years after the RT. The close follow-up of patients receiving RT is of utmost importance because treatment survival is linked to early diagnosis and resection with negative surgical margins. We must not forget that, even if years have passed since receiving RT, these patients may present with such tumors.

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**[382]**
**TITULO / TITLE:** Intracranial metastasis from a “giant” nonoperated sacrococcygeal chordoma. An underestimated metastatic potential?
**RESUMEN / SUMMARY:** [Enlace al Resumen / Link to its Summary](#)
  ●● Enlace al texto completo (gratuito o de pago) [3109/02688697.2013.847175](#)
**AUTORES / AUTHORS:** Della Pepa GM; Visocchi M
**INSTITUCIÓN / INSTITUTION:** Institute of Neurosurgery, Catholic University of Rome, Italy.

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**[383]**
**TITULO / TITLE:** Primary osteosarcoma of clivus: a short report.
**RESUMEN / SUMMARY:** [Enlace al Resumen / Link to its Summary](#)
  ●● Enlace al texto completo (gratuito o de pago) [3109/02688697.2013.841852](#)
**AUTORES / AUTHORS:** Mohindra S; Savardekar A; Mahalingam SS; Mohindra S; Vashista RK
**INSTITUCIÓN / INSTITUTION:** Department of Neurosurgery, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
**RESUMEN / SUMMARY:** Next to multiple myeloma, osteosarcoma is the commonest primary malignant neoplasm of skeletal system. These are aggressive tumours,
composed of spindle cells producing osteoid and rarely occur in calvaria. The authors report a 55-year-old male harbouring clival osteosarcoma, his clinical presentation, radiological findings, management and outcome.

[384]
TITULO / TITLE: Specific ultraviolet-C irradiation energy for functionalization of titanium surface to increase osteoblastic cellular attachment.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: J Biomater Appl. 2013 Nov 27.
AUTORES / AUTHORS: Uchiyama H; Yamada M; Ishizaki K; Sakurai K
INSTITUCION / INSTITUTION: Department of Removable Prosthodontics and Gerodontontology, Tokyo Dental College, Tokyo, Japan.
RESUMEN / SUMMARY: Purpose: The objective of this in vitro study was to examine the influence of the total energy of ultraviolet-C preirradiation on the number and morphology of osteoblastic cells attached to turned or acid-etched titanium surfaces, and physicochemical properties of the surface. Materials and methods: Rat bone marrow-derived osteoblasts were incubated with turned or acid-etched titanium disks preirradiated with ultraviolet-C at 1 or 3 mW/cm², resulting in total energies of 10, 100, 250, 400, 500, 600, 750, or 1000 J/cm². Osteoblast attachment to the surface was evaluated using the WST-1 assay. Physicochemical changes of the titanium were evaluated by measuring water wettability and X-ray photoelectron spectroscopy analysis. Results: Number of attached cells was greater on turned or acid-etched surface preirradiated with 500 or 750 J/cm² of 3 mW/cm² ultraviolet-C than on the nonirradiated surface, respectively. However, the further irradiation energy did not increase the numbers on both types of the surfaces. These phenomena were also seen on the surfaces preirradiated at different ultraviolet-C intensities. Ultraviolet-C irradiation induced superhydrophilicity on both types of surface even with the less irradiation energy. The amount of carbon on ultraviolet-C preirradiated titanium surfaces decreased gradually with an increase in the total irradiation energy. Conclusion: Specific ultraviolet-C energy used to irradiate turned or acid-etched surfaces increased the number of osteoblastic cells attached to each of the surface. This was canceled by overirradiation, despite maintenance of both the acquired superhydrophilicity and the accompanying reduction in carbon on each surface.

[385]
TITULO / TITLE: Osteoid osteoma.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Boscainos PJ; Cousins GR; Kulshreshtha R; Oliver TB; Papagelopoulos PJ
RESUMEN / SUMMARY: Osteoid osteomas consist of a nidus with surrounding sclerotic bone. The differential diagnosis covers a wide range of conditions due to the
variable presentation of osteoid osteoma. The natural history is for regression to occur within 6 to 15 years with no treatment; however, this can be reduced to 2 to 3 years with the use of aspirin and non-steroidal anti-inflammatory drugs. Computed tomography-guided percutaneous techniques, including trephine excision, cryoablation, radiofrequency ablation, and laser thermocoagulation, are described.

[386]
**TÍTULO / TITLE:** Overexpression of CD40 in sacral chordomas and its correlation with low tumor recurrence.

**RESUMEN / SUMMARY:** To determine the degree of CD40 overexpression in sacral chordomas and its correlation with tumor recurrence. METHODS: CD40 or CD31 overexpression was determined by immunohistochemical staining; the microvessel density (MVD) was calculated according to the CD31 expression. The correlation of CD40 over-expression with tumor recurrence was analyzed. RESULTS: 56% of the specimens from 36 cases of sacral chordomas overexpressed CD40, which is a significantly higher percentage than for the 2 specimens in 10 in normal notochordial tissue (p < 0.05). 36.84% of the specimens of the 19 recurrent cases were CD40 overexpressing, in contrast to less than 76.47% in the non-recurrence group (p < 0.05). Multivariate analysis demonstrated that CD40 overexpression and the resection margins were independent factors contributing to tumor recurrence. The MVD value was 25.71 +/- 8.86 mm(-2) in the sacral chordomas and more than 6.63 +/- 2.45 mm(-2) in the normal embryonic notochord tissue (p < 0.01). The MVD value in the recurrence group (30.08 +/- 7.11 mm(-2)) was significantly higher than that of the non-recurrence group (20.82 +/- 8.18 mm(-2); p < 0.05). But the MVD value was significantly lower in the CD40-overexpressing group than in the CD40-less expressing group (p < 0.05). CONCLUSIONS: CD40 was overexpressed in sacral chordomas, and the overexpression was not dependent on the intratumoral MVD. CD40 overexpression was correlated with low recurrence of the tumor, implying that CD40 plays an important role in the antitumor response against sacral chordomas and in the inhibition of tumor recurrence.

[387]
**TÍTULO / TITLE:** Bone Marrow Metastases From Alveolar Rhabdomyosarcoma With Impressive FDG PET/CT Finding But Less-Revealing Bone Scintigraphy.

**RESUMEN / SUMMARY:** To determine the degree of CD40 overexpression in sacral chordomas and its correlation with tumor recurrence. METHODS: CD40 or CD31 overexpression was determined by immunohistochemical staining; the microvessel density (MVD) was calculated according to the CD31 expression. The correlation of CD40 over-expression with tumor recurrence was analyzed. RESULTS: 56% of the specimens from 36 cases of sacral chordomas overexpressed CD40, which is a significantly higher percentage than for the 2 specimens in 10 in normal notochordial tissue (p < 0.05). 36.84% of the specimens of the 19 recurrent cases were CD40 overexpressing, in contrast to less than 76.47% in the non-recurrence group (p < 0.05). Multivariate analysis demonstrated that CD40 overexpression and the resection margins were independent factors contributing to tumor recurrence. The MVD value was 25.71 +/- 8.86 mm(-2) in the sacral chordomas and more than 6.63 +/- 2.45 mm(-2) in the normal embryonic notochord tissue (p < 0.01). The MVD value in the recurrence group (30.08 +/- 7.11 mm(-2)) was significantly higher than that of the non-recurrence group (20.82 +/- 8.18 mm(-2); p < 0.05). But the MVD value was significantly lower in the CD40-overexpressing group than in the CD40-less expressing group (p < 0.05). CONCLUSIONS: CD40 was overexpressed in sacral chordomas, and the overexpression was not dependent on the intratumoral MVD. CD40 overexpression was correlated with low recurrence of the tumor, implying that CD40 plays an important role in the antitumor response against sacral chordomas and in the inhibition of tumor recurrence.
An F-FDG PET/CT scan was performed in a 26-year-old man with a known alveolar rhabdomyosarcoma for staging. The PET/CT scan showed abnormally increased FDG activity involving almost all bones in the imaged regions. In contrast, Tc-MDP whole-body bone scan demonstrated only very limited bone metastases.
There was an additional large FDG activity in the right atrium. The subsequent studies demonstrated that the patient experienced pulmonary embolism derived from the right atrial myxoma.

[390]

**TÍTULO / TITLE:** - Cardiac Metastasis of a Low-Grade Myofibroblastic Sarcoma.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Oylumlu M; Yildiz A; Ercan S; Oylumlu M; Davutoglu V

**INSTITUCIÓN / INSTITUTION:** - Department of Cardiology, Sehitkamil State Hospital, Gaziantep, Turkey.

**RESUMEN / SUMMARY:** - We aim to present a rare case of low-grade myofibroblastic sarcoma arising in the inguinal region accompanied by cardiac metastasis. A 36-year-old male patient suffering from recurrent inguinal swelling was operated on and the initial histopathological evaluation mistakenly diagnosed the condition as benign. During follow-up, a recurrence of mass was detected in the same region and a pathological examination revealed a low-grade myofibroblastic sarcoma. Cardiac metastasis was diagnosed shortly before rapid disease progression and death. The learning points relevant to this case are as follows: (1) Echocardiographic screening in patients with noncardiac myofibroblastic sarcomas may be helpful in the detection of silent metastasis. (2) Low-grade myofibroblastic sarcomas in the inguinal region may be misdiagnosed as benign after enucleation. Thus, rigorous histopathological examination of myofibroblastic sarcomas is crucial. (3) According to our knowledge, this is the first report of a low-grade myofibroblastic sarcoma to have a potential for cardiac metastasis with potentially fatal course.

[391]

**TÍTULO / TITLE:** - Huge Primary Soft Tissue Sarcoma of the Breast on Bone Scan.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Shen YY; Wu YC; Kao CH; Hsieh TC

**INSTITUCIÓN / INSTITUTION:** - From the *Department of Nuclear Medicine, Shin Kong Wu Ho-Su Memorial Hospital, Taipei City; daggerSchool of Medicine, College of Medicine, Fu Jen Catholic University, Xinzhuang District, New Taipei City; double daggerDepartment of Nuclear Medicine, National Taiwan University Hospital Hsinchu Branch, Hsinchu City; and section signDepartment of Nuclear Medicine and PET Center, China Medical University Hospital, and paragraph signSchool of Medicine, and parallelDepartment of Biomedical Imaging and Radiological Science, China Medical University, Taichung City, Taiwan.

**RESUMEN / SUMMARY:** - A 54-year-old woman had a primary breast sarcoma with rapid enlargement in 3 months. The mass became so huge that it was more than 20 cm in diameter and occupied the entire right breast on presentation. Extraosseous
uptake was present in this mass and demonstrated a unique picture, mimicking the posture of a racing driver who holds a helmet under the armpit, on the bone scan.

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TÍTULO / TITLE: - Myeloid sarcoma with megakaryoblastic differentiation mimicking a sellar tumor.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Novello M; Coli A; Della Pepa GM; Martini M; Doglietto F; De Stefano V; Bellesi S; Pescarmona E; Lauriola L
INSTITUCIÓN / INSTITUTION: - Department of Anatomic Pathology, Catholic University, Rome, Italy.

RESUMEN / SUMMARY: - Myeloid sarcoma (MS) is a localized extra-medullary tumor mass of immature myeloid cells, arising de novo or related to acute myeloid leukemia, of which it can be a forerunner, a coinciding or late event. Less commonly, MS represents an acute blastic transformation of myelodysplastic syndromes or myeloproliferative neoplasms. This rare condition commonly consists of a proliferation of more or less immature cells with a myeloid immunophenotype, very exceptional cases showing a megakaryoblastic or erythroid differentiation. The most common localization of MS is the skin, lymph node, soft tissues and bones, but CNS involvement is exceedingly rare, with no cases reported in the sellar region. We report a 54-year-old man, affected by myeloproliferative neoplasm, JAK2 V617F-positive of 13 years duration, who acutely presented with a third cranial nerve palsy; neuroradiology documented a space-occupying lesion at the level of the sellar, upper clival and right parasellar regions, that was sub-totally removed with a trans-sphenoidal approach. The histological examination documented a proliferation of large, blastic cells, frequently multinucleated; a diagnosis of MS with megakaryoblastic differentiation, arising in a background of chronic idiopathic myelofibrosis, was suggested by immunohistochemistry, owing to CD42b, CD45, CD61 and LAT (linker for activation of T cells) positivity. In addition, homozygous JAK2 V617F mutation was detected from the myeloid sarcoma specimen. A few weeks after surgery, an acute blastic leukemic transformation occurred and, despite chemotherapy, the patient died 2 months after surgery. To the best of our knowledge, this is the first MS case with megakaryoblastic differentiation arising within the CNS.

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TÍTULO / TITLE: - Primitive Myxoid Mesenchymal Tumor of Infancy With Rosettes: A New Finding and Literature Review.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cipriani NA; Ryan DP; Nielsen GP

RESUMEN / SUMMARY: - Primitive myxoid mesenchymal tumor of infancy (PMMTI) is a relatively recently described tumor arising in infants and demonstrating a unique histomorphology. We present an unusual case of PMMTI with rosettes, a hitherto
undescribed finding in the reported cases. We also present the cytogenetic and ultrastructural findings of this tumor and review the literature. As awareness of PMMTI increases, additional clinical data and histopathologic findings will aid in the morphologic and behavioral characterization of this neoplasm.

[394]
**TITULO / TITLE:** - Sarcomas, example of a pathologist network organization.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Neuville A; Coindre JM
**INSTITUCIÓN / INSTITUTION:** - Universite Victor-Segalen, institut Bergonie, departement de pathologie, laboratoire d'anatomie pathologie et Inserm U916, 229, cours de l'Argonne, 33076 Bordeaux Cedex, France.
**RESUMEN / SUMMARY:** - Sarcomas are rare and heterogeneous with many subtypes explaining the high level of diagnostic difficulty with frequent important therapeutic consequences. In 2009, a national network of pathologists has been set up with the main objective to perform a systematic histological review of every new sarcoma, gastro-intestinal stromal tumor (GIST) and desmoid tumor. We describe the network organization and report the results of the first two years of activity. These results clearly show the interest of this organization for the patients as well as for all pathologists. Moreover, data and material collect allows a better knowledge of these tumors and an improvement of the rules for their diagnostic management.

[395]
**TITULO / TITLE:** - Signs of cell-cell interactions in sarcoma 45 tissue under conditions of antitumor effect caused by injection of magnetite nanoparticles.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Gudtskova TN; Zhukova GV; Bragina MI; Garkavi LKh; Mikholap AI; Barteneva TA
**INSTITUCIÓN / INSTITUTION:** - Rostov Institute of Oncology, Ministry of Health of the Russian Federation, Rostov-on-Don, Russia. galya_57@mail.ru.
**RESUMEN / SUMMARY:** - Changes in transplanted sarcoma 45 tissue in outbred albino rats with tumor regression under the effect of magnetite nanoparticles (magnetic fluid) were studied by light and electron microscopy. The ultrastructure and cell death types in regressing tumors and signs of cell-cell interactions with participation of macrophages, lymphocytes, neutrophils, and degranulating mast cells were described. Some possible mechanisms of a pronounced antitumor activity of magnetite nanoparticles were discussed.

[396]
**TITULO / TITLE:** - Genetic analysis of the fused in sarcoma gene in Chinese Han patients with Parkinson’s disease.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Gao K; Zheng W; Deng X; Xiong W; Song Z; Yang Y; Deng H

**INSTITUCIÓN / INSTITUTION:** - Center for Experimental Medicine, The Third Xiangya Hospital, Central South University, Changsha, China.

**RESUMEN / SUMMARY:** - BACKGROUND AND PURPOSE: Exome sequencing in a large essential tremor (ET) family identified a novel nonsense mutation (p.Q290X) in the fused in sarcoma gene (FUS) as the cause of this family. Because of the clinical overlap between ET and Parkinson’s disease (PD), the role of FUS in an independent cohort of PD patients from China mainland was evaluated. METHODS: The entire coding region of FUS in 508 Chinese Han patients with PD and the identified variants in 633 normal controls were evaluated. A variant was further screened in an additional 382 controls for the frequency in our population. RESULTS: A novel variant c.696C > T (p.Y232Y) in 2 sporadic patients with PD and six variants (c.52C > A, p.P18T; c.52C > T, p.P18S; c.147C > A, p.G49G; c.291C > T, p.Y97Y; c.684C > T, p.G228G; c.1176G > A, p.M392I) without significant difference in genotypic and allelic distributions in our PD cohort were identified. CONCLUSION: The FUS gene is not a genetic risk factor for PD in the population of Chinese Han ethnicity.

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**AUTORES / AUTHORS:** - Dadlani R; Ghosal N; Hegde AS; Gupta K

**INSTITUCIÓN / INSTITUTION:** - Department of Neurosurgery, Sri Satya Sai Institute of Higher Medical Sciences, Bangalore, India.

**RESUMEN / SUMMARY:** - We describe to our knowledge the first patient with giant desmoplastic fibroblastoma of the calvarium in a 20-year-old woman whose tumor first appeared at the age of 5 years. We also discuss the histopathological differential diagnosis, management dilemmas and complications of desmoplastic fibroblastoma.

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**AUTORES / AUTHORS:** - Hattab EM; Dvorscak LE; Boaz JC; Douglas AC; Ulbright TM

**INSTITUCIÓN / INSTITUTION:** - Department of Pathology and Laboratory Medicine, Indiana University School of Medicine, Indianapolis, Indiana; Department of Neurological Surgery, Indiana University School of Medicine, Indianapolis, Indiana.

**RESUMEN / SUMMARY:** - Cranial fasciitis is a rare lesion of young children characterized by proliferation of fibroblastic spindle cells. Most are scalp masses and
are only rarely intracranial, where an association with radiation therapy is exceptional. We report a 32-month-old toddler with a facial rhabdomyosarcoma, diagnosed at 3 months of age, and treated with surgery, chemotherapy and brachytherapy. Brain MRI at 28 months revealed a large, left parasagittal, dural-based, T2 hyperintense and T1 hypointense enhancing mass with superior sagittal sinus compression and bony hyperostosis. The mass was completely resected during an open craniotomy. Histologically, the lesion was comprised of loosely and haphazardly arranged bland spindle cells embedded in a myxoid background. Thick hyalinized collagen bundles were especially prominent. The spindle cells reacted for vimentin but not SMA, myogenin, MyoD1 or EMA. A diagnosis of cranial fasciitis was rendered. The role of radiation therapy in the pathogenesis of intracranial cranial fasciitis is discussed.

[399]

TÍTULO / TITLE: - MRP1 overexpression determines poor prognosis in prospectively treated localized high risk soft tissue sarcoma patients of limbs and trunk wall. An ISG/GEIS study.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Martin-Broto J; Gutierrez AM; Ramos RF; Lopez-Guerrero JA; Ferrari S; Stacchiotti S; Picci P; Calabuig S; Collini P; Gambarotti M; Bague S; Dei Tos AP; Palassini E; Luna P; Cruz J; Cubedo R; Martinez-Trufero J; Poveda A; Casali PG; Fernandez-Serra A; Lopez-Pousa A; Gronchi A

INSTITUCIÓN / INSTITUTION: - 1Oncology, University Hospital Son Dureta.

RESUMEN / SUMMARY: - Patients with localized high risk STS of limbs and trunk wall still have a considerable metastatic recurrence rate of more than 50%, in spite of adjuvant chemotherapy. This drug ceiling effect of chemotherapy in sarcoma setting could be explained, at least partially, by MDR mechanisms. The aim of the current study was to ascertain whether mRNA and protein expression of ABCB1 (P-gp), ABCC1 (MRP1) and GSTA1 (GST-pi) was prognostic in localized high risk STS. Immunohistochemistry and RT-PCR studies were performed from biopsies at the time of diagnosis. Patients of this series were prospectively enrolled into a phase III trial which compared 3 vs. 5 cycles of epirubicin plus ifosfamide. The series of 102 patients found 41 events of recurrence and 37 of death with a median follow-up of 68 months. In univariate analysis, variables with a statistically significant relationship with RFS were: MRP1 expression (5-year RFS rate of 23% in positive cases and 63% in negative cases, p=0.029), histology (5-year RFS rate of 74% in UPS and 43% in synovial sarcoma, p=0.028) and ABCC1 expression (5-year RFS rate of 33% in overexpression and 65% in downregulation, p=0.012). Combined ABCC1/MRP1 was the only independent prognostic factor for both RFS (HR 2.704, p=0.005) and OS (HR 2.208, p=0.029). ABCC1/MRP1 expression shows robust prognostic relevance in localized high risk STS patients treated with anthracycline based chemotherapy, which is the standard front line treatment in STS. This finding deserves attention as it points to a new targetable protein in STS.

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RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Puri A; Gulia A; Hawaldar R; Ranganathan P; Badwe RA

INSTITUCIÓN / INSTITUTION: Orthopaedic Oncology, Tata Memorial Hospital, Room No. 45, E Borges Road, Mumbai, India, docpuri@gmail.com.

RESUMEN / SUMMARY: BACKGROUND: Whether current postoperative surveillance regimes result in improved overall survival (OS) of patients with extremity sarcomas is unknown. QUESTIONS/PURPOSES: We hypothesized that a less intensive followup protocol would not be inferior to the conventional followup protocol in terms of OS. We (1) assessed OS of patients to determine if less intensive followup regimens led to worsened survival and asked (2) whether chest radiograph followup group was inferior to CT scan followup group in detecting pulmonary metastasis; and (3) whether less frequent (6-monthly) followup interval was inferior to more frequent (3-monthly) followup in detecting pulmonary metastasis and local recurrence. METHODS: A prospective randomized single-center noninferiority trial was conducted between January 2006 and June 2010. On the basis of 3-year survival of 60% with intensive, more frequent followup, 500 nonmetastatic patients were randomized to demonstrate noninferiority by a margin (delta) of 10% (hazard ratio [HR], 1.36). The primary end point was OS at 3 years. The secondary objective was to compare disease-free survival (DFS) (time to recurrence) at 3 years. At minimum followup of 30 months (median, 42 months; range, 30-81 months), 178 deaths were documented. RESULTS: Three-year OS and DFS for all patients was 67% and 52%, respectively. Three-year OS was 67% and 66% in chest radiography and CT groups, respectively (HR, 0.9; upper 90% confidence interval [CI], 1.13). DFS rate was 54% and 49% in chest radiography and CT groups, respectively (HR, 0.82; upper 90% CI, 0.97). Three-year OS was 64% and 69% in 6-monthly and 3-monthly groups, respectively (HR, 1.2; upper 90% CI, 1.47). DFS was 51% and 52% in 6-monthly and 3-monthly groups, respectively (HR, 1.01; upper 90% CI, 1.2). Almost 90% of local recurrences were identified by patients themselves. CONCLUSIONS: Inexpensive imaging detects the vast majority of recurrent disease in patients with sarcoma without deleterious effects on eventual outcomes. Patient education regarding self-examination will detect most instances of local recurrence although this was not directly assessed in this study. Although less frequent visits adequately detected metastasis and local recurrence, this trial could not conclusively demonstrate noninferiority in OS for a 6-monthly interval of followup visits against 3-monthly visits. LEVEL OF EVIDENCE: Level I, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.

[401]

TÍTULO / TITLE: Does Ifosfamide Therapy Improve Survival of Patients With Dedifferentiated Chondrosarcoma?

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Puri A; Gulia A; Hawaldar R; Ranganathan P; Badwe RA

INSTITUCIÓN / INSTITUTION: Orthopaedic Oncology, Tata Memorial Hospital, Room No. 45, E Borges Road, Mumbai, India, docpuri@gmail.com.

RESUMEN / SUMMARY: BACKGROUND: Whether current postoperative surveillance regimes result in improved overall survival (OS) of patients with extremity sarcomas is unknown. QUESTIONS/PURPOSES: We hypothesized that a less intensive followup protocol would not be inferior to the conventional followup protocol in terms of OS. We (1) assessed OS of patients to determine if less intensive followup regimens led to worsened survival and asked (2) whether chest radiograph followup group was inferior to CT scan followup group in detecting pulmonary metastasis; and (3) whether less frequent (6-monthly) followup interval was inferior to more frequent (3-monthly) followup in detecting pulmonary metastasis and local recurrence. METHODS: A prospective randomized single-center noninferiority trial was conducted between January 2006 and June 2010. On the basis of 3-year survival of 60% with intensive, more frequent followup, 500 nonmetastatic patients were randomized to demonstrate noninferiority by a margin (delta) of 10% (hazard ratio [HR], 1.36). The primary end point was OS at 3 years. The secondary objective was to compare disease-free survival (DFS) (time to recurrence) at 3 years. At minimum followup of 30 months (median, 42 months; range, 30-81 months), 178 deaths were documented. RESULTS: Three-year OS and DFS for all patients was 67% and 52%, respectively. Three-year OS was 67% and 66% in chest radiography and CT groups, respectively (HR, 0.9; upper 90% confidence interval [CI], 1.13). DFS rate was 54% and 49% in chest radiography and CT groups, respectively (HR, 0.82; upper 90% CI, 0.97). Three-year OS was 64% and 69% in 6-monthly and 3-monthly groups, respectively (HR, 1.2; upper 90% CI, 1.47). DFS was 51% and 52% in 6-monthly and 3-monthly groups, respectively (HR, 1.01; upper 90% CI, 1.2). Almost 90% of local recurrences were identified by patients themselves. CONCLUSIONS: Inexpensive imaging detects the vast majority of recurrent disease in patients with sarcoma without deleterious effects on eventual outcomes. Patient education regarding self-examination will detect most instances of local recurrence although this was not directly assessed in this study. Although less frequent visits adequately detected metastasis and local recurrence, this trial could not conclusively demonstrate noninferiority in OS for a 6-monthly interval of followup visits against 3-monthly visits. LEVEL OF EVIDENCE: Level I, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.
RESUMEN / SUMMARY: - BACKGROUND: Dedifferentiated chondrosarcoma remains a significant therapeutic challenge. Studies performed to date have not identified efficacious chemotherapy regimens for this disease. QUESTIONS/PURPOSES: We sought to (1) evaluate the disease-specific survival at 2 and 5 years of patients with dedifferentiated chondrosarcoma; (2) assess the prognostic variables (both patient- and treatment-related), including the use of chemotherapy with ifosfamide, that relate to survivorship; and (3) assess specific toxicities associated with ifosfamide use.

METHODS: Data from 41 patients with dedifferentiated chondrosarcoma diagnosed and treated at the University of Texas MD Anderson Cancer Center from 1986 to 2010 were analyzed for demographics, treatments, oncologic outcomes, and prognostic variables. There were 14 women and 27 men. The mean age at diagnosis was 58 years (range, 26-86 years). Seven patients presented with metastasis. Surgical resection alone was performed in 11 patients; resection and chemotherapy in 26 patients; resection and radiotherapy in two patients; and resection, chemotherapy, and radiotherapy in two patients. Ifosfamide-based regimens were used for 16 patients. In general, ifosfamide was used when the tumor was located in the trunk or if cisplatin was discontinued as a result of toxicity. Minimum followup was 8 months (median, 68 months; range, 8-281 months). Survival was estimated using Kaplan-Meier plots and analyzed by using the Cox proportional hazards model.

RESULTS: Disease-specific survival rates at 2 and 5 years were 33% and 15%, respectively. Multivariate analysis revealed that treatment without ifosfamide-based chemotherapy was the only independent negative prognostic factor for disease-specific survival (hazard ratio, 0.4; 95% confidence interval, 0.17-0.92; p = 0.03). Ifosfamide was discontinued in a patient as a result of renal dysfunction and was decreased in dose in another patient who developed encephalopathy.

CONCLUSIONS: In this small retrospective study, it appeared that ifosfamide-based adjuvant chemotherapy combined with surgical resection offered a treatment advantage compared with patients who did not receive the drug in patients with dedifferentiated chondrosarcoma, although disease-specific survival for patients who have this rare tumor remains dismal.

LEVEL OF EVIDENCE: Level IV, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.
**RESUMEN / SUMMARY:** - BACKGROUND AND AIMS: Multiple osteochondromas (MO), an autosomal dominant skeletal disease, is characterized by the presence of multiple cartilage-capped bone tumors (exostoses). Two genes with mutations that are most commonly associated with MO have been identified as EXT1 and EXT2, which are Exostosin-1 and Exostosin-2. In this study, a variety of EXT1 and EXT2 gene mutations were identified in ten Chinese families with MO. METHODS: We investigated ten unrelated Chinese families involving a total of 46 patients who exhibited typical features of MO. The coding exons of EXT1 and EXT2 were sequenced after PCR amplification in ten probands. Radiological investigation was conducted simultaneously. RESULTS: Nine mutations were identified, five in EXT1 and four in EXT2, of which three were de novo mutations and six were novel mutations. One proband carried mutations in both EXT1 and EXT2 simultaneously, and three probands, including one sporadic case and two familial cases, had no detectable mutations. CONCLUSIONS: Our findings are useful for extending the mutational spectrum in EXT1 and EXT2 and understanding the genetic basis of MO in Chinese patients.

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**TÍTULO / TITLE:** - Long-term clinical outcome of sacral chondrosarcoma treated by total en bloc sacrectomy and reconstruction of lumbosacral and pelvic ring using intraoperative extracorporeal irradiated autologous tumor-bearing sacrum A case report with ten years follow-up.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Nishizawa K; Mori K; Saruhashi Y; Takahashi S; Matsusue Y

**INSTITUCIÓN / INSTITUTION:** - Department of Orthopaedic Surgery, Shiga University of Medical Science, Otsu, Shiga, Japan. Electronic address: grotto@belle.shiga-med.ac.jp.

**RESUMEN / SUMMARY:** - BACKGROUND CONTEXT: Primary malignant tumors of the sacrum are rare. Chondrosarcoma is one of the common malignant tumors arising from the sacrum. Chondrosarcoma is often invasive and there is a high propensity for local recurrence. Surgical resection is often the only effective treatment; however, the treatment of malignant sacral tumors can be challenging, both because of the anatomy of the spinopelvic complex and the frequently large tumor size. PURPOSE: We report a case of sacral chondrosarcoma, which was successfully treated by total en bloc sacrectomy and reconstruction of lumbosacral and pelvic ring using intraoperative extracorporeal irradiated autologous tumor-bearing sacrum STUDY DESIGN: A case report with ten years follow-up. METHODS: A 51-year-old male presented with right lower leg pain. Plain radiographs and computed tomography (CT) showed an osteolytic lesion at the sacrum, which extended to the sacroiliac joint. Magnetic resonance imaging (MRI) demonstrated that the tumor mass was localized from S1 to S2 with epidural lesion at L5-S1 disc level. Histopathological evaluation by open biopsy revealed that the lesion was chondrosarcoma. Total en bloc sacrectomy of the tumor-bearing sacrum was performed. The removed tumor-bearing sacrum was extracorporeally irradiated at 200 Gy during the operation, and returned to the original
position as a bone graft and fixed with instruments thereafter. RESULTS: We needed two revision surgeries during the first three years because of the implant failures, however, ten years after the initial surgery, CT revealed that the irradiated sacrum had remodeled by living bone and integrated with surrounding iliac bone without radiological evidence of the tumor recurrence. The patient ambulates without any support and there was no clinical and radiological evidence of tumor recurrence.

CONCLUSIONS: The advantages of our method include the following: availability of high dose of radiation because of extracorporeal irradiation, excellent fit between graft and host bone, reduction of the dead space, no immunological rejection, no need for a bone bank, availability of the sacrum not only for the augmentation of the large defect but also for the scaffold for the other bone grafts. Our report is only one case; however, we consider that it could be one option for the treatment of sacral malignant bone tumors such as chondrosarcoma.
the influential factors for relapse and the results were: preoperative ALP, $b = 0.023$, $p = 0.029$; surgery protocol, $b = -7.597$, $p = 0.007$; tumor size, $\geq 3/\leq 3$, $b = 24.805$, $p < 0.0005$; age, $b = 0.054$, $p = 0.632$; and pathology type, $b = 1.998$, $p = 0.34$.

CONCLUSIONS: Tumor size, preoperative ALP and CT images were helpful for distinguishing AO from CO. The difference in intraoperative blood loss between CO and AO is mainly attributed to the size of the lesion. Preoperative ALP, surgery protocol and tumor size ($\geq 3/\leq 3$) were considered to significantly influence relapse of spinal OBL.

[406]
**TÍTULO / TITLE:** - Left atrial myxoma in a patient with paroxysmal atrial fibrillation.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
  ●● Enlace al texto completo (gratuito o de pago) 1007/s00059-013-3997-6
**AUTORES / AUTHORS:** - Knur R; Ozse J
**INSTITUCIÓN / INSTITUTION:** - Department of Cardiology and Angiology, Allgemeines Krankenhaus Viersen, Hoserkirchweg 63, 41747, Viersen, Germany, drrknur@gmx.de.

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[407]
**TÍTULO / TITLE:** - Giant Cell Tumor of Bone in Childhood: Clinical Aspects and Novel Therapeutic Targets.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**REVISTA / JOURNAL:** - Paediatr Drugs. 2013 Oct 10.
  ●● Enlace al texto completo (gratuito o de pago) 1007/s40272-013-0051-3
**AUTORES / AUTHORS:** - Federman N; Brien EW; Narasimhan V; Dry SM; Sodhi M; Chawla SP
**INSTITUCIÓN / INSTITUTION:** - Department of Pediatrics, Hematology/Oncology, Mattel Children’s Hospital at University of California, Los Angeles, USA, nfederman@mednet.ucla.edu.
**RESUMEN / SUMMARY:** - Giant cell tumor of bone (GCTB) is a rare primary bone tumor that primarily affects young adults, but can be seen in children. The primary modality of treatment is surgical resection; however, this is not always possible given the location and extent of the neoplasm. Recent developments in the understanding of the underlying molecular pathogenesis of disease have pointed to interactions between the stromal component producing receptor activator of nuclear factor-kappaB (RANK) and RANK-ligand (RANKL) causing the formation of osteoclast-like giant cells that drive bone destruction. The development of a monoclonal humanized antibody to RANKL, denosumab, has been shown to reduce skeletal-related events from osteoporosis and from bony metastases from solid tumors. Recent phase II clinical trials with denosumab in skeletally mature adolescents over age 12 years and adults with GCTB, have shown both safety and efficacy, leading to its accelerated US FDA approval on 13 June 2013. In children who are skeletally immature, safety and efficacy has not been established, and there has been only published anecdotal use.

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[408]
TÍTULO / TITLE: Contralateral referred pain in a patient with intramedullary spinal cord metastasis from extraskeletal small cell osteosarcoma.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Kang K; Lee JH; Kim HG
context: Referred pain has been observed in some patients after cordotomy, wherein noxious stimulus applied to a region rendered analgesic by cordotomy produces pain at a spot different from the one where the noxious stimulus is applied. We report a patient who had intramedullary spinal cord metastasis of extraskeletal small cell osteosarcoma, a rare form of metastatic disease, and experienced contralateral referred pain. FINDINGS: Initially, the patient had a mass in the left posterior neck region and later developed a large extradural mass at the C3-C7 level. The masses were excised, and the histological findings led to a diagnosis of small cell osteosarcoma. He underwent chemotherapy and radiation therapy. He experienced numbness in his left leg; subsequently, the numbness slowly spread up the thigh to the left side of the abdomen. When pinched in the numb area on the left side of the body, he felt as though he had been pinched in both that area and the corresponding area on the right side. A magnetic resonance imaging scan showed an enhancing lesion in the right side of the cord at the C6-C7 level. Conclusion/clinical relevance: An intramedullary spinal cord metastasis can arise from primary extraskeletal small cell osteosarcoma and cause contralateral referred pain, especially in a mirror-image location. Contralateral referred pain may be caused by a subsidiary pathway comprising ascending chains of short neurons that link the dorsal horn neurons longitudinally and latitudinally.

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TÍTULO / TITLE: Chondrosarcoma in a patient with osteogenesis imperfecta.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Mandziak DG; Clayer M
INSTITUCIÓN / INSTITUTION: Department of Orthopaedics and Trauma, Royal Adelaide Hospital, Adelaide, South Australia, Australia.

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TÍTULO / TITLE: Natural killer cell therapy and aerosol interleukin-2 for the treatment of osteosarcoma lung metastasis.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Guma SR; Lee DA; Yu L; Gordon N; Hughes D; Stewart J; Wang WL; Kleinerman ES
INSTITUCIÓN / INSTITUTION: Division of Pediatrics, The University of Texas MD Anderson Cancer Center, Houston, Texas.
RESUMEN / SUMMARY: - BACKGROUND: Survival of patients with osteosarcoma lung metastases has not improved in 20 years. We evaluated the efficacy of combining natural killer (NK) cells with aerosol interleukin-2 (IL-2) to achieve organ-specific NK cell migration and expansion in the metastatic organ, and to decrease toxicity associated with systemic IL-2. PROCEDURE: Five human osteosarcoma cell lines and 103 patient samples (47 primary and 56 metastatic) were analyzed for NKG2D ligand (NKG2DL) expression. Therapeutic efficacy of aerosol IL-2 + NK cells was evaluated in vivo compared with aerosol IL-2 alone and NK cells without aerosol IL-2. RESULTS: Osteosarcoma cell lines and patient samples expressed various levels of NKG2DL. NK-mediated killing was NKG2DL-dependent and correlated with expression levels. Aerosol IL-2 increased NK cell numbers in the lung and within metastatic nodules but not in other organs. Therapeutic efficacy, as judged by tumor number, size, and quantification of apoptosis, was also increased compared with NK cells or aerosol IL-2 alone. There were no IL-2-associated systemic toxicities. CONCLUSION: Aerosol IL-2 augmented the efficacy of NK cell therapy against osteosarcoma lung metastasis, without inducing systemic toxicity. Our data suggest that lung-targeted IL-2 delivery circumvents toxicities induced by systemic administration. Combining aerosol IL-2 with NK cell infusions, may be a potential new therapeutic approach for patients with osteosarcoma lung metastasis. Pediatr Blood Cancer © 2013 Wiley Periodicals, Inc.

TÍTULO / TITLE: - Endothelin-1 gene polymorphisms and risk of chemoresistant pediatric osteosarcoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Zhou Y; Liu B; Wang M; Ni J

INSTITUCIÓN / INSTITUTION: - Department of Orthopaedics, The Second Xiangya Hospital, Central South University, Changsha, Hunan, China.

RESUMEN / SUMMARY: - BACKGROUND: Osteosarcoma (OS) is the most common childhood bone cancer. Chemoresistance is the principal reason for poor survival and disease recurrence in OS patients, and ET-1 reportedly plays an important role in the development of chemoresistance in OS cells. In the present study, we for the first time explored the association of endothelin-1 (ET-1) SNPs and haplotypes with the risk of chemoresistant pediatric OS. PROCEDURE: We genotyped three SNPs (rs1800541, rs2070699, and rs5370) in the ET-1 gene in a case-control study, using 350 pairs of age, sex, and tumor location and stage matched pediatric patients with OS. Patients who showed <90% tumor necrosis after neochemotherapy were defined as poor responders (cases), and those who showed >/=90% tumor necrosis were defined as good responders (controls). RESULTS: The G allele at rs1800541 and the G allele at rs2070699 were associated with reduced and increased risk of chemoresistant OS, respectively. The rs1800541-rs2070699 haplotypes TG and GT were respectively associated with increased (P = 0.012; adjusted OR, 1.82; 95% CI, 1.10-5.65) and reduced (P = 0.009; adjusted OR, 0.25; 95% CI, 0.14-0.84) risk of chemoresistant OS. The TG and the GT haplotypes have a gene-dosage effect on increasing and decreasing the ET-1 expression in primary OS tumor cells from chemoresistant pediatric OS subjects, respectively. CONCLUSIONS: This study provides the first
evidence of an association between the ET-1 gene SNPs and haplotypes and the risk of chemoresistant pediatric OS, potentially adding new insights into the pathophysiology and treatment of chemoresistant OS. Pediatr Blood Cancer © 2013 Wiley Periodicals, Inc.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Dotlic S; Gatalica Z; Wen W; Ghazalpour A; Mangham C; Babic D; Zekan J; Vranic S
INSTITUCIÓN / INSTITUTION: - *Department of Pathology and Cytology parallelDepartment of Obstetrics and Gynecology, University Hospital Center Zagreb section signDepartment of Pathology, University of Zagreb Medical School, Zagreb, Croatia daggerCaris Life Sciences, Phoenix, AZ double daggerRobert Jones and Agnes Hunt Orthopaedic and District Hospital, Oswestry and Royal Orthopaedic Hospital, Birmingham, UK paragraph signDepartment of Pathology, Clinical Center of the University of Sarajevo, Sarajevo, Bosnia and Herzegovina.
RESUMEN / SUMMARY: - Extraskeletal myxoid chondrosarcoma (EMC) is a rare mesenchymal neoplasm, rarely reported in the genitourinary tract with only 5 cases reported in the vulva. We investigated 2 cases of vulvar sarcomas whose morphologic appearance and immunohistochemical profiles were consistent with EMC using fluorescence in situ hybridization (FISH), reverse-transcription polymerase chain reaction, and a whole genome expression array. FISH and reverse-transcription polymerase chain reaction assays showed no EWSR1 and NR4A3 loci rearrangements. Microarray-based analysis also revealed no changes in NR4A3 and EWSR1 gene transcription levels. Microarray data showed a significant downregulation of the muscle-related genes (eg, myosin heavy chain family, actins, myoglobin, desmin, creatine kinase, troponins) and cytokeratins (KRT6A, 6B, 13, 14, and 78), upregulation of several neuron-specific genes [neural cell adhesion molecule 1 (NCAM-1/CD56), neurofilament (NEFH)], along with some well-characterized tumor biomarkers [carbonic anhydrase IX (CA-9), topoisomerase I alpha (TOP2A), matrix metalloproteinases (MMP-7, MMP-9), CDKN2 gene (p16-INK4a), checkpoint homolog 2 (CHEK2)]. Notably, both tumors showed upregulation of the pleomorphic adenoma gene 1 (PLAG1), and in 1 case PLAG1 gene rearrangement was detected by break-apart FISH. Some vulvar tumors with morphologic and immunohistochemical characteristics of EMC may represent a molecular genetic entity separate from EMCs arising in other locations. PLAG1 gene activation appears to be involved in the development of these neoplasms.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Zinc finger X-chromosomal protein (ZFX) is a member of the zinc finger family of proteins. The importance of ZFX in several cancer types, including prostate cancer, laryngeal squamous cell carcinoma, and glioma, has been addressed. However, the role of ZFX in human osteosarcoma remains unknown. Here we investigated the phenotype of ZFX knockdown on cell proliferation and in vitro tumorigenesis using lentivirus-mediated loss-of-function strategy. The results demonstrated that the proliferation and colony formation ability of human osteosarcoma Saos-2 and MG63 cells was impaired by ZFX small interfering RNA (siRNA)-expressing lentivirus. Moreover, loss of ZFX led to G0/G1 phase cell cycle arrest and a significant increase of cells in the sub-G1 fraction, indicating that ZFX functions as an oncogene in the malignant proliferation process in osteosarcoma. Furthermore, ZFX siRNA may have an antitumorigenic effect on osteosarcoma cells. Our findings hold important significance for RNA interference-mediated cancer gene therapy for human osteosarcoma.

[414]

Loss of MEF2D expression inhibits differentiation and contributes to oncogenesis in rhabdomyosarcoma cells.

BACKGROUND: Rhabdomyosarcoma (RMS) is a highly malignant pediatric cancer that is the most common form of soft tissue tumors in children. RMS cells have many features of skeletal muscle cells, yet do not differentiate. Thus, our studies have focused on the defects present in these cells that block myogenesis. METHODS: Protein and RNA analysis identified the loss of MEF2D in RMS cells. MEF2D was expressed in RD and RH30 cells by transient transfection and selection of stable cell lines, respectively, to demonstrate the rescue of muscle differentiation observed. A combination of techniques such as proliferation assays, scratch assays and soft agar assays were used with RH30 cells expressing MEF2D to demonstrate the loss of oncogenic growth in vitro and xenograft assays were used to confirm the loss of tumor growth in vivo. RESULTS: Here, we show that one member of the MEF2 family of proteins required for normal myogenesis, MEF2D, is largely absent in RMS cell lines representing both major subtypes of RMS as well as primary cells derived from an embryonal RMS model. We show that the down regulation of MEF2D is a major cause for the failure of RMS cells to differentiate. We find that MyoD and myogenin are bound with their dimerization partner, the E proteins, to the promoters of muscle specific genes in RMS cells. However, we cannot detect MEF2D binding at any promoter tested. We find that exogenous MEF2D expression can activate muscle
specific luciferase constructs, up regulate p21 expression and increase muscle specific gene expression including the expression of myosin heavy chain, a marker for skeletal muscle differentiation. Restoring expression of MEF2D also inhibits proliferation, cell motility and anchorage independent growth in vitro. We have confirmed the inhibition of tumorigenicity by MEF2D in a tumor xenograft model, with a complete regression of tumor growth. CONCLUSIONS: Our data indicate that the oncogenic properties of RMS cells can be partially attributed to the loss of MEF2D expression and that restoration of MEF2D may represent a useful therapeutic strategy to decrease tumorigenicity.

[415]
**TÍTULO / TITLE:** - Spinal angiolipoma: etiology, imaging findings, classification, treatment, and prognosis.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](1007/s00586-013-3073-1)
**REVISTA / JOURNAL:** - Eur Spine J. 2013 Nov 5.
**AUTORES / AUTHORS:** - Si Y; Wang Z; Pan Y; Lin G; Yu T
**INSTITUCIÓN / INSTITUTION:** - Department of Neurosurgery, Peking University Third Hospital, No. 49. North Garden Street, HaiDian District, Beijing, 100191, People’s Republic of China, siyudr@yahoo.com.
**RESUMEN / SUMMARY:** - PURPOSE: To summarise our experience treating patients with spinal angiolipomas (SAs) and to evaluate factors relating to its prognosis.
METHODS: We retrospectively reviewed the records of patients diagnosed with SAs who received surgical treatment from January 2001 to February 2013. RESULTS: Twenty-one patients were described. We divide SAs into two types: “intraspinal” and “dumbbell-shaped”. The former were further subclassified as “with lipomatosis” and “without lipomatosis”. Overweight people are more likely to get the “with lipomatosis” type which needs different surgical strategy and/or a diet therapy to get better outcomes. CONCLUSION: Diagnosis of SAs should be made with reference to clinical, radiological, and pathological findings. Application of different methods is needed to treat SAs.

[416]
**TÍTULO / TITLE:** - Myositis Ossificans of the Quadriceps Femoris Mimicking Sarcoma as a Diagnostic and Therapeutic Problem - Case Report and Literature Review.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](2478/pjs-2013-0081)
**AUTORES / AUTHORS:** - Nowaczyk P; Murawa D; Zmudzinska M; Wasiewicz J
**RESUMEN / SUMMARY:** - Abstract Myositis ossificans (MO) may be included in the group of lesions described as pseudosarcomas. Its clinical and histological picture frequently mimics a malignant neoplasm and therefore, ultimate diagnosis and implementation of adequate treatment requires the cooperation of interdisciplinary team of physicians. The paper presents the case of 20-year old female patient suffering from severe pain in the right thigh. The patient was initially diagnosed with the lower
limb overload. Rest and administration of non-steroidal anti-inflammatory drugs (NSAID) were recommended. Due to the lack of the efficacy of the recommended conservative treatment and detection of tumorous mass on ultrasound examination, the patient was referred to the cancer centre. The diagnostic procedures were extended and an open biopsy of the lesion was performed which revealed the presence of MO. The patient underwent a surgical procedure during which the pathological mass was entirely removed. Follow up examinations conducted upon the conclusion of the rehabilitation indicate no pathologies in the operated area.

[417]

TÍTULO / TITLE: - Optimizing tyrosine kinase inhibitor therapy in gastrointestinal stromal tumors: exploring the benefits of continuous kinase suppression.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Le Cesne A; Blay JY; Reichardt P; Joensuu H

INSTITUCIÓN / INSTITUTION: - Department of Medicine, Institut Gustave Roussy, Villejuif, France;

RESUMEN / SUMMARY: - The oral tyrosine kinase inhibitor (TKI) imatinib has revolutionized the treatment of gastrointestinal stromal tumors (GISTs), most of which harbor oncogenic mutation in genes that encode the receptor tyrosine kinases KIT or PDGFA. Imatinib is the standard of care for patients with advanced GIST and for patients with primary GIST at significant risk of recurrence after surgery. Design. This review discusses data supporting continuous kinase suppression with imatinib and key issues, including response to imatinib reintroduction, effect of treatment interruption on secondary resistance to imatinib, and prognostic factors associated with sustained response to imatinib. Results. Long-term follow-up results of the B2222 study and updated results of the BFR14 trial demonstrate that continuous imatinib treatment in patients with advanced GIST is associated with reduced risk of progression. For patients progressing on or intolerant of imatinib, continuing therapy with TKIs sunitinib followed by regorafenib is recommended. In the adjuvant setting, final results of the trial by the Scandinavian Sarcoma Group and the Sarcoma Group of the Arbeitsgemeinschaft Internistische Onkologie demonstrate that 3 years of adjuvant imatinib, compared with 1 year, significantly reduces the risk of recurrence and improves overall survival of patients with KIT-positive GIST at high risk of recurrence. Conclusions. Maintenance of therapy with TKIs is the key to successful treatment of GIST. Results from recent studies provide a strong rationale for continuous imatinib treatment for 3 years following surgical resection and long-term continuous administration in advanced or metastatic GIST.

[418]

TÍTULO / TITLE: - Surgical treatment of craniofacial fibrous dysplasia in adults.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Le Cesne A; Blay JY; Reichardt P; Joensuu H

INSTITUCIÓN / INSTITUTION: - Department of Medicine, Institut Gustave Roussy, Villejuif, France;

RESUMEN / SUMMARY: - The oral tyrosine kinase inhibitor (TKI) imatinib has revolutionized the treatment of gastrointestinal stromal tumors (GISTs), most of which harbor oncogenic mutation in genes that encode the receptor tyrosine kinases KIT or PDGFA. Imatinib is the standard of care for patients with advanced GIST and for patients with primary GIST at significant risk of recurrence after surgery. Design. This review discusses data supporting continuous kinase suppression with imatinib and key issues, including response to imatinib reintroduction, effect of treatment interruption on secondary resistance to imatinib, and prognostic factors associated with sustained response to imatinib. Results. Long-term follow-up results of the B2222 study and updated results of the BFR14 trial demonstrate that continuous imatinib treatment in patients with advanced GIST is associated with reduced risk of progression. For patients progressing on or intolerant of imatinib, continuing therapy with TKIs sunitinib followed by regorafenib is recommended. In the adjuvant setting, final results of the trial by the Scandinavian Sarcoma Group and the Sarcoma Group of the Arbeitsgemeinschaft Internistische Onkologie demonstrate that 3 years of adjuvant imatinib, compared with 1 year, significantly reduces the risk of recurrence and improves overall survival of patients with KIT-positive GIST at high risk of recurrence. Conclusions. Maintenance of therapy with TKIs is the key to successful treatment of GIST. Results from recent studies provide a strong rationale for continuous imatinib treatment for 3 years following surgical resection and long-term continuous administration in advanced or metastatic GIST.
Craniofacial fibrous dysplasia (FD) is a rare disorder that may require neurosurgical expertise for definitive management; however, surgical management of FD in adult patients is uncommon. Although other therapies have been shown to slow progression, the only definitive cure for adult craniofacial FD is complete resection with subsequent reconstruction. The authors review the biological, epidemiologic, clinical, genetic, and radiographic characteristics of adult FD, with an emphasis on surgical management of FD. They present a small series of three adult patients with complex FD that highlights the surgical complexity required in some adult patients with FD. Because of the complex nature of these adult polyostotic craniofacial cases, the authors used neurosurgical techniques specific to the different surgical indications, including a transsphenoidal approach for resection of sphenoidal sinus FD, a transmaxillary approach to decompress the maxillary branch of the trigeminal nerve with widening of the foramen rotundum, and complete calvarial craniectomy with cranioplasty reconstruction. These cases exemplify the diverse range of skull base techniques required in the spectrum of surgical management of adult FD and demonstrate that novel variations on standard neurosurgical approaches to the skull base can provide successful outcomes with minimal complications in adults with complex craniofacial FD.

[419]
TÍTULO / TITLE: - Intracardiac leiomyomatosis complicated by pulmonary embolism: a multimodality imaging case of a rare entity.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Ribeiro V; Almeida J; Madureira AJ; Lopez E; Machado L; Albuquerque R; Pinho P
INSTITUCIÓN / INSTITUTION: - Cardiology Department, Sao Joao Hospital, Medicine Faculty of Porto University, Porto, Portugal. Electronic address: ribeiro_vania@hotmail.com.
RESUMEN / SUMMARY: - We present a case of intravenous leiomyomatosis with intracaval and right ventricle extension that was misdiagnosed as venous thrombus. Part of the mass had split and embolized the pulmonary artery, requiring urgent surgery. Although the mass fragments were removed from the inferior vena cava, right ventricle, and pulmonary artery successfully, this case clearly shows the importance of prompt surgery.

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[420]
TÍTULO / TITLE: - Cardiac myxoma with glandular elements: A clinicopathological and immunohistochemical study of five new cases with an emphasis on differential diagnosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
RESUMEN / SUMMARY: - This paper reported five new cases of cardiac myxoma with glandular components, known as glandular cardiac myxoma. The goals of this study were to analyze the clinicopathological features of this disease and to explore new features for differential diagnosis. The patient series included three women and two men. All tumors were located in the left atrium without invasion of the adjacent myocardium. Patients presented with cardiac-related or embolization symptoms. Histologically, neoplasms consisted of well-formed glandular structures and typical myxoma areas. No nuclear atypia, mitosis, or necrosis was identified in the glandular structures. Glandular lining cells were strongly positive for pan-cytokeratin, epithelial membrane antigen, CAM5.2 and cytokeratin 7, but were negative for some organ-specific markers, such as thyroid transcription factor-1, calretinin, estrogen receptor, progesterone receptor, gross cystic disease fluid protein, prostate-specific antigen, prostate-specific acid phosphatase, cytokeratin 20 and caudal type homeobox 2. In conclusion, glandular cardiac myxoma is a rare disease which shows characteristics similar to those of classical cardiac myxoma. Because of its rarity, glandular cardiac myxoma must be distinguished from adenocarcinoma metastatic to the heart. The combination of histopathological features and immunohistochemical profiles should improve the diagnostic accuracy of glandular cardiac myxoma.

[421]

TÍTULO / TITLE: - Synovial sarcoma of the spine: a case involving paraspinal muscle with extensive calcification and the surgical consideration in treatment.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Kim J; Lee SH; Choi YL; Bae GE; Kim ES; Eoh W

INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, 50 Irwon-dong, Gangnam-gu, Seoul, 135-710, Republic of Korea, bwv1041@skku.edu.

RESUMEN / SUMMARY: - INTRODUCTION: Synovial sarcoma is a rare malignant tumor of the spine. This tumor may present as a painless mass of the spine or slowly enlarge, causing pain or neurologic deficits. As it is difficult to differentiate this lesion from other soft tissue tumors, synovial sarcoma requires histologic confirmation for definite diagnosis. Thus, the treatment strategy is often planned in the final step depending on the pathologic results. Despite its rare incidence, a few cases of primary or metastatic synovial sarcoma involving the spinal cord, foramen, vertebral body, or paraspinal muscles have been reported in the literature. MATERIALS AND METHODS: We present the case of a 29-year-old man with a synovial sarcoma in the paraspinal muscle of the cervical spine. The patient was evaluated radiologically and histologically. Plain radiography, computed tomography, and magnetic resonance imaging were performed as part of the preoperative workup, and immunohistochemical...
and cytogenetic studies were additionally performed to identify the histologic features of the tumor. The patient underwent marginal resection followed by adjuvant radiation therapy. The patient has been followed up for 2 years. CONCLUSIONS: This article highlights the features of synovial sarcoma of the spine via a comprehensive review. Synovial sarcoma of the spine is uncommon, but it is a challenging issue in both diagnostic and therapeutic aspects. The currently available evidence suggests the use of a multidisciplinary approach in the treatment of synovial sarcoma, which includes complete resection and radiation therapy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Deneve JL; Messina JL; Bui MM; Marzban SS; Letson GD; Cheong D; Gonzalez RJ; Sondak VK; Zager JS
INSTITUCIÓN / INSTITUTION: - Department of Cutaneous Oncology, Moffitt Cancer Center, Tampa, FL 33612, USA. Jonathan.Zager@Moffitt.org.
RESUMEN / SUMMARY: - BACKGROUND: Cutaneous leiomyosarcoma is primarily a low-grade malignancy that affects elderly male Caucasians. It is a rare dermal-based tumor for which treatment algorithms have been poorly defined. METHODS: We retrospectively reviewed the use of a median 1-cm margin for resection to treat patients with cutaneous leiomyosarcoma referred for treatment between 2005 and 2010. RESULTS: Thirty-three patients with cutaneous leiomyosarcoma were treated. Of these, 76% were male, 97% were Caucasian (median age: 63.5 years), and 67% of tumors were located on the extremities. Preoperative staging was negative for distant metastasis in all patients. A majority of the tumors (88%) were low grade (median size: 1.3 cm). All of the tumors were positive for smooth-muscle actin. A total of 94% of patients underwent primary surgical resection with a median margin of 1 cm. Final resection margin was negative in 97% of patients. Adjuvant radiotherapy was used in 15%. No metastatic spread or recurrences were present, and 100% of patients were alive at last follow-up (median: 15.5 months). CONCLUSIONS: Good oncological control and excellent outcomes are possible with a 1-cm resection margin in most cases of cutaneous leiomyosarcoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Moraes OJ
INSTITUCIÓN / INSTITUTION: - Neurosurgery Department, Santa Marcelina's University Hospital-Medical School, R. Maestro Cardim, 591. 11 Floor, Sao Paulo, SP, 0541-300, Brazil, osmarmoraes@gmail.com.

[424]
TÍTULO / TITLE: - Sunitinib Treatment for Multiple Brain Metastases from Jejunal Gastrointestinal Stromal Tumor: Case Report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Takeuchi H; Koike H; Fujita T; Tsujino H; Iwamoto Y
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Yamashiro Public Hospital.
RESUMEN / SUMMARY: - Gastrointestinal stromal tumors (GISTs) are rare malignant tumors and only a few reported cases of brain metastases can be found. Introduction of a new molecular targeted agent, imatinib mesylate in the last decade has dramatically changed the treatment strategy and prognosis. However, imatinib is usually ineffective for brain metastasis from GISTs. The authors present the case of multiple brain metastases from jejunal GIST. The brain metastasis in the right prefrontal gyrus was detected 20 months after resection of the primary lesion when left hemiparesis began although the patient was on imatinib. Then the patient began taking sunitinib instead of imatinib, and the lesion shrunk and the symptom improved. However, after the dose reduction due to side effects, a new brain metastasis was found and this time, stereotactic radiation was effectively done. Sunitinib is one of the promising receptor tyrosine kinase inhibitors used for metastatic renal cell carcinomas or imatinib-refractory GISTs. Sunitinib is thought to penetrate blood-brain barrier, and recent reports indicate effectiveness to brain metastasis. To the authors' knowledge, this is the first report of brain metastases from jejunal GIST responding to sunitinib therapy.

[425]
TÍTULO / TITLE: - High-dose chemotherapy with stem cell rescue in the primary treatment of metastatic and pelvic osteosarcoma: Final results of the ISG/SSG II study.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Boye K; Del Prever AB; Eriksson M; Saeter G; Tienghi A; Lindholm P; Fagioli F; Skjeldal S; Ferrari S; Hall KS
INSTITUCIÓN / INSTITUTION: - Department of Oncology, The Norwegian Radium Hospital, Oslo University Hospital, Oslo, Norway.
RESUMEN / SUMMARY: - BACKGROUND: Patients with metastatic osteosarcoma at diagnosis or axial primary tumors have a poor prognosis. The aim of the study was to evaluate the feasibility and efficacy of intensified treatment with high-dose chemotherapy (HDCT) and stem cell rescue in this group. METHODS: From May 1996 to August 2004, 71 patients were included in a Scandinavian-Italian single arm phase II study. Preoperative chemotherapy included methotrexate, doxorubicin, cisplatin and ifosfamide, and postoperative treatment consisted of two cycles of doxorubicin, one
cycle of cyclophosphamide and etoposide and two courses of high-dose etoposide and carboplatin with stem cell rescue. RESULTS: Twenty-nine patients (43%) received two courses and 10 patients (15%) received one course of HDCT. HDCT was associated with significant toxicity, but no treatment-related deaths were recorded. Fourteen patients (20%) had disease progression before completion of the study protocol, and only 29/71 patients (41%) received the full planned treatment. Median event-free survival (EFS) was 18 months, and estimated 5-year EFS was 27%. Median overall survival (OS) was 34 months, and estimated 5-year OS was 31%. When patients who did not receive HDCT due to disease progression were excluded, there was no difference in EFS (P = 0.72) or OS (P = 0.49) between patients who did or did not receive HDCT. CONCLUSIONS: The administration of high-dose chemotherapy with stem cell rescue was feasible, but associated with significant toxicity. Patient outcome seemed comparable to previous studies using conventional chemotherapy. We conclude that HDCT with carboplatin and etoposide should not be further explored as a treatment strategy in high-risk osteosarcoma. Pediatr Blood Cancer © 2013 Wiley Periodicals, Inc.

[426]
TÍTULO / TITLE: - Dermatofibroma of the Face: A Clinicopathologic Study of 20 Cases.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Estela JR; Rico MT; Perez A; Unamuno B; Garcias J; Cubells L; Alegre V
RESUMEN / SUMMARY: - INTRODUCTION: Dermatofibroma is one of the most common benign skin tumors. It typically develops on the lower limbs between the third and fifth decade of life and is more common in women. Clinical diagnosis is often straightforward. Dermatofibromas are associated with a very low rate of local recurrence following excision. OBJECTIVES: To describe the clinical and histologic features of dermatofibroma of the face based on our experience. MATERIALS AND METHODS: Descriptive retrospective study of the clinicopathologic features of dermatofibromas of the face diagnosed at the dermatology department of Hospital General Universitario de Valencia between 1990 and 2012. RESULTS: Twenty cases of dermatofibroma of the face (1.11% of all dermatofibromas diagnosed) were studied. The age at onset varied widely, from 28 to 84 years. The mean age at onset was 57.15 years and the median was 54 years. There were 11 women and 9 men. Mean follow-up was 83 months and there were no local recurrences. All the tumors were confined to the papillary and reticular dermis and the storiform pattern was the most common growth pattern observed. CONCLUSIONS: This study of facial dermatofibromas diagnosed at our hospital over a period of 22 years suggests that the face is an uncommon site but that dermatofibromas in this location behave similarly to those occurring elsewhere on the body.
[427] **TÍTULO / TITLE:** - Chondrosarcoma of the head of the fifth metacarpal treated with an iliac crest bone graft and concurrent Swanson's arthroplasty.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1016/j.bjps.2013.10.023

**AUTORES / AUTHORS:** - Hills AJ; Tay S; Gateley D

**INSTITUCIÓN / INSTITUTION:** - St George’s Hospital, Blackshaw Road, London SW17 0QT, UK. Electronic address: ajh502@gmail.com.

**RESUMEN / SUMMARY:** - Chondrosarcomas are rare malignant tumours of the bone with hyaline cartilage differentiation - only 1.5% affect the hands. Currently there is a limited range of techniques available to reconstruct the metacarpophalangeal joints affected by such neoplasias. We report a 30-year-old lady who presented with a grade 2 chondrosarcoma in the epiphseal region of her fifth metacarpal who underwent enbloc resection of the affected metacarpal and immediate reconstruction, using a Swansons arthroplasty and non-vascularised iliac crest bone graft. Our findings are presented after follow-up of 9 years and we review the current reconstructive options available.

[428] **TÍTULO / TITLE:** - Clinical profiles of 710 premenopausal women with adenomyosis who underwent hysterectomy.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1111/jog.12211

**AUTORES / AUTHORS:** - Li X; Liu X; Guo SW

**INSTITUCIÓN / INSTITUTION:** - Department of Gynecology, Fudan University, Shanghai, China; Shanghai Key Laboratory of Female Reproductive Endocrine-Related Diseases, Shanghai OB/GYN Hospital, Fudan University, Shanghai, China.

**RESUMEN / SUMMARY:** - AIM: The aim of this study was to determine the frequency of various symptoms and their associated characteristics in women with adenomyosis who underwent hysterectomy, and to determine which symptoms are likely to go with which others in these patients. MATERIAL AND METHODS: In 2007, 1697 consecutive patients underwent hysterectomy in our hospital. Among them, 734 (43.3%) were histologically confirmed to have adenomyosis, and 710 of them were premenopausal. The medical charts of all 734 patients were retrieved, and their demographic, clinical information and postoperative findings were recorded. We used the Verbal Descriptor Scale to measure the preoperative severity of dysmenorrhea. The Apriori Algorithm was used for mining the association of different symptoms. RESULTS: Among the 710 premenopausal patients, only 4.5% of them had no symptoms. Dysmenorrhea was the most common complaint, occurring in 81.7% of patients. Dysmenorrhea co-occurred most frequently with menorrhagia. The presence of adhesion, presence of endometriosis, complaint of menorrhagia, longer duration of disease, gravidity, palpable pain during pelvic examination, and diffuse adenomyosis were positively associated with the severity of dysmenorrhea. Age, severity of dysmenorrhea, and complaint of metrorrhagia were positively associated with the risk
of menorrhagia. CONCLUSIONS: Dysmenorrhea is the most common complaint in women with adenomyosis, which often goes with that of menorrhagia. Adenomyosis often co-occurs with endometriosis and leiomyomas. Various factors are associated with the risk of having different symptoms.

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TÍTULO / TITLE: Surface proteomic analysis of differentiated versus stem-like osteosarcoma human cells.
RESUMEN / SUMMARY: Cancer stem cell characterization represents a breakthrough in cancer research. Despite evidence showing the existence and the role of cancer stem cells in osteosarcoma (OS) onset and progression, little is known about their specific surface phenotype. To address this issue, we carried out a cytometric analysis with an antibody-array comprising 245 membrane proteins comparing the stem and differentiated OS cells. As experimental model, we chose the stem-like cell line 3aminobenzamide-OS and its parental, differentiated, cell line MG63. We identified 50 differentially expressed, 23 homogeneously expressed, and 172 not expressed proteins in the two cell line models, thus defining a surface protein signature specific for each of them. Furthermore, we selected ERK1/2 (p44/42 mitogen-activated protein kinases) as a potential pathway correlated with processes that characterize tumorigenic potential and stemness of 3aminobenzamide-OS cells.

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TÍTULO / TITLE: Low level of baseline circulating VEGF-A is associated with better outcome in patients with vascular sarcomas receiving sorafenib: an ancillary study from a phase II trial.
RESUMEN / SUMMARY: We have carried out a stratified phase II study of sorafenib (So) in patients with advanced angiosarcoma (n = 32) and epithelioid hemangioendothelioma (n = 13). This report concerns the correlative analysis of the predictive values of circulating pro/anti-angiogenic biomarkers. Using the ELISA method (R&D Systems), circulating biomarkers (VEGF-A, in picograms per milliliter), thrombospondin-1 (TSP1, in micrograms per milliliter), stem cell factor (SCF, in
picograms per milliliter), placental growth factor (PIGF, in picograms per milliliter), VEGF-C (in picograms per milliliter), and E-selectin (in nanograms per milliliter) were measured before So treatment and after 7 days. VEGF-A (mean value 475 vs. 541, p = 0.002), TSP1 (16 vs. 24, p = 0.0002), and PIGF (20.9 vs. 40.7, p = 0.0001) significantly increased during the treatment. Treatment did not affect the levels of SCF, VEGF-C, and E-selectin. Only two biomarkers were associated with better outcome as follows: VEGF-A and PIGF. Best objective response and non-progression at 180 days were associated with low level of VEGF-A at baseline (p = 0.04 and 0.03, respectively). There was a correlation between the circulating level of VEGF-A and time to progression (TTP) (r = -0.47, p = 0.001). Best objective response and non-progression at 180 days were not associated with baseline level of PIGF, but there was a correlation between the circulating level of PIGF at baseline and TTP. Low level of VEGF-A at baseline (<500) was significantly associated with better outcome.

[431]
TÍTULO / TITLE: - Multicentric osteoid osteoma in C5 vertebra and parietal bone.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
- Enlace al texto completo (gratuito o de pago) 1016/j.spinee.2013.08.006
AUTORES / AUTHORS: - Amendola L; Cappuccio M; De lure F
INSTITUCIÓN / INSTITUTION: - Department of Spine Surgery, Ospedale Maggiore “C.A. Pizzardi”, Largo B. Nigrisoli 2, 40133 Bologna, Italy.

[432]
TÍTULO / TITLE: - Breakages at YWHAE, FAM22A, and FAM22B loci in uterine angiosarcoma: A case report with immunohistochemical and genetic analysis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
- Enlace al texto completo (gratuito o de pago) 1016/j.prp.2013.09.009
AUTORES / AUTHORS: - Suzuki S; Tanioka F; Minato H; Ayhan A; Kasami M; Sugimura H
INSTITUCIÓN / INSTITUTION: - Division of Pathology and Division of Laboratory Medicine, Iwata City Hospital, Japan. Electronic address: shiosuzuki-path@umin.net.
RESUMEN / SUMMARY: - Described herein is the first reported case of a uterine angiosarcoma with breakages at three loci, YWHAE (17p13), FAM22A (10q23) and FAM22B (10q22). A 62-year-old postmenopausal woman was found to have endometrial thickening of her uterus. An endometrial biopsy indicated a malignant, spindle cell neoplasm. A total hysterectomy with bilateral salpingooopherectomy was performed. Histologic examination of the uterine specimen showed a malignant tumor consisting of irregular rudimentary vascular channels and solid small nests diffusely infiltrating to the middle of the myometrial wall. The tumor cells were epithelioid, and displayed eosinophilic cytoplasm and vesicular nuclei in some areas of the tumor. Immunohistochemically, the tumor cells showed vascular differentiation; they were diffusely positive for CD31 and D2-40 but were negative for factor VIII and CD34. In the course of the procedure of differential diagnoses, we included fluorescence in situ
hybridization analysis for detection of a FAM22B-YWHAE fusion gene resulting from \( t(10;17)(q22;p13) \), recently reported in a series of endometrial stromal sarcoma, and unexpectedly identified breakages at three loci, i.e. YWHAE (17p13), FAM22A (10q23) and FAM22B (10q22). Collectively, these findings suggest that abnormality in the loci of YWHAE, FAM22A and FAM22B, which are known to be associated with oncogenesis of endometrial stromal sarcoma, may contribute to the development of uterine angiosarcoma.

[433]

**TITULO / TITLE:** Osteosarcomas of the jaws differ from their peripheral counterparts and require a distinct treatment approach. Experiences from the DOESAK Registry.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Baumhoer D; Brunner P; Eppenberger-Castori S; Smida J; Nathrath M; Jundt G

**INSTITUCIÓN / INSTITUTION:** Bone Tumor Reference Center and DOESAK Reference Registry (German-Austrian-Swiss Working Group of Tumors of the Face and Jaws) at the Institute of Pathology, University Hospital Basel, Basel, Switzerland; Institute of Pathology, University Hospital Basel, Basel, Switzerland; Clinical Cooperation Group Osteosarcoma, Helmholtz Zentrum Muenchen, German Research Center for Environmental Health, Neuherberg, Germany. Electronic address: dbaumhoer@mac.com.

**RESUMEN / SUMMARY:** OBJECTIVE: We aim to emphasize crucial differences between osteosarcomas of the jaws (OSj) and those of the peripheral skeleton (OSp) and to question current therapeutic concepts in presenting a comprehensive study on 214 patients. BACKGROUND: OSj account for only 6% of all osteosarcomas (OS) but seem to represent a clinically and prognostically distinct subgroup. Due to the limited experience with this rare disease it is still a matter of debate if (neo-)adjuvant chemotherapy can improve the outcome of patients like in OSp or if OSj patients can be cured by surgical treatment only. METHODS: 214 well characterized OSj patients with long-term follow up are presented and the influence of clinico-pathological parameters affecting the prognosis of patients is discussed. RESULTS: The OSj patients in our series showed metastatic spread far less frequently (17.6% of cases) and later in the course of the disease (26 months after diagnosis on average) compared to OSp. Consequently, complete resection of the tumors resulted in an excellent long-term survival (83.2% after 10 years). Neoadjuvant or adjuvant treatment applied in a smaller subset of patients, furthermore, failed to show any additional favorable effect. CONCLUSION: Whereas OSp is regarded as systemic disease at the time of diagnosis in which >90% of patients develop lung metastases without multimodality treatment, the vast majority of OSj patients seem to be curable by complete resection only. Based on the findings presented here, multimodality treatment should be critically scrutinized in OSj patients.
Enlace al texto completo (gratuito o de pago)
emission tomography-positive patients were estrogen receptor negative and the five positron emission tomography-negative patients were estrogen receptor positive (P = 0.073). The Ki-67 index was 10% or higher in the four positron emission tomography-positive patients, but less than 5% in the five positron emission tomography-negative patients (P = 0.003). Three patients with positron emission tomography-positive tumors received more aggressive treatment (e.g. cytotoxic chemotherapy and additional surgery) than did those with positron emission tomography-negative tumors. One patient who died of disease had positron emission tomography-positive tumors, was negative for estrogen and progesterone receptors, and had a 20% Ki-67 index. CONCLUSION: 18 F-Fluorodeoxyglucose uptake was associated with tumor biology of recurrent or metastatic endometrial stromal sarcoma. 18 F-fluorodeoxyglucose-positron emission tomography was useful for developing treatment strategies for recurrent or metastatic endometrial stromal sarcoma.

[436]
**TÍTULO / TITLE:** Analysis of serum insulin growth factor-1 concentrations in localized osteosarcoma: A children’s oncology group study.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Borinstein SC; Barkauskas DA; Bernstein M; Goorin A; Gorlick R; Krailo M; Schwartz CL; Wexler LH; Toretsky JA

**INSTITUCIÓN / INSTITUTION:** Department of Pediatrics, Division of Pediatric Hematology/Oncology, Vanderbilt University, Nashville, TN, Tennessee.

**RESUMEN / SUMMARY:** To investigate the role of insulin-like growth factor-1 (IGF-1), in localized osteosarcoma, serum levels of IGF-1, IGFBP-2, and IGFBP-3 were measured in 224 similarly treated, newly diagnosed patients. We demonstrated that younger patients had lower concentrations of IGF-1 and IGFBP-3 compared to older (P < 0.001) along with lower IGFBP-3:IGF-1 and IGFBP-2:IGF-1 ratios (P < 0.001). IGFBP-2 did not correlate with age (P = 0.16), yet IGFBP-2:IGF-1 ratios were higher in the younger population (P < 0.001). These findings show that older patients have higher concentrations of free IGF-1. None of IGF-1, IGFBP-2, nor IGFBP-3 concentrations were associated with event-free nor overall survival. Pediatr Blood Cancer © 2013 Wiley Periodicals, Inc.

[437]
**TÍTULO / TITLE:** A rare case of malignant epithelioid angiomyolipoma in multiple locations: multifocal disease or metastases?

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** Ann Ital Chir. 2013 Nov 4;84. pii: S0003469X13021106.

**AUTORES / AUTHORS:** Crocetti D; Pedulla G; Tarallo MR; De Gori A; Paliotta A; Cavallaro G; De Toma G

**RESUMEN / SUMMARY:** BACKGROUND: Perivascular epithelioid cell tumors (PEComas), make up a family of extremely rare mesenchymal neoplasms, with characteristic morphological, immunohistochemical and molecular findings. Malignant PEComas and gastrointestinal epithelioid angiomyolipoma (E-AML) are especially rare.
To the best of our knowledge E-AML have not been found in the breast. The difficulty in determining what constitutes optimal therapy for PEComas, owing to the sparse literature available, led us to report this rare case. METHODS: We report a case of a 44-year-old woman, with a family history of multiple endocrine neoplasia syndrome (MEN) (gastrinoma, medullary thyroid cancer and parathyroid hyperplasia), affected by PEComa located in the kidney, stomach, ileum, liver and breast. RESULTS: The renal, gastric, ileal and mammalian tumors were completely resected, with no evidence of local disease. Liver lesions were biopsied. The morphological and immunohistochemical findings confirm the diagnosis of PEComa. CONCLUSION: On this basis it is difficult to determine if some E-AML are multifocal tumors or metastatic disease. KEY WORDS: Breast Gastrointestinal; Malignant epithelioid angiomylipoma; PEComas.

[438]
**TÍTULO / TITLE:** Fibroid infected with Escherichia coli requiring surgical removal following uterine artery embolization.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Sterling L; Boutet M; Colak E; Lefebvre G

**INSTITUCIÓN / INSTITUTION:** Department of Obstetrics and Gynecology, St. Michael’s Hospital, Toronto ON.

**RESUMEN / SUMMARY:** BACKGROUND: Uterine fibroid necrosis and infection is a rare but potentially serious event following uterine artery embolization (UAE). We describe a case of surgical removal of an infected necrotic uterine fibroid. CASE: A 31-year-old Jehovah’s Witness with severe anemia presented with sepsis following UAE. The uterus was preserved by performing transvaginal surgical removal. Final pathology demonstrated Escherichia Coli infection of the necrotic fibroid. The patient improved postoperatively. CONCLUSION: Surgical removal of an infected necrotic fibroid may be a preferred option for women wishing to avoid hysterectomy following UAE. Appropriate case selection and optimization of hemoglobin concentration before UAE is important to minimize complications.

[439]
**TÍTULO / TITLE:** Solitary Epicranial Neurofibroma With NF1-Related Germline Mutation: Case Report.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Sugiyama N; Tsutsumi S; Akiba C; Nakanishi H; Ogino I; Yasumoto Y; Arai H; Ito M

**INSTITUCIÓN / INSTITUTION:** Department of Neurological Surgery, Juntendo University Urayasu Hospital.

**RESUMEN / SUMMARY:** A 33-year-old male became aware of a painless soft mass in the left occipital region. His medical and family history were unremarkable for neurofibromatosis type 1 (NF1) or other genetic disorders. Physical examination showed no signs of NF1. Neurological and ophthalmological examinations found no abnormality. Cranial computed tomography showed an isodense mass located
subcutaneously with irregular deformities in the adjacent occipital bone. Magnetic resonance (MR) imaging demonstrated that the lesion, 7.5 x 5.5 cm in diameter, was hypointense both on T1- and T2-weighted images and intensely enhanced after gadolinium infusion. The patient requested to remove the large mass. The subcutaneous tumor was well circumscribed, encapsulated, and less vascular, and resected en bloc. The histological diagnosis was neurofibroma without findings of cell atypia, whereas genomic exploration identified abnormal gains in NF1 gene, and resultant absence of neurofibromin, a protein coded on NF1 gene. Solitary neurofibromas in “clinically” non-NF1 patients may originate from the genomic changes in NF1 gene.

[440]
**TITULO / TITLE:** - Sino-orbital Spelunking: Stalagmite Formation in Fibrous Dysplasia with Dystrophic Calcification.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - Orbit. 2013 Oct 8.

**AUTORES / AUTHORS:** - Katz SE; Das S; Wakely PE Jr; Kirsch C

**INSTITUCIÓN / INSTITUTION:** - Department of Ophthalmology.

**RESUMEN / SUMMARY:** - ABSTRACT Fibrous dysplasia (FD) involving the mid-face may be associated with cystic and other secondary changes that make diagnosis more difficult. We present a case of FD associated with a remote history of blunt facial trauma and extensive cystic changes involving the medial orbit and sinuses. An endoscopic exploration revealed “stalagmites” along the floor of the cystic cavity that were consistent with dystrophic calcification. This case was unusual given the degree of dystrophic calcification and the presence of sino-orbital stalagmites within the cystic cavity. Radiologic and pathologic features of the stalagmites are characterized.

[441]
**TITULO / TITLE:** - Dramatic response to sirolimus in lymphangioleiomyomatosis.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - Can Respir J. 2013 Oct 17. pii: 15685.

**AUTORES / AUTHORS:** - Rozenberg D; Thenganatt J

[442]
**TITULO / TITLE:** - Proton pump inhibitor chemosensitization in human osteosarcoma: from the bench to the patients' bed.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Ferrari S; Perut F; Fagioli F; Brach Del Prever A; Meazza C; Parafioriti A; Picci P; Gambarotti M; Avnet S; Baldini N; Fais S

**RESUMEN / SUMMARY:** - BACKGROUND: Major goals in translational oncology are to reduce systemic toxicity of current anticancer strategies and improve effectiveness. An extremely efficient cancer cell mechanism to avoid and/or reduce the effects of highly
cytotoxic drugs is the establishment of an acidic microenvironment, an hallmark of all malignant tumors. The H+ -rich milieu that anticancer drugs meet once they get inside the tumor leads to their protonation and neutralization, therefore hindering their access into tumor cells. We have previously shown that proton pump inhibitors (PPI) may efficiently counterattack this tumor advantage leading to a consistent chemosensitization of tumors. In this study, we investigated the effects of PPI in chemosensitizing osteosarcoma. METHOD: MG-63 and Saos-2 cell lines were used as human osteosarcoma models. Cell proliferation after pretreatment with PPI and subsequent treatment with cisplatin was evaluated by using erythrosin B dye vital staining. Tumour growth was evaluated in xenograft treated with cisplatin after PPI pretreatment. Subsequently, a multi-centre historically controlled trial, was performed to evaluate the activity of a pre-treatment administration of PPIs as chemosensitizers during neoadjuvant chemotherapy based on methotrexate, cisplatin, and adriamycin. RESULTS: Preclinical experiments showed that PPI sensitize both human osteosarcoma cell lines and xenografts to cisplatin. A clinical study subsequently showed that pretreatment with PPI drug esomeprazole leads to an increase in the local effect of chemotherapy, as expressed by percentage of tumor necrosis. This was particularly evident in chondroblastic osteosarcoma, an histological subtype that normally shows a poor histological response. Notably, no significant increase in toxicity was recorded in PPI treated patients. CONCLUSION: This study provides the first evidence that PPI may be beneficially added to standard regimens in combination to conventional chemotherapy.

[443]
TÍTULO / TITLE: - Serum VEGF-D concentration as a biomarker of lymphangioleiomyomatosis severity and treatment response: a prospective analysis of the Multicenter International Lymphangioleiomyomatosis Efficacy of Sirolimus (MILES) trial.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Young LR; Lee HS; Inoue Y; Moss J; Singer LG; Strange C; Nakata K; Barker AF; Chapman JT; Brantly ML; Stocks JM; Brown KK; Lynch JP 3rd; Goldberg HJ; Downey GP; Swigris JJ; Taveira-Dasilva AM; Krischer JP; Trapnell BC; McCormack FX
INSTITUCIÓN / INSTITUTION: - Divisions of Pediatric Pulmonary Medicine and Allergy, Pulmonary and Critical Care, Vanderbilt University School of Medicine, Nashville, TN, USA (L R Young MD); Pediatrics Epidemiology Center, Department of Pediatrics, University of South Florida, Tampa, FL, USA (H-S Lee PhD, Prof J P Krischer PhD); Department of Diffuse Lung Diseases and Respiratory Failure, Clinical Research Center, National Hospital Organization Kinki-Chuo Chest Medical Center, Osaka, Japan (Y Inoue MD); National Heart Lung and Blood Institute, National Institutes of Health, Bethesda, MD, USA (J Moss MD, A M Taveira-DaSilva MD); Division of Respirology, Department of Medicine, University of Toronto, Toronto, ON, Canada (L G Singer MD); Division of Pulmonary and Critical Care Medicine, Medical University of South Carolina, Charleston, SC, USA (Prof C Strange MD); Bioscience Medical Research Center, Niigata University Medical and Dental Hospital, Niigata, Japan (Prof
RESUMEN / SUMMARY: 
- BACKGROUND: VEGF-D is a lymphangiogenic growth factor that has a key role in tumour metastasis. Serum VEGF-D concentrations are increased in most patients with lymphangioleiomyomatosis, a rare neoplasm associated with mTOR-activating tuberous sclerosis gene mutations, lymphadenopathy, metastatic spread, and pulmonary cyst formation. We used data from the Multicenter International Lymphangioleiomyomatosis Efficacy of Sirolimus (MILES) trial to assess the usefulness of serum VEGF-D concentration as a marker of severity and therapeutic response to sirolimus in patients with lymphangioleiomyomatosis. 
- METHODS: In the MILES trial, patients with lymphangioleiomyomatosis who had forced expiratory volume in 1 second (FEV1) of 70% or less of predicted were randomly assigned (1:1) to 12 months masked treatment with sirolimus or placebo. Serum VEGF-D concentrations were measured at baseline, 6 months, and 12 months. We used a linear regression model to assess associations of baseline VEGF-D concentrations with markers of disease severity, and a linear mixed effects model to assess the associations of VEGF-D concentrations with between-group differences in clinical, physiological, and patient-reported outcomes. 
- FINDINGS: We included 42 patients from the placebo group and 45 from the sirolimus group in our analysis. Baseline VEGF-D concentrations in individual patients varied from 0.34 ng/mL to 16.7 ng/mL. Baseline VEGF-D concentrations were higher in patients who needed supplemental oxygen than in those who did not need supplemental oxygen (1.7 ng/mL [IQR 0.99-3.36] vs 0.84 ng/mL [0.52-1.39]; p<0.0001) and in those who had a bronchodilator response than in those who did not (2.01 ng/mL [0.99-2.86] vs 1.00 ng/mL [0.61-2.15]; 0.0273). Median serum VEGF-D concentrations were similar at baseline in the sirolimus and placebo groups, and fell from baseline at 6 and 12 months in the sirolimus group but remained roughly stable in the placebo group. Each one-unit increase in baseline log(VEGF-D) was associated with a between-group difference in baseline-to-12-month FEV1 change of 134 mL (p=0.0007). In the sirolimus group, improvement in baseline-to-12-month FEV1 occurred in 15 of 23 (65%) VEGF-D responders (ie, those in whom baseline-to-12-month VEGF-D concentrations decreased by more than they did in any patients in the placebo group) and four of 15 (27%) VEGF-D non-responders (p=0.0448). 
- INTERPRETATION: Serum VEGF-D is a biologically plausible and useful biomarker in lymphangioleiomyomatosis that correlates with disease severity and treatment response. Measurement of serum VEGF-D concentrations could inform the risk-benefit
analysis of sirolimus therapy in patients with lymphangioleiomyomatosis and reduce the numbers of patients needed for clinical trials.

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[444]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Saito Y; Shimada M; Utsunomiya T; Morine Y; Imura S; Ikemoto T; Mori H; Hanaoka J; Sugimoto K; Iwahashi S; Yamada S; Asanoma M; Ishibashi H
INSTITUCIÓN / INSTITUTION: - Department of Surgery, Institute of Health Biosciences, the University of Tokushima Graduate School.
RESUMEN / SUMMARY: - A patient with hepatic epithelioid angiomyolipoma (Epi-AML) with arterioporal venous shunting, who was successfully treated by a laparoscopic left lateral sectionectomy, is presented herein. AML is an uncommon benign neoplasm of the liver. Tumors composed predominantly of epithelioid cells have been subcategorized into Epi-AML, and the treatment strategy for Epi-AML is currently undetermined. There are no reports describing Epi-AML with arterioporal venous shunting to date. An arterioporal venous shunting of the liver tumor was suggested to be one of the malignant signs of the liver tumor. It would be important to differentiate Epi-AML with arterioporal venous shunting from hepatocellular carcinoma and hypervascular metastatic tumors. Minimally invasive resection, such as laparoscopic hepatectomy, for patients having Epi-AML with arterioporal venous shunting may be recommended. J. Med. Invest. 60: 262-266, August, 2013.

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[445]
TÍTULO / TITLE: - Analysis report for osteosarcoma expression profile.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Li C; Zhan C; Chen Y; Fu Q; Zhu XD; He DW; Li M; Wang ZW
INSTITUCIÓN / INSTITUTION: - Department of Orthopedics, Changhai Hospital, Second Military Medical University, Shanghai, China. mlnzhwwang@hotmail.com.
RESUMEN / SUMMARY: - BACKGROUND: Osteosarcoma is a kind of highly malignant primary bone tumor which most common in the teenage, and holds strong aggressive, earlier organs metastases mainly to lung, prone to postoperative recur. Therefore for osteosarcoma, invasion and transfer mechanism and related factors' interaction remains to be a key research subject. AIM: We aim to find biological molecules marker can be used for osteosarcoma diagnosis through contrast of osteosarcoma sample and normal tissue samples. MATERIALS AND METHODS: This analysis using human osteosarcoma expression profile data and three lesions normal tissue samples (liver, kidneys, lymph) expression data and compare them, and find significant specifically expressed genes, according to their function. RESULTS: Research shows that the cancer cell proliferation, invasion, transfer and recurrent process involve many factors interaction, of which angiogenesis is the necessary condition of tumor growth, transfer and the recurrence. CONCLUSIONS: Now the most important positive regulatory factor of angiogenesis is VEGF (vascular endothelial growth factor) and bFGF (basic
fibroblast growth factor). Both of them are with a wide variety and close relationship of tumor angiogenesis and progress.

[446]

**TITULO / TITLE:** Endoscopic endonasal approach in the management of skull base chordomas-clinical experience on a large series, technique, outcome, and pitfalls.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** Neurosurg Rev. 2013 Nov 19.

**AUTORES / AUTHORS:** Chibbaro S; Cornelius JF; Froelich S; Tigan L; Kehrli P; Debry C; Romano A; Herman P; George B; Bresson D

**INSTITUCIÓN / INSTITUTION:** Neurosurgery Department, Strasbourg University Hospital, Strasbourg, France, schibbaro@hotmail.com.

**RESUMEN / SUMMARY:** Skull base chordomas represent very interesting neoplasms, due to their rarity, biological behavior, and resistance to treatment. Their management is very challenging. Recently, the use of a natural corridor, through the nose and the sphenoid sinus, improved morbidity and mortality allowing also for excellent removal rates. Prospective analysis of 54 patients harboring a skull base chordoma that were managed by extended endonasal endoscopic approach (EEA). Among the 54 patients treated (during a 72 months period), 21 were women and 33 men, undergoing 58 procedures. Twenty-two cases (40 %) were recurrent and 32 (60 %) newly diagnosed chordomas. Among the 32 newly diagnosed chordomas, a gross total resection was achieved in 28 cases (88 %), a near total (>95 % of tumor) in 2 cases (6 %), a partial (>50 % of tumor) in 2 cases (6 %). Among the 22 recurrent chordomas, resection was complete in 7 cases (30 %), near total in 7 (30 %), and partial in 8 (40 %). The global gross total resection rate was 65 % (35/54 cases). Four patients (11 %) recurred and 4 (11 %) progressed within a mean follow-up of 34 months (range 12-84 months). Four patients (11 %) were re-operated; one patient (1.8 %) died due to disease progression, one patient (1.8 %) died 2 weeks after surgery due to a massive bleeding from an ICA pseudo aneurysm. CSF leakage occurred in four patients (8 %), and meningitis in eight cases (14 %). No new permanent neurological deficit occurred. The EEA management of skull base chordomas requires a long and gradual learning curve that once acquired offers the possibility of either similar or better resection rates as compared to traditional approaches while morbidity is improved.

[447]

**TITULO / TITLE:** Malignant solitary fibrous tumor of the lumbar spinal root mimicking schwannoma: a case report.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Nagano A; Ohno T; Nishimoto Y; Oshima K; Shimizu K

**INSTITUCIÓN / INSTITUTION:** Department of Orthopaedic Surgery, Graduate School of Medicine, Gifu University, 1-1 Yanagido, Gifu 501-1194, Japan.
RESUMEN / SUMMARY: BACKGROUND CONTEXT: Malignant solitary fibrous tumors (SFTs) arising from the spinal cord are extremely rare and poorly understood mesenchymal neoplasms. To date, only one malignant SFT located in the spinal canal of the sacrum has been described, but none arising from the lumbar nerve root have been reported. Although most SFTs with benign histological features can be treated by complete surgical excision alone, malignant SFTs may require adjuvant therapy. However, systemic chemotherapy and radiotherapy have not been shown effective in patients with malignant SFTs. PURPOSE: To describe a patient with a malignant SFT arising from the lumbar nerve root. STUDY DESIGN: A case report and review of literature. METHODS: We describe the clinical course of the patient and the radiological and pathological findings of the tumor. The effect of systemic chemotherapy was evaluated and the relevant literature was reviewed. This work has no disclosure of funding and was approved by the Institutional Review Board of Gifu University. RESULTS: The tumor had been resected previously at another hospital, but it recurred and showed multiple metastatic lesions on both lungs within 3 months. Although the patient received systemic chemotherapy, both primary and metastatic lesions were found to be stable disease according to Response Evaluation Criteria in Solid Tumors. The patient died due to cachexia 6 months after her first visit. CONCLUSION: This patient presented with a highly unusual tumor. Even if a tumor is a dumbbell-shaped mass, similar to a neural tumor, SFT should be considered in the differential diagnosis.

[448]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Murakami M; Hirai M; Sakakibara T; Yamaki T; Kusuzaki K
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Kyoto Kujo Hospital.
RESUMEN / SUMMARY: - A 50-year-old female presented with more than 20-year history of a large subcutaneous mass in the left parieto-occipital portion. Magnetic resonance (MR) imaging revealed the lipomatous mass to show a high signal intensity in both T1- and T2-weighted images. A part of the lipomatous lesion progressed into the underlying hyperostosis and skull. The preoperative diagnosis was skull invasion of a well-differentiated liposarcoma. The tumor was removed completely, including the underlying hyperostosis and skull. Microscopy confirmed a lipoma without any lipoblasts, which was firmly attached to the reactive hyperostosis, and islands of lipoma were involved in the underlying hyperostosis and skull cortex. A pathological diagnosis of parosteal lipoma with reactive hyperostosis was made. Long-term progression of parosteal lipoma may cause to involve the underlying hyperostosis and skull, and led to the diagnosis of invasion of a malignant tumor on MR imaging.

[449]
TÍTULO / TITLE: - A case of spontaneous rupture perforation of sarcomatoid carcinoma in the urinary bladder.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

RESUMEN / SUMMARY: Desmoplastic fibroma (DF) of the bone is a rare locally aggressive tumor usually occurring in adolescents and young adults. These tumors most commonly occur in the mandibles and metaphyses of long bones but are extremely rare in small bones, often resulting in diagnostic problems. The occurrence of these tumors in the foot is especially limited. We report the clinical, radiographic, and histologic features of DF arising in the distal phalanx of the great toe and a review of the published data.

RESUMEN / SUMMARY: The value of coronary angiography in the work-up of atrial myxomas.

RESUMEN / SUMMARY: Atrial myxomas are the most common primary cardiac tumors. They are highly vascular with evidence of neovascularization and a characteristic "tumor blush" in approximately half of the cases. Although the visualization of feeding vessels has several clinical and therapeutic implications, there is still no consensus on the indication of preoperative coronary angiography to assess tumor vascularity except in patients with angina or those older than 40 years to rule out coronary artery disease. Herein, I present a case of an incidentally discovered right atrial mass receiving vascular supply from the right coronary artery. The mass was successfully excised and the diagnosis of cardiac myxoma was confirmed via histopathology. A review of the value of coronary angiography in detecting myxoma neovascularization is provided, which suggests that it can offer additional valuable information that can alter the surgical approach and therefore may be considered prior to myxoma resection.
Osteoblastoma Is a Metabolically Active Benign Bone Tumor on 18F-FDG PET Imaging.

We describe a case of a 9-y-old girl who on 18F-FDG PET imaging was found to have a highly metabolically active sacral tumor with an average standardized uptake value of 6.2. The tumor was proven to be osteoblastoma by pathologic examination. Osteoblastoma is a relatively rare benign primary bone tumor and occurs predominantly in patients younger than 20 y. The most common area of involvement is the spine. Osteoblastoma has been reported to be metabolically active on 18F-FDG PET imaging, with an average standardized uptake value of 3.2, which renders 18F-FDG PET imaging unable to differentiate benign from malignant primary bone tumors. To our knowledge, only 5 cases of osteoblastoma evaluated by 18F-FDG PET imaging have been reported in the literature; all were metabolically active on 18F-FDG PET imaging. The objective of this case report is to show that a metabolically active primary bone tumor on 18F-FDG PET imaging might be benign and not necessarily malignant.

Giant left atrial myxoma with dual coronary supply presenting with recurrent stroke.

We describe a case of a left atrial myxoma with dual coronary supply presenting with recurrent stroke.

Lipoma in the right atrium.

We describe a case of a lipoma in the right atrium.

Imaging manifestation of mammary fibromatosis.

We describe an imaging manifestation of mammary fibromatosis.

**AUTORES / AUTHORS:** Xu Y; Liu P; Lu H; Zhang S; Zhu Y

**INSTITUCIÓN / INSTITUTION:** Department of Breast Imaging, Tianjin Medical University Cancer Institute and Hospital, National Clinical Research Center of Cancer, Key Laboratory of Cancer Prevention and Therapy, Key Laboratory of Breast Cancer Prevention and Therapy, Tianjin Medical University, Ministry of Education, Tianjin, China.

**TÍTULO / TITLE:** Texture feature extraction based on wavelet transform and gray-level co-occurrence matrices applied to osteosarcoma diagnosis.

**RESUMEN / SUMMARY:** Osteosarcoma is the most common malignant bone tumor among children and adolescents. In this study, image texture analysis was made to extract texture features from bone CR images to evaluate the recognition rate of osteosarcoma. To obtain the optimal set of features, Sym4 and Db4 wavelet transforms and gray-level co-occurrence matrices were applied to the image, with statistical methods being used to maximize the feature selection. To evaluate the performance of these methods, a support vector machine algorithm was used. The experimental results demonstrated that the Sym4 wavelet had a higher classification accuracy (93.44%) than the Db4 wavelet with respect to osteosarcoma occurrence in the epiphysis, whereas the Db4 wavelet had a higher classification accuracy (96.25%) for osteosarcoma occurrence in the diaphysis. Results including accuracy, sensitivity, specificity and ROC curves obtained using the wavelets were all higher than those obtained using the features derived from the GLCM method. It is concluded that, a set of texture features can be extracted from the wavelets and used in computer-aided osteosarcoma diagnosis systems. In addition, this study also confirms that multi-resolution analysis is a useful tool for texture feature extraction during bone CR image processing.

[456]

[457]


**AUTORES / AUTHORS:** Hu S; Xu C; Guan W; Tang Y; Liu Y

**INSTITUCIÓN / INSTITUTION:** Department of Biomedical Engineering, ZhongShan School of Medicine, Sun Yat-Sen University, GuangZhou, 510060, PR China.

**RESUMEN / SUMMARY:** Osteosarcoma is the most common malignant bone tumor among children and adolescents. In this study, image texture analysis was made to extract texture features from bone CR images to evaluate the recognition rate of osteosarcoma. To obtain the optimal set of features, Sym4 and Db4 wavelet transforms and gray-level co-occurrence matrices were applied to the image, with statistical methods being used to maximize the feature selection. To evaluate the performance of these methods, a support vector machine algorithm was used. The experimental results demonstrated that the Sym4 wavelet had a higher classification accuracy (93.44%) than the Db4 wavelet with respect to osteosarcoma occurrence in the epiphysis, whereas the Db4 wavelet had a higher classification accuracy (96.25%) for osteosarcoma occurrence in the diaphysis. Results including accuracy, sensitivity, specificity and ROC curves obtained using the wavelets were all higher than those obtained using the features derived from the GLCM method. It is concluded that, a set of texture features can be extracted from the wavelets and used in computer-aided osteosarcoma diagnosis systems. In addition, this study also confirms that multi-resolution analysis is a useful tool for texture feature extraction during bone CR image processing.


**AUTORES / AUTHORS:** Cabete J; Lencastre A; Fidalgo A; Lobo L; Joao A; Serrao V

**INSTITUCIÓN / INSTITUTION:** Dermatology Department, Hospital de Santo Antonio dos Capuchos - Centro Hospitalar de Lisboa Central, Lisbon, Portugal.
ARTÍCULO 1

TÍTULO / TITLE: - Analysis of GNAS1 mutations in myxoid soft tissue and bone tumors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Walther I; Walther BM; Chen Y; Petersen I
INSTITUCIÓN / INSTITUTION: - Institute of Pathology, University Hospital Jena, Friedrich-Schiller-University Jena, Ziegelmuhlenweg 1, 07740 Jena, Germany.
RESUMEN / SUMMARY: - The aim of this study was to characterize the prevalence of GNAS1 mutations in various tumor types, including intramuscular myxomas, fibrous dysplasias, and other myxoid tumors and implications for malignant transformation. PCR and direct sequencing were applied to analyze GNAS1 mutation status using genomic DNA isolated from 97 formalin-fixed and paraffin-embedded samples, including 63 intramuscular myxomas, 19 various myxoid lesions, 8 cases of sporadically occurring fibrous dysplasia, and 7 cases of atrial myxoma. Mutations of GNAS1 were detected in 23 out of 63 (36.5%) intramuscular myxoma patients, with mutational hotspots R201H and R201C being equally affected. GNAS1 mutations in codon 201 were found in 5 out of 8 fibrous dysplasias (62.5%), while no mutations of GNAS1 were detected in the other studied entities, including atrial myxomas. GNAS1 mutation analysis has diagnostic value in screening patients with intramuscular myxoma and patients with fibrous dysplasia.

ARTÍCULO 2

TÍTULO / TITLE: - Artificial dermis (Matriderm®) followed by skin graft as an option in dermatofibrosarcoma protuberans with complete circumferential and peripheral deep margin assessment.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Bertolli E; Campagnari M; Molina AS; Macedo MP; Pinto CA; Cunha IW; Duprat Neto JP
INSTITUCIÓN / INSTITUTION: - Skin Cancer Department, Hospital AC Camargo, Sao Paulo, Brazil.
RESUMEN / SUMMARY: - Dermatofibrosarcoma protuberans (DFSP) is a locally invasive neoplasia with a pattern of infiltrative growth that leads to extended resections. To avoid unnecessary resections and spare tissues, its treatment requires an adequate assessment of the margins. We present a case where artificial dermis (Matriderm®) was used followed by skin graft for reconstruction. We present a 50-year-old woman with a DFSP in the occipital region. She was referred to us after a first surgery with positive margins. A wide local excision with a 2-cm margin was performed and periosteal tissue was also removed, which led to exposure of the skull. Matriderm was placed on the bone surface and dressings were changed every other day. Meanwhile, margins were evaluated by the complete circumferential and peripheral deep margin assessment (CCPDMA) and were positive for DFSP in the superior margin. After 4
weeks the area was completely covered by granulation tissue and a new resection followed by reconstruction with a skin graft was performed. With regard to the difficulties in the margin assessment in DFSP, we present artificial dermis (Matriderm) as an option for reconstructive surgery in these patients, especially when a skin graft cannot be performed as a first option.

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[460]
**TÍTULO / TITLE:** Paraspinal desmoid-type fibromatosis as a cause of low back pain.
**RESUMEN / SUMMARY:** [Enlace al Resumen / Link to its Summary](#)

**AUTORES / AUTHORS:** Furlan JC; Valiante T; Dickson B; Kiehl TR
**INSTITUCIÓN / INSTITUTION:** Division of Neurology, Department of Medicine, University of Toronto, 1 King’s College Cir, Toronto, ON M5S 1A8, Canada; Department of Genetics and Development, Toronto Western Research Institute, 399 Bathurst St, McL 12-407 Toronto, ON M5T 2S8, Canada; Lyndhurst Centre, Toronto Rehabilitation Institute, 520 Sutherland Dr, Toronto, ON M4G 3V9, Canada.

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[461]
**TÍTULO / TITLE:** Small Molecule Inhibition of PAX3-FOXO1 through AKT Activation Suppresses Malignant Phenotypes of Alveolar Rhabdomyosarcoma.
**RESUMEN / SUMMARY:** [Enlace al Resumen / Link to its Summary](#)
**REVISTA / JOURNAL:** Mol Cancer Ther. 2013 Nov 27.

**AUTORES / AUTHORS:** Jothi M; Mal M; Keller C; Mal AK
**INSTITUCIÓN / INSTITUTION:** Authors’ Affiliations: 1Department of Cell Stress Biology, Roswell Park Cancer Institute, Buffalo, New York; and 2Pediatric Cancer Biology Program, Pape Family Pediatric Research Institute, Department of Pediatrics, Oregon Health and Science University, Portland, Oregon.
**RESUMEN / SUMMARY:** Alveolar rhabdomyosarcoma comprises a rare highly malignant tumor presumed to be associated with skeletal muscle lineage in children. The hallmark of the majority of alveolar rhabdomyosarcoma is a chromosomal translocation that generates the PAX3-FOXO1 fusion protein, which is an oncogenic transcription factor responsible for the development of the malignant phenotype of this tumor. Alveolar rhabdomyosarcoma cells are dependent on the oncogenic activity of PAX3-FOXO1, and its expression status in alveolar rhabdomyosarcoma tumors correlates with worst patient outcome, suggesting that blocking this activity of PAX3-FOXO1 may be an attractive therapeutic strategy against this fusion-positive disease. In this study, we screened small molecule chemical libraries for inhibitors of PAX3-FOXO1 transcriptional activity using a cell-based readout system. We identified the Sarco/endoplasmic reticulum Ca2+-ATPases (SERCA) inhibitor thapsigargin as an effective inhibitor of PAX3-FOXO1. Subsequent experiments in alveolar rhabdomyosarcoma cells showed that activation of AKT by thapsigargin inhibited PAX3-FOXO1 activity via phosphorylation. Moreover, this AKT activation appears to be...
associated with the effects of thapsigargin on intracellular calcium levels. Furthermore, thapsigargin inhibited the binding of PAX3-FOXO1 to target genes and subsequently promoted its proteasomal degradation. In addition, thapsigargin treatment decreases the growth and invasive capacity of alveolar rhabdomyosarcoma cells while inducing apoptosis in vitro. Finally, thapsigargin can suppress the growth of an alveolar rhabdomyosarcoma xenograft tumor in vivo. These data reveal that thapsigargin-induced activation of AKT is an effective mechanism to inhibit PAX3-FOXO1 and a potential agent for targeted therapy against alveolar rhabdomyosarcoma. Mol Cancer Ther; 12(12); 1-12. ©2013 AACR.

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[462]

TÍTULO / TITLE: - Classification of rhabdomyosarcoma and its molecular basis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
- Enlace al texto completo (gratuito o de pago) 1097/PAP.0b013e3182a92d0d
AUTORES / AUTHORS: - Parham DM; Barr FG
INSTITUCIÓN / INSTITUTION: - *Department of Pathology, University of Oklahoma Health Science Center, Oklahoma City, OK daggerLaboratory of Pathology, National Cancer Institute, Bethesda, MD.
RESUMEN / SUMMARY: - Rhabdomyosarcoma (RMS), the most common soft tissue sarcoma in children, has traditionally been classified into embryonal rhabdomyosarcoma (ERMS) and alveolar rhabdomyosarcoma (ARMS) for pediatric oncology practice. This review outlines the historical development of classification of childhood RMS and the challenges that have been associated with it, particularly problems with the diagnosis of “solid variant” ARMS and its distinction from ERMS. In addition to differences in clinical presentation and outcome, a number of genetic features underpin separation of ERMS from ARMS. Genetic differences associated with RMS subclassification include the presence of reciprocal translocations and their associated fusions in ARMS, amplification of genes in ARMS and its fusion subsets, chromosomal losses and gains that mostly occur in ERMS, and allelic losses and mutations usually associated with ERMS. Chimeric proteins encoded in most ARMS from the fusion of PAX3 or PAX7 with FOXO1 are expressed, result in a distinct pattern of downstream protein expression, and appear to be the proximate cause of the bad outcome associated with this subtype. A sizeable minority of ARMS lacks these fusions and shares the clinical and biological features of ERMS. A battery of immunohistochemical tests may prove useful in separating ERMS from ARMS and fusion-positive ARMS from fusion-negative ARMS. Because of limitation of predicting outcome solely based on histologic classification, treatment protocols will begin to utilize fusion testing for stratification of affected patients into low-risk, intermediate-risk, and high-risk groups.

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[463]

TÍTULO / TITLE: - Surgical Removal of an Intravenous Leiomyoma with Intracardiac Extension and Pulmonary Benign Metastases.
**RESUMEN / SUMMARY:** Intravenous leiomyomatosis (IVL) with right intracardiac extension or pulmonary benign metastases (PBM) is rare. We report a case of 51 year-old woman, who underwent successful extensive double stage surgical removal of the intracardiac IVL extension associated with a pulmonary limited resection where the cystic bullae and PBM were found, and a month later gynaecological operation. To our knowledge this is the first reported case of such a combination.

**TÍTULO / TITLE:** Intraparenchymal leiomyoma of the male breast.

**RESUMEN / SUMMARY:**


**AUTORES / AUTHORS:** Strader LA; Galan K; Tenofsky PL

**INSTITUCIÓN / INSTITUTION:** Department of Surgery, The University of Kansas School of Medicine - Wichita, Wichita, Kansas.

**RESUMEN / SUMMARY:**

**REVISTA / JOURNAL:** Expert Opin Pharmacother. 2013 Nov 25.

**AUTORES / AUTHORS:** Martin-Liberal J; Benson C; Judson I

**INSTITUCIÓN / INSTITUTION:** The Royal Marsden Hospital, Sarcoma Unit, Fulham Road SW3 6JJ, London, UK +44 20 7808 2200 ; +44 20 7808 2113; juan.martin@rmh.nhs.uk.

**RESUMEN / SUMMARY:**

**REVISTA / JOURNAL:** - Introduction: Adult sarcomas are rare tumors characterized, in general, by their poor prognosis and the paucity of effective treatments. However, the deeper understanding of their underlying molecular pathology, signaling pathways and key effectors has permitted the development of a number of drugs able to inhibit important processes in sarcoma pathogenesis. Some of these novel compounds have been assessed in clinical trials with successful results. Areas covered: The latest reported trials are comprehensively reviewed. Thus, the Phase III studies with pazopanib, regorafenib, muramyl tripeptide (MTP) and ridaforolimus are extensively discussed as well as the biological rationale for the use of these compounds. In addition, the most promising drugs that still are in earlier stages of development such as CDK4 and MDM2 inhibitors, cediranib, eribulin and crizotinib are also discussed. Expert opinion: It is crucial for the correct identification of active drugs in sarcomas that new clinical trials are focused on specific subtypes and/or molecular alterations. The
results of these studies should improve the prognosis of the patients affected by sarcoma in forthcoming years.

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[466]
**TÍTULO / TITLE:** - Surgical management of retroperitoneal leiomyosarcoma arising from the inferior vena cava.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Dull BZ; Smith B; Tefera G; Weber S

**INSTITUCIÓN / INSTITUTION:** - Division of General Surgery, Department of Surgery, University of Wisconsin, Madison, WI, USA, bzurebzcandull@uwhealth.org.

**RESUMEN / SUMMARY:** - Retroperitoneal leiomyosarcomas are uncommon tumors, with approximately 300 documented cases in the literature. Management necessitates complete surgical resection in order to offer patients a chance at long-term cure. Resection often presents a challenge as these tumors are often large, involving adjacent structures, and may require reconstruction of the inferior vena cava (IVC). In this article, we will present background information on retroperitoneal leiomyosarcomas and the technical aspects of surgical resection and vascular reconstructive options of the IVC.

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[467]
**TÍTULO / TITLE:** - Angioleiomyoma Uterus in an Adolescent Girl: A Highly Unusual Presentation.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Sharma C; Sharma M; Chander B; Soni A; Soni PK

**INSTITUCIÓN / INSTITUTION:** - DR RPGMC Kangra at Tanda (HP), India.

**RESUMEN / SUMMARY:** - BACKGROUND: Uterine angioleiomyoma is a very rare gynecologic tumor; only a few such cases have been reported to date and we have found no such tumor reported in an adolescent girl. CASE: We report the case of a uterine angioleiomyoma in an adolescent girl. It presented as a huge abdomino-pelvic mass. The girl had severe menorrhagia and severe anemia. Intra-operatively no distinct planes were found between myoma and myometrium. Due to severe hemorrhage, she underwent sub-total abdominal hysterectomy. SUMMARY AND CONCLUSION: Uterine angioleiomyoma is an extremely rare tumor since only 16 such cases have been reported to date. Its appearance in an adolescent girl seems to be the first case of its kind. So it is being reported not only to familiarize the managing physicians with the possibility of such a tumor and its variable presentation, but also to highlight the need for inclusion of this tumor in WHO classification of tumors of the female genital tract.
El Título es: Extraskeletal Ewing’s Sarcoma: insight into a ten years follow-up.

Resumen: Extraskeletal Ewing’s sarcoma es un tumor maligno raro de tejido suave, clasificado como parte de los tumores de la familia de los sarcomas de Ewing. Mientras que el sarcoma de Ewing clásico afecta principalmente la bóveda durante la infancia, el histotipo extrasquelético difiere en edad, localización y pronóstico. La incidencia pico ocurre durante la adolescencia y la localización típica en los miembros. Se describe un caso de una mujer de 30 años de edad con un resultado positivo después de diez años desde el primer diagnóstico de Extraskeletal Ewing’s sarcoma. El tratamiento consistió en resección quirúrgica adyuvante con radioterapia y quimioterapia. Conclusion: Nuestro reporte representa un caso atípico debido a la edad de presentación, la ubicación del neoplasm y la larga supervivencia alcanzada. Durante los últimos decenios, varios ensayos clínicos han demostrado que la supervivencia a largo plazo se puede alcanzar con el tratamiento combinado de cirugía y tratamiento multi-agente adyuvante.

Resumen: Atypical fibroxanthoma con características pseudoangiomatosas: un mímico histológico e inmunohistoquímico del sarcoma angiolinfóblico cutáneo.

Resumen: Atypical fibroxanthoma and pleomorphic dermal sarcoma pueden ser difíciles de separar de cutaneous angiosarcoma. Nos propusimos estudiar el espectro morfológico de características pseudoangiomatosas en estos tumores y el valor de la inmunohistoquímica para los marcadores endoteliales CD31, CD34, FLI1, y ERG. Once atypical fibroxanomas and 3 pleomorphic dermal sarcomas were identified. All tumors arose on sun-damaged skin of elderly men. Atypical fibroxanomas were nodular and confined to the dermis, whereas pleomorphic dermal sarcoma invaded into underlying fascia. All tumors were composed of pleomorphic epithelioid and spindle cells showing blood-filled spaces and intratumoral hemorrhage. Intracytoplasmic vacuoles (n = 4), hemosiderin deposition (n = 2), and keloidal stromal change (n = 1) were also noted. Immunohistochemically, CD31 was expressed in 43% of cases, FLI1 in 79% and smooth muscle actin in 50%. Staining for CD34, ERG, S100, HMB-45, desmin, p63 and cytokeratins was negative. Follow up (median, 43.1 months; range 1-100), available for 10 patients, showed no adverse outcome. Pseudoangiomatosous features and aberrant expression of CD31 and FLI1 in atypical fibroxanthoma and pleomorphic.
dermal sarcoma may lead to an erroneous diagnosis of cutaneous angiosarcoma. Negativity for CD34 and ERG, in particular, is a reliable differentiating feature in this setting.

[470]

**TITULO / TITLE:** - Giant gluteal lipoma presenting as a sciatic hernia.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - Hernia. 2013 Nov 15.

**AUTORES / AUTHORS:** - Dulskas A; Poskus E; Jurevicius S; Strupas K

**INSTITUCIÓN / INSTITUTION:** - Clinic of Gastroenterology, Nephrourology and Surgery, Vilnius University Hospital, Santariskiu Clinics, 2 Santariskiu Street, 08661, Vilnius, Lithuania, audrius.dulskas@gmail.com.

**RESUMEN / SUMMARY:** - BACKGROUND: Sciatic hernia is considered to be the rarest hernia of pelvic floor with less than one hundred reports published worldwide. Lipoma in the hernia sac is even more unique pathology with only few cases reported in the literature. We report a case of gluteal lipoma protruding into pelvis, displacing rectum with bladder and presenting as a sciatic hernia. CASE PRESENTATION: A 53-year-old male presented with an expanding, slightly reducible, right gluteal painful mass, dull pressure in lower abdomen and perianal region, back pain, urge to urinate and defecation. Lower back pain lasts for more than 7 years, other symptoms - 6 months. No spinal pathology was found on X-ray. On examination patient seemed well nourished, BMI 29, abdomen was soft, without palpable masses or signs of peritonitis. Digital rectal examination showed no pathology. There was a reducible lump on the lateral side of right gluteus. Computer tomography (CT scan) demonstrated a large intra- and extra-pelvic fatty mass traversing the greater sciatic foramen. The tumor was surgically removed through lower middle laparotomy approach. Subsequent pathological examination revealed lipoma. The patient recovered uneventfully, was discharged 8 days later. MRI scan was advised following 1 year after the surgery. CONCLUSION: The presence of a gluteal mass should always suggest the possibility of a sciatic hernia.

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**TITULO / TITLE:** - Lipoblastoma: an interesting differential of paediatric lipoma.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Lim RS; Flatman S; Sigston E; Longano A

**INSTITUCIÓN / INSTITUTION:** - Ear, Nose and Throat Department, Southern Health, Melbourne, Victoria, Australia.

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**TITULO / TITLE:** - RESPONSE TO LETTER TO THE EDITOR: CEREBELLOPONTINE ANGLE LIPOMA WITH MILD BRAINSTEM COMPRESSION IN A 13-YEAR-OLD FEMALE.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Crowson MG; Symons SP; Chen JM
INSTITUCIÓN / INSTITUTION: - Department of Otolaryngology-Head and Neck Surgery University of Toronto Sunnybrook Health Sciences Centre Toronto, Ontario, Canada

TÍTULO / TITLE: - A rare synovial sarcoma of the spine in the thoracic vertebral body.
RESUMEN / SUMMARY: - INTRODUCTION: Synovial sarcomas of the spine are very rare, most rare of which was occurring in the thoracic vertebral body. The diagnosis of synovial sarcomas was very difficult. It depends on the radiological examination, immunohistochemical examination and gene examination. The best treatment to them was completely surgical resection with negative margins. Other treatments such as radiation therapy and chemotherapy were just adjuvant. The prognosis of synovial sarcomas was disappointing. CASE PRESENTATION: A 26-year-old male patient had low back pain. The radiological examination showed bony erosion of the T7 vertebral body and no soft tissue mass around the spine. He underwent T7 resection en bloc and internal fixation with two levels above T7 and two levels below T7. Then histopathological and gene examination revealed high malignant synovial sarcoma. So he was treated by chemotherapy and external beam radiation therapy after surgery. CONCLUSION: Primary vertebral body synovial sarcoma is very rare and difficult to diagnose and treat.

TÍTULO / TITLE: - CEREBELLOPONTINE ANGLE LIPOMA WITH MILD BRAINSTEM COMPRESSION IN A 13-YEAR-OLD PATIENT.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Buyukkaya R; Buyukkaya A; Ozturk B
TÍTULO / TITLE: - Hibernoma of the chest wall: to excise or not to excise?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Seifman MA; White DC

INSTITUCIÓN / INSTITUTION: - Plastic, Reconstructive and Hand Surgery Unit, Eastern Health, Melbourne, Victoria, Australia.

TÍTULO / TITLE: - Tumor-Suppressing Effects of miR451 in Human Osteosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Xu H; Mei Q; Shi L; Lu J; Zhao J; Fu Q

INSTITUCIÓN / INSTITUTION: - Department of Orthopedics of Jinling Hospital, Nanjing University, School of Medicine, 305 Zhongshan East Road, Nanjing, 210002, China.

RESUMEN / SUMMARY: - Osteosarcoma is the 3rd most common human cancer in childhood and young adults, and is the leading cause of mortality. Recent studies suggest that miRNAs could regulate the growth and progression of osteosarcoma, indicating some novel targets for therapy. In our study, we demonstrated that miR-451 was down-regulated in human osteosarcoma U2OS, SAOS, and MG63 cells lines as well as in tumor tissue surgically resected compared with the normal tissues. Overexpression of miR-451 inhibited cell proliferation and resulted in cell apoptosis in osteosarcoma cells. G1 cell cycle arrest was also induced by miR-451. Repressed by miR-451, PGE2 and CCND1 reversed the inhibitory effects of miR-451 on proliferation. In conclusion, miR-451 played a tumor-suppressing role through modulating the expression of PGE2 and CCND1, suggesting a novel target for the diagnosis and treatment of osteosarcoma.

TÍTULO / TITLE: - Sinonasal tract chondrosarcoma: 18-Year experience at a single institution.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Guo L; Liu J; Sun X; Wang D
INSTITUCIÓN / INSTITUTION: - Department of Otolaryngology, Eye Ear Nose and Throat Hospital, Shanghai Medical College of Fudan University, 83 Fen Yang Road, Shanghai 200031, PR China.

RESUMEN / SUMMARY: - OBJECTIVE: Sinonasal tract chondrosarcomas are considered rare malignancies. Few large series evaluated the clinicopathological characteristics of these tumors. The aim of this study was to describe clinical findings, management and outcome of 24 cases of sinonasal tract chondrosarcoma from a single institution and to focus on the validity and advantage of endoscopic technique compared with conventional surgery. METHODS: A retrospective analysis of clinical information was performed on 24 patients diagnosed as the sinonasal tract chondrosarcomas between 1994 and 2011. RESULTS: There were 10 males and 14 females (age range from 7 months to 67 years; mean age, 34.9 years) in this study. The main complaints were nasal obstruction and swelling/mass. The most common affected sites were maxillary sinus and sphenoid sinus. Except one case of myxoid chondrosarcoma and two cases of mesenchymal chondrosarcoma, 17 patients (70.8%) and 4 patients (16.7%) were, respectively, grade I and II. Three patients were misclassified as other tumors at other hospitals. Two cases had a history of radiation. Five cases superimposed upon a preexisting benign bony conditions. Twenty-three of 24 patients were treated with wide surgical excision, including 15 patients with conventional surgeries by external approach and 8 patients with endoscopic surgeries. Local recurrence was observed in 12 patients. The mean interval of recurrence with endoscopic surgery (37.8 months) was longer than conventional approach (21.9 months), but it did not achieve statistical significance due to small sample size. The 5-year disease-specific survival rate was 83.3%. CONCLUSION: Chondrosarcomas of the sinonasal tract are rare. The patients with earlier diagnosis and adequate surgical treatment have a more favorable prognosis. Uncontrollable local disease resulting in compression of adjacent critical structures is the most common cause of death.

[478]

TÍTULO / TITLE: - Pancreatic metastasis from a solitary fibrous tumor of the kidney: A rare cause of acute recurrent pancreatitis.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Patel YA; Dhalla S; Olson MT; Lennon AM; Khashab MA; Singh VK

INSTITUCIÓN / INSTITUTION: - Department of Medicine, Johns Hopkins Medical Institutions, Baltimore, MD, USA. Electronic address: ypatel2@jhmi.edu.

RESUMEN / SUMMARY: - Solitary fibrous tumors are unusual spindle cell neoplasms that uncommonly originate from the kidney. We report a case of a 43-year old male who presented with acute recurrent pancreatitis secondary to a mass in the head of the pancreas. Endoscopic ultrasound with fine needle aspiration (EUS-FNA) was performed. Cytology revealed solitary fibrous tumor of the kidney. This is the first reported case of solitary fibrous tumor metastasizing to the pancreas and presenting as acute recurrent pancreatitis.
CASE REPORT: A 25-year-old woman with right subacute sinusitis, complained about discomfort in her right eye. Clinical manifestations and computed tomography were suggestive of sub-periosteal orbital ethmoid wall abscess, for which the patient underwent urgent drainage. A solid tumor was found, with a positive biopsy for alveolar rhabdomyosarcoma. Complete remission and resolution of orbital symptoms were achieved with chemotherapy and radiation therapy. DISCUSSION: Alveolar orbital rhabdomyosarcoma in adults is uncommon. Rhabdomyosarcoma has a high risk of spreading. It can simulate a sinusitis, as in our patient, early diagnosis and early treatment being especially important in these patients.
Angiosarcoma of the adrenal gland.

Uterine carcinosarcoma.

Colo-colonic intussusception owing to lipoma of transverse colon.

Hungry Bone Syndrome: Persistent Hypocalcemia Related to Osteoblastic Bone Metastases of Prostate Cancer.

Multifocal right atrial myxoma with multiple pulmonary embolism.
INSTITUCIÓN / INSTITUTION: - Gobind Ballabh Pant Hospital, Maulana Azad Medical College, New Delhi, India.

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TÍTULO / TITLE: - Benign “metastasizing” leiomyoma presenting as cavitating lung nodules.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Loukeri A; Pantazopoulos I; Tringidou R; Giampoudakis P; Valaskatzi A; Loukeri P; Kampolis C

INSTITUCIÓN / INSTITUTION: - Respiratory Intensive Care Unit, Athens Chest Hospital “Sotiria”.
RESUMEN / SUMMARY: - Benign “metastasizing” leiomyoma (BML) was initially used to describe single or multiple pulmonary nodules composed of proliferating smooth muscle cells, lacking cellular atypia, in premenopausal females 3 months to 20 years after hysterectomy for uterine leiomyoma. The lung is the most commonly involved site, thus including many malignant and benign entities in the differential diagnosis. The present article refers to a 47-year-old premenopausal woman with a history of subtotal hysterectomy for a uterine leiomyoma presenting with bilateral cavitating pulmonary nodules. A number of nodules were resected by video-assisted thoracoscopic surgery. The histological findings in correlation with the immunohistochemical results were consistent with the diagnosis of BML. The patient was subjected to bilateral salpingo-oophorectomy combined with complete removal of the remained cervix. One year later, the patient remains asymptomatic and the pulmonary nodules stable in terms of number, size, location and morphology.

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TÍTULO / TITLE: - Ischemic stroke and incidental finding of a right atrial lipoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Censi S; Squeri A; Baldelli M; Parizi ST

INSTITUCIÓN / INSTITUTION: - aCardiology Unit bRadiology Unit, GVM Care & Research-Maria Cecilia Hospital, Cotignola (RA), Italy.
RESUMEN / SUMMARY: - A young man presented with recurrent ischemic stroke under antiplatelet therapy. A patent foramen ovale (PFO) was diagnosed by transesophageal echocardiography (TEE) and the patient was referred to our institution for percutaneous closure. An echogenic mass in the right atrium was detected during the intraprocedural TEE. The interventional team decided to perform transcatheter closure of PFO under fluoroscopy and TEE guide, without complications. Subsequent cardiac magnetic resonance (CMR) imaging confirmed an encapsulated and hyperintense mass located in the roof of the right atrium. The signal intensity pattern and the absence of gadolinium contrast uptake allowed a confident diagnosis of lipoma. Cardiac lipoma accounts for about 10% of primary cardiac tumors and frequently rises
from the epicardial fat tissue. Echocardiographic images can remain equivocal about the nature of the mass and CMR offers a substantial contribution to a correct diagnosis. The tumor usually appears encapsulated and asymptomatic, but dyspnea, atrial and ventricular arrhythmias and, rarely, peripheral embolization have been reported. To our knowledge, this is the second case reported on paradoxical embolization associated with right atrial lipoma. Although the relationship of cardiac lipoma with stroke is not well defined, the potential proembolic significance of this lesion cannot be excluded, especially when a PFO coexists.

[490] TÍTULO / TITLE: - The recurrent primary retroperitoneal liposarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Nagy V; Bober J; Zavacky P; Jr OB; Svajdler M
RESUMEN / SUMMARY: - Aim: Describe a patient with multiple recurrences of the primary recurrent liposarcoma. Clinical case: A 60-years-old man complained of weight loss (BMI 18.4) with a palpable huge retroperitoneal tumour, which displaced left kidney, and was confirmed on USG and CT. Laboratory examination showed anaemia and pathological blood tests. Chest X-ray initially showed a negative finding. A complete transperitonealy surgical extirpation of the tumour with left side nephrectomy was performed on June 28, 2007. The tumour mass weight was 1900 g. It was lying on the posterior face of the kidney in diameters 170x120x120 mm, completely capsulated by thin grey-pink capsula with peripheral fat tissue on the section grey-pink, lobulary shaped, in (3/4) parts with central necrotic changes. Histopathologicaly was confirmed the primary dedifferentiated (non-lipogenous) liposarcoma low grade of malignancy. Nephrectomy specimen was confirmed as age related finding. There was no evidence of positives surgical margins. Despite oncological and surgical treatment, followed repeated recurrence with eight transperitoneal surgeries in the retroperitoneum and abdomen with extirpation of the metastases, left side hemicolecotomy, splenectomy and repeated extirpation tumour metastases from abdomen and radix mesenterii. Last tumour weighed 2900 grams. Patient died on January 9, 2011, after the eight surgeries on multiorgans failure due to hemorrhagic shock and persistent atrial fibrilaton by cardiopulmonary insufficiency. As a speciality, he was treated without transfusion because as Jehovah s witness he refused blood derivates. Conclusion: Despite complex surgical and oncological treatment, the prognosis in patient with recurrent liposarcoma was fatal (Tab. 1, Fig. 5, Ref. 50). Keywords: primary retroperitoneal liposarcoma, recurrence, surgery, chemotherapy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Ippolito E; Farsetti P; Boyce AM; Corsi A; De Maio F; Collins MT
BACKGROUND: Fibrous dysplasia of bone is a skeletal dysplasia with a propensity to affect the femur in its polyostotic form, leading to deformity, fracture, and pain. The proximal femur is most commonly involved with a tendency to distal progression, thereby producing the typical shepherd’s crook deformity. However, there are few data on the spectrum and progression of femoral deformities in polyostotic fibrous dysplasia.

QUESTIONS/PURPOSES: The purposes of this study were (1) to develop a radiographic classification for polyostotic fibrous dysplasia; (2) to test this classification’s intra- and interobserver reliability; and (3) to characterize the radiographic progression of polyostotic fibrous dysplasia in a population of patients with the condition who were treated with a variety of approaches at several centers.

METHODS: We retrospectively reviewed radiographs of 127 femurs from 84 adult patients affected by polyostotic fibrous dysplasia. Fifty-nine femurs had undergone one or more operations. The radiographs were evaluated in the coronal plane for neck-shaft angle and angular deformities along the whole femoral shaft down to the distal epiphysis. Four observers evaluated each film two times at intervals; intra- and interobserver reliability testing was performed using the kappa statistic. Eighty-nine femurs (70%) were available for followup to evaluate for progression at a mean of 10 years (range, 6-20 years).

RESULTS: Six reproducible patterns of deformity were identified in both untreated and operated femurs: type 1 (24%), normal neck-shaft angle with altered shape of the proximal femur; type 2 (6%), isolated coxa valga with neck-shaft angle > 140 degrees; type 3 (7%), isolated coxa vara with neck-shaft angle < 120 degrees; type 4 (20%), lateral bowing of the proximal half of the femur associated with normal neck-shaft angle; type 5 (14%), like in type 4 but associated with coxa valga; and type 6 (29%), like in type 4 but associated with coxa vara. Interobserver and intraobserver kappa values were excellent, ranging from 0.83 to 0.87. In 46 of the 89 femurs (52%) for which longitudinal radiographic documentation was available, there was progressive worsening of the original deformity, although the pattern remained the same; types 1 and 2 tended not to progress, whereas types 3 to 6 did.

CONCLUSIONS: A reproducible radiographic classification of polyostotic fibrous dysplasia-associated femoral deformities is proposed, which can serve as a tool for assessing and treating these deformities. After reviewing the radiographs of 127 femurs, we identified six reproducible patterns of femoral deformities.

LEVEL OF EVIDENCE: Level III, diagnostic study. See Guidelines for Authors for a complete description of levels of evidence.
RESUMEN / SUMMARY: - INTRODUCTION: Behcet's disease (BD) is a multi-system, chronic and relapsing disorder classified as "vasculitic syndrome". It typically affects young adult females between 20 and 40 years of age. There are some typical clinical manifestations associated with this disease, however, at times; rare sign and symptoms pose a challenge to the treating physician and making a definitive diagnosis. Presentations with cardiac symptoms are one of the extremely rare manifestations of the Behcet's disease. METHODS: The authors present clinical, laboratory and imaging findings of a patient who presented with a cardiac mass which was the first presenting feature or manifestation of Behcet's disease. RESULTS: A 19-year-old boy was admitted to our hospital for the investigation of "fever of unknown origin", weight loss, shortness of breath and a scrotal ulcer of recent on-set. X-ray chest and electrocardiograms were inconclusive. Transthoracic echocardiography revealed a right ventricular (RV) mass attached to the interventricular septum measuring 1.5 x 1.5 cms (Panel A). Cardiac MRI identified it as a RV Myxoma. In addition, on CT scan of the chest pulmonary embolism was noted. The patient underwent excision biopsy of the tumor under cardiopulmonary bypass via right atriotomy (Panel B). Histopathology of the mass described it as "an organizing thrombus with a few groups of interrupted myocardial fibers and some infiltration of lymphocytes and plasma cells". Moreover his HLA typing was found positive for HLA-B51 (5). In view of the above findings and associated lesions, the patient was diagnosed as a case of Behcet's disease. The medical management included immunosuppressant and anticoagulation. CONCLUSION: Behcet's disease, even in the absence of the typical clinical features, should be considered in the differential diagnosis of right ventricular mass, especially when dealing with young adults from the Mediterranean basin and the Middle-East.

[493]
TÍTULO / TITLE: - A case of sarcoma most consistent with the desmoplastic small round cell tumor arising from the ileal mesentery.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Maeda M; Morita S

[494]
TÍTULO / TITLE: - Radiofrequency ablation for the treatment of recurrent bone and soft-tissue sarcomas in non-surgical candidates.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Yamakado K; Matsumine A; Nakamura T; Nakatsuka A; Takaki H; Matsubara T; Asanuma K; Sudo A; Sugimura Y; Sakuma H
INSTITUCIÓN / INSTITUTION: - Department of Interventional Radiology, Mie University School of Medicine, Edobashi 2-174, Tsu, Mie, 514-8507, Japan, yama@clin.med.mie-u.ac.jp.
AIMS: The aim of this study was to evaluate the safety and clinical utility of radiofrequency ablation (RFA) retrospectively in non-surgical candidates with recurrent bone and soft-tissue sarcomas. METHODS: Percutaneous RFA was used in 52 patients (21 female, 31 male; mean age, 52.2 +/- 21.1 years; range 10-87 years) with recurrent bone and soft-tissue sarcomas. The number of tumors was 3 or fewer in 23 patients (44.2 %) and 4 or more in the others, with a mean maximum tumor diameter of 3.0 +/- 3.7 cm (range 0.5-18 cm). Safety, tumor control, and prognosis were evaluated. RESULTS: All tumors were ablated after RFA in 21 patients (40.4 %, 21/52), although 14 experienced re-recurrence. Tumors were controlled in 8 cases of recurrence by repeat RFA (n = 7) and surgical intervention (n = 1). Therefore, 15 patients (28.8 %, 15/52) were tumor-free at the end of follow-up (mean follow-up 25.5 +/- 24.2 months; range 3.9-117 months). Residual tumors were found after RFA in the other 31 patients (59.6 %, 31/52). Overall survival rates were 73.4 % (95 % CI 61.0-85.9 %) at 1 year, 39.3 % (95 % CI 23.6-54.9 %) at 3 years, and 34.3 % (95 % CI 18.0-50.7 %) at 5 years in all patients. Recurrence-free interval (p = 0.040), tumor number (p = 0.0094), and complete tumor ablation (p < 0.0001) were detected as significant prognostic factors in univariate analysis. The latter two factors were significant in multivariate analysis. The rate of major complications was 0.9 %. CONCLUSIONS: RFA is a safe and useful therapeutic option for treatment of recurrent bone and soft-tissue sarcomas. Prognostic factors found in this study will help to identify those patients who would benefit from RFA.

Hepatic metastasis with heterologous rhabdomyoblastic differentiation in a patient with gastrointestinal stromal tumor treated with imatinib. Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the intestinal tract. In patients with locally advanced and/or metastatic GIST, the introduction of tyrosine kinase inhibitor, imatinib mesylate, has transformed the management of this previously untreatable neoplasm into a treatable entity. Approximately 80% of advanced metastatic GISTs respond to imatinib treatment. However, the majority of patients suffer disease progression at a median of 2 years due to drug resistance. Generally progressing GISTs retain their typical morphology. Herein, we report an extremely rare case of progressive metastatic GIST with heterologous rhabdomyoblastic differentiation after, imatinib mesylate treatment. We also review the relevant literature.

Efficacy of sorafenib in patients with gastrointestinal stromal tumors in the third- or fourth-line treatment: A retrospective multicenter experience.
Resumen / Summary: Sorafenib es un inhibidor de múltiples quinases de tipo tirosina receptor utilizado para tratar a los pacientes con tumores estromales gastrointestinales avanceados (GISTs). El presente estudio evaluó la eficacia y tolerabilidad del tratamiento con sorafenib para los pacientes con GISTs. Entre enero de 2001 y noviembre de 2012, 25 pacientes, de múltiples centros, que habían recibido sorafenib como la tercera o cuarta línea terapéutica de GISTs fueron investigados retrospectivamente. De estos pacientes, 17 eran varones y ocho eran mujeres. La edad media fue de 54.0 años (rango, 16-82 años). De los pacientes, 21 habían recibido imatinib por más de seis meses y cuatro por menos de seis meses. El beneficio clínico del sorafenib fue del 40.0%. Los eventos adversos relacionados con el tratamiento fueron informados en el 72% de los pacientes. Estos eventos adversos generalmente fueron de intensidad leve a moderada. La supervivencia libre de progresión (PFS) y supervivencia global (OS) de los pacientes que recibieron sorafenib fue de 7.2 y 15.2 meses, respectivamente. La duración de uso de imatinib fue un factor pronóstico independiente para PFS y OS. Sorafenib es un tratamiento efectivo en pacientes con GISTs mostrando un beneficio clínico del 40.0% y una tolerabilidad aceptable.
protein-like 4-anaplastic lymphoma kinase fusion gene had transformed into an undifferentiated sarcoma. This case suggests that echinoderm microtubule-associated protein-like 4-anaplastic lymphoma kinase fusion is an oncogenic event in not only carcinomas but also sarcomas originating from stromal cells.

[498]
TÍTULO / TITLE: - Atypical presentation of Kaposi’s sarcoma in an HIV-infected patient.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Moloi MW; Zhou F; Baliki K; Kayembe MK; Cainelli F; Vento S
INSTITUCIÓN / INSTITUTION: - Department of Medicine, Princess Marina Hospital, Gaborone, Botswana.

[499]
TÍTULO / TITLE: - Common gene variants in RAD51, XRCC2 and XPD are not associated with clinical outcome in soft-tissue sarcoma patients.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Szkandera J; Absenger G; Liegl-Atzwanger B; Pichler M; Stotz M; Gerger S; Zacherl M; Renner W; Haijun M; Leithner A; Gerger A
INSTITUCIÓN / INSTITUTION: - Division of Clinical Oncology, Research Unit Genetic Epidemiology and Pharmacogenetics, Department of Medicine, Medical University of Graz, Graz, Austria.

RESUMEN / SUMMARY: - BACKGROUND: DNA repair mechanisms play a major role in cancer risk and progression. Germline variants in DNA repair genes may result in altered gene function and/or activity, thereby causing inter-individual differences in a patient’s tumor recurrence capacity. In genes of the DNA repair pathway the gene variants RAD51 rs1801320 G>C, XRCC2 rs3218536 G>A and XPD rs13181 A>C have been previously related to genetic predisposition and prognosis of various cancer entities. In this study we investigated the association between these polymorphisms and time to recurrence (TTR) and overall survival (OS) in soft-tissue sarcoma (STS) patients after curative surgery. METHODS: Two hundred sixty STS patients were included in this retrospective study. Germine DNA was genotyped by 5'-exonuclease (TaqMan) technology. Kaplan Meier curves and multivariate Cox proportional models were calculated for TTR and OS. RESULTS: A statistically significant association was observed between tumor grade and adjuvant radiotherapy and TTR and between tumor grade and OS. No association was found between RAD51 rs1801320 G>C, XRCC2 rs3218536 G>A and XPD rs13181 A>C and TTR and OS in univariate and multivariate analysis. CONCLUSION: Our results underline a prognostic effect of tumor grade and adjuvant radiotherapy in STS patients but indicate no association between RAD51 rs1801320 G>C, XRCC2 rs3218536 G>A and XPD rs13181 A>C and clinical outcome in STS patients after curative surgery.

[500]
Salvage treatment experience in advanced synovial sarcoma: a multicenter retrospective analysis of the anatolian society of medical oncology.

The Primary Extra-gastrointestinal Stromal Tumor of Pleura: A Case Report and a Literature Review.

Background: We aimed to evaluate prognostic factors and response rates to various treatment approaches to patients with synovial sarcoma in an advanced setting. Materials and Methods: We retrospectively reviewed the medical records of 55 patients (18 pts; 32.7% women) diagnosed with synovial sarcomas. Twenty had metastatic disease at the time of diagnosis while the remainder of the study group consisted of patients who developed metastatic or inoperable locally advanced disease during follow up. Results: The median follow up time was 15 months (range: 1-53). Regarding outcomes for the 55 patients, 3 and 5 year overall survival rates were 26% and 14%, respectively. In univariate analyses among demographic factors female gender was associated with a better outcome (p=0.030). Patients with early progressing disease (<2 years) had a worse prognosis when compared to patient group with late relapse, but this difference did not reach statistical significance (p=0.056). According to multivariate Cox regression analysis patients who had undergone metastasectomy had a significant survival advantage (p=0.044). The overall response rate to different salvage chemotherapy regimens given as second line treatment was around 42.9-53.9% for all regimens. There were no statistically significant differences between chemotherapy regimens given in either second or third line settings in terms of overall survival. Conclusions: We observed no major differences in terms of response rate and survival between different salvage chemotherapy regimens. Although metastatic disease still carries a poor prognosis, metastasectomy was found to be associated with improved survival.

The gastrointestinal stromal tumor is the most common mesenchymal neoplasm of the gastrointestinal tract. The gastrointestinal stromal tumor universally expresses KIT and DOG-1 and frequently harbors oncogenic mutations in the KIT gene. While the gastrointestinal stromal tumor usually arises in the alimentary...
tract, it is rarely found in the extragastrointestinal area. When it is, it is called an extragastrointestinal stromal tumor. Although the pathogenesis, prognostic factors and outcomes of gastrointestinal stromal tumors are well known, those of extragastrointestinal stromal tumors have not been fully studied. We report, herein, a unique primary extragastrointestinal stromal tumor from the pleura in a 73-year-old woman who presented with pleural mass. The extragastrointestinal stromal tumor was surgically resected and confirmed by means of an immunohistochemical study and a molecular analysis.

[502]

**TÍTULO / TITLE:** Concomitant occurrence of EGFR (epidermal growth factor receptor) and KRAS (V-Ki-ras2 Kirsten rat sarcoma viral oncogene homolog) mutations in an ALK (anaplastic lymphoma kinase)-positive lung adenocarcinoma patient with acquired resistance to crizotinib: a case report.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Rossing HH; Grauslund M; Urbanska EM; Melchior LC; Rask CK; Costa JC; Skov BG; Sorensen JB; Santoni-Rugi E

**RESUMEN / SUMMARY:** BACKGROUND: Anaplastic lymphoma kinase-positive non-small cell lung carcinoma patients are generally highly responsive to the dual anaplastic lymphoma kinase and MET tyrosine kinase inhibitor crizotinib. However, they eventually acquire resistance to this drug, preventing the anaplastic lymphoma kinase inhibitors from having a prolonged beneficial effect. The molecular mechanisms responsible for crizotinib resistance are beginning to emerge, e.g., in some anaplastic lymphoma kinase-positive non-small cell lung carcinomas the development of secondary mutations in this gene has been described. However, the events behind crizotinib-resistance currently remain largely uncharacterized. Thus, we report on an anaplastic lymphoma kinase-positive non-small cell lung carcinoma patient with concomitant occurrence of epidermal growth factor receptor and V-Ki-ras2 Kirsten rat sarcoma viral oncogene homolog mutations upon development of crizotinib-resistance.

**CASE PRESENTATION:** A 61-year-old Caucasian never-smoking male was diagnosed with anaplastic lymphoma kinase-positive pulmonary adenocarcinoma, stage T4N3M1b. Treatment with crizotinib initially resulted in complete objective response in the thorax and partial response in the abdomen, but after 8 months of therapy the patient acquired resistance and progressed. Biopsies from new metastases revealed development of epidermal growth factor receptor and V-Ki-ras2 Kirsten rat sarcoma viral oncogene homolog mutations concomitant with the original anaplastic lymphoma kinase gene rearrangement and without signs of anaplastic lymphoma kinase fusion gene amplification or secondary anaplastic lymphoma kinase mutations.

**CONCLUSION:** To our knowledge, this is the first report of an anaplastic lymphoma kinase-positive pulmonary adenocarcinoma, which upon emergence of crizotinib resistance acquired 2 new somatic mutations in the epidermal growth factor receptor and V-Ki-ras2 Kirsten rat sarcoma viral oncogene homolog genes, respectively, concomitant with the original anaplastic lymphoma kinase rearrangement. Thus, these 3 driver mutations, usually considered mutually exclusive, may coexist in advanced non-small cell lung carcinoma that becomes resistant to crizotinib, presumably because
heterogeneous tumor clones utilize epidermal growth factor receptor and/or V-Ki-ras2 Kirsten rat sarcoma viral oncogene homolog signaling to circumvent the inhibition of anaplastic lymphoma kinase-mediated signaling by crizotinib. The identification of new targetable somatic mutations by tumor re-biopsy may help clarify the mechanism behind the development of the acquired crizotinib resistance and pave the way for combined strategies involving multiple targeted therapies.

[503]

TÍTULO / TITLE: - Surgical procedures and prognostic factors for local recurrence of soft tissue sarcomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Sugiura H; Nishida Y; Nakashima H; Yamada Y; Tsukushi S; Yamada K
INSTITUCIÓN / INSTITUTION: - Department of Orthopaedic Surgery, Aichi Cancer Center Hospital, 1-1 Kanokoden, Chikusa-ku, Nagoya, 464-8681, Japan, shidesh3@aichi-cc.jp.
RESUMEN / SUMMARY: - BACKGROUND: Patients with local recurrence of soft tissue sarcomas are predisposed to future recurrences because treatment is challenging and complicated by prior therapy. This study investigated clinical outcomes following surgical procedures for locally recurrent soft tissue sarcomas and risk factors for recurrence and metastasis. METHODS: A retrospective analysis was conducted with 105 patients (52 males, 53 females) who underwent surgical procedures for local recurrence without distant metastasis of soft tissue sarcoma between 1987 and 2009. Patient follow-up ranged from 1 to 12 years (mean 4.9 years). RESULTS: Overall 5- and 10-year survival rates were 83.4 and 67.7 %, respectively. Twenty-one patients (20.0 %) had additional local recurrences, and 23 (21.9 %) had distant metastases. Amputation rate was 10.5 % at the time of surgical procedures and 17.1 % at final follow-up. Locations deep within muscles in the upper limb or trunk and surgical margins <1 cm wide were risk factors for further local recurrence. Locations deep within muscles, tumor sizes >10 cm, high-grade malignancy, and local recurrence after radical surgery were risk factors for distant metastasis. CONCLUSIONS: Surgical margin and location were independent prognostic factors for local control, and a wider margin was especially important for recurrent tumors located in the trunk and upper extremity. For high-grade sarcomas with local recurrence after radical surgery, new approaches are needed to prevent distant metastases.

[504]

TÍTULO / TITLE: - A rare case of laryngotracheal chondrosarcoma in a patient with past history of radioiodine therapy for thyroid cancer.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Mohajeri G; Hekmatnia A; Ahrar H; Hekmatnia F; Nia RB; Afsharmoghadam N; Eftekhar M; Jafarpishe S
RESUMEN / SUMMARY: Tracheal chondrosarcoma is a rare malignant mesenchymal tumor and there are less than 15 reports in the literature. We report a rare case of laryngotraceal chondrosarcoma in a 74-year-old man. He gave a history of radioiodine therapy for thyroid papillary carcinoma about 24 years ago. Diagnostic steps, histological presentation, and therapy are described in detail.

TÍTULO / TITLE: Neutrality of miniSTR D22S1045 marker by Ewing’s sarcoma phenotype.

RESUMEN / SUMMARY: Neutrality investigations of markers with forensic use are important to see if a phenotypic trait is being expressed in relation to the alleles of the marker. MiniSTR marker D22S1045 (locus 22q12.3) is localized near the breakpoint region of the EWS gene (22q12.2), which leads to the development of Ewing’s Sarcoma. Analyzing allele frequencies and linkage disequilibrium in Ewing’s sarcoma patients and non-affected populations, we found that the marker mD22S1045 was neutral when related to Ewing’s Sarcoma.

TÍTULO / TITLE: IGF-II increases markers of osteoblastic activity and reduces bone resorption via osteoprotegerin and RANK-ligand.

RESUMEN / SUMMARY: BACKGROUND: Bone is one of the major target tissues for Insulin-like Growth Factor I (IGF-I). Low doses of IGF-I were able to improve liver-associated osteopenia. In the present work, a model of partial IGF-I deficiency was used in order to provide insight into the mechanisms of the beneficial actions of IGF-I replacement therapy in bone. METHODS: Several proteins involved in osteoblastic/osteocyte and osteoclastic differentiation and activity were studied in the three experimental groups: control (CO) group (wild type mice, Igf +/+, n = 10), heterozygous Igf +/- group with partial IGF-I deficiency (Hz, n = 10), and heterozygous Igf +/- mice treated with IGF-I for 10 days (Hz + IGF-I, n = 10). RESULTS: Data in this paper confirm that the simple partial IGF-I deficiency is responsible for osteopenia, determined by densitometry and histopathology. These findings are associated with a reduced gene expression of osteoprotegerin, sclerostin, calcitonin receptor (CTR),
insulin-like growth factor binding protein 5 and RUNX2. IGF-I replacement therapy normalized CTR gene expression and reduced markers of osteoclastic activity.

CONCLUSIONS: Low doses of IGF-I constituted a real replacement therapy that normalized IGF-I serum levels improving the expression of most of these proteins closely involved in bone-forming, and reducing bone resorption by mechanisms related to osteoprotegerin, RANKL and PTH receptor.
BACKGROUND: Of the biological reconstruction methods for malignant bone and soft tissue tumors, reconstruction with liquid nitrogen has the advantage of maintaining continuity on the distal side of the tumor bone site (pedicle freezing procedure; PFP). This method is expected to result in early blood flow recovery, with early union and low complication rate. The purpose of this study was to compare the outcomes of the PFP and free freezing procedure (FFP) in the lower extremities. METHODS: The study included 20 patients (12 men and 8 women) with frozen autografts (FFP, 13 cases; PFP, 7 cases). The mean age of the subjects was 36.3 years (range 11-79 years), and the mean follow-up period was 56.4 months (range 12-142 months). RESULTS: Final bone union occurred in 11 patients in the FFP group (84.6 %) and in 7 patients in the PFP group (100 %). The mean union period in patients who did not need additional surgery was 9.8 months (range 4-21 months) in the FFP group and 4.8 months (range 2-7 months) in the PFP group. Postoperative complications occurred in 8 cases: infection in 3 cases, fracture in 3 cases, and joint destruction in 2 cases. Six FFP patients, and 2 PFP patients (two cases of fracture), developed postoperative complications. CONCLUSIONS: The union period was shorter and the rate of postoperative complications was lower with the PFP than with the FFP. We considered that early blood flow recovery might have led to the above results in the PFP.
AUTORES / AUTHORS: - Pundir J; Walawalkar R; Seshadri S; Khalaf Y; El-Toukhy T
INSTITUCIÓN / INSTITUTION: - Assisted Conception Unit, Guy's and St. Thomas NHS Trust, London, UK.
RESUMEN / SUMMARY: - The aim of the study was to systematically review and summarise existing evidence related to the perioperative morbidity associated with abdominal myomectomy in comparison with abdominal hysterectomy for uterine fibroids. A review of MEDLINE and EMBASE was carried out. The primary outcome was the major morbidity rate and secondary outcomes were uterine size, estimated blood loss, blood transfusion, operating time and duration of hospital stay. The results identified six observational studies including 1520 participants. All studies scored moderately on the N-OQA scale and were limited to a uterine size of up to 18 weeks. There was no significant difference in the rate of major morbidity (RR 0.94; 95% CI = 0.31, 2.81; p = 0.91) between the two operations. It was concluded that based on variable quality data from retrospective cohort studies, abdominal myomectomy and hysterectomy appear to have similar major morbidity rates for the uterine size up to 16-18 weeks. Well-designed trials with a standardised morbidity outcome and including uterine size greater than 18 weeks are required.

[513]
TÍTULO / TITLE: - Irreversible Electroporation Ablation (IRE) of Unresectable Soft Tissue Tumors: Learning Curve Evaluation in the First 150 Patients Treated.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Philips P; Hays D; Martin RC
INSTITUCIÓN / INSTITUTION: - University of Louisville, Department of Surgery, Division of Surgical Oncology, Louisville, Kentucky, United States of America.
RESUMEN / SUMMARY: - BACKGROUND: Irreversible electroporation (IRE) is a novel technology that uses peri-target discrete probes to deliver high-voltage localized electric current to induce cell death without thermal-induced coagulative necrosis. “Learnability” and consistently effective results by novice practitioners is essential for determining acceptance of novel techniques. This multi-center prospectively-collected database study evaluates the learning curve of IRE. METHODS: Analysis of 150 consecutive patients over 7 institutions from 9/2010-7/2012 was performed with patients treated divided into 3 groups A (1(st) 50 patients treated), B (2(nd) 50) and C (3(rd) 50 patients treated) chronologically and analyzed for outcomes. RESULTS: A total of 167 IRE procedures were performed, with a majority being liver (39.5%) and pancreatic (35.5%) lesions. The three groups were similar with respect to co-morbidities and demographics. Group C had larger lesions (3.9vs3cm,p=0.001), more numerous lesions (3.2vs2.2,p=0.07), more vascular invasion (p=0.001), underwent more associated procedures (p=0.001) and had longer operative times (p<0.001). Despite this, they had similar complication and high-grade complication rates (p=0.24). Attributable morbidity rate was 13.3%(total 29.3%) and high-grade complications were seen in 4.19%(total 12.6%). Pancreatic lesions (p=0.001) and laparotomy (p=0.001) were associated with complications. CONCLUSION: The review represents that single largest review of IRE soft tissue ablation demonstrating initial patient selection and
safety. Over time, complex treatments of larger lesions and lesions with greater vascular involvement were performed without a significant increase in adverse effects or impact on local relapse free survival. This evolution demonstrates the safety profile of IRE and speed of graduation to more complex lesions, which was greater than 5 cases by institution. IRE is a safe and effective alternative to conventional ablation with a demonstrable learning curve of at least 5 cases to become proficient.

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TÍTULO / TITLE: - Feasibility study of gemcitabine plus docetaxel in advanced or recurrent uterine leiomyosarcoma and undifferentiated endometrial sarcoma in Japan.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Takano T; Niikura H; Ito K; Nagase S; Utsunomiya H; Otsuki T; Toyoshima M; Tokunaga H; Kaiho-Sakuma M; Shiga N; Nagai T; Tanaka S; Otsuki A; Kurosawa H; Shigeta S; Tsuji K; Yamaguchi T; Yaegashi N
INSTITUCIÓN / INSTITUTION: - Clinical Research, Innovation, and Education Center, Tohoku University Hospital, 1-1 Seiryo-machi, Aoba-ku, Sendai, Miyagi, 980-8574, Japan, ttakano@med.tohoku.ac.jp.
RESUMEN / SUMMARY: - BACKGROUND: Uterine leiomyosarcoma (LMS) and undifferentiated endometrial sarcoma (UES) are rare, aggressive malignancies. Both are treated similarly; however, few chemotherapy agents are effective. Recently, the combination of gemcitabine (900 mg/m2, days 1 and 8) plus docetaxel (100 mg/m2, day 8) with granulocyte colony-stimulating factor (G-CSF, 150 mug/m2, days 9-15) has been shown to have activity in LMS. In Japan, neither prophylactic G-CSF at a dose of 150 mug/m2 nor docetaxel at a dose of 100 mg/m2 are approved for use. For this reason, we evaluated the combination of 900 mg/m2 gemcitabine plus 70 mg/m2 docetaxel regimen without prophylactic G-CSF support in advanced or recurrent LMS and UES in Japanese patients. METHODS: Eligible women with advanced or recurrent LMS and UES were treated with 900 mg/m2 gemcitabine on days 1 and 8, plus 70 mg/m2 docetaxel on day 8, every 3 weeks. The primary endpoint was overall response rate, defined as a complete or partial response. RESULTS: Of the eleven women enrolled, 10 were evaluated for a response. One complete response and 2 partial responses were observed (30 %) with an additional 4 (40 %) having stable disease. Mean progression-free survival was 5.4 months (range 1.3-24.8 months), and overall survival was 14 months (range 5.3-38.4 months). Grade 4 neutropenia was the major toxicity (50 %). The median number of cycles was 5 (range 2-18). Twenty-two cycles (44 %) employed G-CSF. CONCLUSION: The gemcitabine plus docetaxel regimen without prophylactic G-CSF support was tolerable and highly efficacious in Japanese patients with advanced or recurrent LMS and UES.

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TÍTULO / TITLE: - Benign lipoma of the inferior vena cava.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cabri-Wiltzer M; Danse E
[516] **TÍTULO / TITLE:** - Polyostotic fibrous dysplasia.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Schouten BJ; Suliman HM
**INSTITUCIÓN / INSTITUTION:** - Department of Radiology, Sint-Lucas Andreas Hospital, Amsterdam, The Netherlands.

[517] **TÍTULO / TITLE:** - Chondroblastoma of calcaneus.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Liu J; Xu N; Sun Y
**INSTITUCIÓN / INSTITUTION:** - Department of Radiology, Second Hospital of Kunming Medical University, Kunming, PR China.

[518] **TÍTULO / TITLE:** - Lipoma arborescens.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Van Landeghem A; Arys B; Heyse C; Peters N; Huysse W
**INSTITUCIÓN / INSTITUTION:** - Department of Medical Imaging, University Hospital Ghent, Ghent, Belgium.

[519] **TÍTULO / TITLE:** - Osteoid osteoma.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Keupers M; Vandevenne J; Gielen E; Horvath M; Palmers Y; Vandersteen M
**INSTITUCIÓN / INSTITUTION:** - Department of Radiology, Ziekenhuizen Oost-Limburg, Genk, Belgium.

[520] **TÍTULO / TITLE:** - Reversion effects of curcumin on multidrug resistance of MNNG/HOS human osteosarcoma cells in vitro and in vivo through regulation of P-glycoprotein.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Si M; Zhao J; Li X; Tian JG; Li YG; Li JM
**RESUMEN / SUMMARY:** - BACKGROUND: P-glycoprotein (P-gp) encoded by ATP-binding cassette sub-family B member 1 (ABCB1) gene is a kind of ATP-dependent drug transporter, which plays important roles in multidrug resistance (MDR) of human cancers, such as osteosarcoma. Curcumin is a natural phenolic coloring compound originating from the rhizomes of Curcuma longa, which is proved to possess antitumor biological activities including reversion of MDR. However, the effect and molecular mechanisms of curcumin to osteosarcoma MDR remain unclear.

**MÉTODOS:** We established a human osteosarcoma drug-resistant cell line MNNG/HOS/MTX by pulse exposure to methotrexate (MTX) and verified that the new cell lines were cross-resistant to other anticancer agents. Then, according to the cytotoxicity assay, we reversed MDR of MNNG/HOS/MTX by 30 micromol/L curcumin, and detected the mechanisms of curcumin reversing MDR through Real-time PCR, Western blotting assay, and Rhodamine123 (Rh123) transport test. Finally, we evaluated the effect of curcumin reversing MDR in vivo by MNNG/HOS/MTX cells xenograft-nude mice model.

**RESULTADOS:** MNNG/HOS/MTX was proved to be a human osteosarcoma MDR cell line. MTT tumor chemosensitivity test indicates that 30 micromol/L curcumin attenuates the half maximal inhibitory concentration (IC50) and resistance index (RI) to MTX, diamminedichloroplatinum (DDP), adriamycin (ADM), ifosfamide (IFO), and epirubicin (EPI) in MNNG/HOS/MTX cells (P < 0.05). Real-time PCR and Western blotting assays demonstrated that curcumin down-regulated P-gp expression of MNNG/HOS/MTX cells. Rh123 transport test showed that curcumin inhibited the transport function of P-gp in vitro. In vivo studies showed that curcumin displayed the features of sensitizing antitumor drugs and inhibiting the proliferation, invasion, and metastasis of osteosarcoma MDR cells. CONCLUSION: Down-regulation of P-gp and inhibition of the function of P-gp efflux pump may contribute to MDR reversion induced by curcumin in vitro and in vivo.

[521]

**TÍTULO / TITLE:** - Role of mTOR inhibition in the treatment of patients with renal angiomyolipomas.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Coombs EJ

**INSTITUCIÓN / INSTITUTION:** - Cincinnati Children’s Hospital Medical Center, Cincinnati, Ohio.

**RESUMEN / SUMMARY:** - PURPOSE: To describe the role of mammalian target of rapamycin (mTOR) inhibition in the treatment of tuberous sclerosis complex (TSC) patients with renal angiomyolipoma in relation to available clinical data and clinical practice guidance for the nurse practitioner (NP). DATA SOURCES: A review of the scientific literature, key clinical congresses, and key clinical trials. CONCLUSIONS: TSC-associated renal angiomyolipomas have a propensity to grow over time and predispose patients to serious and life-threatening consequences. Surgery or invasive interventional therapies may not be the most optimal treatments because of the
multiple, bilateral growth pattern of TSC-associated renal angiomyolipomas. Targeted therapies, such as mTOR inhibitors, which have demonstrated efficacy in maintaining and reducing renal angiomyolipoma size, are of great benefit to patients.

IMPLICATIONS FOR PRACTICE: Treatment with everolimus, an oral mTOR inhibitor, offers patients a noninvasive pharmacotherapeutic treatment option. The NP, as a key member of the healthcare team overseeing TSC patients, must be knowledgeable about the safety and efficacy of mTOR inhibitors as their use in the patient population increases.

[TÍTULO / TITLE: - Kaposi sarcoma in an patient with atopic dermatitis treated with ciclosporin.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace a la Editora de la Revista http://bmj.com/search.dtl
●● Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-202171
AUTORES / AUTHORS: - Wall D; McMenamin M; O'Mahony D; Irvine AD
INSTITUCIÓN / INSTITUTION: - Department of Dermatology, St James’s Hospital, Dublin, Ireland.

RESUMEN / SUMMARY: - There are four clinical subtypes of Kaposi sarcoma (KS): classic, endemic, epidemic and iatrogenic. The geographical prevalence of the endemic variant matches areas of human herpes virus type 8 (HHV8) seroprevalence. The iatrogenic variant, seen in immunosuppressed patients, can be associated with significant morbidity and mortality. This is the first report of KS described in the context of atopic dermatitis (AD) treated with ciclosporin (CSA). We report a case of KS in an HHV8 seropositive Congolese patient following immunosuppression with CSA for AD. Treatment has been challenging, protracted and associated with significant morbidity. Immunosuppressive therapies are increasingly used for inflammatory dermatological conditions, including AD. This case highlights the importance of HHV8 screening of patients from endemic regions or those with other risk factors. It also highlights the importance of early recognition of a condition associated with significant morbidity and even mortality to facilitate appropriate treatment.

[TÍTULO / TITLE: - Management and outcome of 239 adolescent and adult rhabdomyosarcoma patients.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o de pago) 1002/cam4.92
AUTORES / AUTHORS: - Dumont SN; Araujo DM; Munsell MF; Salganick JA; Dumont AG; Raymond KA; Linassier C; Patel S; Benjamin RS; Trent JC
INSTITUCIÓN / INSTITUTION: - Hematology Oncology Department Sylvester Comprehensive Cancer Center, University of Miami Miami, Florida ; Department of Sarcoma Medical Oncology MD Anderson Cancer Center, University of Texas Houston, Texas.
**RESUMEN / SUMMARY:** Adult rhabdomyosarcoma (RMS) is a rare tumor that has an inferior outcome compared to younger patient population. The present work aims to study the age-related differences in management of adolescents and adults with RMS. Under an institutional review board-approved protocol, we retrospectively analyzed 239 patients, 10 years of age and greater, diagnosed with RMS at MD Anderson Cancer Center from 1957 through 2003. Of the 239 patients, 163 patients were nonmetastatic with a median overall survival (OS) of 3.8 years (95% CI 2.8-7.6). In the multivariate analysis, age >50 was significantly associated with shorter OS and recurrence-free survival (RFS) for primary patients. Metastases were present in 76 patients, the median OS was 1.4 years. Approximately 13% of metastatic patients <50 years old had a long-term survival exceeding 15 years. Multimodality therapy, including surgery, radiotherapy, and chemotherapy was significantly associated with longer OS in primary and metastatic patients. Use of bi- and triple modality treatment decreased in metastatic patients over 50 years of age compared to younger patients. RMS in adolescents and adults has a poor outcome compared with younger individuals. Increased use of multidisciplinary therapy may improve older patient clinical outcome. Adult rhabdomyosarcoma is a rare entity that has inferior outcome compared to younger patient population. This retrospective study emphasizes the age-related differences in management of patients that may partly explain their poor prognosis. 

[524]

**TÍTULO / TITLE:** Gastrointestinal stromal tumor of small intestine and synchronous bilateral papillary renal cell carcinoma.

**RESUMEN / SUMMARY:** Association of gastrointestinal stromal tumors (GISTs) with other primary malignant neoplasms has previously been reported. In addition, coexistence of unilateral renal cell cancer and a GIST of the stomach has been documented in the literature. We report herein a unique case of a GIST of the small intestine and bilateral papillary renal cell carcinomas in a patient presenting with melena and dizziness. Literature shows that GIST arising from the small intestine is the most common location of GIST accompanied by a second primary neoplasm. However, a unique feature in our GIST patient is the presence of synchronous (bilateral) papillary renal cell carcinomas.

[525]

**TÍTULO / TITLE:** Kaposi’s sarcoma with HHV8 infection and ANCA-associated vasculitis in a hemodialysis patient.

**RESUMEN / SUMMARY:** Association of gastrointestinal stromal tumors (GISTs) with other primary malignant neoplasms has previously been reported. In addition, coexistence of unilateral renal cell cancer and a GIST of the stomach has been documented in the literature. We report herein a unique case of a GIST of the small intestine and bilateral papillary renal cell carcinomas in a patient presenting with melena and dizziness. Literature shows that GIST arising from the small intestine is the most common location of GIST accompanied by a second primary neoplasm. However, a unique feature in our GIST patient is the presence of synchronous (bilateral) papillary renal cell carcinomas.
RESUMEN / SUMMARY: - The association between Kaposi’s sarcoma (KS) and human herpes virus eight (HHV-8) infection is rarely reported in hemodialysis (HD) patients. We report here the rare association of KS, HHV-8 and hepatitis C virus (HCV) infection as well as syphilis in a HD patient. We report the case of a 72-year-old woman who presented with microscopic polyangiitis with alveolar hemorrhage and pauci-immune necrosing and crescentic glomerulonephritis as well as renal failure requiring HD. Biological tests showed positive HCV and syphilis tests. The patient was treated by HD and intravenous pulse, followed by oral corticosteroids and six cyclophosphamide monthly pulses with remission of the alveolar hemorrhage, but without renal functional recovery as the patient remained HD dependent. Five months after the first treatment administration, she developed extensive purpuric lesions on her lower limbs, abdomen, face and neck. A skin biopsy showed KS. The HHV-8 test was positive, with positive polymerase chain reaction-HHV8 in the serum and skin. After immunosuppression withdrawal, the KS skin lesions regressed rapidly without relapse after 12 months of follow-up, but alveolar hemorrhage relapsed after 16 months of follow-up. Our case showed that the immunosuppressed state related to multiple factors such as aging, vasculitis, HHV-8, HCV, syphilis, immunosuppressive therapy and HD may all have contributed to the development of KS in our patient.

[526]

TÍTULO / TITLE: - Standardization of rehabilitation after limb salvage surgery for sarcomas improves patients’ outcome.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Shehadeh A; Dahleh ME; Salem A; Sarhan Y; Sultan I; Henshaw RM; Aboulafia AJ

INSTITUCIÓN / INSTITUTION: - Section of Orthopedic Oncology, King Hussein Cancer Center, Amman, Jordan. Electronic address: ashehadeh@khcc.jo.

RESUMEN / SUMMARY: - BACKGROUND AND OBJECTIVE: The purpose of this study is to establish a standardized postoperative rehabilitation protocol following limb salvage surgery (LSS) in patients with primary bone sarcoma in five major anatomical locations: distal femur, proximal tibia, proximal and total femur, humerus and shoulder girdle and pelvic resections. SETTING AND DESIGN: Retrospective study. PATIENTS AND METHODS: All LSSs were performed by an orthopedic oncology surgeon, and rehabilitation of all patients was based on a devised standardized rehabilitation protocol. Patient outcomes were measured using the modified Musculoskeletal Tumor Society-International Symposium on the Limb Salvage (MSTS-ISOLS) scoring system. RESULTS: A total of 59 patients received LSS in the above mentioned locations; endoprostheses were used in 49, bone allograft in five, while no replacements were made in five patients. At a mean follow-up of 24 months, the mean modified MSTS-ISOLS score for all patients was 87% (95% CI; 0.85-0.89). The highest scores were encountered for patients with distal femur replacement: 93% (95% CI; 0.91-0.95). Seven patients had interruption of more than six weeks in their rehabilitation and had a mean score of 71% (95% CI; 0.64-0.82). CONCLUSION: The proposed rehabilitation
protocol is a comprehensive, organized and applicable guideline to be used after performing LSS at the above mentioned anatomical locations. The use of standardized rehabilitation protocol resulted in improved patient functional outcome.
**TÍTULO / TITLE:** - Case report of cutaneous histiocytic sarcoma: diagnostic and therapeutic dilemmas.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Trevisan F; Xavier CA; Pinto CA; Cattete FG; Stock FS; Martins ML

**INSTITUCIÓN / INSTITUTION:** - Faculty of Medicine of Jundiaí, Jundiaí SP, Brazil.

**RESUMEN / SUMMARY:** - Histiocytic sarcoma is a rare hematologic malignant neoplasia originating from histiocytic or dendritic cell clones. The lesions may be in nodal or extranodal sites, most commonly in the gastrointestinal tract. A small number of cases presents as unique cutaneous lesions. The definitive diagnosis is made by positivity for the immunohistochemical markers CD163, CD68, CD4 and lysozyme. The treatment is controversial, often with combined systemic chemotherapy. This is a case of cutaneous histiocytic sarcoma in an 82-year-old patient presenting two nodular lesions in the breast and right arm which were treated with simple excision and multidisciplinary follow-up, avoiding aggressive management and exhaustive investigations. Although most studies report aggressive evolution, the patient had good and stable clinical status during the twelve-month follow-up period.

[530]

**TÍTULO / TITLE:** - A rare location for a common bone tumor, meta-diaphyseal giant cell tumor of bone in an adult patient.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Darioush MB; H Sami S; Fallah E

**INSTITUCIÓN / INSTITUTION:** - Department of Orthopedic Surgery, Mostafa Hospital, Shahed University, Tehran.

**RESUMEN / SUMMARY:** - Primary bone tumors can be either benign or malignant considering their natural history and cellular morphology. Benign bone tumors are much more frequent than malignant ones although some of them like giant cell tumor of bone can behave just like a malignant one that means has the capacity for massive local destruction and remote metastasis. Giant cell tumor of bone in adult people has a very strong and diagnostic predilection for epiphysial location in long bones. Very few cases have been so far reported for a giant cell tumor of bone with non-epiphysial location in a long bone.

[531]

**TÍTULO / TITLE:** - Multidisciplinary collaborative therapy for 30 children with orbital rhabdomyosarcoma.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Ge X; Huang DS; Shi JT; Ma JM
INSTITUCIÓN / INSTITUTION: - Key Laboratory of Beijing Ophthalmology and Visual Science, Beijing Tongren Eye Center, Affiliated Beijing Tongren Hospital of Capital Medical University, Beijing, China E-mail : jmma@sina.com.

RESUMEN / SUMMARY: - OBJECTIVE: To explore clinical experience and propose new ideas for treating children diagnosed with orbital rhabdomyosarcoma (RMS).

METHODS: We retrospectively analyzed the clinical data for 30 patients (16 males and 14 females, with a median age of 6.2 years) with primary orbital RMS who were enrolled in the Department of Eye Oncology and Pediatrics of our hospital from November 2004 to December 2012. International Rhabdomyosarcoma Organization Staging Standards indicated that among the 30 patients, 4 cases were in phase II, 20 were in phase III, and 6 were in phase IV. All patients underwent a multidisciplinary collaborative model of comprehensive treatment (surgery, chemotherapy, external radiotherapy, 125I radioactive particle implantation, and autologous peripheral blood stem-cell transplantation). RESULTS: Follow-up was conducted until March 2013, with a median follow-up time of 47.2 months (5 to 95 months), and 7 deaths occurred. The 2-year estimated survival rate reached 86.1%, the >/=3-year estimated survival rate was 77%, and the 5-year estimated survival rate was 70.6%. CONCLUSIONS: The multidisciplinary collaborative model can be a safe and effective approach to the comprehensive treatment of children with orbital RMS. It has clinical significance in improving the tumor remission rate.

[532]

TÍTULO / TITLE: - Orbital solitary fibrous tumor with multinucleate giant cells: Case report of an unusual finding in an uncommon tumor.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Mulay K; Honavar SG

INSTITUCIÓN / INSTITUTION: - Ocular Pathology Service, L. V. Prasad Eye Institute, Banjara Hills, Hyderabad, Andhra Pradesh, India.

RESUMEN / SUMMARY: - Solitary fibrous tumor (SFT) is a rare soft-tissue neoplasm which may occur at any site although it is more frequent in the pleura, mediastinum and lung. Orbital involvement by SFT is uncommon. Giant cells are extremely rare to be seen in a SFT and have been described to be immunoreactive for CD34. We present a case of orbital SFT with multinucleate giant cells expressing CD68 and lacking immunoreactivity for CD34. The differential diagnosis is discussed.

[533]

TÍTULO / TITLE: - Oral pemphigus as first sign of an inflammatory myofibroblastic tumour in an 18-year-old male patient.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


Therapeutic Doses of Nonsteroidal Anti-Inflammatory Drugs Inhibit Osteosarcoma MG-63 Osteoblast-Like Cells Maturation, Viability, and Biomineralization Potential.

Upregulation of SOX9 in osteosarcoma and its association with tumor progression and patients’ prognosis.
SOX9 in human osteosarcoma. METHODS: SOX9 mRNA and protein expression levels were detected by RT-PCR and Western blot assays, respectively, using 30 pairs of osteosarcoma and noncancerous bone tissues. Then, immunohistochemistry was performed to analyze the association of SOX9 expression in 166 osteosarcoma tissues with clinicopathological factors or survival of patients. RESULTS: SOX9 expression at mRNA and protein levels were both significantly higher in osteosarcoma tissues than those in corresponding noncancerous bone tissues (both P < 0.001). Immunohistochemical staining indicated that SOX9 localized to the nucleus and high SOX9 expression was observed in 120 of 166 (72.3%) osteosarcoma specimens. In addition, high SOX9 expression was more frequently occurred in osteosarcoma tissues with advanced clinical stage (P = 0.02), positive distant metastasis (P = 0.008) and poor response to chemotherapy (P = 0.02). Osteosarcoma patients with high SOX9 expression had shorter overall survival and disease-free survival (both P < 0.001). Furthermore, the multivariate analysis confirmed that upregulation of SOX9 was an independent and significant prognostic factor to predict poor overall survival and disease-free survival (both P = 0.006). CONCLUSIONS: Our data show for the first time that SOX9 is upregulated in aggressive osteosarcoma tissues indicating that SOX9 may participate in the osteosarcoma progression. More importantly, SOX9 status is a useful prognostic factor for predicting the prognosis of osteosarcoma, suggesting that SOX9 may contribute to the optimization of clinical treatments for osteosarcoma patients. Virtual slides: The virtual slides for this article can be found here: http://www.diagnosticpathology.diagnomx.eu/vs/1318085636110837.

[536]

**TÍTULO / TITLE:** PIK3CA and AKT Gene Polymorphisms in Susceptibility to Osteosarcoma in a Chinese Population.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** He ML; Wu Y; Zhao JM; Wang Z; Chen YB

**INSTITUCIÓN / INSTITUTION:** Division of Spinal Surgery, The First Affiliated Hospital of Guangxi Medical University, Nanning, Guangxi, China E-mail: zhaojinmin@hotmail.com

**RESUMEN / SUMMARY:** Purpose: To explore the association between PIK3CA and AKT single nucleotide polymorphisms (SNP) and osteosarcoma susceptibility. Methods: TaqMan polymerase chain reaction (PCR) was used to detect the genotypes of SNPs (rs7646409, rs6973569 and rs9866361) in peripheral blood samples from 59 patients with osteosarcoma and from 63 healthy controls. Unconditional logistic regression was used to analyze the correlation between SNPs and osteosarcoma risk. Results: No statistically significant difference was found between osteosarcoma patients and healthy controls in the genotype of AKT rs6973569 (P = 0.7). However, after stratified analysis, the genotype AA of AKT rs6973569 carried a higher risk of osteosarcoma metastasis (OR: 2.94, 95% CL: 1.00-8.59); the difference of rs7646409 genotype distributions between the case and control groups was statistically significant (P = 0.032). Taking genotype TT as a reference, the risk of osteosarcoma increased three fold in patients with genotype CC (OR: 3.47, 95% CL: 1.26-9.56). A statistically significant difference was found between the alleles C and T (P = 0.005). Further analysis showed that the risk factor was more pronounced in male patients with Enneking’s stage IIB and osteoblastic osteosarcoma. PIK3CA rs9866361 did not fit...
Hardy-Weinberg equilibrium (P < 0.05). Conclusions: Genotype CC in locus PIK3CA rs7646409 may increase the risk of osteosarcoma in the Chinese population.

[537]  
TITULO / TITLE: - Expression and clinicopathological significance of CD9 in gastrointestinal stromal tumor.  
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary  
●● Enlace al texto completo (gratuito o de pago) 3346/jkms.2013.28.10.1443  
AUTORES / AUTHORS: - Yang H; Shen C; Zhang B; Chen H; Chen Z; Chen J  
INSTITUCIÓN / INSTITUTION: - Department of Gastrointestinal Surgery, West China Hospital, Sichuan University, Chengdu, Sichuan Province, China. ; Department of Gastrointestinal Surgery of the Affiliated Hospital of Guiyang Medical University, Guiyang, Guizhou Province, China.  
RESUMEN / SUMMARY: - This study investigated the expression and clinicopathological significance of CD9 in gastrointestinal stromal tumor (GIST). Immunohistochemistry staining for CD9 was performed on tumor tissues from 74 GIST patients. The correlation with clinicopathological features, risk classification and prognosis was analyzed. CD9-positive staining comprised 59.5% (44/74) of the GIST patients. The CD9-positive expression rate of the sample was significantly associated with diameter (P = 0.028), mitotic counts (P = 0.035), risk classification (P = 0.018) and three-year recurrence-free survival (RFS) (P < 0.001). Cox proportional hazards regression (HR = 0.352; P = 0.015) showed that CD9 is an independent factor for post-operative RFS. The subgroup analysis showed that CD9 expression in gastric stromal tumor (GST) is significantly associated with diameter (P = 0.031), risk classification (P = 0.023) and three-year RFS (P = 0.001). The Cox proportional hazards regression (HR = 0.104; P = 0.006) also showed that CD9 is an independent factor for RFS of GST. However, CD9 expression does not have a statistically significant correlation with clinicopathological features, risk classification, and prognosis in non-GST. In conclusion, CD9 expression in GIST appears to be associated with the recurrence and/or metastasis of GIST patients, especially in GST, which may indicate the important role of CD9 in the malignant biological behavior and prognosis of GST.

[538]  
TITULO / TITLE: - High-dose chemotherapy and autologous peripheral blood stem cell transplantation in the treatment of children and adolescents with Ewing sarcoma family of tumors.  
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary  
●● Enlace al texto completo (gratuito o de pago) 3345/kjp.2013.56.9.401  
AUTORES / AUTHORS: - Seo J; Kim DH; Lim JS; Koh JS; Yoo JY; Kong CB; Song WS; Cho WH; Jeon DG; Lee SY; Lee JA  
INSTITUCIÓN / INSTITUTION: - Department of Pediatrics, Korea Cancer Center Hospital, Seoul, Korea.
RESUMEN / SUMMARY: PurPOSE: We performed a pilot study to determine the benefit of high-dose chemotherapy and autologous peripheral blood stem cell transplantation (HDCT/autoPBSCT) for patients with Ewing sarcoma family of tumors. METHODS: We retrospectively analyzed the data of patients who received HDCT/autoPBSCT at Korea Cancer Center Hospital. Patients with relapsed, metastatic, or centrally located tumors were eligible for the study. RESULTS: A total of 9 patients (3 male, 6 female), with a median age at HDCT/autoPBSCT of 13.4 years (range, 7.1 to 28.2 years), were included in this study. Patients underwent conventional chemotherapy and local control either by surgery or radiation therapy, and had achieved complete response (CR, n=7), partial response (n=1), or stable disease (n=1) prior to HDCT/autoPBSCT. There was no transplant-related mortality. However, the median duration of overall survival and event-free survival after HDCT/autoPBSCT were 13.3 months (range, 5.3 to 44.5 months) and 6.2 months (range, 2.1 to 44.5 months), respectively. At present, 4 patients are alive and 5 patients who experienced adverse events (2 metastasis, 2 local recur, and 1 progressive disease) survived for a median time of 2.8 months (range, 0.1 to 10.7 months). The 2-year survival after HDCT/autoPBSCT was 44.4% +/- 16.6% and disease status at the time of HDCT/autoPBSCT tended to influence survival (57.1% +/- 18.7% of cases with CR vs. 0% of cases with non-CR, P=0.07). CONCLUSION: Disease status at HDCT/autoPBSCT tended to influence survival. Further studies are necessary to define the role of HDCT/autoPBSCT and to identify subgroup of patients who might benefit from this investigational treatment.

TÍTULO / TITLE: Massive atrial myxoma: surgical treatment for an incidentaloma causing dyspnoea.

RESUMEN / SUMMARY: A patient presented having a one-week history of recurrent falls and confusion and weight loss over an unspecified period of time. A chest radiograph revealed bilateral pleural effusions and the patient was treated for community acquired pneumonia. His weight loss and suspicion of malignant disease prompted computer tomography to be conducted. This revealed widespread mediastinal and oesophageal adenopathy. An echocardiogram showed a large hyperechoic mobile mass. Coronary angiography showed complete occlusion of the left anterior descending and right coronary arteries. He underwent two-vessel coronary artery bypass grafting and an atriotomy to excise the myxoma. He experienced no operative complications and no neurological deterioration and was discharged home three weeks later in sinus rhythm. Myxomas can duplicate a broad array of cardiorespiratory symptoms and signs and can often escape detection by being picked up as an incidentaloma.
TÍTULO / TITLE: - Multimodality imaging of right-sided (tricuspid valve) papillary fibroelastoma: recognition of a surgically remediable disease.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


●● Enlace al texto completo (gratuito o de pago) 1159/000355419

AUTORES / AUTHORS: - Srivatsa SV; Adhikari P; Chaudhry P; Srivatsa SS

INSTITUCIÓN / INSTITUTION: - Department of Cardiology and Cardiothoracic Surgery, Community Regional Medical Center, Fresno, Calif., USA.

TÍTULO / TITLE: - Safety and efficacy of stereotactic body radiation therapy in the treatment of pulmonary metastases from high grade sarcoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


●● Enlace al texto completo (gratuito o de pago) 1155/2013/360214

AUTORES / AUTHORS: - Mehta N; Selch M; Wang PC; Federman N; Lee JM; Eilber FC; Chmielowski B; Agazaryan N; Steinberg M; Lee P

INSTITUCIÓN / INSTITUTION: - Department of Radiation Oncology, David Geffen School of Medicine at UCLA, 200 UCLA Medical Plaza, B265, Los Angeles, CA 90095, USA.

RESUMEN / SUMMARY: - Introduction. Patients with high-grade sarcoma (HGS) frequently develop metastatic disease thus limiting their long-term survival. Lung metastases (LM) have historically been treated with surgical resection (metastasectomy). A potential alternative for controlling LM could be stereotactic body radiation therapy (SBRT). We evaluated the outcomes from our institutional experience utilizing SBRT. Methods. Sixteen consecutive patients with LM from HGS were treated with SBRT between 2009 and 2011. Routine radiographic and clinical follow-up was performed. Local failure was defined as CT progression on 2 consecutive scans or growth after initial shrinkage. Radiation pneumonitis and radiation esophagitis were scored using Common Toxicity Criteria (CTC) version 3.0. Results. All 16 patients received chemotherapy, and a subset (38%) also underwent prior pulmonary metastasectomy. Median patient age was 56 (12-85), and median follow-up time was 20 months (range 3-43). A total of 25 lesions were treated and evaluable for this analysis. Most common histologies were leiomyosarcoma (28%), synovial sarcoma (20%), and osteosarcoma (16%). Median SBRT prescription dose was 54 Gy (36-54) in 3-4 fractions. At 43 months, local control was 94%. No patient experienced G2-4 radiation pneumonitis, and no patient experienced radiation esophagitis. Conclusions. Our retrospective experience suggests that SBRT for LM from HGS provides excellent local control and minimal toxicity.
TÍTULO / TITLE: Subserosal leiomyoma of uterus mimicking an ovarian tumor in adolescent patient.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Kaya dibi Y; Ozmen E; Emir H; Emre S; Dervisoglu S; Adaletli I
INSTITUCIÓN / INSTITUTION: Division of Pediatrics, Department of Radiology, Cerrahpasa Medical Faculty, Istanbul University, Kocamustafapasa, Istanbul, 34098, Turkey, yasemin.kurdoglu@istanbul.edu.tr.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Geel JA; Loveland JA; Pitcher GJ; Beale P; Kotzen J; Poole JE
INSTITUCIÓN / INSTITUTION: Division of Paediatric Haematology and Oncology, Charlotte Maxeke Academic Hospital, University of the Witwatersrand, Johannesburg, South Africa. loveland@wol.co.za.

RESUMEN / SUMMARY: BACKGROUND: Undifferentiated embryonal sarcoma of the liver (UESL) is a rare neoplasm, and the third-most common paediatric hepatic malignancy. However, no treatment guidelines exist. No randomised, controlled trials support specific combinations of therapy. OBJECTIVE: To compare presentation and management of UESL with other series, review the literature, and formulate treatment guidelines. METHODS: A retrospective chart review of all hepatic malignancies was conducted from 1996 to 2007 and 5 children with UESL were identified. Management and outcomes were documented. The literature regarding treatment modalities up to September 2012 was reviewed. RESULTS: Over a period of 11 years, 5 patients presented. All underwent surgery and 4 received chemotherapy. One received radiotherapy at relapse. Three are disease-free with follow-up of 58 - 184 months. One died after relapse, as did the patient whose family declined chemotherapy. CONCLUSION: The improved outcomes are consistent with the international experience and are probably related to combined treatment modalities and advances in supportive care. Pre-operative percutaneous biopsy provides no benefit if the lesion is resectable because it may not prove to be diagnostic, and may cause recurrence in the biopsy tract. If resectable, the recommended treatment is primary excision and adjuvant chemotherapy, with radiotherapy in selected cases. If unresectable, open
biopsy is necessary to document histology, and neo-adjuvant chemotherapy is given prior to resection. If deemed unresectable, liver transplantation is considered.

[544]
**TÍTULO / TITLE:** - Sodium thiosulfate protects human aortic smooth muscle cells from osteoblastic transdifferentiation via high-level phosphate.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Zhong H; Liu F; Dai X; Zhou L; Fu P

**INSTITUCIÓN / INSTITUTION:** - Division of Nephrology, West China Hospital, Sichuan University, Chengdu, China.

**RESUMEN / SUMMARY:** - Vascular calcification is recognized as a common complication in some patients, such as chronic renal failure. The purpose of this study was to investigate the role of sodium thiosulfate (STS) for the transdifferentiation of human aortic vascular smooth muscle cells into osteoblast-like cells induced by high-level phosphate. All human aortic vascular smooth muscle cells were divided into STS group 1 (treatment with STS) and STS group 2 (culture in a medium containing a high level of phosphate). STS group 1 included a normal group, a high-level phosphate group, and other subgroups based on treatment with different concentrations of STS. Cells of STS group 2 were cultured in a medium containing a high level of phosphate for 72 hours, and then divided into a high-phosphate control group and other subgroups based on treatment with different concentrations of STS. The mRNA and protein expressions of bone morphogenetic protein-2 (BMP-2), core binding factor alpha-1 (Cbfalpha-1), and matrix Gla protein (MGP) were detected. Meanwhile, calcium concentration and alkaline phosphatase (ALP) activation were measured. In STS group 1, the mRNA levels of BMP-2 and Cbfalpha-1 were elevated significantly in the high-level phosphate group compared with the normal group (p < 0.05). However, both gene expressions were attenuated in the STS-treated groups (vs. normal group, p < 0.05). MGP mRNA levels were reduced in the high-level phosphate group (vs. normal group, p < 0.05). In the STS-treated groups, mRNA expression of MGP was elevated compared to the high-level phosphate group (p < 0.05). In STS group 2, expression of MGP was enhanced significantly (vs. high-phosphate control group, p < 0.05) with both BMP-2 and Cbfalpha-1 reducing in the STS-treated groups (vs. high-phosphate-control group, p < 0.05). STS attenuates calcium concentration and ALP activation. It can reverse osteoblast differentiation of vascular smooth muscle cells and modulate the expressions of calcification-related factors.

[545]
**TÍTULO / TITLE:** - Myxoid liposarcoma in a 91-year-old patient.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Sheffield BS; Nielsen TO
BACKGROUND: Myxoid liposarcoma is a mesenchymal malignancy most commonly presenting in young adults. This tumor is known for its characteristic chromosomal rearrangement at the DDIT3 locus. RESULTS: We report a case of myxoid liposarcoma in a 91-year-old, the oldest known patient with this disease-entity. FISH analysis of the DDIT3 and FUS loci demonstrate the pathognomonic chromosomal alteration in the setting of predominantly round cell histology on biopsy, confirmed by RT-PCR. CONCLUSION: Myxoid liposarcoma affects mostly young adults but can be seen in the elderly population. Molecular and cytogenetic assays are helpful auxiliaries to histology in the setting of unusual histology and clinical presentation.

RESUMEN / SUMMARY: - Spinal coning after lumbar puncture in a patient with undiagnosed giant cervical neurofibroma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


Enlace al texto completo (gratuito o de pago) 4103/0972-2327.116935

AUTORES / AUTHORS: - Krishnan P; Roychowdhury S

INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, National Neurosciences Centre, Peerless Hospital Complex, II Floor, 360 Panchasayar, Kolkata, India.

RESUMEN / SUMMARY: - Lumbar puncture in the presence of an intracranial tumor with raised intracranial pressure is known to have catastrophic consequences due to herniation of intracranial contents through the tentorial hiatus or foramen magnum. There are relatively few case reports about the same sequence of events when lumbar puncture is performed below the level of a complete spinal block. The mechanism of such deterioration is also subject to conjecture as the spinal cord (unlike the uncus or cerebellar tonsils) is tethered by the dentate ligament and roots on either side, and is hence less mobile. We present one such case of spinal coning and review the available literature.

RESUMEN / SUMMARY: - Erlotinib inhibits growth of a patient-derived chordoma xenograft.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


Enlace al texto completo (gratuito o de pago) 1371/journal.pone.0078895

AUTORES / AUTHORS: - Siu IM; Ruzevick J; Zhao Q; Connis N; Jiao Y; Bettegowda C; Xia X; Burger PC; Hann CL; Gallia GL

INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Johns Hopkins University School of Medicine, Baltimore, Maryland, United States of America.

RESUMEN / SUMMARY: - Chordomas are rare primary bone tumors that occur along the neuraxis. Primary treatment is surgery, often followed by radiotherapy. Treatment options for patients with recurrence are limited and, notably, there are no FDA approved therapeutic agents. Development of therapeutic options has been limited by the paucity of preclinical model systems. We have established and previously reported the initial characterization of the first patient-derived chordoma xenograft model. In this study, we further characterize this model and demonstrate that it continues to
resemble the original patient tumor histologically and immunohistochemically, maintains nuclear expression of brachyury, and is highly concordant with the original patient tumor by whole genome genotyping. Pathway analysis of this xenograft demonstrates activation of epidermal growth factor receptor (EGFR). In vitro studies demonstrate that two small molecule inhibitors of EGFR, erlotinib and gefitinib, inhibit proliferation of the chordoma cell line U-CH 1. We further demonstrate that erlotinib significantly inhibits chordoma growth in vivo. Evaluation of tumors post-treatment reveals that erlotinib reduces phosphorylation of EGFR. This is the first demonstration of antitumor activity in a patient-derived chordoma xenograft model and these findings support further evaluation of EGFR inhibitors in this disease.

[548]
TÍTULO / TITLE: - Retrospective analysis of patients with rare-site and metastatic giant cell tumor.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Liu J; Yang H; Sun R; Yang Z; Zhu Z
INSTITUCIÓN / INSTITUTION: - State Key Laboratory of Oncology in South China, Guangzhou 510060, China; ; Department of Medical Oncology, Sun Yat-sen University Cancer Center, Guangzhou 510060, China;
RESUMEN / SUMMARY: - A giant cell tumor occurs mainly in the proximal tibia, humerus, distal radius bone and the pelvic bone. It is rarely observed in such sites as the ribs and the temporal bone. The condition is primarily treated with surgical excision and functional reconstruction. The effect of chemotherapy on lung metastases and locally advanced giant cell tumors has remained unknown. We collected and analyzed the data of six patients with rare giant cell tumors located in the head and neck patients. After an average follow-up of 42.6 months after surgery (14 to 90 months), no local recurrence or metastasis was observed. We also collected and analyzed the data of five patients with metastatic giant cell tumors who were undergoing surgery for the primary tumor before; of three patients who had experienced multiple chemotherapy cycles, one had spontaneous regression, and one survived for long timer despite progression. The other two patients had their major metastatic lesions resected by surgery, and presented long-term survival during the follow up. In addition, this study reports one patient with locally advanced giant cell tumor of the rib, who has undergone successful surgical resection following two cycles of chemotherapy with ifosfamide and liposomal doxorubicin. Complete resection of the lesion at the head and neck is the key to relapse-free survival. The prognosis of lung metastases in patients with giant cell tumors is relatively satisfying. Neoadjuvant chemotherapy is also conducive to the surgery for locally advanced lesions and improvement of the quality of life.

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[549]
TÍTULO / TITLE: - Myeloid sarcoma developing in preexisting hydroxyurea-induced leg ulcer in a polycythemia vera patient.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Enlace al texto completo (gratuito o de pago) 1155/2013/497593
AUTORES / AUTHORS: - Nafil H; Tazi I; Mahmal L
INSTITUCIÓN / INSTITUTION: - Service d'Hematologie, CHU Mohamed VI, Universite Cadi Ayyad Marrakech, Marrakech 40000, Morocco.
RESUMEN / SUMMARY: - Myeloid sarcoma (MS) is an extramedullary tumour consisting of myeloblasts or immature myeloid cells located in an extramedullary site. It may occur at presentation of AML, at relapse, or prior to the onset of frank leukemia. We report a rare case of MS developing in preexisting Hydroxyurea-induced leg Ulcer in a 70-year-old woman.

[550]
TÍTULO / TITLE: - Criminal or bystander: imatinib and second primary malignancy in GIST patients.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Kanda T
INSTITUCIÓN / INSTITUTION: - Sanjo General Hospital, Sanjo City, Niigata, Japan.

[551]
TÍTULO / TITLE: - High fluorodeoxyglucose ((18)F)PET-uptake lymph nodes in a patient with chordoma: Tumor metastasis or sarcoidosis?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Gharavi MH; Wu HH; Toms SA
INSTITUCIÓN / INSTITUTION: - Department of General Surgery, Geisinger Health System, Danville, PA, U.S.A.
RESUMEN / SUMMARY: - Patient: Male, 48 Final Diagnosis: Chordoma Symptoms: - Medication: - Clinical Procedure: - Specialty: Neurology. OBJECTIVE: Challenging differential diagnosis. BACKGROUND: Fluorodeoxyglucose positron emission tomography (FDG-PET) has been used in imaging and staging of malignancies including sacral chordomas. CASE REPORT: The author’s report describes the coincident pathological diagnosis of sarcoidosis in a 48-year-old male patient with a recurrent sacral chordoma. Chordoma is a low grade malignancy with frequent systemic metastases in advanced disease. Both metastases and sarcoidosis may be high FDG uptake. Unexpected PET findings need to be biopsied in order to make appropriate clinical decision in the management of chordoma. CONCLUSIONS: Lymph nodes involvement in sarcoidosis and neoplastic disease can have similar FDG-PET manifestations.

[552]
TÍTULO / TITLE: - Chloroquine and valproic acid combined treatment in vitro has enhanced cytotoxicity in an osteosarcoma cell line.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Chloroquine (CQ) and valproic acid (VPA) have been extensively studied for biological effects. Here, we focused on efficacy of combined CQ and VPA on osteosarcoma cell lines. Viability of osteosarcoma cell lines (U2OS and HOS) was analyzed by MTT assay. Apoptotic assays and colony formation assays were also applied. ROS generation and Western Blotting were performed to determine the mechanism of CQ and VPA combination in the process of apoptosis. The viability of different osteosarcoma cell lines significantly decreased after CQ and VPA combination treatment compared with either drug used alone, and apoptosis was increased significantly. ROS generation was triggered leading to expression of apoptosis related genes being increased and of anti-apoptotic related genes being decreased. From our data shown here, CQ and VPA combination treatment in vitro enhanced cytotoxicity to osteosarcoma cells.
Expression of hormonal receptors in osteosarcomas of the jaw bones: Clinico-pathological analysis of 21 cases.

Background: Sexual hormones have an important role in many hormone-dependant tumors like breast and prostate carcinomas, and also a relationship has been found with bone metabolism and bone tumors. Some studies have demonstrated that the expression of hormonal receptors (HR) in osteosarcomas (OS) of long bones is associated with gender, histological grade, histological type, and a possibly may be connection with pathogenesis and evolution. However, to our knowledge there are no studies of HR in osteosarcomas of craniofacial bones (OS-CF).

Objective: To assess the expression of hormonal receptors in OS-CF. Material and methods: Twenty one cases of OS-CF were included in this study. Clinical outcome was obtained from clinical charts. Histological sections were reviewed, and immunohistochemistry studies for estrogen, progesterone and androgen receptors were performed. Results: A striking female predominance was found (2:1), with a median age of 35 years. The predominant type of OS was osteoblastic (52.4%), and histological grade was high in 86%. Follow-up was obtained in 13 cases and ranged from 6 to 118 months (median 29 months). There were 8 patients (61.5%) dead or alive with progressive disease in the last follow up. Negative expression of HR was found in 19/21 cases; one showed weak nuclear expression for estrogen receptor, and another for androgen receptor. Progesterone receptor was negative in all cases. Conclusions: OS-CF mostly affected females, most of them were of the osteoblastic type and of high grade. Hormonal expression was practically negative in osteosarcoma of craniofacial bones.
Enlace al texto completo (gratuito o de pago) 10.4103/0377-4929.120358

AUTORES / AUTHORS: Long XH; Zhang ZH; Liu ZL; Huang SH; Luo QF

INSTITUCIÓN / INSTITUTION: Department of Orthopedics, First Affiliated Hospital of Nanchang University, Nanchang, 330006, P. R. China.

RESUMEN / SUMMARY: Background and Aim: The strategies of targeting valosin-containing protein (VCP) may have therapeutic potential for treating cancer metastasis. In this study, we aim to investigate the correlation of VCP protein expression in osteosarcoma (OS) tissues with pulmonary metastasis and its possible molecular mechanism. Materials and Methods: Expression of VCP in 60 OS specimens was detected by immunohistochemistry (IHC) and the relationship with metastasis was analyzed. An artificial micro ribonucleic acid, targeting VCP, was performed to silence the expression of VCP in U2-OS cells. Cell mobility was detected by wound healing and Transwell assays. Western blot and real-time polymerase chain reaction were performed to investigate the expression of VCP in U2-OS cells. Furthermore, the protein of pAKT (phosphorylated serine/threonine protein kinase) and nuclear factor of kappa B protein65 were measured by western blot to evaluate the effect of silencing VCP on AKT/nuclear factor of kappa B (NF-kB) signaling pathway. Results: The results showed that the expression level of VCP protein in cases with pulmonary metastases was significantly higher than that in those without metastasis (P = 0.004). The invasion and migration of U2-OS cells were suppressed by silencing VCP. Furthermore, silencing VCP could down-regulate the phosphorylation of AKT and nuclear transfer of NF-kB. Conclusions: Our findings suggested that inhibition of VCP could suppress OS cells invasion and migration through down-regulating AKT/NF-kB signaling pathway.

Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: Di Veroli A; Micarelli A; Cefalo M; Ceresoli E; Nasso D; Cicconi L; Mauramati S; Ottaviani F; Venditti A; Amadori S

INSTITUCIÓN / INSTITUTION: Department of Hematology, “Tor Vergata” University, Viale Oxford 81, 00133 Rome, Italy.

RESUMEN / SUMMARY: Granulocytic sarcoma (GS) is a rare extramedullary solid tumor defined as an accumulation of myeloblasts or immature myeloid cells. It can cooccur with or precede the acute myeloid leukemia (AML) as well as following treated AML. The incidence of GS in AML patients is 3-8% but it significantly rises in M2 FAB subtype AML. This variety of AML harbors t(8;21) in up to 20-25% of cases (especially in children and black ones of African origin) and, at a molecular level, it is characterized by the generation of a fusion gene known as RUNX1-RUNX1T1. Approximately 10% of M2 AML patients will develop GS, as a consequence, the t(8;21) and the relative transcript represent the most common cytogenetic and molecular abnormalities in GS.
FLT3-ITD mutation was rarely described in AML patients presenting with GS. FLT3 ITD is generally strongly associated with poor prognosis in AML, and is rarely reported in patients with t(8;21). GS presentation is extremely variable depending on organs involved; in general, cranial bones and sinus are very rarely affected sites. We report a rare case of GS occurring as a recurrence of a previously treated t(8;21), FLT3-ITD positive AML, involving mastoid bones and paravertebral tissues.

[558]
**TÍTULO / TITLE:** - An unusual case of pulmonary granulocytic sarcoma treated with combined chemotherapy and radiation.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)

**AUTORES / AUTHORS:** - Sathyanarayanan V; Sirsath NT; Das U; Malathi M; Reddy SL; Srivatsa KS; Thumallapalli A; Appaji L; Kumari BA
**INSTITUCIÓN / INSTITUTION:** - Department of Medical Oncology, Kidwai Memorial Institute of Oncology, Bangalore, Karnataka 560029, India.

**RESUMEN / SUMMARY:** - We report an unusual case of a 6-year-old male child who presented with fever and a cough of one month’s duration. A bone marrow aspiration and cytogenetics were suggestive of acute myeloid leukaemia with t(8;21)(q22;q22). A chest x-ray and computed tomography of the thorax showed a soft tissue lesion in the right lung. The fine needle aspiration cytology (FNAC) of this lesion was suggestive of pulmonary granulocytic sarcoma. The patient was successfully treated with induction chemotherapy (cytosine arabinoside + daunomycin), followed by consolidation with high-dose cytosine arabinoside. In view of the persistent lesion in the right lung, the patient was given external beam radiotherapy (EBRT), which resulted in near total resolution of the lung granulocytic sarcoma. We report this case in view of its rarity and clinical importance, and to highlight the treatment options in this scenario.

[559]
**TÍTULO / TITLE:** - Sclerosing Rhabdomyosarcoma of a Chest Wall in an Adult: A Case Report and Review of the Literature.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)
**AUTORES / AUTHORS:** - Mikubo M; Ikeda S; Hoshino T; Yokota T; Fujii A; Mori M
**INSTITUCIÓN / INSTITUTION:** - Department of Thoracic Surgery, Pathology, Mitsui Memorial Hospital, Chiyoda-ku, Tokyo, Japan.

**RESUMEN / SUMMARY:** - Sclerosing rhabdomyosarcoma (SRMS) is a newly recognized and rare variant of rhabdomyosarcoma. This soft tissue tumor has not yet been reported as a thoracic lesion. We report a case of a 26-year-old woman who presented with a large chest wall tumor. The tumor originated from the right anterior chest wall and protruded into the intra- and extrapleural cavity. A transcutaneous needle biopsy revealed spindle cells in an abundant hyalinized and fibrous stroma. Although the tumor was considered as a malignant soft-tissue neoplasm, a definitive diagnosis could not be established. A wide excision of the chest wall including the second, third and fourth rib and a part of sternum was performed. Histologically, cytoplasmic cross-striations were found in a portion of the tumor cells. The tumor cells
were positive for muscle markers, and the tumor was diagnosed as rhabdomyosarcoma consistent with a sclerosing type of rhabdomyosarcoma. Eighteen months after the complete resection, the patient has pleural disseminations but is alive and undergoing chemotherapy. This case highlights the histologic features of a rare form of rhabdomyosarcoma, and emphasizes the importance of awareness of its existence and the utility of skeletal muscle markers in distinguishing sclerosing rhabdomyosarcoma from its mimics.

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**TÍTULO / TITLE:** Knockdown of autophagy-related protein 5, ATG5, decreases oxidative stress and has an opposing effect on camptothecin-induced cytotoxicity in osteosarcoma cells.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Hollomon MG; Gordon N; Santiago-O Farrill JM; Kleinerman ES

**RESUMEN / SUMMARY:** BACKGROUND: Autophagy induction can increase or decrease anticancer drug efficacy. Anticancer drug-induced autophagy induction is poorly characterized in osteosarcoma (OS). In this study, we investigated the impact of autophagy inhibition on camptothecin (CPT)-induced cytotoxicity in OS. METHODS: Autophagy-inhibited DLM8 and K7M3 metastatic murine OS cell lines were generated by infection with lentiviral shRNA directed against the essential autophagy protein ATG5. Knockdown of ATG5 protein expression and inhibition of autophagy was confirmed by immunoblot of ATG5 and LC3I/II proteins, respectively. Metabolic activity was determined by MTT assay and cell viability was determined by trypan blue exclusion. Acridine orange staining and immunoblotting for LC3I/II protein expression were used to determine autophagy induction. Oxidative stress was assessed by staining cells with HE and DCFH-DA followed by flow cytometry analysis. Mitochondrial membrane potential was determined by staining cells with TMRE followed by flow cytometry analysis. Immunoblotting was used to detect caspase activation, Parp cleavage and p53 phosphorylation. RESULTS: Autophagy inhibition caused a greater deficit in metabolic activity and cell growth in K7M3 cells compared to DLM8 cells. K7M3 cells exhibited higher basal autophagy levels than DLM8 cells and non-transformed murine MCT3 osteoblasts. Autophagy inhibition did not affect CPT-induced DNA damage. Autophagy inhibition decreased CPT-induced cell death in DLM8 cells while increasing CPT-induced cell death in K7M3 cells. Autophagy inhibition reduced CPT-induced mitochondrial damage and CPT-induced caspase activation in DLM8 cells. Buthionine sulfoximine (BSO)-induced cell death was greater in autophagy-competent DLM8 cells and was reversed by antioxidant pretreatment. Camptothecin-induced and BSO-induced autophagy induction was also reversed by antioxidant pretreatment. Significantly, autophagy inhibition not only reduced CPT-induced oxidative stress but also reduced basal oxidative stress. CONCLUSIONS: The results of this study indicate that autophagy inhibition can have an opposing effect on CPT-induced cytotoxicity within OS. The cytoprotective mechanism of autophagy inhibition observed in DLM8 cells involves reduced CPT-induced oxidative stress and...
not reduced DNA damage. Our results also reveal the novel finding that knockdown of ATG5 protein reduces both basal oxidative stress and drug-induced oxidative stress.

[561]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Gupta A; Ramakrishna B
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Christian Medical College, Vellore, Tamil Nadu, India.
RESUMEN / SUMMARY: - Angiomyolipomas are rare benign tumors derived from perivascular epithelioid cells that occur frequently in kidney and rarely in the liver. We present a case of hepatic angiomyolipoma, with various clinical and radiological differential diagnoses. A 34-year-old male was found to have a palpable mass on the left side of the abdomen. Imaging showed heterogeneously enhancing mass lesion 15 cm Chi 7 cm, in the left lobe and segment 4 of the liver. Various benign and malignant diagnoses were entertained. The needle biopsy did not reveal any evidence of malignancy. Patient underwent resection of the lesion in view of the large size, which was diagnosed as angiomyolipoma. Angiomyolipoma is a benign tumor and accurate diagnosis is important to prevent overtreatment. Histology and immunohistochemistry may help in clinching the diagnosis, in proper clinical setting.

[562]
TITULO / TITLE: - Protein kinase C epsilon and genetic networks in osteosarcoma metastasis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Goudarzi A; Gokgoz N; Gill M; Pinnaduwage D; Merico D; Wunder JS; Andrulis IL
INSTITUCIÓN / INSTITUTION: - Department of Molecular Genetics, University of Toronto, 1 King’s College Circle, Toronto, ON M5S 1A8, Canada.
at.a.goudarzi@utoronto.ca
RESUMEN / SUMMARY: - Osteosarcoma (OS) is the most common primary malignant tumor of the bone, and pulmonary metastasis is the most frequent cause of OS mortality. The aim of this study was to discover and characterize genetic networks differentially expressed in metastatic OS. Expression profiling of OS tumors, and subsequent supervised network analysis, was performed to discover genetic networks differentially activated or organized in metastatic OS compared to localized OS. Broad trends among the profiles of metastatic tumors include aberrant activity of intracellular organization and translation networks, as well as disorganization of metabolic networks. The differentially activated PRKCepsilon-RASGRP3-GNB2 network, which interacts with the disorganized DLG2 hub, was also found to be differentially expressed among OS cell lines with differing metastatic capacity in xenograft models.
PRKCε transcript was more abundant in some metastatic OS tumors; however the difference was not significant overall. In functional studies, PRKCε was not found to be involved in migration of M132 OS cells, but its protein expression was induced in M112 OS cells following IGF-1 stimulation.

[563]

**TÍTULO / TITLE:** Desmoplastic fibroma of bone in a toe: radiographic and MRI findings.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Kim OH; Kim SJ; Kim JY; Ryu JH; Choo HJ; Lee SJ; Lee IS; Suh KJ

**INSTITUCIÓN / INSTITUTION:** Department of Radiology, Inje University, Haeundae Paik Hospital, Busan 612-896, Korea.

**RESUMEN / SUMMARY:** Desmoplastic fibroma is a rare benign primary bone tumor that is histologically similar to the soft tissue desmoid tumor. It most often involves the mandible, large long bone or iliac bone. Desmoplastic fibroma in a toe has been extremely rarely reported. Authors report a rare case of desmoplastic fibroma of bone occurring in the distal phalanx of a foot, with descriptions of the radiographic and MRI findings, correlation of the radiologic and pathologic findings, and discussion on the differential diagnosis of the tumor.

[564]

**TÍTULO / TITLE:** Photodynamic Hyperthermal Chemotherapy with Indocyanine Green in 16 Cases of Malignant Soft Tissue Sarcoma: A Novel Cancer Therapy.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Onoyama M; Tsuka T; Imagawa T; Osaki T; Minami S; Azuma K; Kawashima K; Ishi H; Ogawa N; Okamoto Y

**RESUMEN / SUMMARY:** Sixteen cases of malignant soft tissue sarcoma (STS) (10 canine and 6 feline) were treated with a novel triple therapy that combined photodynamic therapy, hyperthermia using indocyanine green with a broad-band light source, and local chemotherapy after surgical tumor resection. This triple therapy was designated photodynamic hyperthermal chemotherapy (PHCT). In all cases, the surgical margin was insufficient. In one feline case, PHCT was performed without surgical removal. PHCT was performed over an interval of 1 to 2 weeks and was repeated 3 to 21 times. No severe side effects, including severe skin burns, necrosis, or rupture of skin sutures, were observed in any of the animals. No recurrence was observed in 7 of 10 (70.0%) dogs and 3 of 6 (50.0%) cats over follow-up periods ranging from 286 to 1901 days. These results suggest that PHCT decreases the risk of recurrence. PHCT should therefore be considered an adjuvant therapy for STS in companion animal medicine.

[565]
Sporadic versus Radiation-Associated Angiosarcoma: A Comparative Clinicopathologic and Molecular Analysis of 48 Cases.

Angiosarcomas are aggressive tumors of vascular endothelial origin, occurring sporadically or in association with prior radiotherapy. We compared clinicopathologic and biologic features of sporadic angiosarcomas (SA) and radiation-associated angiosarcomas (RAA). Methods. From a University of Michigan institutional database, 37 SA and 11 RAA were identified. Tissue microarrays were stained for p53, Ki-67, and hTERT. DNA was evaluated for TP53 and ATM mutations. Results. Mean latency between radiotherapy and diagnosis of RAA was 11.9 years: 6.7 years for breast RAA versus 20.9 years for nonbreast RAA (P = 0.148). Survival after diagnosis did not significantly differ between SA and RAA (P = 0.590). Patients with nonbreast RAA had shorter overall survival than patients with breast RAA (P = 0.03). The majority of SA (86.5%) and RAA (77.8%) were classified as high-grade sarcomas (P = 0.609). RAA were more likely to have well-defined vasoformative areas (55.6% versus 27%, P = 0.127). Most breast SA were parenchymal in origin (80%), while most breast RAA were cutaneous in origin (80%). TMA analysis showed p53 overexpression in 25.7% of SA and 0% RAA, high Ki-67 in 35.3% of SA and 44.4% RAA, and hTERT expression in 100% of SA and RAA. TP53 mutations were detected in 13.5% of SA and 11.1% RAA. ATM mutations were not detected in either SA or RAA. Conclusions. SA and RAA are similar in histology, immunohistochemical markers, and DNA mutation profiles and share similar prognosis. Breast RAA have a shorter latency period compared to nonbreast RAA and a significantly longer survival.

PKPD Modeling of VEGF, sVEGFR-2, sVEGFR-3, and sKIT as Predictors of Tumor Dynamics and Overall Survival Following Sunitinib Treatment in GIST.

The predictive value of longitudinal biomarker data (vascular endothelial growth factor (VEGF), soluble VEGF receptor (sVEGFR)-2, sVEGFR-3, and soluble stem cell factor receptor (sKIT)) for tumor response and survival was assessed based on data from 303 patients with imatinib-resistant gastrointestinal stromal tumors (GIST) receiving sunitinib and/or placebo treatment. The longitudinal
tumor size data were well characterized by a tumor growth inhibition model, which included, as significant descriptors of tumor size change, the model-predicted relative changes from baseline over time for sKIT (most significant) and sVEGFR-3, in addition to sunitinib exposure. Survival time was best described by a parametric time-to-event model with baseline tumor size and relative change in sVEGFR-3 over time as predictive factors. Based on the proposed modeling framework to link longitudinal biomarker data with overall survival using pharmacokinetic-pharmacodynamic models, sVEGFR-3 demonstrated the greatest predictive potential for overall survival following sunitinib treatment in GIST. CPT: Pharmacometrics & Systems Pharmacology (2013) 2, e84; doi:10.1038/psp.2013.61; advance online publication 20 November 2013.

[567]
**TITULO / TITLE:** Langerhans cell sarcoma of the nasopharynx: a rare case.  
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary  
**AUTORES / AUTHORS:** Keklik M; Sivgin S; Kontas O; Abdulrezzak U; Kaynar L; Cetin M  
**INSTITUCIÓN / INSTITUTION:** Doctor, Department of Hematology, Faculty of Medicine, Erciyes Stem Cell Transplantation Hospital, Erciyes University, Turkey.  
**RESUMEN / SUMMARY:** Langerhans cell sarcoma, a tumour with markedly malignant cytological features that originates from Langerhans cells, is a very rare disease. We report the first case of 39-year-old male with Langerhans cell sarcoma arising in the nasopharynx. We chose the 2-chlorodeoxyadenosine (2-CDA) regimen as first-line chemotherapy, and clinical improvement of Langerhans cell sarcoma was obtained. After the fourth cycle of 2-CDA therapy, however, disease progression was observed, and we administered ESHAP regimen (etoposide, carboplatin, cytarabine, methylprednisolone) as a second-line therapy. After we administered two cycles of ESHAP, however, the patient developed aggressive progression and he died. The importance of immunohistochemical findings is obvious in Langerhans cell sarcoma diagnosis. Considering that Langerhans cell sarcoma behaves in a very malignant fashion, a more aggressive treatment approach is necessary for patients with Langerhans cell sarcoma.

[568]
**TITULO / TITLE:** Diagnosis and successful surgical treatment of an unusual inguinal liposarcoma in a pet ferret (Mustela putorius furo).  
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary  
**REVISTA / JOURNAL:** Can Vet J. 2013 Aug;54(8):739-42.  
**AUTORES / AUTHORS:** Gardhouse S; Eshar D; Fromstein J; Smith DA  
**INSTITUCIÓN / INSTITUTION:** Ontario Veterinary College Health Sciences Centre (Gardhouse, Eshar) and Department of Pathobiology (Fromstein, Smith), University of Guelph, Guelph, Ontario.  
**RESUMEN / SUMMARY:** A 4½-year-old female spayed ferret (Mustela putorius furo) was presented for a rapidly growing mass in the inguinal region. Following a complete clinical evaluation, the unusual mass was surgically removed and the histopathological
diagnosis was an inguinal liposarcoma. No post-operative complications were observed over a 14-month follow-up period.

[569]

**TÍTULO / TITLE:**  - The chromatin modification by SUMO-2/3 but not SUMO-1 prevents the epigenetic activation of key immune-related genes during Kaposi’s sarcoma associated herpesvirus reactivation.

**RESUMEN / SUMMARY:**  - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:**  - Chang PC; Cheng CY; Campbell M; Yang YC; Hsu HW; Chang TY; Chu CH; Lee YW; Hung CL; Lai SM; Tepper CG; Hsieh WP; Wang HW; Tang CY; Wang WC; Kung HJ

**RESUMEN / SUMMARY:**  - BACKGROUND: SUMOylation, as part of the epigenetic regulation of transcription, has been intensively studied in lower eukaryotes that contain only a single SUMO protein; however, the functions of SUMOylation during mammalian epigenetic transcriptional regulation are largely uncharacterized. Mammals express three major SUMO paralogues: SUMO-1, SUMO-2, and SUMO-3 (normally referred to as SUMO-1 and SUMO-2/3). Herpesviruses, including Kaposi’s sarcoma associated herpesvirus (KSHV), seem to have evolved mechanisms that directly or indirectly modulate the SUMO machinery in order to evade host immune surveillance, thus advancing their survival. Interestingly, KSHV encodes a SUMO E3 ligase, K-bZIP, with specificity toward SUMO-2/3 and is an excellent model for investigating the global functional differences between SUMO paralogues. RESULTS: We investigated the effect of experimental herpesvirus reactivation in a KSHV infected B lymphoma cell line on genomic SUMO-1 and SUMO-2/3 binding profiles together with the potential role of chromatin SUMOylation in transcription regulation. This was carried out via high-throughput sequencing analysis. Interestingly, chromatin immunoprecipitation sequencing (ChIP-seq) experiments showed that KSHV reactivation is accompanied by a significant increase in SUMO-2/3 modification around promoter regions, but SUMO-1 enrichment was absent. Expression profiling revealed that the SUMO-2/3 targeted genes are primarily highly transcribed genes that show no expression changes during viral reactivation. Gene ontology analysis further showed that these genes are involved in cellular immune responses and cytokine signaling. High-throughput annotation of SUMO occupancy of transcription factor binding sites (TFBS) pinpointed the presence of three master regulators of immune responses, IRF-1, IRF-2, and IRF-7, as potential SUMO-2/3 targeted transcriptional factors after KSHV reactivation. CONCLUSION: Our study is the first to identify differential genome-wide SUMO modifications between SUMO paralogues during herpesvirus reactivation. Our findings indicate that SUMO-2/3 modification near protein-coding gene promoters occurs in order to maintain host immune-related gene unaltered during viral reactivation.

[570]

**TÍTULO / TITLE:**  - Right Native Lung Pneumonectomy Due to over Inflation Three Years after Left Single Lung Transplantation for Pulmonary Lymphangioleiomyomatosis.

**RESUMEN / SUMMARY:**  - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Liu F; Ruan Z; Wang S; Lin Q
INSTITUCIÓN / INSTITUTION: - Department of Thoracic Surgery, The First People’s Hospital Affiliated to Shanghai Jiao Tong University, Shanghai, China.
RESUMEN / SUMMARY: - Native lung hyperinflation (NLH) is one of the known complications after single lung transplantation (SLT). Generally, satisfactory results are achieved in patients undergoing SLT when simultaneous (or second stage) volume reduction of the contralateral native lung is performed. Contralateral native lung pneumonectomy after SLT is rarely reported. In this article, we report a case of a successful, right pneumonectomy of the native lung, 3 years after a left single lung transplant for pulmonary lymphangioleiomyomatosis (PLAM). The patient’s pulmonary function and quality of life improved significantly after a right pneumonectomy of the native lung.

[571]
TÍTULO / TITLE: - Targeting androgen receptor/src complex impairs the aggressive phenotype of human fibrosarcoma cells.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Castoria G; Giovannelli P; Di Donato M; Hayashi R; Arra C; Appella E; Auricchio F; Migliaccio A
INSTITUCIÓN / INSTITUTION: - Department of Biochemistry, Biophysics, and General Pathology, 2nd University of Naples, Naples, Italy.
RESUMEN / SUMMARY: - BACKGROUND: Hormones and growth factors influence the proliferation and invasiveness of human mesenchymal tumors. The highly aggressive human fibrosarcoma HT1080 cell line harbors classical androgen receptor (AR) that responds to androgens triggering cell migration in the absence of significant mitogenesis. As occurs in many human cancer cells, HT1080 cells also express epidermal growth factor receptor (EGFR). EXPERIMENTAL: Findings: We report that the pure anti-androgen Casodex inhibits the growth of HT1080 cell xenografts in immune-depressed mice, revealing a novel role of AR in fibrosarcoma progression. In HT1080 cultured cells EGF, but not androgens, robustly increases DNA synthesis. Casodex abolishes the EGF mitogenic effect, implying a crosstalk between EGFR and AR. The mechanism underlying this crosstalk has been analyzed using an AR-derived small peptide, S1, which prevents AR/Src tyrosine kinase association and androgen-dependent Src activation. Present findings show that in HT1080 cells EGF induces AR/Src Association, and the S1 peptide abolishes both the assembly of this complex and Src activation. The S1 peptide inhibits EGF-stimulated DNA synthesis, cell matrix metalloproteinase-9 (MMP-9) secretion and invasiveness of HT1080 cells. Both Casodex and S1 peptide also prevent DNA synthesis and migration triggered by EGF in various human cancer-derived cells (prostate, breast, colon and pancreas) that express AR. CONCLUSION: This study shows that targeting the AR domain involved in AR/Src association impairs EGF signaling in human fibrosarcoma HT1080 cells. The EGF-elicited processes inhibited by the peptide (DNA synthesis, MMP-9 secretion and invasiveness) cooperate in increasing the aggressive phenotype of HT1080 cells.
Therefore, AR represents a new potential therapeutic target in human fibrosarcoma, as supported by Casodex inhibition of HT1080 cell xenografts. The extension of these findings in various human cancer-derived cell lines highlights the conservation of this process across divergent cancer cells and identifies new potential targets in the therapeutic approach to human cancers.

[572]
TÍTULO / TITLE: - Primary chondrosarcoma of male breast: a rare case.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Badyal RK; Kataria AS; Kaur M
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Government Medical College, Circular Road, Amritsar, 143001 Punjab India.
RESUMEN / SUMMARY: - Sarcomas of the breast are relatively rare and account for 1% of all primary malignant tumors of the breast. Pure and primary chondrosarcoma of the male breast is an extremely rare tumor. It may arise either from the breast stroma itself or from underlying bone or cartilage. Differential diagnoses include cystosarcoma phylloides and breast metaplastic carcinoma with chondroid differentiation.

[573]
TÍTULO / TITLE: - Diagnosis and multimodal therapy for extragastrointestinal stromal tumor of the prostate: A case report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Ou Z; Cao Z; He Y; Tang D
INSTITUCIÓN / INSTITUTION: - Department of Urology, Xiangya Hospital, Central South University, Changsha, Hunan 410008;
RESUMEN / SUMMARY: - Extragastrointestinal stromal tumors (EGISTs), which are neoplasms outside the digestive tract, are predominantly observed in the greater omentum and retroperitoneum. The clinicopathological and molecular characteristics of EGISTs are similar to those of gastrointestinal stromal tumors (GISTs). EGISTs originating from the prostate are extremely rare. In this study, we report a case of a prostatic EGIST in a 39-year-old male, who presented with frequency, urgency, dysuria and a prostatic mass. A 10-core transrectal ultrasound-guided prostate biopsy was performed, and the histological and immunohistochemical results confirmed the diagnosis of EGIST. The patient received a radical prostatectomy, followed by targeted therapy with imatinib (400 mg, daily) for 1 year. Neither recurrence nor metastasis was detected at a 24-month follow-up.

[574]
TÍTULO / TITLE: - Raised CA19.9 and hepatic space occupying lesion after teriparatide therapy in a case of polyostotic fibrous dysplasia.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
**TÍTULO / TITLE:** Pulmonary sarcomatoid carcinoma: a clinicopathologic study and prognostic analysis of 51 cases.  
**RESUMEN / SUMMARY:** BACKGROUND: Pulmonary sarcomatoid carcinoma is a diagnostically challenging group of tumors. It’s a rare histologic subtype of non-small cell lung cancer. There are five subgroups of pulmonary sarcomatoid carcinoma, they are identified as pleomorphic carcinoma, spindle cell carcinoma, giant cell carcinoma, carcinosarcoma, and pulmonary blastoma. We explored the clinicopathologic features and prognostic factors of this tumor. METHODS: We analyzed retrospectively the clinicopathological data of 51 patients with pulmonary sarcomatoid carcinoma who were treated in the First Affiliated Hospital of Zhengzhou University, Henan Cancer Hospital and Henan People Hospital from January 2005 to December 2012. The correlation between prognosis and age, sex, smoking history, tumor size, TNM staging, and treatment modality was analyzed by the statistical software SPSS 17.0. The survival analysis was conducted using the Kaplan-Meier method. The factors influencing survival were analyzed using univariate (Log-rank) and multivariate (Cox) models. RESULTS: The overall survival rates at 1, 2, 3 and 5 years were 45.5%, 35.8%, 28.2% and 20.1%, respectively. Cox univariate analyses revealed that age, tumor size, T stage, M stage, surgery or not, and postoperative chemotherapy or not, were prognostic factors. Cox multivariate analysis found that tumor size and M stage were independent prognostic factors for PSC. CONCLUSIONS: Due to its rarity and the lack of large-scale clinical trial evidence, few studies about PSC have been reported, its clinical and pathological characteristics remain unclear, and its preoperative diagnosis and investigation of novel treatment approaches are imperative. In our study, the main factors affecting the prognosis of tumor size and M staging are the crucial prognostic factors for PSC. Surgical resection and postoperative adjuvant chemotherapy might result in better prognosis.  

**TÍTULO / TITLE:** Transglutaminase-2 Is Involved in Expression of Osteoprotegerin in MG-63 Osteosarcoma Cells.  
**RESUMEN / SUMMARY:** BACKGROUND: Pulmonary sarcomatoid carcinoma is a diagnostically challenging group of tumors. It’s a rare histologic subtype of non-small cell lung cancer. There are five subgroups of pulmonary sarcomatoid carcinoma, they are identified as pleomorphic carcinoma, spindle cell carcinoma, giant cell carcinoma, carcinosarcoma, and pulmonary blastoma. We explored the clinicopathologic features and prognostic factors of this tumor. METHODS: We analyzed retrospectively the clinicopathological data of 51 patients with pulmonary sarcomatoid carcinoma who were treated in the First Affiliated Hospital of Zhengzhou University, Henan Cancer Hospital and Henan People Hospital from January 2005 to December 2012. The correlation between prognosis and age, sex, smoking history, tumor size, TNM staging, and treatment modality was analyzed by the statistical software SPSS 17.0. The survival analysis was conducted using the Kaplan-Meier method. The factors influencing survival were analyzed using univariate (Log-rank) and multivariate (Cox) models. RESULTS: The overall survival rates at 1, 2, 3 and 5 years were 45.5%, 35.8%, 28.2% and 20.1%, respectively. Cox univariate analyses revealed that age, tumor size, T stage, M stage, surgery or not, and postoperative chemotherapy or not, were prognostic factors. Cox multivariate analysis found that tumor size and M stage were independent prognostic factors for PSC. CONCLUSIONS: Due to its rarity and the lack of large-scale clinical trial evidence, few studies about PSC have been reported, its clinical and pathological characteristics remain unclear, and its preoperative diagnosis and investigation of novel treatment approaches are imperative. In our study, the main factors affecting the prognosis of tumor size and M staging are the crucial prognostic factors for PSC. Surgical resection and postoperative adjuvant chemotherapy might result in better prognosis.
Osteoprotegerin (OPG) es un proteína glicoproteína que es miembro de la superfamilia del receptor de factor de necrosis tumoral. Suele funcionar en el remodelado óseo, inhibiendo la formación de osteoclastos través de la interacción con el receptor activador del factor nuclear kappaB (RANKL). Transglutaminasas-2 (Tgase-2) es un grupo de enzimas multifuncionales que desempeña un papel en la metástasis de células cancerosas y la formación de hueso. No obstante, la relación entre OPG y Tgase-2 no ha sido estudiada. Por lo tanto, investigamos la involucración de 12-O-Tetradecanoilphorbol 13-acetato en la expresión de OPG en células MG-63 de osteosarcoma. Interleukin-1beta tiempondependientemente indujo la expresión de OPG y Tgase-2 en líquidos de células y medios de MG-63 por un Western blot. Adicionalmente, un band de 110 kda fue encontrado en el medio de MG-63. 12-O-Tetradecanoilphorbol 13-acetato también indujo la expresión de OPG y Tgase-2. Sin embargo, no se encontró el band de 110 kda en el medio de MG-63 tratado con TPA. Cystamine, un inhibidor de Tgase-2, dosedependientemente suprimió la expresión de OPG en MG-63. La silenciación génica de Tgase-2 también suprimió significativamente la expresión de OPG en MG-63. Enseguida, examinamos si el band de 110 kda de OPG contiene un isopeptido, un indicador de la acción de Tgase-2, mediante una inmunodetección específica para el isopeptido. Sin embargo, no encontramos el isopeptido en el band de 110 kda, sino en 77 kda, que es supuesto que sea la posición del band de Tgase-2. Esto sugiere que el band de 110 kda no es el producto directo de la acción de Tgase-2. En conjunto, OPG y Tgase-2 se inducen por IL-1beta o TPA en células MG-63 y Tgase-2 está implicado en la expresión de OPG en MG-63.
INSTITUCIÓN / INSTITUTION: - Department of Orthopedic Surgery, Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama University, Okayama, Japan ; Department of Molecular Genetics, Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama University, Okayama, Japan.

RESUMEN / SUMMARY: - Synovial sarcoma is a relatively rare high-grade soft tissue sarcoma that often develops in the limbs of young people and induces the lung and the lymph node metastasis resulting in poor prognosis. In patients with synovial sarcoma, specific chromosomal translocation of t(X; 18) (p11.2;q11.2) is observed, and SS18- SSX fusion protein expressed by this translocation is reported to be associated with pathogenesis. However, role of the fusion protein in the pathogenesis of synovial sarcoma has not yet been completely clarified. In this study, we focused on the localization patterns of SS18-SSX fusion protein. We constructed expression plasmids coding for the full length SS18-SSX, the truncated SS18 moiety (tSS18) and the truncated SSX moiety (tSSX) of SS18-SSX, tagged with fluorescent proteins. These plasmids were transfected in synovial sarcoma SYO-1 cells and we observed the expression of these proteins using a fluorescence microscope. The SS18-SSX fusion protein showed a characteristic speckle pattern in the nucleus. However, when SS18-SSX was co-expressed with tSSX, localization of SS18-SSX changed from speckle patterns to the diffused pattern similar to the localization pattern of tSSX and SSX. Furthermore, cell proliferation and colony formation of synovial sarcoma SYO-1 and YaFuSS cells were suppressed by exogenous tSSX expression. Our results suggest that the characteristic speckle localization pattern of SS18-SSX is strongly involved in the tumorigenesis through the SSX moiety of the SS18-SSX fusion protein. These findings could be applied to further understand the pathogenic mechanisms, and towards the development of molecular targeting approach for synovial sarcoma.

[579]
TITULO / TITLE: - Giant Solitary Fibrous Tumor of the Pleura Causing Respiratory Insufficiency: Report of 3 Cases.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Abe M; Nomori H; Fukazawa M; Sugimura H; Narita M; Takeshi A
INSTITUCIÓN / INSTITUTION: - Department of Thoracic Surgery, Kameda Medical Center, Kamogawa, Chiba, Japan.
RESUMEN / SUMMARY: - We present 3 cases of solitary fibrous tumors (SFTs) occupying entire hemithorax and resulting in respiratory insufficiency. All patients were treated by complete resection, resulting in immediate re-expansion of the lungs and recovery from respiratory insufficiency. Although, two patients remain alive without recurrence, one patient had pleural recurrences three times over a 20-year period, all of which were treated by surgical resection. All of the primary tumors exhibited areas of hypercellularity, hemorrhage, or necrosis. All of the recurrent tumors in the recurrent case displayed large areas of hypercellularity, similar to the part of primary tumor. Although, the MIB-1 index in primary tumors was less than 5%, the index of the recurrent tumors increased up to 11% with repeated recurrence. Giant SFTs usually display hypercellularity, hemorrhage, or necrosis. Tumors with hypercellularity could recur. MIB-1 index could display malignant characteristics of recurrent tumors. Long-
term follow-up for more than 10 years after surgery is necessary, particularly for tumors with areas of hypercellularity.

[580]
**TÍTULO / TITLE:** - Case 5/2013 - a four-year-old boy with a rhabdomyoma-type cardiac tumor in both ventricles and repeated ventricular tachycardia.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - Arq Bras Cardiol. 2013 Oct;101(4):e74-e76.

**AUTORES / AUTHORS:** - Atik E

[581]
**TÍTULO / TITLE:** - Co-existence of intramuscular spindle cell lipoma with an intramuscular ordinary lipoma: report of a case.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Laliotis A; De Bree E; Vasilaki S; Papadakis M; Melissas J

**INSTITUCIÓN / INSTITUTION:** - Aggelos Laliotis MD, Department of Surgical Oncology, University Hospital of Heraklion, Crete, Greece, tel. +306944303294, fax +302810392382, e-mail: laliotisac@gmail.com.

**RESUMEN / SUMMARY:** - Spindle cell lipoma is a relatively rare adipocytic neoplasm, which usually occurs in the posterior neck, shoulder or upper back of male patients aged 45-65 years. We report here an unusual coexistence of ordinary and spindle cell lipoma. The patient presented with a painless mass in the area of the right scapula. Imaging was suggestive of a lipomatous mass, possibly liposarcoma. Histological examination revealed the concurrent existence of an intramuscular spindle cell lipoma and an ordinary lipoma. In the literature there are only fourteen cases of intramuscular spindle cell lipoma and only in four cases there was a coexisting mature lipoma. As exclusion of malignancy remains clinicians main concern, diagnosis and treatment of deep seated lipomatous tumors remains challenging.

[582]
**TÍTULO / TITLE:** - Cardiac Papillary Fibroelastoma: Report of Three Cases.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Seto T; Takano T; Otsu Y; Terasaki T; Wada Y; Fukui D; Amano J

**INSTITUCIÓN / INSTITUTION:** - Department of Surgery, Shinshu University School of Medicine, Matsumoto, Nagano, Japan.

**RESUMEN / SUMMARY:** - Primary cardiac tumors are uncommon, and papillary fibroelastoma is considered relatively rare compared to myxoma and lipoma in the primary cardiac tumors. We experienced three cases of fibroelastoma. The patients’ age was 28-75 years, and one patient was female. Two patients were presented with cerebral infarction and cardiac ischemia although, the other patient had no symptoms. Echocardiography revealed a hyperechoic mass in the left atrium, ventricle and on the
aortic valve and helpful for differential diagnosis from myxoma. Surgical excision of the tumor was successfully performed in all patients and post-operative course was uncomplicated.

[583]
TÍTULO / TITLE: - An unusual case of common digital nerve compression caused by a lipoma arising from the flexor tenosynovium.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Suginaka H; Hara A; Kudo T
INSTITUCIÓN / INSTITUTION: - Orthopaedic Surgery, Juntendo University Urayasu Hospital, Chiba 279-0021, Japan.
RESUMEN / SUMMARY: - Lipoma of the hand is a common lesion, but lipoma arising from the flexor tenosynovium is a very rare tumor that induces peripheral nerve disorders. Only four cases of synovial lesions that comprised mature fat at the wrist and imitated carpal tunnel syndrome have been described in the literature. We herein report an unusual case of a lipoma arising from the flexor tenosynovium at the level of the ring finger just proximal to the A1 pulley that was responsible for a sensory disturbance of the ring and middle fingers secondary to compression of the common digital nerve at the palm. The patient was completely relieved of the symptoms after lipoma excision.

[584]
TÍTULO / TITLE: - Giant liposarcoma of the omentum mimicking an ovarian tumor. A case report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Soufi M; Mdaghri J; Benamr S; Lahlou MK; Massrouri R; Essadel A; Mohammadine el H; Taghy A; Settaf A; Chad B
INSTITUCIÓN / INSTITUTION: - Faculty of medicine, University Mohammed first, Oujda, Morocco.
RESUMEN / SUMMARY: - Liposarcomas are common soft tissue of the retroperitoneum and the limbs. They are rarely found in the greater omentum. Once in the peritoneum, these can become enormous and can be mistaken for ovarian mass. The authors report a case of giant omental liposarcoma revealed by an abdominal mass and genital prolapse due to the compression syndrome. A complete macroscopic resection of the omental tumour was performed. A post operative chemotherapy was also prescribed. A description of this clinical presentation in the preoperative assessment, the characteristics of this tumour, together with the use of adjuvant chemotherapy are discussed in this report.

 Bookmark not found.
Rapid Screening of Novel Agents for Combination Therapy in Sarcomas.

For patients with sarcoma, metastatic disease remains very difficult to cure, and outcomes remain less than optimal. Treatment options have not largely changed, although some promising gains have been made with single agents in specific subtypes with the use of targeted agents. Here, we developed a system to investigate synergy of combinations of targeted and cytotoxic agents in a panel of sarcoma cell lines. Agents were investigated alone and in combination with varying dose ratios. Dose-response curves were analyzed for synergy using methods derived from Chou and Talalay (1984). A promising combination, dasatinib and triciribine, was explored in a murine model using the A673 cell line, and tumors were evaluated by MRI and histology for therapy effect. We found that histone deacetylase inhibitors were synergistic with etoposide, dasatinib, and Akt inhibitors across cell lines. Sorafenib and topotecan demonstrated a mixed response. Our systematic drug screening method allowed us to screen a large number of combinations of sarcoma agents. This method can be easily modified to accommodate other cell line models, and confirmatory assays, such as animal experiments, can provide excellent preclinical data to inform clinical trials for these rare malignancies.

Does immunohistochemistry provide additional prognostic data in gastrointestinal stromal tumors?

To investigate the predictive and prognostic effects of clinicopathologic and immunohistochemical (IHC) features in patients with gastrointestinal stromal tumours (GISTs). MATERIALS AND METHODS: Fifty-six patients who were diagnosed with GIST between 2002 and 2012 were retrospectively evaluated. Relationships between clinicopathologic/immunohistochemical factors and prognosis were investigated. RESULTS: Median overall survival (OS) of the whole study group was 74.9 months (42.8-107.1 months), while it was 95.2 months in resectable and 44.7 months in metastatic patients respectively (p=0.007). Epithelioid tumor morphology was significantly associated with shortened OS as compared to...
other histologies \( (p=0.001) \). SMA(+) tumours were significantly correlated with low \((<10/50\text{HPF})\) mitotic activity \( (p=0.034) \). Moreover, SMA(+) patients tended to survive longer and had significantly longer disease-free survival (DFS) times than SMA (-) patients \((37.7 \text{ months vs } 15.9 \text{ months}; p=0.002) \). High Ki-67 level \((>=30\%)\) was significantly associated with shorter OS \((34 \text{ vs } 95.2 \text{ months}; 95\%\text{CI}; p=0.001) \). CD34 (-) tumours were significantly associated with low proliferative tumours \((\text{Ki-67}<10\%)\) \( (p=0.026) \). Median PFS (progression-free survival) of the patients who received imatinib was 36 months \((27.7\text{-}44.2 \text{ months})\). CD34 (-) patients had significantly longer PFS times than that of negative tumours \((50.8 \text{ vs } 29.8 \text{ months}; p=0.045) \). S100 and desmin expression did not play any role in predicting the prognosis of GISTs. Multivariate analysis demonstrated that \(>=10/50\text{HPF}\) mitotic activity/HPF was the only independent factor for risk of death in GIST patients. CONCLUSIONS: Despite the negative prognostic and predictive effect of high Ki-67 and CD34 expression, mitotic activity remains the strongest prognostic factor in GIST patients. SMA positivity seems to affect GIST prognosis positively. However, large-scale, multicenter studies are required to provide supportive data for these findings.

[587]
**TÍTULO / TITLE:** The Inflammatory Kinase MAP4K4 Promotes Reactivation of Kaposi’s Sarcoma Herpesvirus and Enhances the Invasiveness of Infected Endothelial Cells.

**RESUMEN / SUMMARY:** Kaposi’s sarcoma (KS) is a mesenchymal tumour, which is caused by Kaposi’s sarcoma herpesvirus (KSHV) and develops under inflammatory conditions. KSHV-infected endothelial spindle cells, the neoplastic cells in KS, show increased invasiveness, attributed to the elevated expression of metalloproteinases (MMPs) and cyclooxygenase-2 (COX-2). The majority of these spindle cells harbour latent KSHV genomes, while a minority undergoes lytic reactivation with subsequent production of new virions and viral or cellular chemo- and cytokines, which may promote tumour invasion and dissemination. In order to better understand KSHV pathogenesis, we investigated cellular mechanisms underlying the lytic reactivation of KSHV. Using a combination of small molecule library screening and siRNA silencing we found a STE20 kinase family member, MAP4K4, to be involved in KSHV reactivation from latency and to contribute to the invasive phenotype of KSHV-infected endothelial cells by regulating COX-2, MMP-7, and MMP-13 expression. This kinase is also highly expressed in KS spindle cells in vivo. These findings suggest that MAP4K4, a known mediator of inflammation, is involved in KS aetiology by regulating KSHV lytic reactivation, expression of MMPs and COX-2, and, thereby modulating invasiveness of KSHV-infected endothelial cells.
TÍTULO / TITLE: - Primary embryonal rhabdomyosarcoma of the liver in a young male.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Haider N; Nadim MS; Piracha MN

INSTITUCIÓN / INSTITUTION: - Department of Oncology, Combined Military Hospital, Rawalpindi.

RESUMEN / SUMMARY: - Rhabdomyosarcoma (RMS) occurs infrequently in the liver. Rhabdomyosarcomas are malignant tumours that display features of striated muscle differentiation. They are the most common soft-tissue sarcomas among children. In adults however, these are very rare. We report a case of a primary embryonal rhabdomyosarcoma of the liver in a 17 years old boy. This was confirmed by histological examination using immunohistochemical analysis (LCA negative, desmin positive, myogenin focally positive and cytokeratin negative) and site was confirmed by PET CT scan. He received multiple chemotherapies including (doxorubicin, ifosfamide, dacarbazine; gemcitabine, paclitaxel; vincristine, actinomycin D, cyclophosphamide) but longest sustained stable disease was seen with gemcitabine-paclitaxel regimen. The patient died 31 months after the first presentation, secondary to complicated abundant abdominal progressive disease. The poor prognosis and early death of most previously reported cases imply the need for investigation of a more effective treatment method of this uncommon tumour.

TÍTULO / TITLE: - Prevalence of self-reported medical diagnosis of uterine leiomyomas in a Brazilian population: demographic and socioeconomic patterns in the Pro-Saude Study.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Boclin Kde L; Faerstein E

RESUMEN / SUMMARY: - INTRODUCTION: Uterine leiomyomas (UL) are considered the most common tumors of the female reproductive system. However, there are few epidemiological studies about this condition in Brazil. AIM: To estimate the prevalence of self-reported history of UL according to demographic and socioeconomic characteristics, and to markers of access to health care. METHODS: We analyzed data from 1,733 university employees who participated at the baseline waves of the Pro-Saude Study (1999-2001), in relation to three outcomes: (1) self-reported medical diagnosis of UL, (2) UL with symptoms prior to diagnosis, and (3) cases with hysterectomy due to UL. Prevalence and 95% confidence intervals (95% CI) were estimated in relation to strata of variables related to demographic (age, color/race) and socioeconomic characteristics (education, income) and of markers of access to health care (Pap smear, breast clinical exam and private health insurance status). RESULTS:
The prevalence of medically-diagnosed UL was 23.3% (95% CI - 21.3, 25.2), the UL with symptoms prior to diagnosis of 13.3% (95% CI - 11.7, 15.0) and hysterectomy due to UL, 8.4% (95% CI - 7.5, 10.3). Among participants younger than 45 years old, higher prevalence was observed among women with worse socioeconomic conditions and of black color/race. Among those with 45 years or more, there was higher prevalence among women with better access to health care. CONCLUSION: In this study population of Brazilian women, UL is a relevant health problem, and its prevalence and associated socio-demographic gradients are similar to those observed in other countries.

TÍTULO / TITLE: Low income and rural county of residence increase mortality from bone and joint sarcomas.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Cheung MR
INSTITUCIÓN / INSTITUTION: 275 South Bryn Mawr Ave, Bryn Mawr, PA E-mail: cheung.r100@gmail.com.
RESUMEN / SUMMARY: Background: This is a part of a larger effort to characterize the effects on socio-economic factors (SEFs) on cancer outcome. Surveillance, Epidemiology and End Result (SEER) bone and joint sarcoma (BJS) data were used to identify potential disparities in cause specific survival (CSS). Materials and Methods: This study analyzed SEFs in conjunction with biologic and treatment factors. Absolute BJS specific risks were calculated and the areas under the receiver operating characteristic (ROC) curve were computed for predictors. Actuarial survival analysis was performed with Kaplan-Meier method. Kolmogorov-Smirnov’s 2-sample test was used to for comparing two survival curves. Cox proportional hazard model was used for multivariate analysis. Results: There were 13501 patients diagnosed BJS from 1973 to 2009. The mean follow up time (SD) was 75.6 (90.1) months. Staging was the highest predictive factor of outcome (ROC area of 0.68). SEER stage, histology, primary site and sex were highly significant pre-treatment predictors of CSS. Under multivariate analysis, patients living in low income neighborhoods and rural areas had a 2% and 5% disadvantage in cause specific survival respectively. Conclusions: This study has found 2-5% decrement of CSS of BJS due to SEFs. These data may be used to generate testable hypothesis for future clinical trials to eliminate BJS outcome disparities.

TÍTULO / TITLE: Sclerosing stromal tumour in young women: clinicopathologic and immunohistochemical spectrum.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Kaygusuz EI; Cesur S; Cetiner H; Yavuz H; Koc N
INSTITUCIÓN / INSTITUTION: Pathologist, Department of Pathology, Zeynep Kamil Hospital , Burhanettin Ustunel Cad. No.10 Uskudar, Isanbul, Turkey.
RESUMEN / SUMMARY: - Aim: Sclerosing stromal tumor is a benign tumor of ovary. We aimed to review the clinical findings and immunohistochemical results of SSTs through the 7 diagnosed cases in our hospital. Material and Methods: As immunohistochemical, blocks were applied with estrogen receptor, progesterone receptor, inhibin, calretinin, melan-A, CD10, smooth muscle actin, desmine, vimentin, CD34, S-100, C-kit, cytokeratin, cytokeratin7. Results: Macroscopically, while 5 tumors had solid appearance, 2 tumors were composed of solid and cystic areas. All the tumors were in shape of ovarian masses with good limits. Microscopically, two types of cells were observed as fusiform fibroblast-like cells and theca-like cells with vacuolised cytoplasm. Immunohistochemical results: vimentin, smooth muscle actin, desmine, progesterone receptor, calretinin, inhibin were positive in all the cases; S-100, cytokeratin, cytokeratin7, estrogen receptor were negative in all the cases; CD-10 was positive in 2 cases; C-kit was positive in 5 cases; melan-A was positive in 4 cases. Conclusions: The significance of these tumors is that it is necessary to distinguish the histopathology in the frozen section in order to protect the other adnexa because of the characteristics to be observed at early ages (2(nd) and 3(rd) decades). Our findings support the conclusion that sclerosing stromal tumors are benign-character tumors that stem from over stroma and are hormonally active tumors because of the detected clinical and immunohistochemical results, although no hormonal effect that could be supported with laboratory tests was observed.

[592]

TÍTULO / TITLE: - Electrochemotherapy for the treatment of recurring aponeurotic fibromatosis in a dog.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Spugnini EP; Di Tosto G; Salemme S; Pecchia L; Fanciulli M; Baldi A
INSTITUCIÓN / INSTITUTION: - S.A.F.U. Department (Stabilimento Allevatore, Fornitore, Utilizzatore), Regina Elena Cancer Institute - Via delle Messi d’Oro 156, 00158 Rome, Italy (Spugnini, Fanciulli); Ambulatorio Veterinario Tor de Schiavi, Via Tor de Schiavi, 00194 Rome, Italy (Di Tosto, Salemme, Pecchia); Department of Environmental, Biological and Pharmaceutical Sciences and technologies, Second University of Naples, Italy (Baldi).
RESUMEN / SUMMARY: - This paper reports the clinical findings, histopathology, and clinical outcome of a rare case of aponeurotic fibromatosis in a dog. The dog was treated with 4 courses of electrochemotherapy using the drugs cisplatin and bleomycin. There was complete remission and the dog was still disease-free after 18 months.

[593]
TÍTULO / TITLE: - Identification of sequence polymorphisms in the D-loop region of mitochondrial DNA as risk biomarkers for malignant fibrous histiocytoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Xun J; Li Z; Song X; Wang X
INSTITUCIÓN / INSTITUTION: - Department of Osteology, The Fourth Hospital of Hebei Medical University, Shijiazhuang, P.R. China.
RESUMEN / SUMMARY: - Abstract Single nucleotide polymorphisms (SNPs) in the mitochondrial DNA Displacement-loop (D-loop) region particularly in a highly polymorphic homopolymeric C stretch named D310 have been reported to be associated with cancer risk in several types of cancer. In order to evaluate the frequency of D-loop SNPs in a large series of malignant fibrous histiocytoma (MFH) and establish correlations with cancer risk, we sequenced the D-loop of 92 MFH patients and analyzed their use as predictive biomarkers for MFH risk. The minor alleles of nucleotides 73G, 151T were associated with an increased risk for MFH patients, whereas the alleles of nucleotides 16,298C, 152C, and insertion of C at the site 315 (located within the D310) were associated with a decreased risk for MFH patients. These results suggest that SNPs in the mitochondrial D-loop should be considered as a biomarker which may be useful for the early detection of MFH in individuals at risk of this cancer.

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TÍTULO / TITLE: - Initial staging and treatment monitoring of right atrial sarcoma with F-18 fluorodeoxyglucose positron emission tomography/computed tomography.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Manohar K; Kashyap R; Bhattacharya A; Mittal BR
INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine and PET, Postgraduate Institute of Medical Education and Research, Chandigarh, India.

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TÍTULO / TITLE: - How could imaging reduce therapy-associated morbidity in rhabdomyosarcoma of the bladder or prostate?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Kuru TH; Roethke M; Fenchel M; Hadaschik BA
INSTITUCIÓN / INSTITUTION: - Department of Urology, University Hospital Heidelberg, Heidelberg, Germany.

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TÍTULO / TITLE: - Effects of fibronectin and type IV collagen on osteosarcoma cell apoptosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Incesu Z; Hatipoglu I; Sivas H; Ergene E; Ciftci GA
INSTITUCIÓN / INSTITUTION: - Department of Biochemistry, Faculty of Farmacy, Anadolu University, Eskisehir, Turkey. zseller@anadolu.edu.tr
RESUMEN / SUMMARY: - The aims of this study are the investigation of the effects of fibronectin and type IV collagen extracellular matrix proteins and the role of caspase-3
and -9 on cis-platin induced U2-OS apoptosis were studied. First the cytotoxic effects of cis-platin on cell system were investigated by colorimetric method and than morphological and ELISA analysis were used for determination of cell apoptosis when induced with cis-platin. In addition, after adhering the cells to fibronectin or type IV collagen proteins, the apoptotic rate and the effects of caspase-3 and -9 were also investigated by ELISA in presence of specific inhibitors. U2-OS cells showed 20% cytotoxicity after treatment with 2.4 microM of cis-platin for 48 h. Morphological and the numerical data showed that cis-platin was able to induced apoptosis on cells as a dose-dependent manner. Caspase-3 and -9 inhibitors inhibited cis-platin-induced apoptosis in U2-OS cells, respectively. The binding of cells to 10 microg/mL of fibronectin but not type IV collagen enhanced the apoptosis about 2.5 fold that effects inhibited with caspase-3 inhibitor. The caspase-3 and -9 are involved in the apoptotic signals induced by cis-platin in U2-OS. The binding to fibronectin, but not type IV collagen enhanced the apoptotic response of U2-OS and fibronectin-dependent apoptosis was activated by caspase-3. These finding might be useful for patients to fight against osteosarcoma.
RESUMEN / SUMMARY: Osteosarcoma (OS) is a malignant mesenchymal tumor, which rarely occurs in the maxilla. Although variable histologic and radiographic features of OS have been reported previously, in the majority of the cases painful swelling of the jaw is mentioned as the first clinical presentation. Furthermore, early diagnosis and wide surgical resection of the tumor are the most important determinant factors of prognosis. Therefore, the unusual clinical presentations of OS should be considered meticulously to expedite the diagnosis process. We describe a case of OS of the maxilla with extremely unusual presentation in a 42-year-old female, that was initially designated as “epulis fissuratum”. Here, we highlight the importance of combining the clinical, radiographic and histopathologic examination to obtain a definitive diagnosis and also the significance of early effective surgical intervention in evaluation of pathologic lesions.

TÍTULO / TITLE: Angiofibroma of soft tissue: clinicopathologic study of 2 cases of a recently characterized benign soft tissue tumor.

RESUMEN / SUMMARY: Angiofibroma of soft tissue is a very recently characterized, histologically distinctive benign mesenchymal neoplasm of unknown cellular origin composed of 2 principal components, the spindle cell component and very prominent stromal vasculatures. It usually occurs in middle-aged adults, with a female predominance. Herein, we describe the clinical and pathologic details of 2 other examples of this benign tumor. Both patients were middle-aged male and presented with a slow-growing, painless mass located in the deep-seated soft tissue of thigh and left posterior neck region, respectively. Grossly, both tumors were well-demarcated, partial encapsulated of a grayish-white color with firm consistence. Histologically, one case showed morphology otherwise identical to those have been described before, whereas the other case showed in areas being more cellular than most examples of this subtype tumor had, with the lesional cells frequently exhibiting short fascicular, vaguely storiform and occasionally swirling arrangements, which posed a challenging differential diagnosis. Immunostains performed on both tumors did not confirm any specific cell differentiation with lesional cells only reactive for vimentin and focally desmin and negative for all the other markers tested. This report serves to broaden the morphologic spectrum of angiofibroma of soft tumor. Awareness of this tumor is important to prevent misdiagnosis as other more aggressive soft tissue tumor.
Hypoxia-inducible factors: mediators of cancer progression; prognostic and therapeutic targets in soft tissue sarcomas.

RESUMEN / SUMMARY: Soft-tissue sarcomas remain aggressive tumors that result in death in greater than a third of patients due to either loco-regional recurrence or distant metastasis. Surgical resection remains the main choice of treatment for soft tissue sarcomas with pre- and/or post-operational radiation and neoadjuvant chemotherapy employed in more advanced stage disease. However, in recent decades, there has been little progress in the average five-year survival for the majority of patients with high-grade soft tissue sarcomas, highlighting the need for improved targeted therapeutic agents. Clinical and preclinical studies demonstrate that tumor hypoxia and up-regulation of hypoxia-inducible factors (HIFs) is associated with decreased survival, increased metastasis, and resistance to therapy in soft tissue sarcomas. HIF-mediated gene expression regulates many critical aspects of tumor biology, including cell survival, metabolic programming, angiogenesis, metastasis, and therapy resistance. In this review, we discuss HIFs and HIF-mediated genes as potential prognostic markers and therapeutic targets in sarcomas. Many pharmacological agents targeting hypoxia-related pathways are in development that may hold therapeutic potential for treating both primary and metastatic sarcomas that demonstrate increased HIF expression.

Myofibroblastoma: an unusual rapidly growing benign tumour in a male breast.

RESUMEN / SUMMARY: Myofibroblastoma is an unusual benign tumour of the breast predominantly seen in men in their sixth to seventh decade. The gross appearance is that of a well circumscribed nodule, characteristically small, seldom exceeding 3 cm. We present a case of an unusually large myofibroblastoma, which mimicked a malignant breast tumour. A 40 years old male, known case of tetralogy of Fallot, was operated in infancy in abroad, presented with a rapid enlargement of right breast over 5 - 6 weeks. Examination revealed a firm 10 cm hemispherical lump occupying the whole of the right breast with normal overlying skin. Since core biopsy was inconclusive, a subcutaneous mastectomy was performed to remove the tumour, which weighed 500 gms. Histopathology and immunocytochemistry revealed a mixed...
classical and collagenised type of myofibroblastoma. The patient is well with no evidence of recurrence.

[602]
TITULO / TITLE: - A Novel Chordoma Xenograft Allows In Vivo Drug Testing and Reveals the Importance of NF-kappaB Signaling in Chordoma Biology.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Trucco MM; Awad O; Wilky BA; Goldstein SD; Huang R; Walker RL; Shah P; Katuri V; Gul N; Zhu YJ; McCarthy EF; Paz-Priel I; Meltzer PS; Austin CP; Xia M; Loeb DM
INSTITUCIÓN / INSTITUTION: - Sidney Kimmel Comprehensive Cancer Center, Johns Hopkins University, Baltimore, Maryland, United States of America.
RESUMEN / SUMMARY: - Chordoma is a rare primary bone malignancy that arises in the skull base, spine and sacrum and originates from remnants of the notochord. These tumors are typically resistant to conventional chemotherapy, and to date there are no FDA-approved agents to treat chordoma. The lack of in vivo models of chordoma has impeded the development of new therapies for this tumor. Primary tumor from a sacral chordoma was xenografted into NOD/SCID/IL-2R gamma-null mice. The xenograft is serially transplantable and was characterized by both gene expression analysis and whole genome SNP genotyping. The NIH Chemical Genomics Center performed high-throughput screening of 2,816 compounds using two established chordoma cell lines, U-CH1 and U-CH2B. The screen yielded several compounds that showed activity and two, sunitinib and bortezomib, were tested in the xenograft. Both agents slowed the growth of the xenograft tumor. Sensitivity to an inhibitor of IkappaB, as well as inhibition of an NF-kappaB gene expression signature demonstrated the importance of NF-kappaB signaling for chordoma growth. This serially transplantable chordoma xenograft is thus a practical model to study chordomas and perform in vivo preclinical drug testing.

[603]
TITULO / TITLE: - Anterior mediastinal alveolar rhabdomyosarcoma in an infant: rare site for a common paediatric tumour.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Chu WP
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Princess Margaret Hospital, Laichikok, Hong Kong (currently at the Department of Radiology, Tseung Kwan O Hospital, Tseung Kwan O, Hong Kong).
RESUMEN / SUMMARY: - Rhabdomyosarcoma is a common paediatric soft tissue tumour. However, the anterior mediastinum is an extremely rare site for its occurrence. This report describes the imaging and histological findings of such a tumour in a 4-month-old boy.
TÍTULO / TITLE: - Cutaneous epithelioid clear cells angiosarcoma in a young woman with congenital lymphedema.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Tabareau-Delalande F; de Muret A; Miquelestorena-Standley E; Decouvelaere AV; de Pinieux G

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Tours University Hospital, 37044 Tours, France.

RESUMEN / SUMMARY: - Angiosarcomas are rare aggressive neoplasms that can occur secondary to chronic lymphedema (Stewart-Treves syndrome). Although secondary angiosarcomas are commonly described after mastectomy and/or after radiotherapy, few cases have been reported in association with chronic lymphedema of congenital origin. We report the clinical, pathological, and cytogenetic findings in a case of cutaneous epithelioid clear cells angiosarcoma that occurred in a 21-year-old woman with hemibody congenital lymphedema. Surgical biopsies of the tumor mass revealed diffuse epithelioid proliferation of clear atypical cells, for which immunophenotyping highlighted the vascular differentiation. Despite en bloc resection of the tumor, the patient died of metastatic disease three months after diagnosis. This case illustrates the clinical and pathology characteristics of angiosarcoma that is a rare entity secondary to chronic lymphedema. It is the first reported case for which the c-MYC amplification status was assessed. The diagnostic value of this amplification should be further evaluated in this specific context.

TÍTULO / TITLE: - External beam radiation therapy for locally advanced and metastatic gastrointestinal stromal tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Cuaron JJ; Goodman KA; Lee N; Wu AJ

INSTITUCIÓN / INSTITUTION: - Department of Radiation Oncology, Memorial Sloan Kettering Cancer Center, 1275 York Avenue, New York, NY 10065, USA.

RESUMEN / SUMMARY: - BACKGROUND: The role of radiation therapy (RT) in the management of gastrointestinal stromal tumors (GIST) is not well described. Here we report our institutional experience for patients with locally advanced or metastatic GIST treated with RT. METHODS: Between 1997 and 2012, 15 patients with 22 GISTs were treated with RT at our center. The median age was 68 (range, 41-86). Fourteen patients had stage IV disease and 1 patient had stage IIIB disease, per the American Joint Committee on Cancer (AJCC), 7th Edition staging. Tumors were in a variety of locations, and were most commonly referred for palliative treatment. Eighteen of 22 tumors were symptomatic. Prior to RT, 14 of 15 patients received systemic therapy in the form of tyrosine kinase inhibitors (TKIs) (n = 11), chemotherapy (n = 4), or both (n =
1). TKIs were used concurrently for nine tumors (40.9%). No tumors were treated with concurrent chemotherapy. Several fractionation schemes were used, most commonly 3 Gy x 10 (n = 8). Local progression-free survival and overall survival were estimated using the Kaplan-Meier method. Acute toxicity was graded per Common Terminology Criteria for Adverse Events (CTCAE) v4.0. RESULTS: The median follow-up was 5.1 months (range, 1.3-28.3). At the time of analysis, 12 patients have died (80%). The estimated 6-month local progression-free survival and overall survival were 57.0% and 57.8%, respectively. Among the 18 symptomatic tumors, at least partial palliation was achieved in 17 (94.4%), and symptoms were completely palliated in eight (44.4%). Treatment was well tolerated, with no Grade 4 or 5 toxicities. There was no Grade >/=3 toxicity associated with concurrent TKI use. CONCLUSIONS: In this largest series to date of GISTs treated with RT, a high rate of palliation was achieved for symptomatic tumors in a cohort of advanced stage, heavily pretreated patients. Treatment was well tolerated, and concurrent use of tyrosine kinase inhibitor therapy was not associated with additional toxicity. While follow-up was short, durable control is possible for some patients, providing evidence that GIST is not universally radioresistant and that RT can provide an important benefit in patients with progressive or metastatic disease.

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[606]

TÍTULO / TITLE: - Sonographic assessment of pregnancy co-existing with uterine leiomyoma in Owerri, Nigeria.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Eze C; Odumeru E; Ochie K; Nwadike U; Agwuna K

INSTITUCIÓN / INSTITUTION: - Department of Medical Radiography and Radiological Sciences, Faculty of Health Sciences and Technology, University of Nigeria, Enugu Campus, Enugu State, Nigeria.

RESUMEN / SUMMARY: - BACKGROUND: Uterine myomas co-existing with pregnancy could cause obstetric complications. OBJECTIVES: To assess sonographically the frequency of occurrence and effect of uterine myomas co-existing with pregnancy. METHODS: A longitudinal study was conducted during a period of 23 months. A convenience sample of 816 consecutive consenting pregnant women who met the inclusion criteria was evaluated during routine prenatal ultrasound scan. The women were referred for prenatal sonography. One hundred of the subjects who had myoma co-existing with pregnancy and another 100 subjects without myoma were selected for follow-up. These groups were followed up till delivery and obstetric complications and outcomes were documented. Any changes in size and growth rate of myoma were documented. RESULTS: Subjects with myoma co-existing with pregnancy were 12.3%. This was commoner with increasing maternal age. An increase was observed in myoma mean size from 60mm to 63mm from the 1(st) scan sequence to the 2(nd) scan sequence and a reduction from 63mm to 59mm in the 3(rd) scan sequence. Myoma growth rate was 0.667mm per week. Myomas in pregnancy especially large ones caused more complications during delivery when compared to pregnancies without myomas. CONCLUSION: Routine sonography is important in pregnancy management of uterine myomas co-existing with pregnancy.
TÍTULO / TITLE: Human umbilical cord blood-derived CD34 positive endothelial progenitor cells stimulate osteoblastic differentiation of cultured human periosteal-derived osteoblasts.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Byun JH; Lee JH; Oh SH; Hah YS; Cho HY; Kim JH; Park BW; Kang YH; Choi MJ; Shin JK; Rho GJ; Jeon RH; Lee HC; Kim GC; Kim UK; Kim JR; Lee CI

INSTITUCIÓN / INSTITUTION: Gyeongsang National University School of Medicine, Oral and Maxillofacial Surgery, Chilam-dong 90, Jinju, Korea, Republic of, 660-702, 82-55-750-8258, 82-55-761-8024; surbyun@hanmail.net.

RESUMEN / SUMMARY: The aim of this study was to examine the effects of human umbilical cord blood-derived CD34 positive endothelial progenitor cells (CD34+ EPCs) on osteoblastic differentiation of cultured human periosteal-derived osteoblasts (POs). CD34+ cells from human umbilical cord blood were sorted to purify more EPCs in characterization. These sorted cells showed CD31, VE-cadherin and KDR expression as well as CD34 expression, and formed typical tubes in Matrigel. These sorted cells were referred to as human cord blood-derived CD34+ EPCs. In in vivo bone formation using a miniature pig model, the newly formed bone was clearly examined in defects filled with polydioxanone/pluronic F127 (PDO/Pluronic F127) scaffolds containing either human umbilical cord blood-derived CD34+ EPCs and POs, or HUVEC and POs, however, the new bone had the greatest density in the defect treated with CD34+ EPCs and POs. Osteoblastic phenotypes of cultured human POs using ALP activity and von Kossa staining were also more clearly found in CD34+ EPC-conditioned medium than CD34 negative (CD34-) cell-conditioned medium, whereas HUVEC-conditioned medium had an intermediate effect. PCR array for common cytokines and growth factors showed that secretion of IL-1beta was significantly higher in CD34+ EPCs than in HUVEC, followed by level in CD34- cells. In addition, IL-1beta also potently and dose-dependently increased ALP activity and mineralization of POs in culture. These results suggest that human umbilical cord blood-derived CD34+ EPCs stimulates osteoblastic differentiation of cultured human POs. The functional role of human umbilical cord blood-derived CD34+ EPCs in increasing the osteogenic phenotypes of cultured human POs may depend on IL-1beta secreted from human umbilical cord blood-derived CD34+ EPCs.

TÍTULO / TITLE: Ossifying fibroma of the middle turbinate revealed by infection in a young child.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Collin M; Roman S; Fernandez C; Triglia JM; Nicollas R
INTRODUCCIÓN: El fibroma óseo (OF) es un raro tumor fibro-óseo benigno, principalmente ubicado en la región craneal y facial. A menudo, afecta la mandíbula, pero su involucramiento en los senos paranasales ha sido reportado, asociado con un comportamiento más agresivo.

Caso reportado: Reportamos el caso de un niño de 8 años con OF del turbinado medio, revelado por etmoiditis. La resección total fue realizada por un enfoque endoscópico. El paciente no mostró recurrencia clínica o radiológica a 3 años de seguimiento. Es el paciente más joven con OF del turbinado medio hasta la fecha reportado en la literatura internacional.

DISCUSIÓN: El diagnóstico presumptivo se establece mediante el examen clínico y la tomografía computarizada (ubicación, masa ovalada, tumor heterogeneous con un círculo óseo). El diagnóstico definitivo se basa en la examinación histológica (bodies psammomatosos, círculo óseo osteoblastico, hueso trabecular). El tratamiento de los OF en los senos paranasales es quirúrgico, preferentemente por un enfoque endoscópico. La resección debería ser tan completa como sea posible para minimizar el riesgo de recurrencia, especialmente en las localizaciones sinusalas, conocidas como más agresivas. La etmoiditis en una edad inusual debe sugerir una etiología tumoral.
Videoendoscopic surgery for the treatment of esophagus’ leiomyoma.

INTRODUCTION: Leiomyomas are the commonest benign esophageal neoplasms. Surgical treatment is the therapy of choice for such tumors. Open enucleation via thoracotomy has long been the standard procedure. With the emergence of thoracoscopic and laparoscopic approaches, minimally invasive surgery represent interesting alternatives to open surgical procedures. AIM: To propose endoscopic technique for the treatment of these myomas avoiding thoracotomy.

TECHNIQUE: Enucleation of leiomyoma by: A) thoracoscopy, for thoracic esophageal tumors, or B) laparoscopy to the ones located in abdominal esophagus. A) The operations are performed under general anesthesia with selective intubation of the left lung. Patients are placed in the left lateral decubitus position and mild dorsiflexion. Four work trocars are used, two of 11 mm and two of 5 mm. One of the 11 mm is put in the 6th intercostal space in the posterior axillary line to use the 30 degrees endoscope; another, at the same hemi-clavicular line, to take the lung away off surgical site. Other two trocars of 5 mm are installed for working tools of the surgeon, one in the 4th space in the posterior axillary line, and another in the 7th, also in the posterior axillary line. Operations are always initiated by opening the mediastinal pleura, dissection of the tumor with opening the muscle of the esophageal wall, simple enucleation of the tumor and closure of esophageal parietal muscular layer. B) The interventions are done with patients undergoing general anesthesia and placed in the French position. The approach is the same performed to correct the hiatal hernia, and enucleation is done without difficulty. CONCLUSION: Videosurgery for leiomyomas resection is safe and feasible and provides results similar to open procedure, but with a significant reduction in morbidity.
column or at the base of the skull. The association between the brachyury Gly177Asp single nucleotide polymorphism (SNP) and the risk of skull base chordoma in Chinese populations is currently unknown. We investigated the genotype distribution of this SNP in 65 skull-base chordoma cases and 120 healthy subjects. Comparisons of the genotype distributions and allele frequencies did not reveal any significant difference between the groups. Our data suggest that the brachyury Gly177Asp SNP is not involved in the risks of skull-base chordoma, at least in the Chinese population.

[612]
TITULO / TITLE: Silibinin improves the cytotoxicity of methotrexate in chemo resistant human rhabdomyosarcoma cell lines.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Hussain SA; Marouf BH
INSTITUCIÓN / INSTITUTION: Department of Pharmacology and Toxicology, College of Pharmacy, University of Baghdad, Baghdad, Iraq. Tel. +964 (790) 1712624. E-mail: saad_alzaidi@yahoo.com.
RESUMEN / SUMMARY: OBJECTIVE: To investigate whether silibinin (SDH) could overcome chemoresistance of methotrexate (MTX)-resistant human rhabdomyosarcoma (hRD). METHODS: This study was conducted at the Department of Pharmacology and Toxicology, College of Pharmacy, University of Baghdad, Baghdad, Iraq from October 2012 to March 2013. In this in vitro study, resistance to MTX was induced in hRD cell line, the cells were treated with different concentrations of MTX or SDH alone, and in combination. Cell viability was determined by tetrazolium assay. RESULTS: The SDH in a concentration-dependent pattern, enhanced the sensitivity of MTX-resistant cells to the maximum cytotoxic concentration of MTX, and decreased the IC50 (concentration resulting in 50% inhibition of cell growth) of MTX by 17.8 fold. The decrease in IC50 of MTX was negatively correlated with increasing SDH concentrations with R2 = 0.78 and p=0.04. CONCLUSION: The SDH improves the sensitivity of MTX-resistant hRD cell lines to the cytotoxic activity of MTX in concentration-dependent pattern.

[613]
TITULO / TITLE: Hematoporphyrin monomethyl ether-mediated photodynamic therapy selectively kills sarcomas by inducing apoptosis.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Zeng H; Sun M; Zhou C; Yin F; Wang Z; Hua Y; Cai Z
INSTITUCIÓN / INSTITUTION: Department of Orthopedics, Shanghai First People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai, China.
RESUMEN / SUMMARY: We investigated the antitumor effect and mechanism of hematoporphyrin monomethyl ether-mediated photodynamic therapy (HMME-PDT) in sarcomas. Intracellular uptake of HMME by osteosarcoma cells (LM8 and K7) was time- and dose-dependent, while this was not observed for myoblast cells (C2C12) and fibroblast cells (NIH/3T3). HMME-PDT markedly inhibited the proliferation of sarcoma
cell lines (LM8, MG63, Saos-2, SW1353, TC71, and RD) (P<0.05), and the killing effect was improved with increased HMME concentration and energy intensity. Flow cytometry analysis revealed that LM8, MG63, and Saos-2 cells underwent apoptosis after treatment with HMME-PDT. Additionally, apoptosis was induced after HMME-PDT in a three-dimensional culture of osteosarcoma cells. Hoechst 33342 staining confirmed apoptosis. Cell death caused by PDT was rescued by an irreversible inhibitor (Z-VAD-FMK) of caspase. However, cell viability was not markedly decreased compared with the HMME-PDT group. Expression levels of caspase-1, caspase-3, caspase-6, caspase-9, and poly (ADP-ribose) polymerase (PARP) proteins were markedly up-regulated in the treatment groups and increased with HMME concentration as determined by western blot analysis. In vivo, tumor volume markedly decreased at 7-16 days post-PDT. Hematoxylin and eosin staining revealed widespread necrotic and infiltrative inflammatory cells in the HMME-PDT group. Immunohistochemistry analysis also showed that caspase-1, caspase-3, caspase-6, caspase-9, and PARP proteins were significantly increased in the HMME-PDT group. These results indicate that HMME-PDT has a potent killing effect on osteosarcoma cells in vitro and significantly inhibits tumor growth in vivo, which is associated with the caspase-dependent pathway.

[614]
TITULO / TITLE: - Sarcomatoid renal cell carcinoma with rhabdoid features.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Al-Saidi NA; Akhtar M
INSTITUCIÓN / INSTITUTION: - Dr. Mohammed Akhtar, King Faisal Specialist Hospital and Research Center, Pathology & Laboratory Medicine, DPLM, MBC 10 PO Box 3354 Riyadh 11211 Saudi Arabia, T: +966-11-4424280, F: +966-11-4424280, makhatar69@kfshrc.edu.sa.
RESUMEN / SUMMARY: - Sarcomatoid renal cell carcinoma (SRCC) with rhabdoid features is a rare tumor with aggressive behavior and poor prognosis. We report a case of a 71-year-old man with a large left-sided renal mass. Nephrectomy specimen revealed clear cell carcinoma with sarcomatoid and rhabdoid tumor cells. The rhabdoid cells were immunoreactive for mesenchymal markers such as vimentin, epithelial markers such as cytokeratin, and epithelial membrane antigen. These cells were also positive for p53 and had a high proliferation index. The rhabdoid component also demonstrated the loss of immunostaining for integrase interactor 1 (INI1), which stained the other components of the tumor. Only a few cases are available in the published reports documenting rhabdoid cells in SRCC. None of these cases were studied by INI1 immunostain.

[615]
TITULO / TITLE: - Detection of maternal transmission of a splicing mutation in the TSC2 gene following prenatal diagnosis of fetal cardiac rhabdomyomas mimicking congenital cystic adenomatoid malformation of the lung and cerebral tubers and awareness of a family history of maternal epilepsy.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Chen CP; Chang TY; Guo WY; Su YN; Chen YY; Chern SR; Su JW; Wang W
INSTITUCIÓN / INSTITUTION: - Department of Obstetrics and Gynecology, Mackay Memorial Hospital, Taipei, Taiwan; Department of Medical Research, Mackay Memorial Hospital, Taipei, Taiwan; Department of Medicine, Mackay Medical College, New Taipei City, Taiwan; Department of Biotechnology, Asia University, Taichung, Taiwan; School of Chinese Medicine, College of Chinese Medicine, China Medical University, Taichung, Taiwan; Institute of Clinical and Community Health Nursing, National Yang-Ming University, Taipei, Taiwan; Department of Obstetrics and Gynecology, School of Medicine, National Yang-Ming University, Taipei, Taiwan. Electronic address: cpc_mmh@yahoo.com.

RESUMEN / SUMMARY: - OBJECTIVE: To present a prenatal diagnosis of familial tuberous sclerosis complex (TSC). CASE REPORT: A 29-year-old woman was referred to our institution for amniocentesis at 24 weeks of gestation because of congenital anomaly. The fetus had been found to have an intrathoracic echogenic mass, suspicious of type III congenital cystic adenomatoid malformation of the lung (CCAML). The woman presented with a medical history of epilepsy and had received anticonvulsants but did not disclose the disease entity associated with the epilepsy. Amniocentesis revealed a karyotype of 46,XX. A fetal ultrasound examination at 26 weeks of gestation reported the diagnosis of type III CCAML. At 30 weeks of gestation, magnetic resonance imaging showed multiple cortical tubers in the brain along with an intracardiac mass suspicious of cardiac rhabdomyoma, and a diagnosis of fetal TSC was made. A prenatal ultrasound examination at 30 weeks of gestation revealed multiple cardiac tumors and multiple cortical tubers in the brain. The mother admitted that she had been diagnosed to have TSC. Molecular analysis of the cultured amniocytes and the parental blood showed a splicing mutation of c.2639+1G>C in the splice donor site of intron 22 of TSC2 gene in the mother and the fetus. CONCLUSION: Prenatal diagnosis of an intrathoracic lesion with a family history of parental epilepsy should raise a suspicion of fetal cardiac rhabdomyoma and TSC, and prompt magnetic resonance imaging investigation and molecular genetic analysis if necessary.

[616]

TÍTULO / TITLE: - Surgical resection should be taken into consideration for the treatment of small gastric gastrointestinal stromal tumors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Yang J; Feng F; Li M; Sun L; Hong L; Cai L; Wang W; Xu G; Zhang H
INSTITUCIÓN / INSTITUTION: - Department of Digestive Surgery, Xijing Hospital of Digestive Diseases, the Fourth Military Medical University, 127 West Changle Road, 710032, Xi'an, Shaanxi, China. zhanghwfmmu@126.com.
BACKGROUND: The National Comprehensive Cancer Network (NCCN) recommends conservative follow-up for gastric gastrointestinal stromal tumors (GISTs) less than 2 cm. The aim of the present study was to investigate the clinical and pathological features of small gastric GISTs, re-evaluate the risk potential, and discuss the treatment strategy of small gastric GISTs.

METHODS: In this retrospective study, 63 cases of small gastric GISTs (less than 2 cm) were resected surgically from May 2010 to March 2013 in our department. Clinicopathological factors were collected and the malignant potential of small gastric GISTs was analyzed.

RESULTS: The mitotic index of 14 out of 63 cases (22.22%) exceeded 5. The malignant potential of small gastric GISTs was related to tumor location (P = 0.0218). The mitotic index of 4 out of 8 GISTs (50%) located in gastric cardia exceeded 5, 8 out 28 GISTs (28.57%) located in the gastric fundus exceeded 5, and only 2 out of 27 GISTs (7.41%) located in the gastric body exceeded 5. We also discovered a good consistency between mitotic index and Ki-67 expression of small gastric GISTs.

CONCLUSIONS: Gastric GISTs less than 2 cm also have malignant potential. Thus, we recommended surgical resection of all small gastric GISTs once diagnosed.

TÍTULO / TITLE: Multicentric giant cell tumor of bone: synchronous and metachronous presentation.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Wirbel R; Blumler F; Lommel D; Syre G; Krenn V

INSTITUCIÓN / INSTITUTION: Department of Trauma, Hand and Reconstructive Surgery, Verbundkrankenhaus Bernkastel-Wittlich, Koblenzer Street 91, 54516 Wittlich, Germany.

RESUMEN / SUMMARY: A 27-year-old man treated 2.5 years ago for synchronous multicentric giant cell tumor of bone located at the right proximal humerus and the right 5th finger presented now with complaints of pain in his right hip and wrist of two-month duration. Radiology and magnetic resonance revealed multicentric giant cell tumor lesions of the right proximal femur, the left ileum, the right distal radius, and the left distal tibia. The patient has an eighteen-year history of a healed osteosarcoma of the right tibia that was treated with chemotherapy, resection, and allograft reconstruction. A literature review establishes this as the first reported case of a patient with synchronous and metachronous multicentric giant cell tumor who also has a history of osteosarcoma.

TÍTULO / TITLE: A cutaneous angiosarcoma arising from the rhinophyma.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Duzgun S; Pekdemir I; Yilanci S; Bali YY; Singin S; Tapan M

INSTITUCIÓN / INSTITUTION: Department of Plastic and Reconstructive Surgery, Ankara Numune Training and Research Hospital, 06100 Altindag, Ankara, Turkey. serdarduzgun@gmail.com.
RESUMEN / SUMMARY: - In this article, we report a 66-year-old male case of rhinophyma who had a persistent lesion on his nose for two years. Despite steroid therapy, the lesion continued to grow. Histopathological and immunohistochemical findings were consistent with cutaneous angiosarcoma. Rhinophyma-like features should be considered as an unusual clinical manifestation of cutaneous angiosarcoma.

[619]
TÍTULO / TITLE: - Carney Complex with Biatrial Cardiac Myxoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Havrankova E; Stenova E; Olejarova I; Sollarova K; Kinova S
INSTITUCIÓN / INSTITUTION: - Department of Anaesthesiology and Intensive Care, National Institute of Cardiovascular Disease, Bratislava, Slovakia.
RESUMEN / SUMMARY: - Cardiac myxomas make up approximately 50% of all benign cardiac tumors and represented 86% of all surgically treated cardiac tumors. Most of them originated from the left atrium, in some cases from both of atria. We report a case of male patient with biatrial myxomas and other extracardiac involvement: hypophyseal adenoma, enlargement of thyroid gland, tubular adenoma polyp of colon and bilateral large cell calcifying Sertoli cell tumor (LCCSCT) of testis. These findings led to the diagnosis of Carney’s complex, which is a syndrome with multiple neoplasias, cardiac myxomas, lentigines, and endocrine abnormalities. A genetic test confirm this diagnosis.

[620]
TÍTULO / TITLE: - Angiomyolipoma presenting as renal abscesses: a consideration for further evaluation of renal abscess.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Popoola AA; Komolafe OO
INSTITUCIÓN / INSTITUTION: - Department of Surgery University of Ilorin Teaching Hospital, Ilorin Nigeria.
RESUMEN / SUMMARY: - AIMS AND OBJECTIVES: This is to describe an usual presentation of renal angiomyolipoma as renal abscess CASE REPORT: This is a case report of a 51 year old man with clinical and radiological features suggestive of renal abscess. The patient had exploratory laparotomy and a simple nephrectomy for a non functioning kidney which was more like a bag of pus. The histological report however revealed renal abscess coexisting with angiomyolipoma supporting previous reports that renal abscess may be a harbinger of other renal pathologies. CONCLUSION: Abscesses are usually managed by drainage and the dictum of ‘wherever there is pus let it out’ holds generally but this does not always suffice in renal abscesses because renal abscesses are often found in the background of other diseases which may be of more clinical importance.

[621]
TÍTULO / TITLE: - Solitary fibrous tumor of the pleura manifesting as an air-containing cystic mass: radiologic and histopathologic correlation.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Baek JE; Ahn MI; Lee KY

INSTITUCIÓN / INSTITUTION: - Department of Radiology, Seoul St. Mary’s Hospital, College of Medicine, The Catholic University of Korea, Seoul 137-701, Korea.

RESUMEN / SUMMARY: - Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm that typically presents as a well-defined lobular soft tissue mass commonly arising from the pleura. We report an extremely rare case of an SFT containing air arising from the right major fissure in a 58-year-old woman. Chest CT showed an ovoid air-containing cystic mass with an internal, homogeneously enhancing solid nodule. To our knowledge, this is the first case in the literature. The histopathologic findings were correlated with the radiologic findings, and the mechanism of air retention within the tumor is discussed.

[622]

TÍTULO / TITLE: - A Solitary Fibrous Tumour of the Eyelid.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Pecorella I; Cruciani F; Russo V

INSTITUCIÓN / INSTITUTION: - Department of Radiologic, Oncologic and Anatomic Pathology, University of Rome ‘Sapienza’, Rome, Italy.

RESUMEN / SUMMARY: - Objective: To report a case of palpebral solitary fibrous tumour (SFT). Clinical Presentation and Intervention: An elderly man presented with a slow-growing painless mass in the lower conjunctival fornix in the left eye. The lesion was excised and it measured 0.9 cm in maximum diameter. The microscopic features were characteristic of a benign SFT, with immunohistochemical reactivity for vimentin, CD34 and Bcl-2 protein. Nuclear staining for progesterone receptor was also observed. Conclusion: This tumour displayed a benign course, with no recurrence after excision. CD34 immunohistochemistry proved to be a useful adjunct to the microscopic diagnosis. © 2013 S. Karger AG, Basel.

[623]

TÍTULO / TITLE: - Primary pulmonary leiomyosarcoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Shen W; Chen J; Wei S; Wang X; Li X; Zhou Q

INSTITUCIÓN / INSTITUTION: - Department of Lung Cancer Surgery, Tianjin Key Laboratory of Lung Cancer Metastasis and Tumor Microenvironment, Tianjin Medical University General Hospital, Tianjin, China.
Primary pulmonary leiomyosarcoma (PPL) is an extremely rare malignant tumor. In the case presented here, a 52-year-old Chinese female with a lung mass underwent a right upper-middle lobectomy with pulmonary artery sleeve resection and reconstruction, and was thereafter diagnosed with PPL. After 28 months, the patient was well and without local recurrence or distant metastasis.

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**TÍTULO / TITLE:** - Primary chondrosarcoma of breast.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)
**AUTORES / AUTHORS:** - Mujtaba SS; Haroon S; Faridi N
**INSTITUCIÓN / INSTITUTION:** - Department of Histopathology, Liaquat National Postgraduate Medical Institute, Karachi.
**RESUMEN / SUMMARY:** - Mammary sarcomas are heterogeneous group of malignant neoplasms that arise from the mammary stroma. They are uncommon tumours and most of these occur as a component of other tumours. Of the malignant breast mesenchymal tumours, pure sarcomas which lack epithelial component are rarer as these comprise only 0.5% of the breast tumours. Of these, the most common are angiosarcomas, liposarcomas and osteosarcomas. Pure, primary and De novo chondrosarcomas features as one of the rarer types of sarcomas of breast and should be differentiated from Phylloides tumours with chondromatous areas by extensive sampling which also excludes ductal elements in the tumour. This case report describes very rare primary breast sarcoma i.e. chondrosarcoma in a female aged 40 years which was treated by simple mastectomy.

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**TÍTULO / TITLE:** - Minimal fat renal angiomyolipoma with central scar and stellate calcification mimicking a calyceal calculus.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)
**AUTORES / AUTHORS:** - Low E; Tan CH; Ho B; Chong S
**INSTITUCIÓN / INSTITUTION:** - Department of Diagnostic Radiology, Tan Tock Seng Hospital, 11 Jalan Tan Tock Seng, Singapore 308133. low_eugene@hotmail.com.
**RESUMEN / SUMMARY:** - Renal angiomyolipomas are benign neoplasms composed of varying amounts of adipose tissue, smooth muscles and blood vessels. They typically contain macroscopic fat, which is seen as negative attenuation on computed tomography. Calcification and scarring is rarely seen in renal angiomyolipomas. We report the case of a 40-year-old man who was found to have a renal angiomyolipoma with a central stellate scar and focal calcification. The lesion was initially misdiagnosed as a calyceal calculus.

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**TÍTULO / TITLE:** - Atrophic dermatofibroma.
**RESUMEN / SUMMARY:** Dermatofibroma is a benign fibrohistiocytic tumor, common and easily diagnosed when classical clinicopathologic features are present. The atrophic variant of dermatofibroma is of uncertain origin. This lesion is characterized clinically by a flat or atrophic and depressible surface. Histopathological features show reduction of the thickness of the dermis and elastic fibers. We report a typical case of this uncommon and probably underdiagnosed variant.

**TÍTULO / TITLE:** An unusual simultaneous occurrence of gastric adenocarcinoma, leiomyoma and B-cell small lymphocytic lymphoma involving the perigastric lymph nodes and spleen.

**RESUMEN / SUMMARY:** To the Editor, Chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL) is a low-grade, indolent, systemic neoplasm of monomorphic small, round, B-lymphocytes in the peripheral blood, bone marrow and lymph nodes (1). This disease exhibits a variety of immunologic impairments that might increase the risk of second malignancy (1, 2). In fact, there is evidence that the risk of development of non-hematologic malignancies is increased in patients with CLL/SLL, compared with that of the general population. There have also been reports that more aggressive forms of cancers can be observed in the CLL/SLL patients (3-5). We present here a rare case of a simultaneous occurrence of adenocarcinoma of the gastric cardia, submucosal leiomyoma, and CLL/SLL involving the perigastric lymph nodes and spleen.

**TÍTULO / TITLE:** Inflammatory myofibroblastic tumors of the duodenum.
Inflammatory myofibroblastic tumors (IMTs) are rare soft-tissue tumors that can occur at virtually any anatomical site. We report the case of a 58-year-old male with an IMT of the fourth part of the duodenum who presented with signs and symptoms of high intestinal obstruction and bilious vomiting. The patient underwent a surgical resection of the fourth part of the duodenum with end-to-end duodenjejunal anastomosis. The follow-up period of 6 months was uneventful with no evidence of recurrence. According to our knowledge, only six cases of duodenal IMTs have been reported in the literature thus far, and this is the first report of a duodenal IMT sited at the fourth part of the duodenum. The duodenum is among the rarest sites of IMTs. Signs and symptoms resulting from diagnostic imaging investigations are nonspecific and inadequate to obtain diagnosis accurately. In most cases, surgical treatment is considered a cure for IMTs. There is no evidence of deaths caused by duodenal IMT. IMT of the duodenum is a possible diagnosis in differential diagnosis of tumor-like lesions of the duodenum.

Título / Title: Distinct and overlapping sarcoma subtypes initiated from muscle stem and progenitor cells.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Blum JM; Ano L; Li Z; Van Mater D; Bennett BD; Sachdeva M; Lagutina I; Zhang M; Mito JK; Dodd LG; Cardona DM; Dodd RD; Williams N; Ma Y; Lepper C; Linardic CM; Mukherjee S; Grosveld GC; Fan CM; Kirsch DG
INSTITUCIÓN / INSTITUTION: Department of Pharmacology and Cancer Biology, Duke University Medical Center, Durham, NC 27710, USA; Program in Molecular Cancer Biology, Duke University, Durham, NC 27710, USA.
RESUMEN / SUMMARY: Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children, whereas undifferentiated pleomorphic sarcoma (UPS) is one of the most common soft tissue sarcomas diagnosed in adults. To investigate the myogenic cell(s) of origin of these sarcomas, we used Pax7-CreER and MyoD-CreER mice to transform Pax7(+) and MyoD(+) myogenic progenitors by expressing oncogenic Kras(G12D) and deleting Trp53 in vivo. Pax7-CreER mice developed RMS and UPS, whereas MyoD-CreER mice developed UPS. Using gene set enrichment analysis, RMS and UPS each clustered specifically within their human counterparts. These results suggest that RMS and UPS have distinct and overlapping cells of origin within the muscle lineage. Taking them together, we have established mouse models of soft tissue sarcoma from muscle stem and progenitor cells.

Título / Title: Primary multicentric angiosarcoma of bone: true entity or metastases from an unknown primary? Value of comparative genomic hybridization on paraffin embedded tissues.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
Multicentric primary angiosarcoma of bone has been described as a distinct entity from bone metastases from angiosarcoma. Bone angiosarcoma accounts for less than 1% of sarcomas. It has dismal prognosis overall, but the multicentric expression does not confer worse prognosis. We describe the case of an old male with bone angiosarcoma of the extremities with multicentric presentation. He soon after had soft tissue angiosarcoma of the head and neck. Histology and immunohistochemistry were consistent with the diagnosis of high-grade angiosarcoma. Comparative genomic hybridization on paraffin-embedded samples of the bone and head and neck samples suggested additional abnormalities in the bone fragment, thus suggesting that bone lesions were indeed metastatic from his head and neck angiosarcoma; although these preliminary analyses warrant confirmation in other similar rare cases. The patient died after 3 years of relapsed acute leukemia with progressive angiosarcoma.

Enlace al Resumen / Link to its Summary

Autore / Authors: Thariat J; Peyrottes I; Chibon F; Benchetrit M; Saada E; Gastaud L; Dassonville O; Iannessi A; Thyss A
Institución / Institution: Department of Radiation Oncology, Center Antoine-Lacassagne, Nice.

Título / Title: Ossifying Fibroma Tumor Stem Cells Are Maintained by Epigenetic Regulation of a TSP1/TGF-beta/SMAD3 Autocrine Loop.
Resumen / Summary: Enlace al Resumen / Link to its Summary

Enlace al texto completo (gratuito o de pago) 1016/j.stem.2013.08.010
Autore / Authors: Qin H; Qu C; Yamaza T; Yang R; Lin X; Duan XY; Akiyama K; Liu Y; Zhang Q; Chen C; Chen Y; Qi HH; Feng XH; Le AD; Shi S
Institución / Institution: Center for Craniofacial Molecular Biology, Herman Ostrow School of Dentistry, University of Southern California, Los Angeles, CA 90033, USA.

Resumen / Summary: Abnormal stem cell function makes a known contribution to many malignant tumors, but the role of stem cells in benign tumors is not well understood. Here, we show that ossifying fibroma (OF) contains a stem cell population that resembles mesenchymal stem cells (OFMSCs) and is capable of generating OF-like tumor xenografts. Mechanistically, OFMSCs show enhanced TGF-beta signaling that induces aberrant proliferation and deficient osteogenesis via Notch and BMP signaling pathways, respectively. The elevated TGF-beta activity is tightly regulated by JHDM1D-mediated epigenetic regulation of thrombospondin-1 (TSP1), forming a JHDM1D/TSP1/TGF-beta/SMAD3 autocrine loop. Inhibition of TGF-beta signaling in OFMSCs can rescue their abnormal osteogenic differentiation and elevated proliferation rate. Furthermore, chronic activation of TGF-beta can convert normal MSCs into OF-like MSCs via establishment of this JHDM1D/TSP1/TGF-beta/SMAD3 autocrine loop. These results reveal that epigenetic regulation of TGF-beta signaling in MSCs governs the benign tumor phenotype in OF and highlight TGF-beta signaling as a candidate therapeutic target.
Inhibition of the mammalian target of rapamycin leads to autophagy activation and cell death of MG63 osteosarcoma cells.

It has been well documented that the inhibition of the mammalian target of rapamycin (mTOR) induces autophagy in proliferative cells. Therefore, mTOR inhibitors have been proposed for the treatment of cancer. As autophagy plays significant roles in tumor cell survival, the present study aimed to investigate the contribution of autophagy activation to the antitumor effects of cis-diamminedichloroplatinum (CDDP). An MTT assay was used to determine the cytotoxic effects of rapamycin on MG63 osteosarcoma cells. The cell cycle was assessed using a flow cytometry analysis subsequent to staining the DNA with propidium iodide. The mitochondrial membrane potential (Deltapsi) was measured using the fluorescent probe, JC-1. Western blot analysis was used to determine the expression of the proteins that are involved in apoptosis and autophagy, including p53, p62, light chain 3 (LC3) and Beclin-1. The viability of the MG63 cells was inhibited following rapamycin or CDDP treatment. The mitochondrial Deltapsi collapsed following treatment with rapamycin or CDDP. Rapamycin induced cell death and enhanced the effects of the induction of MG63 cell death by CDDP. Western blot analysis detected the induced expression of the p53 and Beclin-1 proteins and the autophagic proteins, LC3 and p62. Rapamycin was observed to induce the death of cancer cells through apoptotic and autophagic mechanisms. Rapamycin may enhance the effects of the activation of autophagy and the induction of apoptosis by CDDP.

Ewing sarcoma cells secrete EWS/Fli-1 fusion mRNA via microvesicles.

Tumours defined as Ewing sarcoma (ES) constitute a group of highly malignant neoplasms that most often affect children and young adults in the first 2 decades of life. The EWS/Fli-1 fusion gene, a product of the translocation t(11;22) (q24; 12), is detected in 95% of ES patients. Recently, it was validated that cells emit a heterogeneous mixture of vesicular, organelle-like structures (microvesicles, MVs) into their surroundings including blood and body fluids, and that these MVs contain a selected set of tumor-related proteins and high levels of mRNAs and miRNAs. In this present study, we detected the Ewing sarcoma-specific EWS/Fli-1
mRNA in MVs from the culture medium of ES cell lines carrying t(11;22) (q24; 12). Also, we detected this fusion gene in approximately 40% of the blood samples from mice inoculated with xenografts of TC135 or A673 cells. These findings indicate the EWS/Fli-1 mRNA in MVs might be a new non-invasive diagnostic marker for specific cases of Ewing sarcoma.

[634]

TÍTULO / TITLE: - Observation of collagen fibrils produced by osteosarcoma cells using atomic force microscopy.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Hoshi O
INSTITUCIÓN / INSTITUTION: - Anatomy and Physiological Science, Graduate School of Health Care Science, Tokyo Medical and Dental University, Tokyo, 113-8519, Japan, o-hoshi.aps@tmd.ac.jp.

RESUMEN / SUMMARY: - The present study examined the three-dimensional process of collagen fibril formation in the human osteosarcoma cell line NOS-1 by conventional scanning electron microscopy (SEM) and atomic force microscopy (AFM). SEM images showed collagen fibril formation on the bottom of culture dishes after 1 week of culture. The collagen fibrils had diameters of 30-100 nm. The surfaces of individual fibrils had characteristic grooves and ridges with periodicities of 60-70 nm. AFM images showed that the newly formed collagen fibrils were 30-300 nm in diameter and possessed characteristic grooves and ridges with periodicities of 60-70 nm. The thicker collagen fibrils contained thinner (approximately 30 nm thick) subfibrils that ran in a helical direction along the long axis of the thicker fibrils. Furthermore, twisted structures of collagen fibrils, which possessed a characteristic rope-like structure, were also identified. The ultrastructure of the collagen fibrils was clearly imaged in liquid medium by AFM, and the process of collagen fibril assembly was successfully analyzed under conditions much closer to the physiological state than those afforded by transmission electron microscopy or SEM. AFM also provided a precise morphological measurement, particularly of the vertical distance, of collagen fibrils with nanometer-scale resolution in liquid conditions.

[635]

TÍTULO / TITLE: - Total spondylectomy following carbon ion radiotherapy to treat chordoma of the mobile spine.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Matsumoto T; Imagama S; Ito Z; Imai R; Kamada T; Shimoyama Y; Matsuyama Y; Ishiguro N
INSTITUCIÓN / INSTITUTION: - Nagoya University Graduate School of Medicine, Department of Orthopaedic Surgery, 65 Tsurumai-cho, Showa-ku, Nagoya city, Aichi 466-8550, Japan.
RESUMEN / SUMMARY: - The main form of treatment of a chordoma of the mobile spine is total en bloc spondylectomy (TES), but the clinical results are not satisfactory. Stand-alone carbon ion radiotherapy (CIRT) for bone and soft-tissue sarcomas has recently been reported to have a high rate of local control with a low rate of local recurrence. We report two patients who underwent TES after CIRT for treating a chordoma in the lumbar spine with good medium-term outcomes. At operation, there remained histological evidence of viable tumour cells in both cases. After the combination use of TES following CIRT, neither patient showed signs of recurrence at the follow-up examination. These two cases suggest that CIRT should be combined with total spondylectomy in the treatment of chordoma of the mobile spine.

[636]

TÍTULO / TITLE: - Temporal trends of incidence and survival of sarcoma of digestive tract including Gastrointestinal Stromal Tumours (GIST) in two areas of the north-east of España in the period 1981-2005: a population-based study.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Rubio-Casadevall J; Borras JL; Carmona C; Ameijide A; Osca G; Vilardell L; Izquierdo A; Galceran J; Marcos-Gragera R

INSTITUCIÓN / INSTITUTION: - Medical Oncology Department, Catalan Institute of Oncology of Girona, Hospital Josep Trueta, Avda Franca s/n, 17005, Gerona, Catalonia, España. jrubioc@iconcologia.net.

RESUMEN / SUMMARY: - INTRODUCTION: The diagnostic approach of Gastrointestinal Stromal Tumours (GIST) was established in 2002. Before this, GIST had been classified with a wide range of histological terms. This fact and the consideration of potential malignity of all these tumours led to a false perception of an increasing incidence. PURPOSE: This study aimed at evaluating the accuracy in registration of sarcoma of digestive tract and GIST and to elucidate the trends of incidence and survival of those. MATERIALS AND METHODS: We used data from two population-based cancer registries in España. In the Girona’s Cancer Registry we previously reclassified all sarcoma of digestive tract performing c-kit to confirm GIST and analysed the time period 1994-2005. In Tarragona’s Cancer Registry, where we analysed the time period 1981-2005, this reclassification was not done. RESULTS: We obtained a significant increasing trend in incidence of all sarcoma of digestive tract in the Tarragona Cancer Registry database, with an annual per cent of change of 3.87 but a non-statistically significant trend in incidence in the Girona Cancer Registry database. The incidence of GIST in Girona Cancer Registry was 1.24 cases/100,000 inhabitants/year. Survival rates did not change in time and was high in less aggressive GIST. The 5-year relative survival for low, intermediate and high risk of malignant behaviour GIST groups were, respectively, 80.5, 85.6 and 64.6 %. CONCLUSIONS: The increase in the incidence of GIST could be explained by the improvement in their diagnosis and registration. The survival of low and intermediate risk of malignant behaviour is high and close to normal population survival.

[637]

TÍTULO / TITLE: - Gene expression of the IGF pathway family distinguishes subsets of gastrointestinal stromal tumors wild type for KIT and PDGFRA.
Gastrointestinal stromal tumors (GISTs) arise from the interstitial cells of Cajal (ICCs) and are the most common mesenchymal neoplasm of the gastrointestinal tract. While the majority of GISTs harbor activating mutations in either the v-kit Hardy-Zuckerman feline sarcoma viral oncogene homolog (KIT) or platelet-derived growth factor receptor alpha (PDGFRA) tyrosine kinases, approximately 10-15% of adult GISTs and 85% of pediatric GISTs lack such mutations. These “wild-type” GISTs have been reported to express high levels of the insulin-like growth factor 1 receptor (IGF1R), and IGF1R-targeted therapy of wild-type GISTs is being evaluated in clinical trials. However, it is not clear that all wild-type GISTs express IGF1R, because studies to date have predominantly focused on a particular subtype of gastric wild-type GIST that is deficient in the mitochondrial succinate dehydrogenase (SDH) complex. This study of a series of 136 GISTs, including 72 wild-type specimens, was therefore undertaken to further characterize wild-type GIST subtypes based on the relative expression of transcripts encoding IGF1R. Additional transcripts relevant to GIST biology were also evaluated, including members of the IGF-signaling pathway (IGF1, IGF2, and insulin receptor [INSR]), neural markers (CDH2 [CDH: Cadherin], neurofilament, light polypeptide, LHX2 [LHX: LIM homeobox], and KIRREL3 [KIRREL: kin of IRRE like]), KIT, PDGFRA, CD34, and HIF1A. Succinate dehydrogenase complex, subunit B protein expression was also assessed as a measure of SDH complex integrity. In addition to the previously described SDH-deficient, IGF1R(high) wild-type GISTs, other SDH-intact wild-type subpopulations were defined by high relative expression of IGF1R, neural markers, IGF1 and INSR, or low IGF1R coupled with high IGF2. These results underscore the complexity and heterogeneity of wild-type GISTs that will need to be factored into molecularly-targeted therapeutic strategies.
enlarged mesenteric lymph nodes. There were multiple variably sized discrete nodules in both lungs. Cavoigraphy showed subtotal occlusion of the inferior vena cava (IVC). She was successfully treated by wide resection and IVC reconstruction with partial cardiopulmonary bypass and metastasectomy.

[639]

**Título / Title:** Surgical treatment of odontogenic myxoma and facial deformity in the same procedure.

**Resumen / Summary:**


**Autores / Authors:** Mayrink G; Luna AH; Olate S; Asprino L; De Moraes M

**Institución / Institution:** Department of Oral and Maxillofacial Surgery, Piracicaba Dental School, State University of Campinas, Brazil.

**Resumen / Summary:** Odontogenic myxoma (OM) is an uncommon benign tumor with aggressive and invasive behavior. Predominant symptoms are usually slow and painless swelling, sometimes resulting in perforation of the cortical borders of the affected bone. In this paper, a case report of a patient with an OM on the right maxillary sinus and a vertical excess of maxilla will be presented. The treatment chosen was tumor resection in association with orthognathic surgery with biomodels assessment for surgical planning. A 3-year follow-up showed disease free and stability of the new position of maxilla. The international literature is evaluated to discuss this case report.

[640]

**Título / Title:** An invasive extragastrointestinal stromal tumor curably resected following imatinib treatment.

**Resumen / Summary:**


**Autores / Authors:** Muto M; Fujiya M; Okada T; Inoue M; Yabuki H; Kohgo Y

**Institución / Institution:** Internal Medicine, Enagaru-Kosei General Hospital, Enagaru, Japan; Email: fjym@asahikawa-med.ac.jp.

**Resumen / Summary:** Extragastrointestinal stromal tumors (EGISTs) are rare tumors located outside the gastrointestinal tract. While curable resection is accepted as a noninvasive EGIST treatment, the therapeutic strategy for invasive EGISTs has not yet been established. The present report is the first to show a case of invasive EGIST completely resected after downsizing the tumor with imatinib treatment. A 69-year-old female had multiple masses adjacent to the stomach and ileocecum. The primary lesion measured 18 cm in size and had invaded the stomach, pancreas and liver. The histological findings of fine-needle aspiration samples revealed a proliferation of dysplastic spindle cells that exhibited immunoreactivity for anti-c-kit antibodies. The masses were therefore diagnosed as multiple GISTs with invasion to other organs, with origin difficult to determine at the time. Nineteen months after the imatinib treatment, the tumors were downsized and distinct from the stomach, pancreas and liver. Accordingly, the tumors were regarded to be EGISTs derived from the mesentery. Because they slightly regressed 26 months after treatment, surgery was applied to remove the EGISTs. The intraoperative findings showed no invasive signs, and the tumors were completely removed. The histological findings revealed the presence of
dysplastic and c-kit-positive spindle cells in the tumor with an MIB-I index of more than 5%, resulting in a final diagnosis of high-risk EGIST derived from the mesentery. No recurrence was detected for 16 months after resection. In conclusion, preoperative treatment with imatinib followed by curable resection is a feasible option to cure invasive EGISTs.

[641]
TÍTULO / TITLE: - An unusual extremely distant noncommunicating uterine horn with myoma and adenomyosis treated with laparoscopic hemihysterectomy.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Morelli M; Venturella R; Mocciaro R; Lico D; Zullo F
INSTITUCIÓN / INSTITUTION: - Department of Obstetrics and Gynecology, Magna Graecia University of Catanzaro, Viale Europa, Loc alita Germaneto, 88100 Catanzaro, Italy.
RESUMEN / SUMMARY: - A 41-year-old woman referred to us with dysmenorrhea and severe pelvic pain although she was previously submitted to right laparotomic adnexectomy for ovarian endometrioma and to a subsequent operative laparoscopy for pelvic adhesions. After ultrasound examination, the patient underwent diagnostic hysteroscopy and operative laparoscopy which confirmed the clinic suspect of an unicornuate uterus. However, it was very unusual to see an extremely distanced right horn, without communication with uterus, without adnexa, and with a small myoma belonging to it. Moreover, omentum and bowel were attached to fundus of right horn and thick adhesions fixed it to rectum and right pelvic wall. Therefore, identification of anatomical structures was difficult, as it was extremely arduous to isolate the ureter, which was involved inside the adhesions surrounding the right uterine horn. Nevertheless, laparoscopic right hemihysterectomy was successfully performed and right horn was sent to our pathologist who recognized hypotrophic endometrium and adenomyosis.

[642]
TÍTULO / TITLE: - Spontaneous Retroperitoneal Hemorrhage (Wunderlich Syndrome) due to Large Upper Pole Renal Angiomyolipoma: Does Robotic-Assisted Laparoscopic Partial Nephrectomy Have a Role in Primary Treatment?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Ploumidis A; Katafigiotis I; Thanou M; Bodozoglou N; Athanasiou L; Ploumidis A
INSTITUCIÓN / INSTITUTION: - Department of Urology, Athens Medical Center, 15125 Athens, Greece.
RESUMEN / SUMMARY: - Spontaneous rapture with consequent retroperitoneal hemorrhage (Wunderlich’s syndrome) is the complication mostly feared from large renal angiomyolipomas (RAMLs). In hemodynamic stable patients, minimal invasive therapies have superseded open surgery as the mainstay of treatment, with
contemporary cases mostly treated by selective arterial embolization. Robotic-assisted laparoscopic partial nephrectomy (RALPN) is an established minimal access treatment that has been used in the past for benign and malignant lesions of the kidney in the elective setting, but rarely in urgent situations as primary treatment. We present a case of a ruptured RAML in a young female treated effectively by RALPN.

[643] **TÍTULO / TITLE:** - The effect of electroacupuncture on osteosarcoma tumor growth and metastasis: analysis of different treatment regimens.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Smeester BA; Al-Gizawi M; O'Brien EE; Ericson ME; Triemstra JL; Beitz AJ
**INSTITUCIÓN / INSTITUTION:** - Department of Medicinal Chemistry, College of Pharmacy, University of Minnesota, Minneapolis, MN 55455, USA.
**RESUMEN / SUMMARY:** - Osteosarcoma is the most common malignant bone tumor found in children and adolescents and is associated with many complications including cancer pain and metastasis. While cancer patients often seek complementary and alternative medicine (CAM) approaches to treat cancer pain and fatigue or the side effects of chemotherapy and treatment, there is little known about the effect of acupuncture treatment on tumor growth and metastasis. Here we evaluate the effects of six different electroacupuncture (EA) regimens on osteosarcoma tumor growth and metastasis in both male and female mice. The most significant positive effects were observed when EA was applied to the ST-36 acupoint twice weekly (EA-2X/3) beginning at postimplantation day 3 (PID 3). Twice weekly treatment produced robust reductions in tumor growth. Conversely, when EA was applied twice weekly (EA-2X/7), starting at PID 7, there was a significant increase in tumor growth. We further demonstrate that EA-2X/3 treatment elicits significant reductions in tumor lymphatics, vasculature, and innervation. Lastly, EA-2X/3 treatment produced a marked reduction in pulmonary metastasis, thus providing evidence for EA's potential antimetastatic capabilities. Collectively, EA-2X/3 treatment was found to reduce both bone tumor growth and lung metastasis, which may be mediated in part through reductions in tumor-associated vasculature, lymphatics, and innervation.

[644] **TÍTULO / TITLE:** - Pazopanib is an active treatment in desmoid tumour/aggressive fibromatosis.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Martin-Liberal J; Benson C; McCarty H; Thway K; Messiou C; Judson I
**RESUMEN / SUMMARY:** - BACKGROUND: Desmoid tumours/aggressive fibromatosis (DT/AF) are infrequent soft-tissue neoplasms. They usually behave as indolent diseases. However, they may grow locally infiltrating or compressing adjacent structures. The role of local treatment is limited and only a few drugs have shown
Cases presentation: We report the outcome of two patients affected by progressive DT/AF treated with the angiogenesis inhibitor pazopanib in two different institutions. Both patients achieved dramatic improvement in their symptoms and radiological signs of response. The clinical benefit lasted for more than 1 year and it is still ongoing. CONCLUSIONS: Pazopanib is an active treatment in DT/AF. It is the first time this has been reported.

[645]

TÍTULO / TITLE: - Neoadjuvant treatment of soft tissue sarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Greto D; Livi L; Saieva C; Bonomo P; Meattini I; Loi M; Di Brina L; Beltrami G; Campanacci D; Scoccianti G; Capanna R; Mangoni M; Paia R; Franchi A; Biti G
INSTITUCIÓN / INSTITUTION: - Radiotherapy Unit, University of Florence, Largo Brambilla 1, 50134, Florence, Italy, daniela.greto@gmail.com.
RESUMEN / SUMMARY: - PURPOSE: The aim of this study was to evaluate disease-free survival (DFS), overall survival and toxicity of patients who underwent preoperative therapy for soft tissue sarcoma. MATERIALS AND METHODS: The data of 38 consecutive patients affected by soft tissue sarcoma were retrospectively analysed. Six (15.8 %) patients were treated only with neoadjuvant radiotherapy, and 32 (84.2 %) with neoadjuvant chemo-radiation therapy. Surgery was performed within 4-6 weeks after the completion of neoadjuvant treatment. RESULTS: Median follow-up was 4.9 years (range 1-13.7 years). All patients received preoperative external beam radiotherapy (RT). Most patients (84.2 %) underwent neoadjuvant chemotherapy treatment associated with radiotherapy. After neoadjuvant treatment, the majority of patients underwent wide excision (32 out of 38) and five patients had marginal surgery; only one patient underwent amputation. Local recurrence was observed in only two patients (5.2 %). Fourteen (36.8 %) patients experienced metastatic relapse. At the time of our analysis 13 patients (34.2 %) had died due to metastatic spread of the disease. In our series, DFS in relation to distant metastases (DM) showed a significant result for lower limb involvement (p = 0.038) and marginal excision (p = 0.024), both predictors of a worse DFS, histology was statistically significant although it was not possible to evaluate the risk for specific histology due to the small number of events in the different subtypes. CONCLUSIONS: The results obtained from our study are encouraging with regard to the feasibility and efficacy of preoperative RT in the treatment of soft tissue sarcoma in view of the results obtained in terms of local control, limb sparing and safety.

[646]

TÍTULO / TITLE: - Abdominal Fibromatosis in a Young Child: A Case Study and Review of the Literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Chu HH; Hwang PH; Jeong YJ; Chung MJ
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Chonbuk National University Medical School, Jeonju, Korea.

RESUMEN / SUMMARY: - Fibromatoses comprise many different entities of well-differentiated fibroblastic proliferation with variable collagen production and form a firm nodular mass. Abdominal fibromatosis is distinguishable from other forms of fibromatosis because of its location and its tendency to occur in women of childbearing age during or following pregnancy. Abdominal fibromatosis in children is an extremely rare condition. A 15-month-old boy presented with an abdominal wall mass that had recently increased in size. Mass excision was performed. The tumor was 4.3x4.1 cm and partly circumscribed. Histologically, the tumor was composed of parallel long fascicles of spindle-cells with a uniform appearance. The edges of the resected mass were infiltrative, and the surgical margins were positive. Mitotic figures were <1/10 high power fields. No cellular atypia or necrosis was present. The tumor cells were positive for vimentin and nuclear beta-catenin staining.

[647]
TÍTULO / TITLE: - Clinicopathologic features and responses to radiotherapy of myeloid sarcoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Chen WY; Wang CW; Chang CH; Liu HH; Lan KH; Tang JL; Tien HF; Kuo SH; Cheng AL

RESUMEN / SUMMARY: - BACKGROUND: To evaluate clinicopathological features, radiotherapeutic parameters, and their associations with responses to radiotherapy (RT) in patients with myeloid sarcoma (MS). METHODS: We reviewed 20 patients receiving RT for MS lesions (in 43 RT courses) and analyzed the patients' clinicopathologic features and radiotherapeutic parameters, and their associations with complete responses (CR) to RT using Fisher’s exact test and univariate logistic regression analysis. Generalized Estimating Equation was used to analyze all 43 irradiated lesions and account for the correlations in RT responses among lesions from the same patient. RESULTS: We found that the underlying hematological diseases of the evaluated patients were acute myeloid leukemia (AML) in 14 patients (70%), chronic myeloid leukemia in 4 patients (20%), myelodysplastic syndrome with AML transformation in one patient (5%), and de novo MS in one patient (5%). Most patients (55%) received RT for MS at the time of relapse following bone marrow transplantation (BMT). The most common cytogenetic abnormality was t(8;21)(q22;q22). The median RT dose of 20 Gy (range 6--35 Gy), administered in 1.5-3.5 Gy fractions, provided a 63% CR rate. RT dose, sex, cytogenetics, and bone marrow status at the time of RT had no significant effect on CR. Younger age (<50 y, P = 0.06), BMT prior to RT (P = 0.05), and underlying AML (P = 0.05) were marginally associated with higher CR to RT. CONCLUSIONS: Our results indicate that a modest RT dose (20-30Gy) achieves good local control of MS. Age, previous BMT, and underlying hematologic disease can affect RT response.

[648]
TÍTULO / TITLE: - Current therapeutic strategies and novel approaches in osteosarcoma.
Osteosarcoma is the most frequent malignant primary bone tumor and a main cause of cancer-related death in children and adolescents. Although long-term survival in localized osteosarcoma has improved to about 60% during the 1960s and 1970s, long-term survival in both localized and metastatic osteosarcoma has stagnated in the past several decades. Thus, current conventional therapy consists of multi-agent chemotherapy, surgery and radiation, which is not fully adequate for osteosarcoma treatment. Innovative drugs and approaches are needed to further improve outcome in osteosarcoma patients. This review describes the current management of osteosarcoma as well as potential new therapies.

[649]

New clinical application of high-intensity focused ultrasound: local control of synovial sarcoma.

Hu X; Cai H; Zhou M; He H; Tian W; Hu Y; Chen L; Deng Y

Department of Surgical Oncology, Second Affiliated Hospital, Zhejiang University College of Medicine, No, 88 Jiefang Road, Hangzhou, PR China. dengyc001@hotmail.com.

High-intensity focused ultrasound (HIFU) is playing an increasingly important role in cancer therapy. Primary synovial sarcomas of the chest wall are extremely rare. We report the first case of noninvasive HIFU therapy for the control of synovial sarcoma. A 51-year-old man was diagnosed with spindle cell sarcoma on the left chest wall through lumpectomy. After four cycles of chemotherapy, local recurrence of the sarcoma was detected. Subsequent extended resection confirmed synovial sarcoma. After five cycles of a new chemotherapy option, the sarcoma relapsed again. Then the patient received five courses of HIFU; this completely ablated the sarcoma without complications. No chemotherapy, radiotherapy, or biological therapy has been applied since. Now the patient is stable and has a high quality of life.

[650]

Role of Topoisomerases in Pediatric High Grade Osteosarcomas: TOP2A Gene Is One of the Unique Molecular Biomarkers of Chemoresponse.

Hu X; Cai H; Zhou M; He H; Tian W; Hu Y; Chen L; Deng Y

Department of Surgical Oncology, Second Affiliated Hospital, Zhejiang University College of Medicine, No, 88 Jiefang Road, Hangzhou, PR China. dengyc001@hotmail.com.

High-intensity focused ultrasound (HIFU) is playing an increasingly important role in cancer therapy. Primary synovial sarcomas of the chest wall are extremely rare. We report the first case of noninvasive HIFU therapy for the control of synovial sarcoma. A 51-year-old man was diagnosed with spindle cell sarcoma on the left chest wall through lumpectomy. After four cycles of chemotherapy, local recurrence of the sarcoma was detected. Subsequent extended resection confirmed synovial sarcoma. After five cycles of a new chemotherapy option, the sarcoma relapsed again. Then the patient received five courses of HIFU; this completely ablated the sarcoma without complications. No chemotherapy, radiotherapy, or biological therapy has been applied since. Now the patient is stable and has a high quality of life.
AUTORES / AUTHORS: - Nguyen A; Lasthaus C; Guerin E; Marcellin L; Pencreach E; Gaub MP; Guenot D; Entz-Werle N

INSTITUCIÓN / INSTITUTION: - Laboratoire de Biochimie et Biologie Moleculaire, CHRU Hautepierre, Avenue Moliere, Strasbourg Cedex 67098, France. Natacha.entz-werle@chru-strasbourg.fr.

RESUMEN / SUMMARY: - Currently, the treatment of pediatric high-grade osteosarcomas systematically includes one topoisomerase IIalpha inhibitor. This chemotherapy is usually adapted to the response to the neo-adjuvant therapy after surgery. The current and unique marker of chemoresponsiveness is the percentage of viable residual cells in the surgical resection. This late patient management marker has to be evaluated earlier in the therapeutic history of the patients on initial biopsy. Therefore, new biomarkers, especially those involved in the topoisomerase IIalpha inhibitor response might be good candidates. Therefore, our study was designed to target TOP1, TOP2A and TOP2B genes in 105 fresh-frozen diagnostic biopsies by allelotyping and real-time quantitative PCR. Our analyses in those pediatric osteosarcomas, homogeneously treated, highlighted the frequent involvement of topoisomerase genes. The main and most important observation was the statistical link between the presence of TOP2A amplification and the good response to neo-adjuvant chemotherapy. Compared to adult cancers, the 17q21 amplicon, including TOP2A and ERBB2 genes, seems to be differentially implicated in the osteosarcoma chemoresponsiveness. Surprisingly, there is no ERBB2 gene co-amplification and the patients harboring TOP2A amplification tend to show a worse survival, so TOP2A analyses remain a preliminary, but a good molecular approach for the evaluation at diagnosis of pediatric osteosarcoma chemoresponsiveness.

[TÍTULO / TITLE: - A case of myxoid adrenocortical neoplasm: computed tomography and magnetic resonance imaging characteristics.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Fukuhara H; Bilim V; Ohtake H; Yahagi Y; Tomita Y

INSTITUCIÓN / INSTITUTION: - Department of Urology, Yamagata University Faculty of Medicine, Yamagata, Japan.

RESUMEN / SUMMARY: - Myxoid adrenocortical neoplasms are rare; to our knowledge, only 56 cases have been reported in the literature. Therefore, distinguishing benign from malignant cases is challenging. Although the histopathological features of myxoid adrenocortical neoplasia have been amply demonstrated, their imaging characteristics are yet to be reported. We describe here these characteristics for such a neoplasm. Our patient, a 70-year-old male, was found to have a 3-cm left adrenal incidentaloma through a non-enhanced computed tomography. Attenuation measurements were 22 Hounsfield units on precontrast imagery, and percentage enhancement washout was 92%. Magnetic resonance imaging showed no loss of signal intensity in T1-weighted out-of-phase images, but high signal intensity on T2-weighted and diffusion-weighted images. Left adrenalectomy was performed and the pathological diagnosis was confirmed as myxoid adrenocortical neoplasm. The imaging characteristics reported here will be beneficial to the differential diagnosis of myxoid adrenocortical neoplasms based upon image analysis and will help distinguish benign from malignant neoplasms.
Histologic and genetic advances in refining the diagnosis of “undifferentiated pleomorphic sarcoma”.

Undifferentiated pleomorphic sarcoma (UPS) is an inclusive term used for sarcomas that defy formal sub-classification. The frequency with which this diagnosis is assigned has decreased in the last twenty years. This is because when implemented, careful histologic assessment, immunohistochemistry, and ultrastructural evaluation can often determine lineage of differentiation. Further attrition in the diagnostic frequency of UPS may arise by using array-comparative genomic hybridization. Gene expression arrays are also of potential use as they permit hierarchical gene clustering. Appraisal of the literature is difficult due to a historical perspective in which specific molecular diagnostic methods were previously unavailable. The American Joint Committee on Cancer (AJCC) classification has changed with different inclusion criteria. Taxonomy challenges also exist with the older term “malignant fibrous histiocytoma” being replaced by “UPS”. In 2010 an analysis of multiple sarcoma expression databases using a 170-gene predictor, re-classified most MFH and “not-otherwise-specified” (NOS) tumors as liposarcomas, leiomyosarcomas or fibrosarcomas. Interestingly, some of the classifier genes are potential molecular therapeutic targets including Insulin-like growth factor 1 (IGF-1), Peroxisome proliferator-activated receptor gamma (PPARgamma), Nerve growth factor beta (NGF beta) and Fibroblast growth factor receptor (FGFR).

Generalized Lymphadenopathy as the First Presentation of Granulocytic Sarcoma: A Diagnostic Challenge.

Granulocytic sarcoma (GS), also known as chloroma or extramedullary myeloblastoma, is a solid tumor composed of primitive precursors of the granulocytic series that include myeloblasts, promyelocytes, and myelocytes. Granulocytic sarcoma is a rare tumor that may develop during acute myeloid leukemia (AML) but less frequently may precede its presentation. Although generalized lymph node enlargement is a presentation for malignant lymphoma, it can
also rarely be the early presenting sign of GS. Methods. We present a case of GS mimicking lymphoma in a 45-year-old male. The patient presented with bilateral neck masses and had widespread, prominent lymphadenopathy secondary to AML as the first presenting manifestation of GS for the last 4 months with concurrent marrow AML.

Result. A clinical diagnosis of lymphoma was suspected; fine needle aspiration cytology findings were also suggestive of lymphoma. However, peripheral blood and bone marrow examination reported as acute myeloid leukemia with monocytic differentiation and histopathology of excised lymph node confirmed it to be a GS not lymphoma. Conclusion. GS is often misdiagnosed as malignant lymphoma because of cytomorphic and histologic similarities of the blasts to large cell lymphoma. A careful search for immature myeloid is a useful clue to the diagnosis accompanied with appropriate immunophenotyping.

[654]
TITULO / TITLE: Peripheral odontogenic myxoma of maxillary gingiva: A rare clinical entity.  
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary  
AUTORES / AUTHORS: Jain VK; Reddy SN  
INSTITUCIÓN / INSTITUTION: Department of Oral and Maxillofacial Pathology, M. R. Ambedkar Dental College, Bangalore, Karnataka, India.  
RESUMEN / SUMMARY: Odontogenic myxoma comprises 3-6% of all odontogenic tumors. Odontogenic myxomas are relatively rare benign mesenchymal tumors found exclusively in the tooth-bearing areas of the jaw and are usually located centrally in the mandible. Soft-tissue localization is rarely seen and is classified as peripheral myxoma. Peripheral myxoma is slow growing and less aggressive, as compared to the central myxoma. It has a low recurrence rate. Till date, only few cases of maxillary gingival myxomas are reported in the literature. Here, we present an unusual case of primary peripheral odontogenic myxoma occurring in the gingiva of anterior maxilla in a 41-year-old female patient.

[655]
TITULO / TITLE: Imatinib mesylate in clinically suspected gastric stromal tumors.  
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary  
AUTORES / AUTHORS: Li ZY; Tang L; Li SX; Shan F; Bu ZD; Ji JF  
INSTITUCIÓN / INSTITUTION: Department of Gastrointestinal Surgery, Beijing Cancer Hospital and Institute, Peking University School of Oncology, Key laboratory of Carcinogenesis and Translational Research (Ministry of Education), Beijing 100142, China;  
RESUMEN / SUMMARY: Gastrointestinal stromal tumors (GISTs) occur most frequently in the stomach. Diagnosis of gastric GIST is not always clear before surgery. Flexible endoscopy may suggest the nature of the lesion (a bulky tumor with preserved mucosa); however, biopsy is rarely diagnostic. Therefore, diagnostic medication with
safe drugs may provide a feasible way under such conditions after an informed consent is obtained. Based on the excellent efficacy of imatinib mesylate (IM) in the treatment of GIST, we successfully applied it in the diagnostic medication of two patients with clinically suspected gastric stromal tumors. In conclusion, the diagnostic medication with IM can be an alternative option for patients with suspected GIST that can not be confirmed pathologically.

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[656]
**TÍTULO / TITLE:** - Clinical activity of lenalidomide in visceral human immunodeficiency virus-related kaposis sarcoma.

**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](http://jama.ama-assn.org/search.dtl)

**REVISTA / JOURNAL:** - JAMA. Acceso gratuito al texto completo.

**AUTORES / AUTHORS:** - Steff M; Joly V; Di Lucca J; Feldman J; Burg S; Sarda-Mantel L; Peytavin G; Marinho E; Crickx B; Raymond E; Lariven S; Maubec E

**INSTITUCIÓN / INSTITUTION:** - Service de Dermatologie, Hopital Bichat, Assistance Publique-Hopitaux de Paris, Paris, France2Universite Paris Diderot Sorbonne Paris Cite, Paris, France.

**RESUMEN / SUMMARY:** - IMPORTANCE Curative treatment of aggressive Kaposi sarcoma (KS) with conventional chemotherapy in human immunodeficiency virus (HIV)-infected patients remains difficult. The administration of thalidomide, an immunomodulatory drug with antiangiogenic effects, is limited by its toxicity. This engenders interest in evaluating thalidomide analogues such as lenalidomide with better toxicity profiles. To our knowledge, we describe for the first time a patient with visceral KS successfully treated with lenalidomide. OBSERVATIONS A man with advanced visceral HIV-related KS progressing after 11 months of highly active antiretroviral therapy (HAART) and 2 lines of conventional chemotherapy (pegylated liposomal doxorubicin and docetaxel) was treated with lenalidomide on a compassionate use basis. He showed a rapid partial response without any substantial adverse effect but experienced relapse after 5 months of treatment, in a context of virologic failure. CONCLUSIONS AND RELEVANCE Similar to our observation, good partial response without toxic effects has been reported in 3 patients with only skin involvement. Because immune reconstitution syndrome may occur in HIV-infected patients with KS undergoing HAART, KS improvement may be partly explained by immune recovery. An ongoing US phase ½ trial will better evaluate the efficacy and tolerance of lenalidomide in patients with HIV-related KS with and without visceral involvement.

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[657]
**TÍTULO / TITLE:** - Gastrointestinal stromal tumors: a case-only analysis of single nucleotide polymorphisms and somatic mutations.

**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](http://jama.ama-assn.org/search.dtl)


**INSTITUCIÓN / INSTITUTION:** - [Enlace a la Editora de la Revista](http://jama.ama-assn.org/search.dtl)

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BACKGROUND: Gastrointestinal stromal tumors are rare soft tissue sarcomas that typically develop from mesenchymal cells with acquired gain-in-function mutations in KIT or PDGFRA oncoproteins. These somatic mutations have been well-characterized, but little is known about inherited genetic risk factors. Given evidence that certain susceptibility loci and carcinogens are associated with characteristic mutations in other cancers, we hypothesized that these signature KIT or PDGFRA mutations may be similarly fundamental to understanding gastrointestinal stromal tumor etiology. Therefore, we examined associations between 522 single nucleotide polymorphisms and seven KIT or PDGFRA tumor mutation types. Candidate pathways included dioxin response, toxin metabolism, matrix metalloproteinase production, and immune and inflammatory response.

METHODS: We estimated odds ratios and 95% confidence intervals for associations between each candidate SNP and tumor mutation type in 279 individuals from a clinical trial of adjuvant imatinib mesylate. We used sequence kernel association tests to look for pathway-level associations.

RESULTS: One variant, rs1716 on ITGAE, was significantly associated with KIT exon 11 non-codon 557--8 deletions (odds ratio = 2.86, 95% confidence interval: 1.71-4.78) after adjustment for multiple comparisons. Other noteworthy associations included rs3024498 (IL10) and rs1050783 (F13A1) with PDGFRA mutations, rs2071888 (TAPBP) with wild type tumors and several matrix metalloproteinase SNPs with KIT exon 11 codon 557--558 deletions. Several pathways were strongly associated with somatic mutations in PDGFRA, including defense response (p = 0.005) and negative regulation of immune response (p = 0.01).

CONCLUSIONS: This exploratory analysis offers novel insights into gastrointestinal stromal tumor etiology and provides a starting point for future studies of genetic and environmental risk factors for the disease.

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TÍTULO / TITLE: TIM-3 expression in human osteosarcoma: Correlation with the expression of epithelial-mesenchymal transition-specific biomarkers.

RESUMEN / SUMMARY: Signals from the T cell Ig- and mucin-domain-containing molecules (TIMs) have been demonstrated to be actively involved in regulating the progression of carcinomas. However, the expression and distribution of these molecules in osteosarcoma, the most common primary bone malignancy with poor prognosis, have not been investigated. In this study, the expression of TIMs was examined in nine invasive human osteosarcomas using immunohistochemistry, and the phenotypes were detected by dual immunofluorescence staining. Using immunohistochemistry, it was observed that only TIM-3, rather than TIM-1 or TIM-4, was expressed in these tumor specimens, where it was localized in the cytoplasm and plasma membrane of tumor cells. Dual immunofluorescence staining revealed that the
expression of TIM-3 was observed in all cell types investigated, including CD68+ macrophages, CD31+ endothelial cells, CK-18+ epithelial cells and PCNA+ tumor cells. Notably, in sarcoma cells, TIM-3 was co-expressed with certain biomarkers of epithelial-mesenchymal transition (EMT), including vimentin, Slug, Snail and Smad. These combined results suggest that TIM-3 triggers tumor cells to acquire features of aggressive EMT and may be involved in the pathogenesis of this malignancy.

[659]
TÍTULO / TITLE: - Analysis of risk factors for recurrence of giant cell tumor of the sacrum and mobile spine combined with preoperative embolization.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Ming Z; Kangwu C; Huilin Y; Genlin W; Jian L; Yiming J; Chunshen W; Chao C
INSTITUCIÓN / INSTITUTION: - The First Affiliated Hospital of Soochow University, Department of Orthopedic Surgery, Suzhou, Jiangsu, China.
RESUMEN / SUMMARY: - AIM: To investigate the factors related to the local recurrence-free survival time (LRFS) after surgical treatment of GCT of the sacrum and mobile spine combined with preoperative embolization. MATERIAL and METHODS: We retrospectively reviewed 28 consecutive patients with GCT of the sacrum and mobile spine who underwent initial surgical excision combined with preoperative embolization between 1995 and 2011. Data regarding age, gender, tumor location, tumor size, tumor extension, radiation therapy, and local recurrences were reviewed and analyzed statistically. RESULTS: All patients underwent intralesional resection. The average duration of follow-up was 86.4 months (range, 15 - 193 months). 8 (28.6%) patients developed local recurrence. The average recurrence time was 35.6 months (range, 5 - 79 months), and the local recurrence-free survival rates at 3 and 5 years were 89.1% and 75.5%, respectively. LRFS was found statistically longer in intracompartmental (T1) tumors as compared with extracompartmental (T2) tumors (P < 0.05), but not for age, gender, tumor location, tumor size, or radiation therapy. CONCLUSION: Intralesional excision with preoperative embolization is a feasible choice for T1 tumors of the sacrum and mobile spine, but for T2 tumors, more aggressive treatment may be required. The choice of surgical treatment should be balanced between the complications and tumor recurrence.

[660]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Amelia Souza A; de Araujo VC; Passador Santos F; Ferreira Martinez E; de Menezes Filho JF; Soares de Araujo N; Soares AB
INSTITUCIÓN / INSTITUTION: - Department of Oral Pathology, Sao Leopoldo Mandic Institute and Research Center, Rua Jose Rocha Junqueira 13 Ponte Preta, 13045-755 Campinas, SP, Brazil.
RESUMEN / SUMMARY: - A case of adult rhabdomyoma is reported. The lesion is a rare benign tumor of skeletal muscle origin which occurs predominantly in the head and neck region. In the present case, the clinical diagnosis favored a benign salivary gland tumor. Histologically, the tumor was composed of large round, oval, and polygonal cells of varying size with abundant pale, eosinophilic, fine, granular cytoplasm with peripherally located nuclei. Immunohistochemically, the lesion was positive for muscle-specific actin, smooth muscle actin, desmin, S100 protein, and Masson’s trichrome. Electron microscopic examination confirmed the presence of numerous myofibrils. The lesion was treated by surgical resection. The clinical, histological, immunohistochemical, and ultrastructural features are discussed in this study.

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TÍTULO / TITLE: - Magnetic resonance imaging of vulvar dermatofibrosarcoma protuberans - report of a case.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Ozmen E; Guney G; Algin O

INSTITUCIÓN / INSTITUTION: - Department of Radiology.

RESUMEN / SUMMARY: - BACKGROUND: Dermatofibrosarcoma protuberans (DFSP) of the vulva is a rare low-grade soft tissue sarcoma. Magnetic resonance imaging (MRI) findings of vulvar DFSP were essentially unreported in the literature. CASE REPORT: We report a DFSP of vulva with its clinical, histological and MRI features. As far we know this is the first case of histologically confirmed vulvar DFSP presenting with MR images. The diagnosis of DFSP is usually made by histopathologic and clinical findings. CONCLUSIONS: MRI is useful both for the diagnosis of DFSP and following up the patients since it has high soft tissue resolution and no risk of radiation exposure. With MRI the relation to the adjacent anatomical structures, extension and depth of the tumour and possible lymph node involvement can also be demonstrated.

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TÍTULO / TITLE: - Granulocytic Sarcoma of Parotid Gland in a 4-Year-Old Child with Subleukemic AML: A Diagnostic Challenge!

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Ingale Y; Patil T; Chaudhari P; Routray S; Agrawal M

INSTITUCIÓN / INSTITUTION: - Department of Dentistry, Yashwantrao Chavan Memorial Hospital, Pimpri, Pune 411018, India.

RESUMEN / SUMMARY: - A 4-year-old male child presented to our outpatient department with large swelling in the parotid region. Routine investigations were all within normal limits, and evaluation of complete blood count was normal except for anaemia. Excisional biopsy as a therapeutic diagnosis was done. Microscopic examination showed monomorphic population of discohesive, hyperchromatic small round cells having high N : C ratio, coarse chromatin, conspicuous nucleoli, and sometimes angulated nuclei lying in sheets. Immunohistochemistry was done to rule
out possible differential diagnosis. Fine needle aspiration from the swelling showed predominant population of blast cells. Myeloperoxidase and PBO were strongly positive, and diagnosis of granulocytic sarcoma was confirmed.

[663]

**TÍTULO / TITLE:** Paraneoplastic autoimmunity associated with testicular myeloid sarcoma and chronic myelomonocytic leukemia.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Craig JW; Lin RJ

**INSTITUCIÓN / INSTITUTION:** Weill Cornell/Rockefeller/Sloan-Kettering Tri-Institutional MD-PhD Program, 1300 York Avenue, Room C-103, New York, NY 10065, USA.

**RESUMEN / SUMMARY:** Myeloid sarcomas are rare extramedullary solid tumors composed of immature myeloid cells. The clinical presentations of these malignant neoplasms are highly variable, ranging from asymptomatic to localized mass effect. Here, we report an unusual case of myeloid sarcoma of the testis found in association with chronic myelomonocytic leukemia where the presenting symptoms were autoimmune pericarditis and migratory arthralgias and myalgias that preceded testicular enlargement by nearly three months. Treatment with both radical orchiectomy and leukemia-directed chemotherapy led to immediate reductions in symptom severity, suggesting that these early symptoms were paraneoplastic in origin. Review of the literature identified the association between hematological malignancies, including chronic myelomonocytic leukemia, and paraneoplastic autoimmune phenomena with features similar to polymyalgia rheumatica and rheumatoid arthritis. Importantly, rheumatologic symptoms related to these disease entities may be easily dismissed as vague or unrelated complaints or treated as purely rheumatologic conditions, thus delaying the formal diagnoses. Clinicians must recognize the common association between possible paraneoplastic rheumatologic symptoms and hematologic malignancies such as chronic myelomonocytic leukemia.

[664]

**TÍTULO / TITLE:** Elastofibroma dorsi management and outcomes: review of 16 cases.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Karakurt O; Kaplan T; Gunal N; Gulbahar G; Kocer B; Han S; Dural K; Sakinci U

**INSTITUCIÓN / INSTITUTION:** Department of Thoracic Surgery, Ankara Numune Teaching and Research Hospital, Ankara, Turkey.

**RESUMEN / SUMMARY:** OBJECTIVES: Elastofibroma dorsi (ED) is a rare, benign lesion arising from connective tissue, usually found at the inferior pole of the scapula. To date, only a few small series have been reported in the English literature and there are few data about the long-term outcomes after surgery. Our goal is to contribute a better understanding of this tumour and to determine the long-term outcomes after surgery. METHODS: Sixteen patients with a diagnosis of ED were identified from the unit’s database. The clinical presentation, diagnosis, pathological evidences and long-
term outcomes were evaluated. RESULTS: There were 11 females and 5 males with a mean age of 61.1 years (range 38-78 years). The tumour was located on the right in 5 (31.2%) patients, on the left in 6 (37.5%) patients and bilaterally in 5 (31.2%). Six patients had painful scapular swelling resulting in restriction of movement of the shoulder whereas 10 reported only painful scapular mass. All 16 patients underwent complete resections. The tumour size ranged from 3 to 15 cm. The mean hospital stay was 3.1 +/- 1.4 days with a morbidity of 18.75% (seroma observed in 3 patients). The mean follow-up was 58.4 +/- 29.5 months (range 11-92 months). In 2 patients (12.5%) a new occurrence on the contralateral side was observed at the follow-up.

CONCLUSIONS: Elastofibroma dorsi is a rare, ill-defined, pseudotumoural lesion of the soft tissues. Surgical treatment can be proposed if the lesion is symptomatic. Furthermore, at the follow-up, the possibility of new occurrences on the contralateral side should be kept in mind.

[665]
**TÍTULO / TITLE:** - What is your diagnosis? Disseminated histiocytic sarcoma.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Thakkar P; Pease AP; Guiot LP; Fitzgerald SD; Adu Addai B; Smedley RC
**INSTITUCIÓN / INSTITUTION:** - Department of Small Animal Clinical Sciences, College of Veterinary Medicine, Michigan State University, Lansing, MI 48910.

[666]
**TÍTULO / TITLE:** - CD133 and Ki-67 expression is associated with gastrointestinal stromal tumor prognosis.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
**AUTORES / AUTHORS:** - Lu C; Liu L; Wu X; Xu W
**INSTITUCIÓN / INSTITUTION:** - Department of General Surgery, Chinese People’s Liberation Army General Hospital, Beijing 100853, P.R. China.
**RESUMEN / SUMMARY:** - CD133+ tumor cells have a greater potential ability for tumorigenesis, proliferation, invasion and metastasis compared with CD133- tumor cells. Ki-67 is associated with cell proliferation in various tumors and has a markedly positive correlation with the prognosis of patients. However, there are a limited number of studies that have investigated the association between the prognosis of gastrointestinal stromal tumors (GISTs) and the two markers. The present study aimed to investigate CD133 and Ki-67 expression in GISTs and to explore their clinicopathological significance in the prognosis of patients with GISTs. A total of 111 GIST patients from the Chinese People’s Liberation Army (PLA) General Hospital were retrospectively followed up and immunohistochemistry was used to detect CD133, Ki-67 and CD117 expression in the tumor samples. The survival rates of the patients were analyzed using the Kaplan-Meier method. The log-rank test, chi² test and Cox’s proportional hazards model were used to determine the association between CD133,
Ki-67, CD117 expression and the prognosis of GIST. The 1-, 3- and 5-year survival rates were 93.0, 89.0 and 82.0%, respectively, in all the patients. However, in the patients with CD133+ or Ki-67+, the 1-, 3- and 5-year survival rates were 81.0, 61.5 and 50.0% and 83.0, 66.6 and 53.0%, respectively. Compared with the negative groups, the survival rates in the positive groups were statistically lower (CD133 log-rank, P=0.028; Ki-67 log-rank, P=0.002). The multivariate Cox analysis revealed that CD133 and Ki-67 expression were considerable factors in the prognosis of GIST patients (CD117, P=0.495; CD133, P=0.036; Ki-67, P=0.003). In conclusion, the positive expression of CD133 and Ki-67 was associated with a poor prognosis of GIST.

[667]  
**TITULO / TITLE:** - Canonical Wnt signaling activates miR-34 expression during osteoblastic differentiation.  
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary  
- Enlace al texto completo (gratuito o de pago) [3892/mmr.2013.1713](3892/mmr.2013.1713)  
**AUTORES / AUTHORS:** - Tamura M; Uyama M; Sugiyama Y; Sato M  
**INSTITUCIÓN / INSTITUTION:** - Department of Biochemistry and Molecular Biology, Graduate School of Dental Medicine, Hokkaido University, Kitaku, Sapporo 060-8586, Japan.  
**RESUMEN / SUMMARY:** - The canonical Wnt signaling pathway is crucial for the regulation of bone mass in humans and for the development of osteoblasts. MicroRNAs (miRs) represent a class of noncoding RNAs, ~22 nucleotides in length, that regulate gene expression by targeting mRNAs for cleavage or translational repression. Several previous studies have demonstrated the involvement of miRNAs in modulating gene expression in osteoblasts and regulating osteoblast differentiation. In the present study, microRNA profiling was conducted using Wnt3aC2C12 cells; C2C12 cells were transfected with a Wnt3a expression plasmid to activate canonical Wnt signaling. miR34b5p and miR34c were identified to be upregulated by the activation of canonical Wnt signaling in C2C12 cells. Expression of mature miR34b/c increased from low levels at day 0 to maximum levels at day 28 of MC3T3E1 cell differentiation. To analyze the effects of these miRNAs on osteoblast differentiation, an antisense inhibitor was transfected into MC3T3E1 cells and osteoblastrelated gene expression was investigated. Knockdown of miR34b/c enhanced osteocalcin mRNA expression; however, alkaline phosphatase mRNA expression and activity were decreased by miR34b/c inhibition. These results indicated that miR34b/c regulates gene expression by targeting regulators of the osteogenic pathways and thereby contributes to osteoblast differentiation.

[668]  
**TITULO / TITLE:** - Expression of hypoxia-related markers in inflammatory myofibroblastic tumors of the head and neck.  
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary  
- Enlace al texto completo (gratuito o de pago) [1186/1477-7819-11-294](1186/1477-7819-11-294)
RESUMEN / SUMMARY: - BACKGROUND: The etiology of inflammatory myofibroblastic tumors (IMTs) is controversial and the prognosis is unpredictable. Previous studies have not investigated the expression of hypoxia-related markers in IMTs. METHODS: Between 2002 and 2012, 12 consecutive patients with histologically proven IMTs were enrolled in the study. Immunohistochemistry was used to detect GLUT-1, HIF-1alpha, PI3K, and p-Akt expression in paraffin-embedded tumor specimens. Associations among GLUT-1, HIF-1alpha, PI3K, and p-Akt protein expression and clinical parameters were investigated. RESULTS: The mean duration of follow-up was 52.1 months (range, 11 to 132 months). Six patients had local recurrence. GLUT-1, HIF-1alpha, PI3K, and p-Akt expression were detected in 41.7%, 50.0%, 33.3%, and 41.7% of patients, respectively. Fisher’s exact test revealed significant correlations between recurrence of IMT and PI3K expression (P = 0.01) and p-Akt expression (P = 0.015). Univariate analyses revealed significant correlations between survival and GLUT-1 expression (P = 0.028), PI3K expression (P = 0.006), and p-Akt expression (P = 0.028). Multivariate analysis did not show a significant relationship between survival and GLUT-1, HIF-1alpha, PI3K, or p-Akt. Spearman rank correlation analysis showed significant correlations between HIF-1alpha and PI3K expression (r = 0.707, P = 0.01) and between p-Akt and PI3K expression (r = 0.837, P = 0.001). CONCLUSIONS: Although our results are inconclusive owing to the small sample size, they suggest that PI3K and p-Akt expression may play a role in the recurrence of IMTs of the head and neck.

TÍTULO / TITLE: - Diagnostic dilemma: late presentation of amelanotic BRAF-negative metastatic malignant melanoma resembling clear cell sarcoma—a case report.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Wagner BP; Epperla N; Medina-Flores R

RESUMEN / SUMMARY: - Clear cell sarcoma is a rare cancer primarily of tendons, fascia, and aponeuroses that can be difficult to discern from primary cutaneous malignant melanoma. The two cancers share several histological markers, with most cases of both cancers staining positively for S-100, HMB-45, and melanin. Primary therapy of both cancers involves wide local excision, but while systemic therapy has proven benefit for malignant melanoma, it has not been established for clear cell sarcoma. We report the case of a 58 year old woman with a large, ulcerated, fungating mass on her left lower leg. Frozen section of the mass showed a malignant epithelioid and spindle cell tumor confined to the subcutaneous tissue. A provisional diagnosis of soft-tissue sarcoma was made. Through in-depth study of initial biopsy with immunohistochemistry for S-100, HMB-45, MART-1, and MITF, along with karyotyping and FISH analysis for EWS gene rearrangement, the diagnosis of amelanotic malignant melanoma was confirmed. The patient then underwent systemic treatment with ipilimumab upon recurrence with good response. Virtual slides: The virtual slide(s) for this article can be found here:

TÍTULO / TITLE: Diagnostic pitfalls in the preoperative F-FDG PET/CT evaluation of a case of giant malignant solitary fibrous tumor of the pleura.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Lococo F; Rapicetta C; Ricchetti T; Cavazza A; Filice A; Treglia G; Tenconi S; Paci M; Sgarbi G
INSTITUCIÓN / INSTITUTION: Unit of Thoracic Surgery, IRCCS Arcispedale Santa Maria Nuova, Reggio Emilia, Italy. Electronic address: filippo_lococo@yahoo.it.

RESUMEN / SUMMARY: Solitary fibrous tumor of the pleura (SFTP) is an uncommon entity, generally with an indolent behavior. Nevertheless, some malignant forms have been rarely reported. These, often have an aggressive biological behavior with pathological findings of invasiveness. The preoperative diagnosis and evaluation of the grade of malignancy are extremely challenging. Herein we report a case of a 64-year-old man who presented with a left giant intra-thoracic mass imaged with fluorine-18 fluordeoxyglucose positron emission tomography/computed tomography (18F-FDG/PET-CT) and sampled via fine-needle aspiration biopsy (FNAB). Imaging and FNAB findings showed suspicion of a benign form of SFTP. Surgical radical resection of the giant mass was performed. The definitive histological diagnosis showed a malignant SFTP. Based on this report, we take the opportunity to briefly discuss the insidious pitfalls concerning the radiological and 18F-FDG/PET-CT features as well as cyto/histological findings in the pre-operative diagnostic work-up examination of this rare entity.

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TÍTULO / TITLE: Rhabdomyosarcoma of prostate presenting as bladder outlet obstruction in a young adult.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Prabhakaran P; Sanjayan R; Somanathan T; Narayanan G
INSTITUCIÓN / INSTITUTION: Department of Medical Oncology, Regional Cancer Centre, Thiruvananthapuram, 695011 Kerala, India.

RESUMEN / SUMMARY: A 19-year-old boy presented with bladder outlet obstruction and on evaluation was found to have prostatomegaly, which on biopsy was diagnostic of embryonal rhabdomyosarcoma (RMS). He had pulmonary metastasis and received chemotherapy with cyclophosphamide, doxorubicin, vincristine, actinomycin D, and radical radiotherapy. At one year, his prostatic tumour has resolved completely. Embryonal RMS of prostate occurs more commonly in infancy and childhood, so occurrence in young adults is rare, and a high index of suspicion is essential for early diagnosis and treatment.
A 27-year-old man who presented with a palpable abdominal mass was diagnosed as having testicular cancer with multiple liver and lung metastases. At 16 months after chemotherapy, a follow-up computed tomographic scan revealed a supraclavicular mass measuring 3 cm in size, which was suspected to be a recurrence. The patient underwent surgical excision, and the mass was pathologically diagnosed as a AFH. The patient has had no local recurrence and no distant metastasis for 12 months after resection. To the best of our knowledge, this is the first case report of AFH as a second tumor in a patient with testicular cancer.

Primary pleural synovial sarcoma: A rare cause of hemorrhagic pleural effusion in a young adult.

This is a case report of a young adult presenting with hemorrhagic pleural effusion. Chest CT scan showed loculated pleural effusion with pleural nodule. Whole body PET scan showed thickening of pleura with multiple enhancing pleural nodules with different metabolic activity. Pleural nodule was biopsied which on histopathology showed pleural synovial sarcoma.

Leiomyoma: A rare tumor in the head and neck and oral cavity: Report of 3 cases with review.
Leiomyomas are benign tumors arising from smooth muscle, most commonly seen in uterine myometrium, gastrointestinal tract, skin and lower extremities of middle-aged women. Leiomyomas are uncommon in the oral cavity with reported incidence of 0.065%, which accounts for 0.42% of all soft-tissue neoplasms in the oral cavity. Leiomyomas of head and neck region account for less than 1% of all leiomyomas. The most common site of leiomyoma in the head and neck region is the lips (27.46%) followed by tongue (18.30%), cheeks and palate (15.49%), gingiva (8.45%) and mandible (5.63%). The purpose of this article is to present three cases of leiomyoma comprising of an intraoral vascular leiomyoma and two solid leiomyomas in the head and neck region. The clinical features, etiology, differential diagnosis and treatment of leiomyoma are discussed with review of the literature.

AUTORES / AUTHORS: - Takahashi H; Nakayama R; Hayashi S; Nemoto T; Murase Y; Nomura K; Takahashi T; Kubo K; Marui S; Yasuhara K; Nakamura T; Sueo T; Takahashi A; Tsutsumiuchi K; Ohta A; Kawai A; Sugita S; Yamamoto S; Kobayashi T; Honda H; Yoshida T; Hasegawa T

INSTITUCIÓN / INSTITUTION: - Graduate School of Horticulture, Chiba University, Matsudo, Chiba, Japan; Graduate School of Bioscience and Biotechnology, Chubu University, Kasugai, Aichi, Japan; Plant Biology Research Center, Chubu University, Kasugai, Aichi, Japan; Division of Genetics, National Cancer Center Research Institute, Tokyo, Japan.

RESUMEN / SUMMARY: - The diagnosis and treatment of soft tissue sarcomas (STSs) has been particularly difficult, because STSs are a group of highly heterogeneous tumors in terms of histopathology, histological grade, and primary site. Recent advances in genome technologies have provided an excellent opportunity to determine the complete biological characteristics of neoplastic tissues, resulting in improved diagnosis, treatment selection, and investigation of therapeutic targets. We had previously developed a novel bioinformatics method for marker gene selection and applied this method to gene expression data from STS patients. This previous analysis revealed that the extracted gene combination of macrophage migration inhibitory factor (MIF) and stearoyl-CoA desaturase 1 (SCD1) is an effective diagnostic marker to discriminate between subtypes of STSs with highly different outcomes. In the present study, we hypothesize that the combination of MIF and SCD1 is also a prognostic marker for the overall outcome of STSs. To prove this hypothesis, we first analyzed microarray data from 88 STS patients and their outcomes. Our results show that the survival rates for MIF- and SCD1-positive groups were lower than those for negative groups, and the p values of the log-rank test are 0.0146 and 0.00606, respectively. In addition, survival rates are more significantly different (p = 0.000116) between groups that are double-positive and double-negative for MIF and SCD1. Furthermore, in vitro cell growth inhibition experiments by MIF and SCD1 inhibitors support the hypothesis. These results suggest that the gene set is useful as a prognostic marker associated with tumor progression.

[678]

TÍTULO / TITLE: - Testicular yolk sac tumor of myxomatous, reticular, and polyvesicular vitelline type in a newborn calf.

RESUMEN / SUMMARY: - Testicular yolk sac tumor of myxomatous, reticular, and polyvesicular vitelline type in a newborn calf.


AUTORES / AUTHORS: - Sakaguchi K; Matsuda K; Suzuki H; Yamamoto N; Kondo Y; Ando T; Koizumi M; Kagawa Y; Taniyama H

INSTITUCIÓN / INSTITUTION: - 1Kazuya Matsuda, Department of Veterinary Pathology, School of Veterinary Medicine, Rakuno Gakuen University, 582 Bunkyodai-Midorimachi, Ebetsu, Hokkaido 069-8501, Japan. kmatsuda@rakuno.ac.jp.

RESUMEN / SUMMARY: - Yolk sac tumors (YSTs) are rare neoplasms of germ cell origin. In humans, the tumors primarily occur in the testes or ovaries, but occasionally...
develop at other sites. The neoplastic cells of YSTs form many histological patterns resembling embryonal structures, and the World Health Organization classification lists 11 such patterns: reticular, macrocystic, endodermal sinus, papillary, solid, glandular-alveolar, myxomatous, sarcomatoid, polyvesicular vitelline, hepatoid, and parietal. Among domestic animals, only 2 cases of YST, which were of testicular and abdominal cavity origin, have been reported in calves. In both cases, neoplastic cells had epithelial properties and disseminated metastases in the abdomen. In the present study, the enlarged testis of a newborn calf, which was subsequently diagnosed as YST and exhibited myxomatous, reticular, and polyvesicular vitelline histological patterns, is described. There was no metastasis in this case, and histological and immunohistochemical features varied from previous cases of YST.

**AUTORES / AUTHORS:** Jia C; Zhao W; Dai C; Wang X; Bu X; Peng S; Xu F; Xu Y; Zhao Y

**INSTITUCIÓN / INSTITUTION:** Department of General Surgery, Shengjing Hospital, China Medical University, Shenyang 110004 Liaoning Province, P R China. daicl_sj@163.com

**RESUMEN / SUMMARY:** Adult primary undifferentiated embryonal sarcoma of the liver (UESL) is a rare disease. While the etiology of UESL remains largely unknown, association with systemic inflammatory disorders has been observed. Here, we report a case of UESL in a 46-year-old woman with systemic lupus erythematosus (SLE) and without chronic hepatitis or liver cirrhosis. Systematic review of the publicly available English language medical literature identified only 27 cases of UESL in patients aged >45 years and none with SLE. Our patient presented with abdominal pain and had a 2-year history of SLE. Abdominal ultrasonography and enhanced computed tomography revealed a solid mass in the right lobe of the liver. Presumptive diagnosis of atypical hepatocellular carcinoma was made and the patient was treated with segmentectomy of S5 and S4a and cholecystectomy. The final diagnosis of UESL was made according to the pathology results. Since SLE patients may be at increased risk of malignancy, it is possible that the SLE pathogenesis may have contributed to the development of UESL in our patient. According to this case, UESL should be considered when SLE patients present with hepatic space-occupying lesions.

[682]

**TÍTULO / TITLE:** A young woman with a giant breast fibrosarcoma: a case report.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Jiao Q; Wu A; Liu P; Tang J; Yang M; Fan X; Zheng L

**INSTITUCIÓN / INSTITUTION:** Department of General Surgery, Zhujiang Hospital, Southern Medical University, Guangzhou 510515, China;

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**RESUMEN / SUMMARY:** We report a case of a 15-year-old female, no family history of huge fibrosarcoma. Computed tomography (CT) showed that there was no clearance between the lump and pectoralis major and that there were pathological fractures in the third and fourth ribs. Fine-needle aspiration result suggested that it might be a phyllodes tumor of the breast. According to the postoperative pathologic and immunohistochemical results, the final diagnosis was breast fibrosarcoma.

[683]

**TÍTULO / TITLE:** Adenocarcinoma of the Ileocolic Junction and Multifocal Hepatic Sarcomas in an Aged Rhesus Macaque (Macaca mulatta).

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Lang CD; Daviau JS; Merton DA; Caraker SM

**INSTITUCIÓN / INSTITUTION:** Laboratory Animal Services, Thomas Jefferson University, Philadelphia, Pennsylvania, USA. cynthia.lang@jefferson.edu
RESUMEN / SUMMARY: - An aged male rhesus macaque in our colony had decreased appetite and a loss of interest in behavioral testing. CBC analysis revealed a regenerative, microcytic, hypochromic anemia with thrombocytosis, consistent with iron deficiency. A fecal occult blood test was positive. Ultrasound imaging revealed numerous, vascularized focal liver lesions that suggested metastases. The macaque’s appetite continued to decrease, and he became more lethargic. At this point, the investigator elected to euthanize the macaque. At necropsy, the ileocolic junction was white and abnormally thickened, and the liver was pale tan with approximately 18 discrete white masses randomly scattered throughout the hepatic parenchyma. Histologically, the mass at the ileocolic junction was identified as an intestinal adenocarcinoma, whereas the liver masses were confirmed to be undifferentiated hepatic sarcomas. This case report describes a rhesus macaque that had 2 unrelated primary neoplasms. A review of the literature indicates that this rhesus macaque is the first reported to have an adenocarcinoma of the ileocolic junction and multiple hepatic sarcomas simultaneously.

[684]
TÍTULO / TITLE: - Gingival Rhabdomyosarcoma in an Adult: A Unique Entity.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Sahni P; Singhvi A; Nayak MT; Deora SS
INSTITUCIÓN / INSTITUTION: - Department of Oral & Maxillofacial Pathology, Vyas Dental College & Hospital, Jodhpur, RAJASTHAN, INDIA.
RESUMEN / SUMMARY: - Rhabdomyosarcoma is a disease that predominantly affects children. Approximately 40 per cent are located in the head and neck region but it is rare in the oral cavity. This article describes an interesting case of an embryonal rhabdomyosarcoma in a 36-year-old male, involving the mandibular gingiva. The lesion showed radiolucency with ill-defined margins that was crossing the midline. The history revealed a similar lesion six months back at the same site and the lesion had been completely excised. The biopsy reports confirmed the diagnosis of embryonal rhabdomyosarcoma after which en-bloc resection of the tumor was performed with administration of chemotherapy and radiotherapy. Due to high recurrence rate of rhabdomyosarcomas in adults, multimodal therapy should be planned for proper care of the patient. Clinical, radiological, histopathological and management aspects are discussed here.

[685]
TÍTULO / TITLE: - Kaposiform Lymphangiomatosis, a Newly Characterized Vascular Anomaly Presenting with Hemothysis in an Adult Woman.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Safi F; Gupta A; Adams D; Anandan V; McCormack FX; Assaly R
Disorders of the pulmonary lymphatic system include macro and microcystic lymphatic malformations, primary or secondary lymphangiectasias, generalized lymphatic anomalies (GLA), diffuse pulmonary lymphangiomatosis (DPL), and combinations of lymphatic and other tissue anomalies including lymphangioleiomyomatosis (LAM). We report a case of a patient with a newly defined entity classified as kaposiform lymphangiomatosis (KLA). A 50 year old nonsmoking Hispanic female presented with a 20 year history of cough, hemoptysis, chyloptysis, and pleuritic chest pain. Laboratory evaluation demonstrated a low normal platelet count, elevated D-Dimer, low normal fibrinogen and elevated fibrin split products. Chest computerized tomography scan showed enlarged hypodense lymph nodes in the mediastinum and hila, and peribronchovascular thickening, without evidence of cystic parenchymal lesions. MRI of the chest showed a cystic, septated appearance of the mediastinal lymph nodes with heterogeneously increased T2 and decreased T1 signal intensity. Fiberoptic bronchoscopy revealed hyperemic mucosa with granular appearance secondary to submucosal infiltrative process. Pathological specimens revealed dilated, malformed lymphatic channels within the pleura, pulmonary septa, and bronchovascular bundles, and foci of perilymphatic and intralymphatic spindle cells which reacted with the PROX-1 immunostain. The morphology and immunohistochemistry results were consistent with a diagnosis of KLA. KLA is a recently described entity amongst lymphatic anomalies. Clinically, patients have a generalized dysplastic lymphatic process with involvement of the mediastinum, lungs, retroperitoneum, spleen, bones, soft tissue, and skin. Mortality is associated with progressive pulmonary disease and serosal hemorrhage. Medical treatments reported include Sirolimus, and anti-angiogenic agents. Our patient received Sirolimus and will be followed in our clinic.
Angiosarcomas are malignant tumours of endovascular origin. They are rare tumours accounting for 0.04-1% of all breast malignancies. Two different forms are described: primary, occurring in young women, and secondary angiosarcoma, which occurs in older women with a history of breast-conserving surgery and radiation therapy. Imaging findings on mammography and ultrasound are non-specific, but magnetic resonance imaging with dynamic contrast enhancement is more informative. We present two cases - one of primary and one of secondary angiosarcoma - and review the imaging findings.

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Zinc chelation: a metallothionein 2A's mechanism of action involved in osteosarcoma cell death and chemotherapy resistance.

Osteosarcoma is the most common primary tumor of bone occurring in children and adolescents. The histological response to chemotherapy represents a key clinical factor related to survival. We previously showed that statins exhibit antitumor effects in vitro, inducing apoptotic cell death, reducing cell migration and invasion capacities and strengthening cytotoxic effects in combination with standard drugs. Comparative transcriptomic analysis between control and statin-treated cells revealed strong expression of several genes, including metallothionein (MT) 2A. MT2A overexpression by lentiviral transduction reduced bioavailable zinc levels, an effect associated with reduced osteosarcoma cell viability and enhanced cell differentiation. In contrast, MT2A silencing did not modify cell viability but strongly inhibited expression of osteoblastic markers and differentiation process. MT2A overexpression induced chemoresistance to cytotoxic drugs through direct chelation of platinum-containing drugs and indirect action on p53 zinc-dependent activity. In contrast, abrogation of MT2A enhanced cytotoxic action of chemotherapeutic drugs on osteosarcoma cells. Finally, clinical samples derived from chemonaive biopsies revealed that tumor cells expressing low MT2A levels correspond to good prognostic (good responder patients with longer survival rate), whereas high MT2A levels were associated with adverse prognosis (poor responder patients). Taken together, these data show that MT2A contributes to chemotherapy resistance in osteosarcoma, an
effect partially mediated by zinc chelation. The data also suggest that MT2A may be a potential new prognostic marker for osteosarcoma sensitivity to chemotherapy.

[689]
**TITULO / TITLE:** Endoscopic resection of giant colonic lipoma: case series with partial resection.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Kim GW; Kwon CI; Song SH; Jin SM; Kim KH; Moon JH; Hong SP; Park PW

**INSTITUCIÓN / INSTITUTION:** Digestive Disease Center, CHA Bundang Medical Center, CHA University, Seongnam, Korea.

**RESUMEN / SUMMARY:** Colonic lipoma, a very rare form of benign tumor, is typically detected incidentally in asymptomatic patients. The size of lipoma is reported variously from 2 mm to 30 cm, with higher likelihood of symptoms as the size is bigger. Cases with symptom or bigger lesion are surgically resected in principle; endoscopic resection, which has developed recently with groundbreaking advance of endoscopic excision technology, is being used more often but with rare report of success due to high chance of complications such as bowel perforation or bleeding. The authors report here, together with a literature review, our experiences of three cases of giant colonic lipomas showing complete remission after aggressive unroofing technique, at certain intervals, using snare catheter at the origin of the lipoma so that the remaining lipoma could be drained out of the exposed surface spontaneously, in order to reduce complications.

[690]
**TITULO / TITLE:** Extracapsular wide resection of a femoral neck osteosarcoma and its reconstruction using a pasteurized autograft-prosthesis composite: A case report.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Yasuda T; Hori T; Suzuki K; Hachinoda J; Matsushita I; Ito Y; Kanamori M; Kimura T

**INSTITUCIÓN / INSTITUTION:** Department of Orthopaedic Surgery, Faculty of Medicine, University of Toyama, Toyama 930-0194, Japan.

**RESUMEN / SUMMARY:** The requirement for an extracapsular resection is indicated for malignant bone tumors that have disseminated intracapsularly. Extracapsular resections are often performed for malignant tumors arising from the knee joint, but there are relatively few studies that have described an extracapsular resection of a tumor arising from the hip joint. The present study describes a case of extracapsular wide resection of the hip joint using rotational acetabular osteotomy. The patient was a 17-year-old female and the diagnosis was an osteoblastic osteosarcoma with a pathological fracture of the femoral neck. The joint was reconstructed using an allograft-implant composite graft and total hip arthroplasty. Although the patient presented a slight Trendelenburg gait, no recurrence or metastases were identified.
during a follow-up period of 3 years. The clinical features and surgical procedure of the case are described.

[691]
Título / Title: Detection of intravenous leiomyomatosis with intracardiac extension by ultrasonography: A case report.
Resumen / Summary: Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o de pago) 3892/ol.2013.1387
Autores / Authors: Liu W; Liu M; Xue J
Institución / Institution: Department of Gynecology, Shandong Provincial Hospital Affiliated to Shandong University, P.R. China.
Resumen / Summary: Intravenous leiomyomatosis (IVL) is characterized by histologically benign tumors that exhibit aggressive clinical behavior. On rare occasions, the tumors may extend into the regional and systemic veins, thus reaching the heart. This may subsequently cause intracardiac leiomyomatosis (ICL), which may lead to congestive heart failure and occasionally, sudden fatalities. Due to its rarity and diffuse symptoms, the misdiagnosis of ICL is common and as a result, the condition may be under-reported. The present study reports a 33-year-old female who was admitted to Shandong Provincial Hospital Affiliated to Shandong University for myomectomy due to a rapidly growing myoma of the uterus. In routine pre-operative abdominal ultrasonography, a moderately sized echoic mass in the right internal iliac vein was observed, which extended to the common iliac vein, the inferior vena cava and the orifice of the right atrium. A presumptive diagnosis of ICL was made. The patient underwent a well-prepared one-stage thoraco-abdominal surgical procedure and the pathological report confirmed ICL. This case illustrates that the early detection of ICL may prevent a potential emergency situation and abdominal ultrasonography may be considered a useful tool in the diagnosis of ICL.

[692]
Título / Title: Cotyledonoid Dissecting Leiomyoma of the Uterus with Intravascular Luminal Growth: A Case Study.
Resumen / Summary: Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o de pago) 4132/KoreanJPathol.2013.47.5.477
Autores / Authors: Kim NR; Park CY; Cho HY
Institución / Institution: Department of Pathology, Gachon University Gil Medical Center, Incheon, Korea.
Resumen / Summary: Here, we report the case of a 43-year-old female who was diagnosed with a cotyledonoid dissecting leiomyoma (CDL) of the uterus. CDL is a recently described and extremely rare variant of a benign leiomyoma that can grossly masquerade as a malignancy. The 13-cm sized tumor was located primarily on the extraterine surface as an intrauterine continuity, which showed dark red, congested, bulbous protuberances. It was multinodular appearance, encasing the bilateral adnexae and the left iliac vein. Microscopically, the nodules were separated by extensive hydropic degeneration. The nodules were composed of cigar-shaped spindle cells with no mitosis, cellular pleomorphism or coagulation necrosis. They also showed
an intravascular luminal growth pattern. CDL with intravascular growth was diagnosed after excluding intravascular leiomyomatosis, disseminated peritoneal leiomyomatosis, and benign metastasizing leiomyoma. The present case is the second reported case of CDL in Korea. Recognition of this rare and bizarre, malignancy-mimicking leiomyoma is crucial to prevent inappropriate treatment.
review for correct diagnosis and discusses the successful management of a recurrent forearm lesion.

[695]
**TITULO / TITLE:** - Inflammatory myofibroblastic tumor of the kidney with viral hepatitis B and trauma: A case report.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Li Z; Wang W; Wang Y; Zhai X; Tian Y; Fu Y; Zhou H

**INSTITUCIÓN / INSTITUTION:** - Center of Urology, First Hospital, Jilin University, Changchun, Jilin 130021, P.R. China.

**RESUMEN / SUMMARY:** - Inflammatory myofibroblastic tumor (IMT) is a rare entity that most commonly involves the lung. However, an IMT of the kidney is extremely rare. The etiology and pathogenesis of IMT remain unknown. The present study describes the case of a 48-year-old female who presented asymptptomatically. Imaging investigations revealed a mass in the left kidney and a pathological examination of the nephrectomy specimen revealed an IMT. The patient had a history of trauma in the left hypochondrium 13 years previously and a history of hepatitis B for 20 years. The latter developed into hepatic cirrhosis, hypersplenism and coagulation disorders, which may have played a significant role in the development of the IMT of the kidney in the present case and also may aid in improving the understanding of the etiology and pathogenesis of IMT of the kidney.

[696]
**TITULO / TITLE:** - IGF signaling pathway analysis of osteosarcomas reveals the prognostic value of pAKT localization.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - van de Luijtgaarden AC; Roeffen MH; Leus MA; Flucke UE; Schreuder BH; van der Graaf WT; Versleijen-Jonkers YM

**INSTITUCIÓN / INSTITUTION:** - Radboud University Medical Centre, Department of Medical Oncology, Internal Postal Code 452, PO Box 9101, 6500 HB Nijmegen, The Netherlands.

**RESUMEN / SUMMARY:** - Aim: The aim of this study was to examine the expression of the IGF signaling pathway components in osteosarcoma samples before and after chemotherapy with special emphasis on their prognostic value. Materials & methods: Tumor material and follow-up data of 58 osteosarcoma patients were analyzed. Immunohistochemical staining was carried out to identify proteins related to the IGF pathway. Changes in protein expression during treatment, correlations between proteins and subsequent influence on survival were tested. Results: Proteins of the IGF signaling system are widely expressed in osteosarcoma samples. We demonstrate a change in expression of intracellular pathway proteins after chemotherapy. Remarkably, cytoplasmic pAKT, but not nuclear pAKT, is associated with poor survival. Conclusion: IGF pathway proteins seem to be widely activated in osteosarcoma, but their expression changes after chemotherapy. This has implications for the timing of
both measuring target expression and pathway interference. Our observations on the prognostic value of cytoplasmic pAKT warrant further investigation while considering the introduction of AKT inhibitors for osteosarcoma treatment.

[697]

TÍTULO / TITLE: Dedifferentiated liposarcoma and pleomorphic liposarcoma: A comparative study of cytomorphology and MDM2/CDK4 expression on fine-needle aspiration.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Marino-Enriquez A; Hornick JL; Dal Cin P; Cibas ES; Qian X

INSTITUCIÓN / INSTITUTION: Department of Pathology, Brigham and Women's Hospital and Harvard Medical School, Boston, Massachusetts.

RESUMEN / SUMMARY: BACKGROUND: Dedifferentiated liposarcoma (DDLPS) and pleomorphic liposarcoma (PLPS) are distinct high-grade liposarcomas. DDLPS is a nonlipogenic sarcoma characterized by amplification of MDM2 and CDK4. PLPS is a high-grade sarcoma containing lipoblasts, characterized by a complex karyotype and a more aggressive clinical course. Rarely, DDLPS shows lipogenic differentiation, mimicking PLPS. The cytomorphologic features of DDLPS and PLPS and the utility of ancillary studies have not been systemically analyzed. METHODS: Cytologic preparations of 25 DDLPS and 13 PLPS, all histologically confirmed, were retrospectively reviewed along with clinical and cytogenetic data. Sample cellularity, vascular architecture, background material, predominant cell morphology, quality of the cytoplasm, and nuclear pleomorphism were compared for both tumor types. Immunohistochemistry for MDM2 and CDK4 was performed on cell blocks and/or core needle biopsies. RESULTS: Fine-needle aspirate smears from both DDLPS and PLPS were variably cellular, composed of cellular clusters and noncohesive cells. Abundant myxoid stroma was present in approximately 25% of DDLPS and PLPS cases, whereas branching curvilinear vessels were more common in DDLPS than in PLPS (7 of 25 versus 2 of 13). Tumors were composed of predominantly spindled (18 of 25 DDLPS versus 3 of 13 PLPS) or epithelioid cells (7 of 25 DDLPS versus 6 of 13 PLPS). Pleomorphic cells were predominant in 3 PLPS, and were frequent in both (13 of 25 DDLPS versus 10 of 13 PLPS). The cytoplasm was mostly fibrillary and often vacuolated in both entities. Other features included necrosis, mitoses, and a prominent inflammatory infiltrate. The main cytologic differences were the presence of marked pleomorphism, abundant lipoblasts, and cells with microvacuolated cytoplasm in most PLPS. A total of 24 (96%) and 20 (80%) cases of DDLPS expressed MDM2 and CDK4, respectively, whereas none of the PLPS expressed both markers. Six DDLPS tested showed ring or giant marker chromosomes and/or MDM2 amplification by fluorescence in situ hybridization; 2 PLPS had complex karyotypes. CONCLUSIONS: DDLPS and PLPS exhibit variable and occasionally overlapping cytologic features. The presence of lipoblasts, cells with microvacuolated cytoplasm, and marked pleomorphism are more suggestive of PLPS, but these characteristics can be present in DDLPS. Coexpression of MDM2 and CDK4 distinguishes DDLPS from PLPS. Cancer (Cancer Cytopathol) 2013. © 2013 American Cancer Society.

[698]
**TÍTULO / TITLE:** - Uterine leiomyoma with spontaneous intraleiomyoma hemorrhage, perforation, and hemoperitoneum in postmenopausal woman: Computed tomography diagnosis.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Alharbi SR

**INSTITUCIÓN / INSTITUTION:** - Department of Radiology and Medical Imaging, King Khalid University Hospital, King Saud University, Riyadh, Kingdom of Saudi Arabia.

**RESUMEN / SUMMARY:** - Spontaneous intraleiomyoma hemorrhage, perforation, and hemoperitoneum are very rare complications of uterine leiomyoma. We report a case of postmenopausal woman who presented with acute abdomen found to have intraleiomyoma hemorrhage, perforation, and hemoperitoneum. Our case also illustrates the computed tomography findings of such complications of uterine leiomyoma.

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**TÍTULO / TITLE:** - A Case of Recurrent Pulmonary Inflammatory Myofibroblastic Tumor with Aggressive Metastasis after Complete Resection.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Moon CH; Yoon JH; Kang GW; Lee SH; Baek JS; Kim SY; Kim HR; Kim CH

**INSTITUCIÓN / INSTITUTION:** - Department of Internal Medicine, Korea Cancer Center Hospital, Korea Institute of Radiological and Medical Sciences, Seoul, Korea.

**RESUMEN / SUMMARY:** - An inflammatory myofibroblastic tumor (IMT) is a rare disease entity reported to arise in various organs. It is thought to be a neoplastic or reactive inflammatory condition, controversially. The treatment of choice for myofibroblastic tumor is surgery, and recurrence is known to be rare. The optimal treatment method is not well-known for patients ineligible for surgery. We report a 47-year-old patient with aggressive recurrent IMT of the lungs. The patient had been admitted for an evaluation of back-pain two years after a complete resection of pulmonary IMT. Radiation therapy was performed for multiple bone recurrences, and the symptoms were improved. However the patient presented again with aggravated back-pain six months later. High-dose steroid and non-steroidal anti-inflammatory drugs were administered, but the disease progressed aggressively, resulting in spinal cord compression and metastasis to intra-abdominal organs. This is a very rare case of aggressively recurrent pulmonary IMT with multi-organ metastasis.

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**TÍTULO / TITLE:** - Tongue rotation for reconstruction after rostral hemiglossectomy for excision of a liposarcoma of the rostral quadrant of the tongue in a dog.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Montinaro V; Boston SE
INSTITUCIÓN / INSTITUTION: - Department of Clinical Studies, Ontario Veterinary College, University of Guelph, Guelph, Ontario N1G 2W1.

RESUMEN / SUMMARY: - A 15-year-old female beagle dog was presented for a lingual liposarcoma. Full staging of the disease did not show any evidence of metastasis. A tongue rotation for reconstruction after rostral hemiglossectomy was performed after removal of the mass. This surgical technique may be useful in tongue reconstruction after trauma or tumor excision.

[701]
TÍTULO / TITLE: - Bcl-2 correlates with localization but not outcome in human osteosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 3892/ol.2013.1395
AUTORES / AUTHORS: - Trieb K; Sulzbacher I; Kubista B
INSTITUCIÓN / INSTITUTION: - Department of Orthopaedics, Klinikum Wels-Grieskirchen, Wels A-4600, Austria.

RESUMEN / SUMMARY: - bcl-2 is a member of the bcl-2 family that inhibits apoptosis, plays a crucial role in cell viability and is expressed in various types of tumors. With respect to inconsistent results in previous studies, the aim of the present study was to generate a clear hypothesis with regards to the value of bcl-2 expression as a predictive or prognostic factor in human osteosarcoma. The expression of bcl-2 was examined immunohistochemically in 49 patients with high-grade osteosarcoma and the results were correlated with localization, histological response to chemotherapy, survival and the occurrence of metastases. In patients with osteosarcoma, 21/49 cases (43%) were positive for bcl-2 expression and the remaining cases were negative. A significantly higher expression of bcl-2 was observed in central tumors located in the pelvis (83 vs. 37% positive; P<0.05). The bcl-2 expression status revealed no statistically significant correlation with response to chemotherapy, with 57% of patients with bcl-2-positive tumors showing a good response and 43% showing a poor response. No significant difference was observed when comparing survival or occurrence in bcl-2-positive and -negative tumors. In conclusion, the results of the present study indicate that, despite higher bcl-2 expression in central osteosarcoma, the expression in high-grade osteosarcoma is not a reliable prognostic or predictive marker.

[702]
TÍTULO / TITLE: - Desmoplastic (collagenous) fibroma of the femur: A case report and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 3892/ol.2013.1535
AUTORES / AUTHORS: - Gao S; Cai Q; Yao W; Wang J; Zhang P; Wang X
INSTITUCIÓN / INSTITUTION: - Department of Orthopedics of Henan Tumor Hospital, The Affiliated Tumor Hospital of Zhengzhou University, Zhengzhou, Henan 450008, P.R. China.
RESUMEN / SUMMARY: - Desmoplastic fibroma is a rare, benign soft-tissue tumor composed of spindled and stellate-shaped cells that are embedded in a dense collagenous stroma. Clinically, desmoplastic fibroma presents as a firm, mobile, slow-growing mass that is located in the subcutaneous tissue or near the deep aspect of the skeletal muscles. The present study describes the case of a 66-year-old female who presented with an inactive, firm, slightly tender mass in the lower medial segment of the right femur. An open biopsy was performed and the result of the pathological examination indicated a desmoplastic fibroma. The patient underwent a radical resection of the tumor and the accompanying bone, which was then reimplemented using devitalized tumor bone, self-ilium graft and homologous allograft bone transplantation, with an internal fixation by locking the compression plate. This was followed by a reconstruction of the anterior and posterior cruciate ligaments and the lateral and medial collateral ligaments. There was no evidence of local recurrence at five years post-surgery.

[703]

TÍTULO / TITLE: - Primary osteosarcoma of breast, a rare case.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Dey S; Chaudhury MK; Basu SK; Manna AK; Dutta SK
INSTITUCIÓN / INSTITUTION: - Demonstrator, N R S Medical College, Kolkata, India.
RESUMEN / SUMMARY: - Mammary sarcomas are very uncommon and make up less than 1% of all primary breast malignancies. Primary osteosarcoma of the breast is extremely rare and represents 12.5% of mammary sarcomas. A secondary lesion from a primary osteosarcoma of the bone should be considered in the differential diagnosis. In addition, the absence of a direct connection between the tumour and the underlying skeleton is mandatory for the diagnosis. We report a case of primary osteosarcoma of the breast occurring in young patient with fatal evolution.

[704]

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - S Amarjit K; Budhiraja S; Chandramouleeswari K; Anita S
INSTITUCIÓN / INSTITUTION: - Professor & Head, Goverment Medical College, Amritsar, Punjab, India.
RESUMEN / SUMMARY: - Intra-articular synovial lipomas are very rare and only few cases have been reported till now. We are reporting a rare case of a unilateral intra-articular lipoma of osteoarthritic knee joint in a 62 years old male. Patient had two episodes of sudden locking of knee joint, which resolved spontaneously. A plain X-ray showed changes which were suggestive of osteoarthritis. Clinically, patient was diagnosed as a case of loose bodies in left knee joint. An arthrotomy was performed. After a Histopathological Examination (HPE) of loose bodies, a diagnosis of an intra-
Articular synovial lipoma was made. Due to wide differentials and varied clinical behaviour of loose bodies, lipoma should be included in differential diagnosis of osteoarthritic patients who complain of episodic locking of knees. Intraarticular lipomas, on arthroscopic guided excision, get cured permanently, with no recurrence. The differentiation of an intra-articular lipoma from a relatively more common entity, Lipoma arborescens, has also been discussed.

[705]
TÍTULO / TITLE: - Lipoma on palmar aspect of thumb: a rare case report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

ENLACE AL TEXTO COMPLETO (GRATUITO O DE PAGO) - 7860/JCDR/2013/5683.3261
AUTORES / AUTHORS: - Kamra HT; Munde SL
INSTITUCIÓN / INSTITUTION: - Associate Professor, Department of Pathology, Bhagat Phool Singh Government Medical College for women , Khanpur Kalan, Sonepat, Haryana, India.

[706]
TÍTULO / TITLE: - Granulocytic sarcoma presenting as an orbital mass: report of two cases.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

ENLACE AL TEXTO COMPLETO (GRATUITO O DE PAGO) - 7860/JCDR/2013/5582.3260
AUTORES / AUTHORS: - Thakur B; Varma K; Misra V; Chauhan S
INSTITUCIÓN / INSTITUTION: - Assistant Professor, Department of Pathology, SGRRIM & HS, Dehradun, India.

RESUMEN / SUMMARY: - Granulocytic sarcoma is a rare variant of a myeloid malignancy, which shows an extra-medullary tumour mass which is composed of myeloblasts and myeloid precursors with varying degrees of differentiation. It occurs most commonly in bone, periosteum, soft tissue, lymph nodes, and skin; although it can occur anywhere throughout the body. Here, we are reporting two cases of orbital granulocytic sarcoma in children, which presented clinically with proptosis and periorbital swellings, which were first diagnosed by Fine Needle Aspiration Cytology (FNAC). Later, peripheral blood and bone marrow aspirate examinations revealed the evidence of Acute Myeloid Leukaemia (AML). These cases are being documented to demonstrate the utility and diagnostic accuracy of FNAC in evaluation of this entity, in cases of unsuspected AML. Recognition of this rare entity is important, because giving an early aggressive chemotherapy can cause regression of the tumour and thus improve the patient survival.
TÍTULO / TITLE: - A case of cutaneous angiolipoleiomyoma (angiomyolipoma) in a budgerigar (Melopsittacus undulatus).
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Jakab C; Balka G; Szabara A; Csaba C; Pazar P
INSTITUCIÓN / INSTITUTION: - a Department of Pathology and Forensic Veterinary Medicine, Faculty of Veterinary Science, Szent Istvan University, Budapest, Hungary.
RESUMEN / SUMMARY: - We report a case of cutaneous angiolipoleiomyoma (angiomyolipoma) found on the anterior wall of the ventral part of the abdomen of a three-year-old female budgerigar (Melopsittacus undulatus). Histologic examination of the well-circumscribed, surgically removed tumour (1.5 cm in diameter) showed a benign admixed proliferation of blood vessels of different size, smooth muscle bundles, and mature adipose tissue, without evidence of malignancy. Endothelial cells of the haemangioma component were positive for claudin-5 endothelium-specific immunohistochemical marker, and the leiomyoma component was positive for alpha-smooth muscle actin. The differentiated lipocytes showed S-100 protein positivity. The Ki-67 labelling index was 2 to 3%. The mesenchymal tumour was negative for HMB45 melanocytic immunohistochemical marker. To the best of our knowledge, this is the first report describing a cutaneous angiolipoleiomyoma in a budgerigar with histological and immunohistochemical analyses.

TÍTULO / TITLE: - Perforated sarcomatoid carcinoma of the jejunum: Case report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Han N; Han QH; Liu YZ; Li ZC; Li J
INSTITUCIÓN / INSTITUTION: - Department of Emergency and Trauma Surgery, East Hospital Affiliated to Tongji University, Shanghai 200120, P.R. China; Department of Orthopaedics, Shanghai Tenth People’s Hospital Affiliated to Tongji University, Shanghai 200072, P.R. China.
RESUMEN / SUMMARY: - Sarcomatoid carcinomas exhibit features that are common to epithelial and mesenchymal tumors. These carcinomas are rare, particularly in the small intestine. In the current case report, we describe a case of an intestinal sarcomatoid carcinoma in a 70-year-old Chinese female. Sarcomatoid carcinoma was confirmed based on light microscopy and immunohistochemical observations. The patient presented with symptoms of acute abdomen, which was due to an intestinal perforation caused by sarcomatoid carcinoma of the small bowel. Patients with sarcomatoid carcinoma are usually associated with a poor prognosis. However, this patient experienced a relatively favorable prognosis, which may be attributed to low positivity for Ki67 in the tumor.
A bladder wall angiomyolipoma as a manifestation of tuberous sclerosis: first case report.

RESUMEN / SUMMARY: A 21-year-old female patient admitted to the emergency department complaining of left side pain. Hypovolemic shock, which was probably caused by retroperitoneal bleeding from left sided renal angiomyolipoma, was developed. Abdominal computed tomography showed multiple fat containing lesions in different, regions including right bladder wall, lower pole of left kidney, and right kidney. Some lesions compatible with tuberous sclerosis such as angiofibromas, Shagreen patches, myocardial, and brain hamartomas were also detected. Bladder wall mass showing intra- and extravesical extensions was seen at exploration. We removed the tumor completely preserving bladder trigone. Angiomyolipoma located at lower pole of left kidney was also removed. Diagnosis of bladder angiomyolipoma was confirmed by the immunohistochemical examination. Recurrent or residual mass was not detected at the three-months-follow-up. We report the first case of bladder angiomyolipoma confirmed by histopathologically as a tuberous sclerosis.

TÍTULO / TITLE: Sarcomatoid carcinoma of the renal pelvis: Experience of multiple cases over a ten-year period.

RESUMEN / SUMMARY: Sarcomatoid carcinoma of the renal pelvis is a rare clinical entity. To the best of our knowledge, only 14 cases of this type of neoplasm have been reported in the literature to date. In the present study, the records at The First Affiliated Hospital, Medicine School of Zhejiang University (Hangzhou, Zhejiang, China) between 2000 and 2010 were reviewed to identify patients with primary renal pelvis sarcomatoid carcinoma (RPSC). A particular emphasis was placed on the treatment, recurrence and survival outcome. Eight patients with RPSC were identified and treated with nephrectomy or nephroureterectomy. All of the patients presented with Grade 3 RPSC. According to the TNM classification system, 2 patients were in stage pT2, 5 in stage pT3 and 1 in stage pT4. Adjuvant chemotherapy was administered to four patients, and the mean follow-up period was 27.5+/41.0 months. In total, 6 patients succumbed to the disease with a mean survival time of 7.7+/5.3 months (range, 1-18 months), while 2 patients were free of disease at 54 and 120 months, respectively, following treatment. The mean disease-specific survival time was 27.5+/41.0 months and the 1-year recurrence-free survival, 1-year survival and overall survival rates were 37.5, 37.5 and 25%, respectively. The present analysis suggests a poor prognosis for the majority of
RPSC patients, most likely resulting from the advanced stage of the disease at diagnosis and a poor response to systemic therapy. To improve the survival rate of RPSC, it is therefore essential to perform an early diagnosis and early radical surgery. Intravesical instillation is not essential following surgery.

[711]

**TÍTULO / TITLE:** - Hereditary nonsyndromic gingival fibromatosis: report of family case series.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) [1155/2013/835989](#)

**AUTORES / AUTHORS:** - Peeran SW; Ramalingam K; Peeran SA; Mugrabi MH; Abdulla KA

**INSTITUCIÓN / INSTITUTION:** - Department of Periodontology and Oral Implantology, Faculty of Dentistry, Sebha University, Sebha, Libya.

**RESUMEN / SUMMARY:** - Hereditary gingival fibromatosis (HGF) is a rare, benign disorder with slowly progressive enlargement of maxillary and mandibular gingiva. Herewith, we report the first case series of HGF presenting among mother and all of her 3 children. Their complaints included unaesthetic appearance due to gingival growth, malocclusion, and difficulty in mastication. Conventional gingivectomy with oral hygiene measures and regular followup is the treatment of choice for such presentation.

[712]

**TÍTULO / TITLE:** - F-FDG PET/CT findings in a case with HIV (-) Kaposi Sarcoma.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - Rev Esp Med Nucl. Acceso gratuito al texto completo a partir de los 2 años de la fecha de publicación.

- Enlace a la Editora de la Revista [http://db.doyma.es](http://db.doyma.es)


- Enlace al texto completo (gratis o de pago) [1016/j.remn.2013.08.003](#)

**AUTORES / AUTHORS:** - Ozdemir E; Poyraz NY; Keskin M; Kandemir Z; Turkolmez S

**INSTITUCIÓN / INSTITUTION:** - Department of Nuclear Medicine, Ataturk Training and Research Hospital, Ankara, Turkey. Electronic address: ecingi@yahoo.com

**RESUMEN / SUMMARY:** - Although mucocutaneous sites are the most frequently encountered sites of involvement, Kaposi Sarcoma (KS) may also occasionally involve the breast and the skeletal, endocrine, urinary and nervous systems. Various imaging modalities may be used to delineate the extent of the disease by detecting unexpected sites of involvement. Herein, we report a case of classical type KS, in whom staging with 18F-FDG PET/CT imaging disclosed widespread disease and unexpected findings of bone and salivary gland involvement.

[713]

**TÍTULO / TITLE:** - Intramuscular lipomas: Large and deep benign lumps not to underestimated. review of a series of 51 cases.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Ramos-Pascua LR; Guerra-Alvarez OA; Sanchez-Herraez S; Izquierdo-Garcia FM; Maderuelo-Fernandez JA

INSTITUCIÓN / INSTITUTION: - Servicio de Cirugia Ortopedica y Traumatologia, Complejo Asistencial Universitario de Leon, Gerencia Regional de Salud de Castilla y Leon (SACYL), Leon, España. Electronic address: luisramospascua@gmail.com.

RESUMEN / SUMMARY: - OBJECTIVE: To review a poorly studied pathology in the scientific literature. MATERIAL AND METHODS: An observational, longitudinal and ambispective study of a series of 51 intramuscular lipomas in 50 patients. The frequency distribution of qualitative variables, and the median and the interquartile range (IQR) for continuous variables were calculated. The relationship between the size of the lipomas (recoded into two values) and the study variables were analyzed using the Fisher exact test. RESULTS: Men made up 62% of the series, and the median age was 61 years, with 55% of the total being overweight. About half of the patients were diagnosed in the upper limb. More than three-quarters (78%) were strictly intramuscular lipomas. Location, clinical and image presentation, treatment and results are described. DISCUSSION: Intramuscular lipomas have their own particular characteristics. Nevertheless, MRI is sometimes unable to distinguish them from well differentiated liposarcomas. Using size as the only criterion for referring a patient with a soft tissue injury to a reference center is still debatable. CONCLUSIONS: Patients with intramuscular lipomas, although they may be typical in their presentation, especially when they are large and show findings that can be confused with a well-differentiated low grade liposarcoma, should be treated in experienced centers.

[714]

TÍTULO / TITLE: - Calcaneal chondrosarcoma: A case report.

RESUMEN / SUMMARY: - Chondrosarcoma is a rare malignant cartilaginous tumour of the bone. It commonly occurs in the pelvis, scapula proximal femur, and shoulder girdle. We present a case of a woman in her 56 years of age with chondrosarcoma of the calcaneum—a rare lesion that accounts for 0.5-2.97% of all chondrosarcomas of other sites. Treatment for chondrosarcoma is generally wide surgical excision. Chemotherapy or radiation is not effective for most of these lesions.

[715]


RESUMEN / SUMMARY: - Chondrosarcoma is a rare malignant cartilaginous tumour of the bone. It commonly occurs in the pelvis, scapula proximal femur, and shoulder girdle. We present a case of a woman in her 56 years of age with chondrosarcoma of the calcaneum—a rare lesion that accounts for 0.5-2.97% of all chondrosarcomas of other sites. Treatment for chondrosarcoma is generally wide surgical excision. Chemotherapy or radiation is not effective for most of these lesions.

AUTORES / AUTHORS: Umeononihu OS; Adinma JI; Obiechina NJ; Eleje GU; Udegbenam OI; Mbachu II

INSTITUCIÓN / INSTITUTION: Department of Obstetrics and Gynecology, Nnamdi Azikiwe University Teaching Hospital, Nnewi, Anambra State, Nigeria. Electronic address: docomene@gmail.com.

RESUMEN / SUMMARY: INTRODUCTION: Uterine inversion is an uncommon complication of parturition which often occurs in the immediate postpartum period. The chronic (non-puerperal) uterine inversion is rarer and most times tumour associated.

PRESENTATION OF CASE: A 51-year old grand multiparous lady presented with a month history of abnormal vaginal bleeding associated with offensive vaginal discharge, lower abdominal pain and dizziness. The initial evaluation suggested severe anaemia secondary to advanced cervical cancer. Examination under anaesthesia (EUA), staging and biopsy was attempted but this was however inconclusive due to profuse haemorrhage. A repeat EUA revealed chronic uterine inversion secondary to fundal submucous uterine leiomyoma. Myomectomy was done with tissue histology confirming benign uterine leiomyoma. Two weeks later, a modified Haultain’s procedure was done followed by simple hysterectomy and posterior colpoperineorrhaphy. She had satisfactory recovery.

DISCUSSION: This is the first reported case of chronic non-puerperal uterine inversion in our hospital. When it occurs, it is usually tumour associated with the commonest tumour being prolapsed myoma and leiomyosarcoma. The diagnosis is based on high index of suspicion.

CONCLUSION: Chronic uterine inversion is a rare gynaecological condition and can be misdiagnosed as advanced cervical cancer or other causes of severe genital haemorrhage in women. A high index of suspicion is needed for its proper diagnosis. Sometimes, an EUA and biopsy was required to determine the cause here and conveniently it could be described as a "gynaecological near miss".

[716]

TÍTULO / TITLE: Pulmonary benign metastasizing leiomyoma: report of three cases.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Jeon HW; Choi SH; Sung SW; Park JK

RESUMEN / SUMMARY: Benign metastasizing leiomyoma is very rare and usually occurs in women who undergo hysterectomy and myomectomy for uterine leiomyoma. This is a benign spindle-shaped smooth muscle cell tumor pathologically but metastasizes to the extraterine organs. Lungs are the most common site of metastasis. We observed three cases of pulmonary benign metastasizing leiomyoma.

[717]

TÍTULO / TITLE: A Unique Case of Classic Kaposi’s sarcoma restricted to the toes.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary

Enlace al texto completo (gratuito o de pago) 3315/jdcr.2013.1146
**BACKGROUND:** Kaposi’s sarcoma associated-herpesvirus causes all forms of Kaposi’s sarcoma, and six major subtypes have been described based on the amino acid sequences of the open reading frame K1. **MAIN OBSERVATION:** A 71-year-old man from China, HIV negative, presented with nodules on the dorsal aspect of his toes. Biopsy confirmed the diagnosis of Kaposi’s sarcoma and virology studies of his blood and saliva confirmed the presence of Kaposi’s sarcoma associated-herpesvirus infection. Viral genotyping was consistent with subtype C3. Intervention has been deferred as our patient has remained clinically asymptomatic and without evident growth of his lesions over a 2-year follow up. **CONCLUSIONS:** We herein report the first known case of Kaposi’s sarcoma restricted to the toes caused by the viral subtype C3 in an HIV-negative patient from Harbin, China.

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**BACKGROUND:** Intussusception is a relatively common cause of intestinal obstruction in children but a rare clinical entity in adults, representing fewer than 1% of intestinal obstructions in this patient population. Colonic lipomas are uncommon nonepithelial neoplasms that are typically sessile, asymptomatic and incidentally found during endoscopy, surgery, or autopsy. **CASE PRESENTATION:** A 55-year-old man visited our emergency department with severe abdominal pain, multiple episodes of vomiting, abdominal distension. Abdominal ultrasound sonography and computed tomography showed a sausage-shaped mass presenting as a target sign, suggestive of intussusception. Surgery revealed a hard elongated mass in the right colon which telescoped in the transverse colon and caused colo-colonic intussusception. Right hemicolectomy was performed and pathology documented a mature submucosal lipoma of the colon. We describe the difficulties in diagnosis and management of this rare cause of bowel obstruction and review the literature on adult intussusceptions. **CONCLUSION:** A large submucosal lipoma is a very rare cause of colon intussusception that presents as intestinal obstruction in patients without malignancy. CT and magnetic resonance imaging remain the methods of choice for studying abdominal lipomas, particularly those rising into the layers of the colonic wall. Surgical resection remains the treatment of choice and produces an excellent prognosis.

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**BACKGROUND:** Gastrointestinal stromal tumor with extensive lymphatic metastasis: a case report.
Gastrointestinal stromal tumor is a rare tumor which arises from the whole gastrointestinal tracts and most of it is detected in the stomach. It is uncommon with small intestine originated gastrointestinal stroma tumor and more uncommon with lymphatic metastasis. We experienced an unusual case of the small bowel gastrointestinal stromal tumor during experimental autopsy. Two primary tumors with central necrosis were detected in the ileum. The sizes of each tumor were 6.1x3.4x4.0 cm and 3.7x4.2x3.2 cm. There was extensive lymphatic metastasis on the greater omentum and mesenteric, iliac lymph nodes were also involved. With histologic findings, the eosinophilic spindle cells were densely distributed. Immunohistochemical findings were CD117 (-), CD34 (+), desmin (-), and S-100 protein (-). Therefore, we diagnosed the tumors as small bowel gastrointestinal stromal tumors with broad lymph node metastasis.

Neurofibromas are benign tumors that originate from the peripheral nerves, including neurites and fibroblasts. Generally, a solitary neurofibroma is located in the skin and rarely in other places. A 72-year-old female suffered from epigastric discomfort for 2 months. Endoscopic findings showed an early gastric cancer type IIc at the antrum. Abdominal computed tomography revealed early gastric cancer with a 1.6 cm-sized metastatic node posterior to the duodenum. Laparoscopic assisted distal gastrectomy and retro-pancreatic dissection were performed uneventfully. Histological examination revealed gastric adenocarcinoma, invading the mucosa without nodal metastasis, and a neurofibroma. Herein, we present a case of a gastric cancer patient with a solitary retroperitoneal neurofibroma which mimicked a distant metastatic node.

Atrial Myxoma-Related to Chronic Immunosuppression: A case report.

Neurofibromas are benign tumors that originate from the peripheral nerves, including neurites and fibroblasts. Generally, a solitary neurofibroma is located in the skin and rarely in other places. A 72-year-old female suffered from epigastric discomfort for 2 months. Endoscopic findings showed an early gastric cancer type IIc at the antrum. Abdominal computed tomography revealed early gastric cancer with a 1.6 cm-sized metastatic node posterior to the duodenum. Laparoscopic assisted distal gastrectomy and retro-pancreatic dissection were performed uneventfully. Histological examination revealed gastric adenocarcinoma, invading the mucosa without nodal metastasis, and a neurofibroma. Herein, we present a case of a gastric cancer patient with a solitary retroperitoneal neurofibroma which mimicked a distant metastatic node.

Atrial Myxoma-Related to Chronic Immunosuppression: A case report.
Although rare, atrial myxoma is the most common primary tumour of the heart. Its relation to immunosuppression in solid organ transplant is presently debateable. We report the case of a 71-year-old male patient who underwent renal transplant 17 years prior. Since that time he continued high dose immunosuppression without physician consultation and presented to us with atrial myxoma and its complications raising the question of any association between immunosuppression and the development of atrial myxoma.

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A Case Report of an Extraintestinal GIST Presenting as a Giant Abdominopelvic Tumor.

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Primary occipital myxoma: A rare case report.

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Pulmonary Lymphangioleiomyomatosis: A Rare Case.
RESUMEN / SUMMARY: Lymphangioleiomyomatosis is an uncommon lung disease primarily affecting women of childbearing age. It is characterized by the progressive proliferation and infiltration of smooth muscle-like cells, which lead to cystic destruction of the lung parenchyma; obstruction of airways, blood vessels, and lymphatics; and loss of pulmonary function. We present the case of a 46-year-old female patient with chest pain, cough, sputum, and dyspnea on exertion for three weeks. Minimal pneumothorax was noted, and the patient was referred to our center for further investigation and treatment. High-resolution computed tomography revealed numerous bilateral thin-walled air cysts and interstitial thickening affecting the central and peripheral part of the upper zone of the lung. We performed an open-lung biopsy to confirm lymphangioleiomyomatosis. Our aim is to discuss the pathogenesis and other lesions noted in the differential diagnosis of this rare disease.
denied any leading symptoms, even with the lesion involving extensively. Radiographic and microscopic similarities to a number of entities make diagnostic interpretation of odontogenic myxoma challenging. Therefore sound knowledge of clinical, radiographic and histopathologic features is important to establish an appropriate treatment aimed at a good clinical course and patient cure.

[727]
**TÍTULO / TITLE:** Sylvian fissure lipoma with angiomatous component and associated brain malformation: A case report.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Thakur S; Sood RG; Jhobta A; Makhaik S; Thakur C

**INSTITUCIÓN / INSTITUTION:** Resident, Department of Radiology, Indira Gandhi Medical College and Hospital, Shimla, Himachal Pradesh, India.

**RESUMEN / SUMMARY:** Intracranial lipomas are congenital malformations. These uncommon lesions have an incidence of 0.1 to 1.7% of all intracranial tumors. Most cases are located at midline and 5% are along the sylvian fissures. If symptomatic, seizures are the most common symptom. These tumors are slow growing and have favorable outcome. We report a case of a 25-year-old man whose CT and MRI revealed a lesion in right sylvian fissure suggesting a lipoma with abnormal vasculature and overlying cortical dysplasia.

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[728]
**TÍTULO / TITLE:** A case of chordoma invading multiple neuroaxial bones: report of ten years follow up.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Aydin AL; Sasani M; Oktenoglu T; Solaroglu I; Ozer AF

**INSTITUCIÓN / INSTITUTION:** Istanbul Physical Therapy and Rehabilitation Hospital, Department of Neurosurgery, Istanbul, Turkey.

**RESUMEN / SUMMARY:** AIM: Chordoma is a rare, slow-growing primary malignant tumor of the axial skeleton, arising from the embryonic cells of primitive notochord. Chordomas may arise at different sites of the vertebral column simultaneously or more probably they may metastasise along the neural axis insidiously. Recurrence despite radical surgery and following adjuvant therapy is possible. MATERIAL AND METHODS: A 46-year-old female patient presented weakness and numbness of the lower extremities. She was operated for clivus chordoma five years ago at another institute. RESULTS: First the patient underwent surgery for resection of the tumor at the cervical region. a second surgery was performed to resect tumor on the foramen magnum and at the C1 level. Histologic examination of the removed vertebra confirmed the diagnosis of chordoma involving the vertebral body. Radiotherapy was administered after the second surgery. Follow-up neurological and radiological examinations revealed no abnormal neurological symptoms 2,5 years after second surgery. There were no distant organ metastases. CONCLUSION: A patient with diagnosed chordoma of the spine must be investigated with MRI of other regions of the
neuraxis to exclude second or even third source of chordoma metastases. In metastatic chordoma cases, radical or gross total resection should be performed for each lesion but if complete surgical resections are impossible, preoperative or postoperative radiation therapy should be planned to improve life expectancy.

Parosteal lipoma of humerus-A rare case.

INTRODUCTION: Parosteal lipoma is an extremely rare benign tumor composed mainly of mature adipose tissue with a bony component.

PRESENTATION OF CASE: This study reports the case of a 65-year old woman with a big mass on the posteromedial aspect of the right upper arm since 1 year. The swelling was a slow growing, painless, nontender, immobile mass which was not fixed to skin. She had no complaints of painful or restricted movements of the shoulder joint. She had no history of trauma to the upper limb. MRI revealed a large 13cmx5cmx8cm well defined, nonenhancing, lobulated, heterointense, predominantly fat intensity lesion with a small area of chondroid component measuring 2cmx1.6cm in posteromedial aspect of proximal right humerus, seen completely separate from the adjacent muscles. DISCUSSION: The patient underwent surgery under general anesthesia. Vertical elliptical incision was taken over the posterior border of the upper arm over the mass. The tumor was below the lower part of deltoid near the upper end of humerus, which formed the roof, and between the long and medial heads of triceps muscles. A part of tumor, measuring 6cmx5cmx5cm, was under the long head of triceps displacing it along with radial nerve and vessels medially while the other part, measuring 7cmx6cmx3cm, was under the medial head of triceps displacing it laterally. The tumor was excised undocking its periosteal attachment. The specimen weighed 250g. On histopathology, the lesion was composed of mature lipocytes that had an intimate relationship with the periosteum. No cellular atypia or lipoblasts were seen.

CONCLUSION: Parosteal lipomas are rare neoplasias with no proven malignant potential. These tumors can be resected without much damage to the adjacent structures, thus preserving the function of the upper limb.

An abdominal extraskeletal osteosarcoma: A case report.

RESUMEN / SUMMARY: - An abdominal extraskeletal osteosarcoma: A case report.


AUTORES / AUTHORS: - Wu Z; Chu X; Meng X; Xu C

INSTITUCIÓN / INSTITUTION: - Department of General Surgery, Shaoxing Hospital, China Medical University, Shaoxing, Zhejiang 312030, P.R. China.

RESUMEN / SUMMARY: - Primary abdominal extraskeletal osteosarcoma (EOS) is a rare carcinoma. The present study reports a case of a primary abdominal EOS
involving the greater omentum and also presents a review of the literature on the etiology, diagnosis, differential diagnosis, pathological features, treatment and prognosis of the disease. The patient in the present study underwent laparoscopic surgery. A pathological examination revealed that the tumor tissues contained malignant and primitive spindle cells with varying amounts of neoplastic osteoid and osseous or cartilaginous tissue. The post-operative follow-up appointments were scheduled at three-month intervals for two years. The tumor recurred three months after the surgery.

[731]
**TITULO / TITLE:** - Fatal esophageal fibrosarcoma associated to parasitism by spirurid nematode Spirocerca lupi in a dog: a case report.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)
**AUTORES / AUTHORS:** - Da Fonseca EJ; Do Amarante EE; de S Abboud LC; Hees SJ; Franco RJ; de A Silva BJ
**INSTITUCIÓN / INSTITUTION:** - Instituto Qualittas de Pos-graduacao em Medicina Veterinaria, Rio de Janeiro, Brazil.
**RESUMEN / SUMMARY:** - A 7-years-old intact female mixed breed dog was referred to the private veterinary clinic because of history of intense food regurgitation. The plain X-ray examination and the contrast-enhanced X-ray using barium sulfate revealed the presence of a radio-opaque mass in the final third of the esophagus. Stool samples were collected and the coproparasitological test was done, which showed thick-shelled eggs from Spirocerca lupi. As the canine died, the necropsy was performed, showing nodular lesions in the esophagus, compatible with the typical lesions of spirocercosis. The histopathological analysis showed the proliferation of spindle-shaped cells, process compatible with fibrosarcoma. This is the first official report of the presence of canine spirocercosis in Guapimirim city, Rio de Janeiro, Brazil, and can serve as a warning to veterinary practitioners, about this probably endemic region for S. lupi.

[732]
**TITULO / TITLE:** - Osteoblastoma of the trapezoid bone and triquetral bone: report of two cases.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)
**AUTORES / AUTHORS:** - Kaya I; Boynuk B; Gunerbuyuk C; Ugras A
**INSTITUCIÓN / INSTITUTION:** - Department of Orthopedics and Traumatology, Haseki Training and Research Hospital, Istanbul, Turkey.
**RESUMEN / SUMMARY:** - Osteoblastoma is a benign local aggressive tumor mostly localized in the vertebra or long bones. Carpal location and recurrence are extremely rare. Treatment options include either curettage or wide en bloc resection which causes functional disability in the hand and wrist and should be reserved only for recurrence. We present a case of recurrent trapezoid osteoblastoma previously treated with curettage of the trapezoid bone and a case of primary triquetral osteoblastoma.

[733]
Osteoblastoma of the Sacrum: Report of 18 Cases and Analysis of the Literature.

Study Design. Retrospective case series

Objective. 1) to analyze clinical and radiographic characteristics, treatment and outcome in patients with sacral osteoblastoma 2) to evaluate progression free survival and local recurrence rate and 3) to identify prognostic factors.

Summary of Background Data.

Osteoblastoma is a rare tumor that has been reported to affect the sacrum from 7% to 17%. Symptoms are various and the diagnosis is often delayed. Methods. From 1980 to 2010, 18 patients with sacral osteoblastoma (16 males and 2 females) were treated at our institution. Lesion involved S1 (2 cases), S1-S2 (3 cases), S2 (1 case), S2-S3 (1 case), S2-S4 (1 case), S3 (2 cases), S3-S4 (5 cases), S4 (1 case) and almost the entire sacrum in 2 cases. According to Enneking’s classification for benign bone tumors, thirteen (72%) were diagnosed at stage 2 and 5 (28%) at stage 3. Mean tumor volume was 64 cm3 (range, 2-441 cm3). Nine patients had preoperative MRI. Five patients had a previous inadequate intralesional surgery elsewhere. Treatment consisted in intralesional surgery (16 cases), intralesional surgery and radiotherapy (one case) and wide resection (one case). Local adjuvants used were phenol (7 patients), cryocoagulation with “iceball” technique (1 case). Embolizations were performed in 7 patients.

Results. At a mean of 8.4 years (range, 1-28 years), 15 patients (83%) remained continuously disease-free while three patients had local recurrence (17%). Progression free survival was 87% at 5 years and 74% at 10 years. No statistical difference was found between patients that received or not local adjuvants (p = 1.254), older or younger than 20 years (p = 0.970), at stage 2 or 3 (p = 0.826), evaluated preoperatively with or without MRI (p = 0.160), primarily treated versus patients with previous intralesional surgery elsewhere (p = 0.131). Conclusion. In our series, curettage was successful in most of the patients. Local adjuvants did not seem to reduce the risk of local recurrence when combined with intralesional surgery.

Intraperitoneal dedifferentiated liposarcoma showing MDM2 amplification: case report.

BACKGROUND: Liposarcoma is the most common type of soft tissue sarcoma (STS). It is divided into five groups according to histological pattern: well-differentiated, myxoid, round cell, pleomorphic, and dedifferentiated.
Dedifferentiated liposarcoma most commonly occurs in the retroperitoneum, while an intraperitoneal location is extremely rare. Only seven cases have been reported in literature. Many pathologists recognize that a large number of intra-abdominal poorly differentiated sarcomas are dedifferentiated liposarcomas. We report a case initially diagnosed as undifferentiated sarcoma that was reclassified as intraperitoneal dedifferentiated liposarcoma showing an amplification of the MDM2 gene. CASE PRESENTATION: A 59-year-old woman with abdominal pain and constipation was referred to the Department of Advanced Biomedical Sciences, University of Naples Federico II, Naples, Italy, in November 2012. On physical examination, a very large firm mass was palpable in the meso-hypogastrium. Computed tomography (CT) scan showed a heterogeneous density mass (measuring 10 x 19 cm) that was contiguous with the mesentery and compressed the third part of the duodenum and jejunum. At laparotomy, a large mass occupying the entire abdomen was found, adhering to the first jejunal loop and involving the mesentery. Surgical removal of the tumor along with a jejunal resection was performed because the first jejunal loop was firmly attached to the tumor. Macroscopic examination showed a solid, whitish, cerebroid, and myxoid mass, with variable hemorrhage and cystic degeneration, measuring 26 x 19 x 5 cm. Microscopic examination revealed two main different morphologic patterns: areas with spindle cells in a myxoid matrix and areas with pleomorphic cells. The case was initially diagnosed as undifferentiated pleomorphic sarcoma. Histological review showed areas of well-differentiated liposarcoma. Fluorescence in situ hybridization (FISH) analysis was performed and demonstrated an amplification of the MDM2 gene. Definitive diagnosis was intraperitoneal dedifferentiated liposarcoma. No adjuvant therapy was given, but 5 months after laparotomy, the patient presented with a locoregional recurrence and chemotherapy with high-dose ifosfamide was started. CONCLUSIONS: No guidelines are available for the management of intraperitoneal dedifferentiated liposarcoma. We report this case to permit the collection of a larger number of cases to improve understanding and management of this tumor. Moreover, this study strongly suggests that poorly differentiated sarcomas should prompt extensive sampling to demonstrate a well-differentiated liposarcoma component and, if possible, FISH analysis.

[TÍTULO / TITLE: - A rare case of a solitary intraocular neurofibroma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Chawla U; Khurana AK; Anand N; Jain P
RESUMEN / SUMMARY: - Background: Solitary neurofibroma in the absence of neurofibromatosis is of rare occurrence and very few cases have been reported till date. Objective: To report a case of a solitary intra-ocular neurofibroma. Case: A 65-year-old man presented to us with a large swelling appearing to arise from right phthisical eye for the past one and a half years. After knowing the extent and origin of mass lesion, right eyeball was enucleated and subjected to histopathological examination which revealed intraocular neurofibroma in the absence of neurofibromatosis which is of very rare occurrence. Conclusion: The isolated...
neurofibroma of intraocular origin can present as an isolated orbital mass without systemic features.

[736]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Arora P; Rehman F; Girish KL; Kalra M
INSTITUCIÓN / INSTITUTION: - Department of Oral Pathology, Jodhpur Dental College, Jodhpur, Rajasthan, India.
RESUMEN / SUMMARY: - Osteosarcoma (OS) is a malignant connective tissue tumor originating from bone and is the most common primary bone malignancy of long bones but seldom arises in jaw bones. Osteosarcoma of jaws is frequently seen arising in the second and third decade as compared to earlier occurrences in other bones and show a slight predilection for body of mandible. It is a highly malignant tumor with varied radiographic features. We present a case with detailed radiographic assessment using intraoral radiograph, computed tomography (CT), 3-D CT, CT angiography techniques and histological evaluation.

[737]
TÍTULO / TITLE: - Three-dimensional ultrasound in diagnosis of adenomyosis: histologic correlation with ultrasound targeted biopsies of the uterus.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Luciano DE; Exacoustos C; Albrecht L; Lamonica R; Proffer A; Zupi E; Luciano AA
INSTITUCIÓN / INSTITUTION: - Department of Obstetrics and Gynecology, University of Connecticut, New Britain, Connecticut. Electronic address: dluciano@aol.com.
RESUMEN / SUMMARY: - STUDY OBJECTIVE: To evaluate the accuracy of 3-dimensional transvaginal sonography (3D TVS) in the diagnosis of adenomyosis by correlating adenomyosis-induced morphologic alterations in the myometrium and the junctional zone (JZ) with histopathologic features of targeted biopsy specimens of the uterus. DESIGN: Prospective study (Canadian Task force classification II-2). SETTING: Private practice associated with a university program. PATIENTS: Symptomatic premenopausal women scheduled to undergo hysterectomy because of benign conditions. INTERVENTIONS: Patients underwent preoperative 3D TVS of the uterus to evaluate alterations to the JZ, to measure the smallest (JZmin) and largest (JZmax) JZ thickness, and to assess for the presence of myometrial heterogeneous and cystic areas, hyperechoic striations, and asymmetry of the myometrial wall. Localization and position of the lesions in the myometrial wall were accurately recorded. Results of the sonographic features were correlated with the histopathologic findings of the ultrasound-based targeted biopsy specimens of the uterus. MEASUREMENTS AND MAIN RESULTS: The study included 54 symptomatic premenopausal women with a
mean age of 42.1 years. Of these, 12 had previously undergone endometrial ablation and 10 were receiving medical therapy, and these patients were considered separately for the statistical analysis. The prevalence of adenomyosis at histology was 66.6% (36/54). Of 32 patients who had received no previous treatment, 26 had adenomyosis on the targeted biopsy specimens of the myometrium. 3D TVS features of adenomyosis with the best specificity (83%) and positive predictive values were JZmax >=8 mm, myometrial asymmetry, and hypoechoic striation. When we considered the presence of at least 2 of the described ultrasound features for the diagnosis of adenomyosis, accuracy was 90% (sensitivity, 92%; specificity, 83%; positive predictive value, 99%; and negative predictive value, 71%). Diagnostic accuracy was decreased to 50% in patients who had previously undergone endometrial ablation, and to 60% in patients receiving medical therapy. CONCLUSION: 3D TVS demonstrates high diagnostic accuracy in detection of site and position of adenomyosis in the uterine walls. Endometrial ablation and medical therapy alter the appearance of the JZ, compromising the accuracy of 3D US in enabling the diagnosis of adenomyosis.

[738]

**TÍTULO / TITLE:** - Inflammatory myofibroblastic tumour in the nasal cavity of a dog.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Swinbourne F; Kulendra E; Smith K; Leo C; Ter Haar G

**INSTITUCIÓN / INSTITUTION:** - Department of Veterinary Clinical Sciences, The Royal Veterinary College, Hertfordshire.

**RESUMEN / SUMMARY:** - A 4.5-year-old, female neutered Leonberger was presented with a 2-month history of sneezing, nasal discharge and epistaxis. A presumptive diagnosis of nasal aspergillosis was made based on a suspected (fungal) granuloma on rhinoscopic examination and fungal hyphae on cytological examination. A poor response to targeted therapy was observed and computed tomography 16 months after initial presentation revealed a progressive, locally invasive mass lesion. Histopathological and immunohistochemical analysis of deep surgical biopsies revealed a spindle cell population and a plasma cell rich inflammatory infiltrate, with diffuse expression of vimentin, supporting a diagnosis of inflammatory myofibroblastic tumour. Complete resolution of the nasal discharge and reduced sneezing frequency was reported 9 months post-surgical debridement via rhinotomy. To the authors’ knowledge, this is the first report of IMT in the nasal cavity of a dog. IMT should be considered when presented with a nasal mass lesion, particularly if histopathological features and clinical course are inconsistent.

[739]

**TÍTULO / TITLE:** - Subcutaneous hemangiosarcoma induced by a foreign body (steel staple) in a cat.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Tan RM; Singh K; Sandman K
INSTITUCIÓN / INSTITUTION: - Veterinary Diagnostic Laboratory, University of Illinois, Urbana, Illinois 61801, USA (Tan, Singh); Animal Emergency Treatment Center, 3927 W. Belmont Avenue, Chicago, Illinois 60618, USA (Sandman).

RESUMEN / SUMMARY: - An 8-year-old, female domestic shorthair cat was presented with a ventral abdominal subcutaneous mass. A radiograph showed that the center of the mass contained what appeared to be steel sutures, presumed to be from an ovariohysterectomy performed 7 years earlier. The excised mass was irregular and contained numerous pockets filled with friable necrotic material and hemorrhages that were dissected by fibrous connective tissue bands. Multiple tangled and fragmented pieces of steel staples were deeply embedded within the mass. Histologically, the mass was non-encapsulated, densely cellular, and infiltrative. Neoplastic cells lined caverns and channels and were factor VIII-positive by immunohistochemistry. The neoplastic cells were oval to round with granular cytoplasm and vesicular nucleus and exhibited moderate cellular and nuclear pleomorphism. A diagnosis of subcutaneous hemangiosarcoma was made. To our knowledge, this is the first report of foreign body associated hemangiosarcoma and the first case of steel staple associated neoplasm in domestic animals.

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RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Vassos N; Lell M; Hohenberger W; Croner RS; Agaimy A

INSTITUCIÓN / INSTITUTION: - Department of Surgery, University Hospital Erlangen, Germany.

RESUMEN / SUMMARY: - BACKGROUND: Hibernoma is a rare benign fat-forming soft tissue tumor that differentiates similar to brown fat, hence an origin from remnants of fetal brown adipose tissue has been proposed. Mainly young adults are affected, usually without significant clinical symptoms. MATERIAL AND METHODS: We report on four patients with hibernomas, who were treated at our hospital during the last 10 years. The clinicopathologic and immunohistochemical features are presented and treatment and follow-up data discussed. RESULTS: Patients were 2 women and 2 men aged 21-67 years (mean: 45 yrs) who presented with a slowly growing, painless mass. The anatomic location was the thigh, upper arm, lateral thoracic wall and paravertebral soft tissue. Two of them were diagnosed preoperatively through a percutaneous core needle biopsy and the other two underwent surgery because of high clinical and radiological suspicion of liposarcoma. The tumor’s size ranged from 7 cm to 15.5 cm (mean: 11 cm). All were deep-seated subfascial intramuscular masses. Histologically, all four tumors were of the typical variant. All patients underwent a R0-surgical resection of the tumor and they were recurrence-free at last follow-up (mean: 47 months; range: 25-87). CONCLUSION: Hibernoma may present as huge deep intramuscular soft tissue mass in adults, closely mimicking well differentiated liposarcoma and should be considered in the differential diagnosis of fatty soft tissue tumors in any location. Surgical excision is the treatment of choice. The tumor has no malignant or recurrence potential.

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**TÍTULO / TITLE:** Measurement of phenolic environmental estrogens in women with uterine leiomyoma.

**RESUMEN / SUMMARY:**

Enlace al Resumen / Link to its Summary


●● Enlace al texto completo (gratuito o de pago) 1371/journal.pone.0079838

**AUTORES / AUTHORS:** Shen Y; Xu Q; Ren M; Feng X; Cai Y; Gao Y

**INSTITUCIÓN / INSTITUTION:** Department of Obstetrics and Gynecology, Zhongda Hospital, School of Medicine, Southeast University, Nanjing, China.

**RESUMEN / SUMMARY:**

OBJECTIVES: To investigate the effect of phenolic environmental estrogens on uterine leiomyoma from the perspective of clinical epidemiology.

METHODS: Urine and blood samples were collected from Han women with uterine leiomyoma and women without uterine leiomyoma, living in Nanjing, China, between September 2011 and February 2013. A total of 156 urine samples and 214 blood samples were collected from the uterine leiomyoma group and 106 urine samples and 126 blood plasma samples from the control group. Bisphenol A (BPA), nonylphenol (NP) and octylphenol (OP) concentrations were determined by solid-phase extraction (SPE) coupled with liquid chromatography-tandem mass spectrometry (HPLC-MS/MS).

RESULTS: PHENOLIC ENVIRONMENTAL ESTROGENS IN THE UTERINE LEIOMYOMA AND CONTROL GROUPS WERE COMPARED BASED ON: gravida>3 and gravida /= 3. In participants with gravida>3, urine OP concentration was significantly (P<0.05) higher in the uterine leiomyoma group than in the control group. In participants with gravida /= 3, urine NP concentration was significantly (P<0.05) higher in the uterine leiomyoma group compared to controls. Despite obstetric history, urine BPA mean exposure concentration was significantly (P<0.05) different between uterine leiomyoma group and control group. The urine BPA concentration was not significantly (P>0.05) different between gravida>3 and gravida /= 3 patients. There was no significant (P>0.05) difference in plasma concentrations of BPA, OP and NP between the leiomyoma group and control group. Mean exposure concentration and range of distribution of BPA, OP and NP plasma concentration differed between the uterine leiomyoma and control group.

CONCLUSION: Exposure level of phenolic environmental estrogens in human was related with leiomyoma tumorigenesis.

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**TÍTULO / TITLE:** Surveillance snapshot: myomectomies and hysterectomies performed for uterine fibroids at military health facilities, active component service women, U.S. Armed Forces, 2000-2012.

**RESUMEN / SUMMARY:**

Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** MSMR. 2013 Sep;20(9):19.

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**TÍTULO / TITLE:** Selective arterial embolisation of bilateral angiomyolipomata in a symptomatic pregnant female.

**RESUMEN / SUMMARY:**

Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** British Medical J (BMJ). Acceso gratuito al texto completo.

●● Enlace a la Editora de la Revista http://bmj.com/search.dtl
RENAL ANGIOMYOLIPOMA (RA) IS A RARE BENIGN TUMOUR THAT CAN EXPAND RAPIDLY DURING PREGNANCY DUE TO OESTROGEN AND OTHER HORMONAL FACTORS. COMPLICATIONS ASSOCIATED WITH EXPANDING RENAL ANGIOMYOLIPOMATA ARE SPONTANEOUS RETROPERITONEAL HAEMORRHAGES SECONDARY TO ACUTE RUPTURE AND THROMBOSIS OF THE RENAL VEIN OR INFERIOR VENA CAVA. MRI IS RECOMMENDED FOR DIAGNOSTIC PURPOSES IN PREGNANCY; HOWEVER, THIS MODALITY IS NOT ALWAYS READILY AVAILABLE. IN THE PRESENT REPORT, WE DESCRIBE THE FIRST CASE OF BILATERAL SELECTIVE ARTERIAL EMBOLISATION FOR RENAL ANGIOMYOLIPOMATA PRESENTING SYMPTOMATICALLY IN A PREVIOUSLY HEALTHY PREGNANT FEMALE.

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TÍTULO / TITLE: A PHASE I STUDY OF THE COMBINATION OF TEMSIROLIMUS WITH IRINOTECAN FOR METASTATIC SARCOMA.

RESUMEN / SUMMARY: mTOR inhibitors are emerging as important anti-neoplastic agents with a wide range of clinical applications. The topoisomerase I inhibitor irinotecan is a potent DNA damaging drug, with a broad spectrum of anticancer activities. mTOR appears to enhance cancer cell survival following DNA damage, thus the inhibition of mTOR after irinotecan could theoretically show synergistic activities in patients. Both mTOR inhibitors and irinotecan have been used as single agents in soft tissue sarcomas with limited efficacy. We completed a phase I trial of the combination of the mTOR inhibitor, temsirolimus, and irinotecan in patients with advanced soft tissue sarcoma. Seventeen patients were recruited. The Phase II recommended dose is 20 mg of temsirolimus and 80 mg/m2 of irinotecan administered on weekly basis for three out of four weeks. Most frequently encountered toxicities include cytopenias, fatigue, and gastrointestinal toxicities. Two patients (one with leiomyosarcoma and one with high grade undifferentiated sarcoma) had stable disease for more than 12 months.

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TÍTULO / TITLE: MULTI-MODALITY IMAGING IN THE ASSESSMENT OF A METASTATIC CARDIAC RHABDOMYOSARCOMA PRESENTING WITH RECURRENT VENTRICULAR TACHYCARDIA.

RESUMEN / SUMMARY: Multi-modality imaging is in assessment of a metastatic cardiac rhabdomyosarcoma presenting with recurrent ventricular tachycardia.
INSTITUCIÓN / INSTITUTION: - Cardiac Investigations Unit, Department of Echocardiography, The Prince Charles Hospital, Rode Rd., Chermside, Brisbane, QLD 4032, Australia.

[746]

TÍTULO / TITLE: - Management of synovial osteochondromatosis of the distal radioulnar joint with imaging features consistent with malignancy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - McInnes CW; Goetz TJ

INSTITUCIÓN / INSTITUTION: - Section of Plastic Surgery, University of Manitoba, Winnipeg, MB, Canada R3A 1R9.

RESUMEN / SUMMARY: - Synovial osteochondromatosis of the distal radioulnar joint is a rare entity with only 14 cases reported in the literature. Malignant transformation of synovial osteochondromatosis is the most worrisome complication of the disease. It has been described in joints such as the hip and knee but never for the distal radioulnar joint. We report a case of synovial osteochondromatosis of the distal radioulnar joint which presented with radiographic features which were worrisome for malignant transformation and required a comprehensive preoperative workup. Discussed are the preoperative management, surgical treatment, and a literature review of this rare disease.

[747]

TÍTULO / TITLE: - Tissue microarray immunohistochemical detection of brachyury is not a prognostic indicator in chordoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Zhang L; Guo S; Schwab JH; Nielsen GP; Choy E; Ye S; Zhang Z; Mankin H; Hornicek FJ; Duan Z

INSTITUCIÓN / INSTITUTION: - Sarcoma Biology Laboratory, Center for Sarcoma and Connective Tissue Oncology, Massachusetts General Hospital, Boston, Massachusetts, United States of America ; Department of Pathology, the Third Affiliated Hospital of Zhengzhou University, Zhengzhou, China.

RESUMEN / SUMMARY: - Brachyury is a marker for notochord-derived tissues and neoplasms, such as chordoma. However, the prognostic relevance of brachyury expression in chordoma is still unknown. The improvement of tissue microarray technology has provided the opportunity to perform analyses of tumor tissues on a large scale in a uniform and consistent manner. This study was designed with the use of tissue microarray to determine the expression of brachyury. Brachyury expression in chordoma tissues from 78 chordoma patients was analyzed by immunohistochemical staining of tissue microarray. The clinicopathologic parameters, including gender, age, location of tumor and metastatic status were evaluated. Fifty-nine of 78 (75.64%) tumors showed nuclear staining for brachyury, and among them, 29 tumors (49.15%) showed 1+ (<30% positive cells) staining, 15 tumors (25.42%) had 2+ (31% to 60%
positive cells) staining, and 15 tumors (25.42%) demonstrated 3+ (61% to 100% positive cells) staining. Brachyury nuclear staining was detected more frequently in sacral chordomas than in chordomas of the mobile spine. However, there was no significant relationship between brachyury expression and other clinical variables. By Kaplan-Meier analysis, brachyury expression failed to produce any significant relationship with the overall survival rate. In conclusion, brachyury expression is not a prognostic indicator in chordoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Peille AL; Brouste V; Kauffmann A; Lagarde P; Le Morvan V; Coindre JM; Chibon F; Bresson-Bepoldin L
INSTITUCION / INSTITUTION: - Institut Bergonie, Comprehensive Cancer Centre, Bordeaux, France; Univ. Bordeaux, Bordeaux, France; INSERM U916 VINCO, Bordeaux, France.
RESUMEN / SUMMARY: - Soft tissue sarcomas (STS) are rare, complex tumors with a poor prognosis. The identification of new prognostic biomarkers is needed to improve patient management. Our aim was to determine the methylation status of the 118 CpG sites in the PLAGL1 tumor-suppressor gene P1 CpG island promoter and study the potential prognostic impact of PLAG1 promoter methylation CpG sites in STS. Training cohorts constituted of 28 undifferentiated sarcomas (US) and 35 leiomyosarcomas (LMS) were studied. PLAGL1 mRNA expression was investigated by microarray analysis and validated by RT-qPCR. Pyrosequencing was used to analyze quantitative methylation of the PLAGL1 promoter. Associations between global promoter or specific CpG site methylation and mRNA expression were evaluated using Pearson's product moment correlation coefficient. Cox univariate and multivariate proportional hazard models were used to assess the predictive power of CpG site methylation status. Sixteen CpG sites associated with PLAGL1 mRNA expression were identified in US and 6 in LMS. Statistical analyses revealed an association between CpG107 methylation status and both overall and metastasis-free survival in US, which was confirmed in a validation cohort of 37 US. The exhaustive study of P1 PLAG1 promoter methylation identified a specific CpG site methylation correlated with mRNA expression, which was predictive for both metastasis-free and overall survival and may constitute the first US-specific biomarker. Such a biomarker may be relevant for identifying patients likely to derive greater benefit from treatment.

[749] TÍTULO / TITLE: - LIM kinase 1 is required for insulin-dependent cell growth of osteosarcoma cell lines.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
RESUMEN / SUMMARY: Osteosarcoma is a type of malignant bone tumor with high metastasis and poor prognosis. Previous studies have demonstrated the involvement of LIM kinase 1 (LIMK1) in the proliferation of osteosarcoma cells. LIMK1 is overexpressed in human osteosarcoma tissues and cell lines. To further study LIMK1-associated mechanisms, we used shRNA targeted to the LIMK1 gene to block its expression in the osteosarcoma cell lines MG63 and U2OS. Insulin promoted the proliferation of MG63 cells in a time- and dose-dependent manner, however, this insulin induced proliferation was significantly inhibited by transfection of shRNA targeted to the LIMK1 gene, as well as by the PI3K inhibitor LY294002, but not by the mitogen activated protein kinase (MAPK) inhibitor PD98059. The level of cofilin phosphorylation was increased significantly following stimulation of insulin for 24 h, indicating the activation of LIMK1. MG63 cell proliferation was also significantly inhibited by 1,25-dihydroxy vitamin D3 (1,25(OH)2D3) in a time-dependent manner. Furthermore, 1,25(OH)2D3 negated the inhibitory effect of LIMK1 shRNA, indicating that LIMK1 is important in the inhibitory pathway of 1,25(OH)2D3. The present study confirms that LIMK1 is important in regulating osteosarcoma cell proliferation via the insulin/PI3K/LIMK1 signaling pathway, thus the development of gene therapy for osteosarcoma targeting LIMK1 is warranted.
were recurrently and homozygously lost in osteoblastoma. Four of them are functionally involved in regulating osteogenesis and/or tumorigenesis. MN1 and NF2 have previously been implicated in the development of leukemia and solid tumors, and ZNRF3 and KREMEN1 are inhibitors of the Wnt/beta-catenin signaling pathway. In line with deletions of the latter two genes, high beta-catenin protein expression has previously been reported in osteoblastoma and aberrations affecting the Wnt/beta-catenin pathway have been found in other bone lesions, including osteoma and osteosarcoma.
Uterine fibroids having the distinct pathological and immunohistochemical features of cotyledonoid dissecting leiomyoma have been reported infrequently. We describe a postmenopausal woman with an incidental finding of an abdominopelvic mass arising from the uterine fundus on routine radiological imaging of the lumbar spine. The imaging was performed for the investigation of chronic radicular lower back pain refractory to usual pain management. However, the woman did not manifest any gynaecological symptoms. Intraoperatively, the pelvic mass appeared malignant and a frozen section suggested uterine sarcoma. As such, the mass was radically resected, resulting in significant resolution of the back pain. To the authors' knowledge, this is the first report of cotyledonoid dissecting leiomyoma presenting solely as chronic lower back pain, and also the first report of this fibroid variant in Australasia. We discuss the diagnostic and operative challenges, emphasising the role of radiological imaging and immunohistopathology in such cases and review current literature.

Benign medullary fibroma of the kidney: a rare diagnostic dilemma.

Renomedullary interstitial cell tumor or medullary fibroma, is a small tumor that commonly presents as an incidental finding, but in rare cases maybe large and symptomatic. Although it is a benign tumor, it is difficult to differentiate this lesion from other malignancies of the kidney on radiological basis and hence many patients undergo radical nephrectomy. We present a case of renal medullary fibroma and various nuances associated with radiological identification of this lesion and its management related dilemmas.

Cavernous hemangioma-like kaposi sarcoma: histomorphologic features and differential diagnosis.

Aim. Cavernous hemangioma-like Kaposi sarcoma is a rare morphologic type of Kaposi sarcoma. So far there are no cases in the literature defining the histological features of this morphologic spectrum in detail. In this study we presented two classical-type cutaneous Kaposi sarcoma cases with histologic findings resembling cavernous hemangioma in company with clinical and histopathological
data. Cases. One hundred and eighty-five classical-type cutaneous Kaposi sarcoma lesions in 79 patients were assessed retrospectively in terms of histopathological features. Findings of two cases showing features of cavernous hemangioma-like Kaposi sarcoma whose clinical data could be accessed were presented in accompany with the literature data. Both cases were detected to have bluish-purple, protruded, irregularly bordered cutaneous lesions. Histopathological examination revealed a lesion formed by cavernous hemangioma-like vascular structures organized in a lobular pattern that became dilated and filled with blood. Typical histological findings of early-stage KS, consisting of mononuclear inflammation, extravasated erythrocytes, and a few immature vascular structures in superficial dermis, were observed. All cases were serologically HIV-1 negative. A positive reaction with HHV-8, CD31, CD34, and D2-40 monoclonal antibodies was identified at both cavernous hemangioma-like areas and in immature vascular structures. Results. Cavernous hemangioma-like Kaposi sarcoma is a rare Kaposi sarcoma variant presenting with diagnostic challenges, that may be confused with hemangioma. As characteristic morphological features may not be observed in every case, it is important for diagnostic purposes to show immunohistochemical HHV-8 positivity in this variant.

[755]
TÍTULO / TITLE: - Renal Leiomyosarcoma: A Diagnostic Challenge.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o de pago) 1155/2013/459282
AUTORES / AUTHORS: - Valery JR; Tan W; Cortese C
INSTITUCIÓN / INSTITUTION: - Community Internal Medicine, Mayo Clinic, 4500 San Pablo Road, Jacksonville, FL 32224, USA.
RESUMEN / SUMMARY: - Renal leiomyosarcoma is a very rare tumor that clinically and radiographically mimics more common renal malignancies. The infrequency of the condition makes it very difficult to diagnose. A 70-year-old male smoker presented with months of hematuria, right-sided flank pain, and weight loss. Imaging revealed a 3.8-centimeter renal mass that had characteristics similar to renal cell carcinoma. Initial biopsy of the mass was negative for malignancy. Two months later, subsequent imaging revealed what appeared to be metastatic bone lesions. Again, a biopsy of one of the lesions was negative for malignancy. Subsequent ureteral pyeloscopy, ureteroscopic renal pelvis biopsy, and brush cytology were negative for malignancy as well. The decision was made to perform nephrectomy for the removal of the mass. Pathologic analysis revealed renal leiomyosarcoma. This case illustrates the difficulty in diagnosing renal leiomyosarcoma. Repeated pathologic sampling was negative because of the tumor heterogeneity. Prompt diagnosis and treatment are very significant as surgical resection at an early stage offers the best prognosis.

[756]
TÍTULO / TITLE: - Prenatal diagnosis of giant cardiac rhabdomyoma with fetal hydrops in tuberous sclerosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Schlaegel F; Takacs Z; Solomayer EF; Abdul-Kaliq H; Meyberg-Solomayer G
INTRODUCTION: fetal rhabdomyoma is the most common fetal cardiac tumor and is often associated with tuberous sclerosis. Usually the tumors are relatively small and show no mediastinal shift. Fetal hydrops and pericardial effusion are rarely seen. CASE: in this case report we present the neonatal clinical course of a case of prenatal diagnosis of giant cardiac rhabdomyomas. CONCLUSION: an early prenatal diagnosis may help for an adequate planning of perinatal monitoring and treatment with involvement of a multidisciplinary team.
crown of an impacted tooth and foci of radiopacity, which led to a misdiagnosis of either an odontogenic lesion or a bone tumour, but proved to be a fibrosarcoma on histopathological and immunohistochemistry investigations.

[759]
TÍTULO / TITLE: - Rb1 family mutation is sufficient for sarcoma initiation.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
** Enlace al texto completo (gratuito o de pago) - 1038/ncomms3650
AUTORES / AUTHORS: - Liu Y; Sanchez-Tillo E; Lu X; Clem B; Telang S; Jenson AB; Cuatrecasas M; Chesney J; Postigo A; Dean DC
INSTITUCIÓN / INSTITUTION: - 1] Molecular Targets Program, James Brown Cancer Center, University of Louisville Health Sciences Center, 529 South Jackson Street, Louisville, Kentucky 40202, USA [2] Department of Ophthalmology and Visual Sciences, University of Louisville Health Sciences Center, 301 East Muhammad Ali Boulevard, Louisville, Kentucky 40202, USA [3] Birth Defects Center, University of Louisville Health Sciences Center, 301 East Muhammad Ali Boulevard, Louisville, Kentucky 40202, USA.
RESUMEN / SUMMARY: - It is thought that genomic instability precipitated by Rb1 pathway loss rapidly triggers additional cancer gene mutations, accounting for rapid tumour onset following Rb1 mutation. However, recent whole-genome sequencing of retinoblastomas demonstrated little genomic instability, but instead suggested rapid epigenetic activation of cancer genes. These results raise the possibility that loss of the Rb1 pathway, which is a hallmark of cancers, might be sufficient for cancer initiation. Yet, mutation of the Rb1 family or inactivation of the Rb1 pathway in primary cells has proven insufficient for tumour initiation. Here we demonstrate that traditional nude mouse assays impose an artificial anoikis and proliferation barrier that prevents Rb1 family mutant fibroblasts from initiating tumours. By circumventing this barrier, we show that primary fibroblasts with only an Rb1 family mutation efficiently form sarcomas in nude mice, and a Ras-ZEB1-Akt pathway then causes transition of these tumours to an invasive phenotype.

[760]
TÍTULO / TITLE: - Targeting wild-type and mutationally activated FGFR4 in rhabdomyosarcoma with the inhibitor ponatinib (AP24534).
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
** Enlace al texto completo (gratuito o de pago) - 1371/journal.pone.0076551
AUTORES / AUTHORS: - Li SQ; Cheuk AT; Shern JF; Song YK; Hurd L; Liao H; Wei JS; Khan J
INSTITUCIÓN / INSTITUTION: - Oncogenomics Section, Pediatric Oncology Branch, Center for Cancer Research, National Cancer Institute, National Institutes of Health, Bethesda, Maryland, United States of America.
RESUMEN / SUMMARY: - Rhabdomyosarcoma (RMS) is the most common childhood soft tissue sarcoma. Despite advances in modern therapy, patients with relapsed or metastatic disease have a very poor clinical prognosis. Fibroblast Growth Factor Receptor 4 (FGFR4) is a cell surface tyrosine kinase receptor that is involved in normal
myogenesis and muscle regeneration, but not commonly expressed in differentiated muscle tissues. Amplification and mutational activation of FGFR4 has been reported in RMS and promotes tumor progression. Therefore, FGFR4 is a tractable therapeutic target for patients with RMS. In this study, we used a chimeric Ba/F3 TEL-FGFR4 construct to test five tyrosine kinase inhibitors reported to specifically inhibit FGFRs in the nanomolar range. We found ponatinib (AP24534) to be the most potent FGFR4 inhibitor with an IC50 in the nanomolar range. Ponatinib inhibited the growth of RMS cells expressing wild-type or mutated FGFR4 through increased apoptosis. Phosphorylation of wild-type and mutated FGFR4 as well as its downstream target STAT3 was also suppressed by ponatinib. Finally, ponatinib treatment inhibited tumor growth in a RMS mouse model expressing mutated FGFR4. Therefore, our data suggests that ponatinib is a potentially effective therapeutic agent for RMS tumors that are driven by a dysregulated FGFR4 signaling pathway.

[761]

TÍTULO / TITLE: Ultrasound-Guided High-Intensity Focused Ultrasound vs Laparoscopic Myomectomy for Symptomatic Uterine Myomas.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Wang F; Tang L; Wang L; Wang X; Chen J; Liu X; Gong Y
INSTITUCIÓN / INSTITUTION: Department of Gynecology and Obstetrics, First Affiliated Hospital of Chongqing Medical University, Chongqing, China (all authors).
RESUMEN / SUMMARY: STUDY OBJECTIVE: To assess the quality of life (QoL) of women at 1 and 12 months after ultrasound-guided high-intensity focused ultrasound (USgHIFU) treatment as compared with laparoscopic myomectomy for treatment of symptomatic uterine myomas. DESIGN: Nonrandomized prospective clinical trial (Canadian Task Force classification II-2). SETTING: Urban university-based hospital in China. INTERVENTIONS: One hundred thirty premenopausal women underwent USgHIFU (n = 89) or laparoscopic myomectomy (n = 41) for treatment of symptomatic uterine myomas. MEASUREMENTS AND MAIN RESULTS: Eighty-three patients in the HIFU group and 39 in the surgical group were followed up at 1 and 12 months. QoL was assessed using the Medical Outcomes Study 36-Item Short-Form General Health Survey, which showed no significant differences between groups in any of the 8 subscales at the 12-month follow-up visit. Symptom score, willingness to recommend the treatment to a friend, hospital stay, and recovery period were compared between the 2 groups. In the HIFU group, hospital stay was shorter (mean [SD] 2.9 [1.5] days vs 6.2 [2.7] days; p <.001) and patients resumed normal activities sooner (4.5 [1.5] days vs 10.9 [3.8] days; p <.001). Significant clinical complications and adverse events after each treatment were documented and compared, and HIFU yielded significantly better results. CONCLUSIONS: Compared with laparoscopic myomectomy, HIFU treatment of symptomatic uterine myomas leads to comparable QoL and symptom improvement, fewer significant clinical complications and adverse events, shorter hospital stay, and faster recovery. Randomized studies with long-term follow-up are needed to reach definitive conclusions insofar as HIFU treatment of uterine myomas.

[762]
Nectandrin A Enhances the BMP-Induced Osteoblastic Differentiation and Mineralization by Activation of p38 MAPK-Smad Signaling Pathway.

RESUMEN / SUMMARY: Osteoblastic activity of nectandrin A was examined in C2C12 cells. Nectandrin A enhances the BMP-induced osteoblastic differentiation and mineralization, manifested by the up-regulation of differentiation markers (alkaline phosphatase and osteogenic genes) and increased calcium contents. In C2C12 cells co-transfected with expression vector encoding Smad4 and Id1-Luc reporter, nectandrin A increased Id1 luciferase activity in a concentration-dependent manner, when compared to that in BMP-2 treated cells, indicating that Smad signaling pathway is associated with nectandrin A-enhanced osteoblastic differentiation in C2C12 cells. In addition, nectandrin A activated p38 mitogen-activated protein kinase (MAPK) in time- and concentration-dependent manners, and phosphorylated form of pSmad1/5/8 and alkaline phosphatase activity were both decreased when the cells were pretreated with SB203580, a p38 MAPK inhibitor, suggesting that p38 MAPK might be an upstream kinase for Smad signaling pathway. Taken together, nectandrin A enhances the BMP-induced osteoblastic differentiation and mineralization of C2C12 cells via activation of p38 MAPK-Smad signaling pathway, and it has a therapeutic potential for osteoporosis by promoting bone formation.

Saphenous vein graft pseudoaneurysm mimicking cardiac myxoma.

RESUMEN / SUMMARY: A Saphenous vein graft pseudoaneurysm mimicking cardiac myxoma is reported.

EXTENSIVELY OSSIFYING ORAL LEIOMYOMA: A RARE HISTOLOGIC FINDING.

RESUMEN / SUMMARY: Extensively ossifying oral leiomyoma is a rare histologic finding.
identified a total of three cases of extensively ossified leiomyoma in the head and neck in the literature including lesions in the lateral pterygoid muscle and orbit. To the best of our knowledge, only one case of extensively calcified leiomyoma has been reported in the oral cavity. We present two such rare cases of oral leiomyoma with extensive intratumoral calcifications and ossification. Ossified leiomyoma should be considered in the differential diagnosis of calcified or hard/firm soft tissue masses in the oral cavity.

[765]
**TÍTULO / TITLE:** - Solitary plexiform neurofibroma of the gingiva: unique presentation in the oral cavity.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - S S; Shashikumar P; H S S; Kumar G S

**INSTITUCIÓN / INSTITUTION:** - Post Graduate Student, Department of Periodontology, JSS Dental College and Hospital, Mysore, Karnataka, India.

**RESUMEN / SUMMARY:** - Neurofibromas are benign tumours originating from the nerve sheath. Amongst the histological variants, plexiform types are considered exclusive. These are poorly circumscribed, locally invasive and may exhibit sarcomatous potential. Plexiform neurofibromas are key features of Neurofibromatosis - 1 and their solitary intra-oral presentation is uncommon. The following case report describes a unique case of an isolated solitary plexiform neurofibroma of the maxillary anterior gingival region in a middle aged female patient.

[766]
**TÍTULO / TITLE:** - Abnormal uterine bleeding as a presenting symptom is related to multiple uterine leiomyoma: an ultrasound-based study.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Fonseca-Moutinho JA; Barbosa LS; Torres DG; Nunes SM

**INSTITUCIÓN / INSTITUTION:** - Faculty of Health Sciences, School of Medicine, Beira Interior University, Covilha, Portugal; Child and Women Department, Cova da Beira Academic Medical Center, Covilha, Portugal.

**RESUMEN / SUMMARY:** - PURPOSE: To determine the prevalence of uterine leiomyomas, diagnosed by ultrasound, in a private health care setting located in the central eastern region of Portugal, and to explore the demographic and clinical factors related to diagnosis and symptomatology. PATIENTS AND METHODS: The files of 624 patients attending a private clinic in Covilha, Portugal, from January 2 to December 31, 2010 were retrieved for evaluation. Pelvic ultrasound record, age, weight, height, age at menarche, number of pregnancies and deliveries, marital status, menstrual cycles characteristic, and contraceptive method at consultation were included in the analysis. RESULTS: Uterine leiomyoma (UL) was diagnosed by ultrasonography in 161 (25.8%) patients. A single UL was diagnosed in 80 (49.7%) patients. In 79 (49.1%) patients, the largest leiomyoma had a dimension <20 mm. Prevalence of UL was age dependent: at 11.0% for women 20-39 years old; 45.4% for
those aged 40-59 years; and 19.5% for women 60 years or older. Metrorrhagia was the
most distressing presenting symptom. When menorrhagia was the presenting
symptom, the probability of having an ultrasound diagnosis of UL was 73.3%.
Metrorrhagia or menorrhagia, as presenting symptom, was significantly related to the
ultrasound diagnosis of multiple ULs. CONCLUSION: UL was especially prevalent in
women aged between 40 and 59 years. Patients with multiple ULs had significantly
more abnormal uterine bleeding. In patients with menorrhagia or metrorrhagia, special
attention should be taken in searching for the presence of multiple ULs during ultrasound.

[767]
TÍTULO / TITLE: - Breast metastasis from rhabdomyosarcoma of the anus in an
adolescent female.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Breast Cancer. 2013 Sep;16(3):345-8. doi:
AUTORES / AUTHORS: - Jung SP; Lee Y; Han KM; Lee SK; Kim S; Bae SY; Kim J; Kim M; Kim S; Kil WH; Koo HH; Nam SJ; Bae JW; Lee JE
INSTITUCIÓN / INSTITUTION: - Division of Breast and Endocrine Surgery, Department
of Surgery, Korea University Anam Hospital, Korea University College of Medicine,
Seoul, Korea.
RESUMEN / SUMMARY: - Rhabdomyosarcoma (RMS) of the breast is rare and there is
scant information about the clinical behavior and treatment strategies. We report an
adolescent female patient with metastatic RMS of the breast from the anus. An 18-
year-old female patient was referred to our clinic due to palpable mass in the left
breast. At age seven, she was diagnosed with acute lymphoblastic leukemia and
treated with chemoradiation therapy. After 10 years of complete remission state, she
presented with anal mass which was diagnosed as RMS and she received
chemoradiation therapy. After 1 year of complete remission state, she noticed a
palpable mass in her left breast. The breast mass was diagnosed as metastatic RMS
based on core needle biopsy specimen. The RMS in breast was excised for the
decreasing tumor burden despite of another metastatic lesion. Although rarely
reported, metastasis of RMS should be considered as a cause of breast mass. Tissue
biopsy is recommended when clinically suspected lesion is detected.

[768]
TÍTULO / TITLE: - Management of great vessels and nerves in limb-salvage surgery for
bone and soft tissue tumors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Lun DX; Hu YC; Huang HC
INSTITUCIÓN / INSTITUTION: - Department of Spine Surgery, Weifang People’s
Hospital, Weifang City, Shandong Province, China.
RESUMEN / SUMMARY: - In recent years, limb-salvage surgery has gradually replaced
amputations and become one of the main treatment strategies for patients with bone
and soft tissue tumors of the extremities. The goals of tumor resection in limb-salvage
surgery are to reduce the recurrence rate and preserve as much limb function as possible. However, depending on the size and specific location of the tumor, large neurovascular bundles may be involved. In addition, management of large nerves and vessels can make wide marginal resection more difficult. Sites where these problems commonly arise include the sciatic and tibial common peroneal nerve, artery and vein in the lower limbs.

[769]
**TÍTULO / TITLE:** Sarcomatoid carcinoma of male urethra with bone and lung metastases presenting as urethral stricture.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Badhiwala N; Chan R; Zhou HJ; Shen S; Coburn M

**INSTITUCIÓN / INSTITUTION:** Washington University School of Medicine, St. Louis, MO 63110, USA.

**RESUMEN / SUMMARY:** A 57-year-old man who presented with urinary retention was found to have a sarcomatoid carcinoma of the urethra. Evaluation with CT scan of the abdomen and pelvis revealed multiple pulmonary nodules and osteolytic lesions of left posterior ribs. After external beam radiation therapy and six cycles of systemic chemotherapy, patient underwent a surgical resection of the urethral cancer. After his surgery, patient was also found to have multiple brain metastases and underwent whole brain radiation therapy, nine months after his initial diagnosis. Sarcomatoid carcinomas of the genitourinary tract are extremely rare tumors that require a very aggressive, multimodal treatment approach.

[770]
**TÍTULO / TITLE:** Efficacy and morbidity of temporary low-dose-rate 125I brachytherapy in pediatric rhabdomyosarcomas.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** Hentz C; Barrett W

**INSTITUCIÓN / INSTITUTION:** Department of Radiation Oncology, University of Cincinnati College of Medicine, Cincinnati, OH. Electronic address: hentzcl@mail.uc.edu.

**RESUMEN / SUMMARY:** PURPOSE: Rhabdomyosarcomas (RMSs) are the most common soft tissue tumors in the pediatric population. The American Brachytherapy Society provides recommendations for the use of brachytherapy (BRT) in the treatment of soft tissue sarcomas; yet, there are no clearly defined recommendations for the use of adjuvant BRT in treating RMSs in particular. Radiation therapy has an important role in maximizing local control, and BRT has the advantage over external beam radiation therapy of providing a high dose of radiation to the most susceptible area of recurrence, while delivering a lower dose to the surrounding normal tissue. METHODS AND MATERIALS: This study examines a group of 8 pediatric patients with RMSs who were treated with temporary low-dose-rate 125I BRT and investigates the efficacy and
side effects of such treatment. RESULTS: The results demonstrate a local recurrence rate of 12.5%, with minimal side effects occurring in the patients who had no prior radiation history. Each patient’s side effects are discussed. CONCLUSIONS: The high efficacy and ease of radiation protection for visitors establishes this as an effective treatment that is logistically convenient for patients and families. This is the first report of patients exclusively with RMSs being treated exclusively with 125I BRT and demonstrates promising results.
considering this entity as differential diagnosis even when the lesion is purely intradural with no bony involvement.
tumor in the present case was diagnosed as an ameloblastic fibroma. This is the first report of ameloblastic fibroma in guinea pigs.

[775]
**TÍTULO / TITLE:** - Safety and accuracy of core biopsy in retroperitoneal sarcomas.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)
  ● Enlace al texto completo (gratuito o de pago) 1111/ajco.12125
**AUTORES / AUTHORS:** - Hwang SY; Warrier S; Thompson S; Davidson T; Yang JL; Crowe P
**INSTITUCIÓN / INSTITUTION:** - Prince of Wales Hospital Clinical School, University of New South Wales, Sydney, New South Wales, Australia; Department of Surgery, Prince of Wales Hospital, Sydney, New South Wales, Australia.
**RESUMEN / SUMMARY:** - AIM: Retroperitoneal sarcomas (RPSs) are large, rare tumors. The role of core biopsy for retroperitoneal masses identified by preoperative imaging is unclear and we report the safety and accuracy of core biopsies at a specialized sarcoma unit in Sydney, Australia. METHODS: A retrospective analysis of a prospectively collected database was performed to identify the safety and accuracy of core biopsies in patients who were confirmed to have RPS. RESULTS: Twenty-two patients underwent biopsies with no recorded morbidity or tumor seeding. RPS was correctly identified in 82%. Median follow-up was 19 months. CONCLUSION: We demonstrate that core biopsy is safe and can identify RPSs.

[776]
**TÍTULO / TITLE:** - A Chimeric RNA Characteristic of Rhabdomyosarcoma in Normal Myogenesis Process.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](#)
**REVISTA / JOURNAL:** - Cancer Discov. 2013 Nov 21.
  ● Enlace al texto completo (gratuito o de pago) 1158/2159-8290.CD-13-0186
**AUTORES / AUTHORS:** - Yuan H; Qin F; Movassagh M; Park H; Golden W; Xie Z; Zhang P; Sklar J; Li H
**INSTITUCIÓN / INSTITUTION:** - 1Department of Pathology and 2University of Virginia Cancer Center, University of Virginia, Charlottesville, Virginia; and 3Department of Pathology, Yale University, New Haven, Connecticut.
**RESUMEN / SUMMARY:** - Gene fusions and their chimeric products are common features of neoplasia. Given that many cancers arise by the dysregulated recapitulation of processes in normal development, we hypothesized that comparable chimeric gene products may exist in normal cells. Here, we show that a chimeric RNA, PAX3-FOXO1, identical to that found in alveolar rhabdomyosarcoma, is transiently present in cells undergoing differentiation from pluripotent cells into skeletal muscle. Unlike cells of rhabdomyosarcoma, these cells do not seem to harbor the t(2;13) chromosomal translocation. Importantly, both PAX3-FOXO1 RNA and protein could be detected in the samples of normal fetal muscle. Overexpression of the chimera led to continuous expression of MYOD and MYOG-two myogenic markers that are overexpressed in rhabdomyosarcoma cells. Our results are consistent with a developmental role of a specific chimeric RNA generated in normal cells without the corresponding chromosomal rearrangement at the DNA level seen in neoplastic cells presumably of the same lineage.
TÍTULO / TITLE: Management of Spigelian hernia caused by necrobiotic fibroma of the uterus in a pregnant woman.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Kassir R; Tarantino E; Lacheze R; Brek A; Di Bartolomeo A; Tiffet O

INSTITUCIÓN / INSTITUTION: Department of Digestive Surgery, CHU Hospital, Jean Monnet University, Saint Etienne, France. Electronic address: Radwankassir42@hotmail.Fr.

RESUMEN / SUMMARY: INTRODUCTION: Spigelian hernias are a rare type of hernia through the Spigelian aponeurosis. Spigelian hernias are very uncommon and constitute only 0.12% of all abdominal wall hernias. These hernias are located in the aponeurosis of the internal oblique muscle and transverse abdominal muscle.

PRESENTATION OF CASE: A 30-year-old woman at 28 weeks' gestation was admitted to the obstetrics department due to pain and swelling in the anterior abdominal right region. On inspection, we suspected either a lipoma, a spontaneous hematoma, a tumor of the abdominal wall, or a Spigelian hernia. A Doppler USG and abdominal and pelvic Magnetic Resonance Imaging revealed necrobiotic fibroma of the uterus in Spigelian hernia. The patient was started on dual analgesic and corticotherapy. Overall, the patient improved one week after the acute episode and had no further pain during her gynecologic follow-up. DISCUSSION: We have reported a first case of Spigelian hernia that was complicated by uterine fibroid. The clinical presentation varies, depending on the contents of the hernial sac and the degree of herniation. MRI is the preferred method for accurately identifying masses of the abdominal wall. Our treatment options were based on the extent of the acute-phase reaction and the venous thrombosis. CONCLUSION: It is important to differentiate this rare Spigelian hernia from other hernias as the treatment for this hernia is medical rather than surgical. Before the final choice of treatment is made, digestive surgeons should bear this rare hernia in mind.

TÍTULO / TITLE: Analgesic Effects of Microwave Ablation of Bone and Soft Tissue Tumors Under Local Anesthesia.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: Kastler A; Alnassan H; Pereira PL; Alemann G; Barbe DA; Aubry S; Tiberghien F; Kastler B

INSTITUCIÓN / INSTITUTION: Neuroradiology Department, CLUNI, University Hospital, Grenoble, France; I4S Laboratory-EA 4268-IFR 133, Franche Comte University, Besancon, France.

RESUMEN / SUMMARY: OBJECTIVE: To assess the feasibility and efficacy of microwave ablation (MWA) of painful refractory bone and soft tissue tumors performed under local anesthesia. STUDY DESIGN: A retrospective study between 2011 and
2013. **SETTING:** A single center, Academic Interventional Pain Management Unit.

**SUBJECTS:** Fifteen patients with 25 refractory painful bone (N = 19) or soft tissue (N = 6) tumors treated with MWA were consecutively included. **METHOD:** Local Institutional Review Board approval was obtained, and written informed consent was waived. Lesions included spinal (N = 3), sacral (N = 4), and extraspinal (N = 18) locations. Pain was measured on a visual analog scale (VAS) from 0 to 10 before and immediately after procedure, at 1 week, and on a monthly basis following procedure. MWA procedures were always performed under computed tomography guidance and local anesthesia along with nitrous oxide inhalation. **RESULTS:** Mean ablation time was 4.09 minutes (range 1-11) with an average of 4.2 cycles with a mean ablation power of 60 W. Preprocedure mean VAS score was 7.2 +/- 0.97 (range 6-9). Follow-up postprocedure VAS scores were as follows: day 0: 1.64 +/- 1.86, day 7: 1.82 +/- 1.79, month 1: 2.05 +/- 2.03 (14/15 patients), month 3: 2.13 +/- 1.81, month 6: 2.36 +/- 2.17; and were statistically significant (P < 0.001). Mean pain relief was 5.5 months. **CONCLUSION:** MWA is feasible, safe, and effective in the management of painful refractory bone and soft tissue tumors. It may therefore be considered as a potential alternative to existing percutaneous ablation techniques in the management of bone and soft tissue tumors.

**TÍTULO / TITLE:** Reverse posterior interosseous artery flap for reconstruction of the wrist and hand after sarcoma resection.

**RESUMEN / SUMMARY:**


**AUTORES / AUTHORS:** Wang JQ; Cai QQ; Yao WT; Gao ST; Wang X; Zhang P

**INSTITUCIÓN / INSTITUTION:** Department of Orthopaedic Surgery, Henan Cancer Hospital, Zhengzhou, China.

**RESUMEN / SUMMARY:**

**OBJECTIVE:** To report our experience of posterior interosseous artery flap reconstruction of the wrist and hand after soft tissue sarcoma resection. **METHODS:** Thirteen patients who presented to our institution for treatment of soft tissue sarcomas of the wrist and hand from February 2007 to January 2009 were prospectively enrolled. After tumor resection, the soft tissue defects were covered with posterior interosseous artery flaps. Relevant patient characteristics, results of creation of flap and functional outcomes were reviewed. Relevant clinical characteristics were recorded prior to surgery. The size, pedicle length and thickness of flaps were measured intraoperatively and complications evaluated two weeks after surgery. Functional outcomes and aesthetic results were evaluated 6 to 24 months after surgery, using Enneking’s Musculoskeletal Tumor Society (MSTS) 93 functional scoring system. **RESULTS:** The patients’ ages ranged from 14 to 82 years (54.92 +/- 19.09 years), including six aged >65 years. Flap sizes varied from 5 cm x 3 cm to 8 cm x 5 cm. Flap survival rate was 100%. One patient had partial necrosis of the Z-shaped incision, which healed with routine wound care. Mean Musculoskeletal Tumor Society functional score was 27.31 +/- 3.68. Eleven patients were alive and disease-free at a mean follow-up of 36 months. One patient died of pulmonary metastases 18 months after surgery. One patient developed recurrence and underwent amputation 12 months after surgery. **DISCUSSION:** Reverse posterior interosseous flaps are reliable flaps for reconstruction of the wrist and hand after soft tissue sarcoma resection. They are...
suitable for elderly patients. The one-stage surgery and primary healing allow for timely postoperative adjuvant therapy following this type of reconstruction.

[780]
TITULO / TITLE: - Lipoma arborescens associated with osseous/chondroid differentiation in subdeltoid bursa.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Kim RS; Kim YT; Choi JM; Shin SH; Kim YJ; Kim L
INSTITUCIÓN / INSTITUTION: - Department of Orthopaedic Surgery, School of Medicine, Inha University, Incheon, Korea.
RESUMEN / SUMMARY: - Lipoma arborescens (LA) is a rare benign lesion of unknown etiology. It is characterized histologically by villous proliferation of the synovial membrane and diffuse replacement of the subsynovial tissue by mature fat cells. This condition affects the knee joint most commonly. Cases involving other locations including glenohumeral joint,[1] hip,[2] elbow,[3] hand[4] and ankle[5] have been rarely described. Involvement of the subdeltoid bursa has also been reported, but to date no case has described LA with osseous/chondroid differentiation of this bursa. Another significant finding in our case was the coexistence of LA with intermuscular lipoma, SLAP lesion and labral cyst.

[781]
TITULO / TITLE: - Osteoid differentiation in mesodermal (mullerian) adenosarcoma of ovary.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Patel T; Gupta A; Trivedi P; Shah M
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Gujarat Cancer and Research Institute, M.P. Shah Cancer Hospital, Ahmedabad, Gujarat, India.
RESUMEN / SUMMARY: - A 55-year-old female presented with abdominal pain and 10 cm mass per abdominal examination. Computerized tomography scan of abdomen and pelvis revealed a heterogeneously enhancing solid cystic mass right ovarian mass and mild ascites. Surgery was performed. Specimens were sent for examination. Microscopic examination revealed an admixture of benign but occasionally atypical appearing mullerian type glands with sarcomatous stroma. Solid area showed undifferentiated tumour cells. Atypical mitoses and necrosis were also seen. Areas with extensive benign osteoid surrounded by fibroblastic stroma were also present. Glandular component showed positivity for CK-7, AE-1 and EMA while sarcomatous areas showed positivity for vimentin only. Mullerian adenosarcoma of ovary with sarcomatous overgrowth (SO) having heterologous component was confirmed. Postoperative 3 cycles of chemotherapy was given and the patient was well till date (three months after surgery).

[782]
Juvenile aggressive cemento-ossifying fibroma of the sphenoid sinus with proptosis: a surgical dilemma.

The term fibro-osseous lesion has currently grown in popularity as an overall designation for a number of rare, histologically benign head and neck lesions that are made up of bone, fibrous tissue and cementum. Cemento-ossifying fibroma is a variant of cementifying fibroma and is a fibro-osseous disease. They are usually small innocuous lesions which follow a slow benign course and are commonly seen in the skull bone rather than the sphenoid. It is rare for these tumours to attain large size, behave aggressively, destroy bone and require a radical surgery. One such rapidly growing juvenile cemento-ossifying lesion of sphenoid in our 10-year-old young patient causing proptosis and impaired vision is reported here because of its uncommon nature and its surgical dilemma. Selection of surgical approach to resect this tumour becomes difficult because it is deeply seated and needs a multidisciplinary approach.

MAXILLARY SINUS OSTEOMA: FROM INCIDENTAL FINDING TO SURGICAL MANAGEMENT.

Due to the frequent use and availability of the orthopantomogram (OPG), dental practitioners are more frequently confronted with incidental findings such as osteomas located in the maxillary-/mandibular bone or inside the maxillary sinuses. Osteomas are benign slow-growing osteogenic tumors, which frequently develop in the mandible. In the midface, osteomas appear frequently in the frontoethmoidal sinuses. Maxillary sinus osteoma is a rare entity. Also in asymptomatic patients, cranio-facial osteomas need to be further investigated for a precise diagnosis. The clinical importance of osteomas lies in their differentiation from a malignant lesion such as the osteosarcoma. In patients with multiple osteomas, Gardner’s syndrome (GS) as an underlying disease needs to be excluded. In this report, we present the case of a solitary maxillary sinus osteoma, incidentally found on the OPG. The surgical technique for the removal of the osteoma is presented. In this case, the patient was free of the stigmas associated with GS.
TÍTULO / TITLE: Can proliferation biomarkers reliably predict recurrence in world health organization 2003 defined endometrial stromal sarcoma, low grade?
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Feng W; Malpica A; Skaland I; Gudlaugsson E; Robboy SJ; Dalen I; Hua K; Zhou X; Baak JP
INSTITUCIÓN / INSTITUTION: Department of Gynecology and Shanghai Key Laboratory of Female Reproductive Endocrine-Related Diseases, Obstetrics and Gynecology Hospital of Fudan University, Shanghai, China.
RESUMEN / SUMMARY: An estimated 1500-3000 invasive Endometrial Stromal Sarcomas (ESS) cases annually occur worldwide. Before 2003, ESS was divided as low and high grade ESS based on mitotic activity. In 2003, the WHO changed the names, excluded mitoses and made nuclear atypia and necrosis the essential diagnostic criteria to distinguish ESS, Low Grade (ESS-LG, recurrence-free survival >90%) and Undifferentiated Endometrial Sarcoma (UES, poor prognosis). We have evaluated in WHO2003 defined ESS-LG whether proliferation biomarkers predict recurrence. Using survival analysis, the prognostic value of classical mitosis counts (Mitotic Activity Index, MAI) in haematoxylin-eosin (H&E) sections, and immunohistochemical proliferation biomarkers (Ki-67 and PhosphoHistone-3 (PPH3)) were examined in 24 invasive endometrial stromal sarcomas. Three of 24 (12.5%) ESS-LG recurred. The MAI, PPH3 and Ki-67 were all prognostic (P = 0.001, 0.002 and 0.03). MAI values were >3 in the recurrent cases, but never exceeded 10 (the classical threshold for low and high grade). Non-recurrent cases had 0</=MAI</=3. PPH3 and Ki67 counts can be easier to perform than MAI and therefore helpful in the diagnosis of ESS, Low Grade. In conclusion, in this small study of WHO2003 defined ESS-LG, high levels of proliferation as measured by MAI, PPH3 and Ki-67 are predictive of recurrence. Larger studies are required to confirm these results.

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[785]
TÍTULO / TITLE: Inhibitory effects of tamoxifen and doxorubicin, alone and in combination, on the proliferation of the MG63 human osteosarcoma cell line.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Ouyang ZX; Li XA
INSTITUCIÓN / INSTITUTION: Department of Orthopaedics, Hunan Provincial Tumor Hospital and Tumor Hospital of Xiangya School of Medicine, Central South University, Changsha, Hunan 410013, P.R. China.
RESUMEN / SUMMARY: The present study aimed to compare the combined effect of tamoxifen (TAM) and doxorubicin (ADM) with the individual effects of TAM and ADM alone on the MG63 human osteosarcoma cell line. Estrogen receptor (ER) expression was detected in the MG63 cells using reverse transcription PCR. The morphological changes during the inhibition of cell growth were observed using an inverted microscope and a 3-(4, 5-diethyl-2-thiazol-2-yl)-2,5-diphenyl-tetrazolium bromide (MTT) colorimetric assay following the individual or combined addition of TAM and
ADM. ERalpha and ERbeta expression was detected in the MG63 cells. The typical apoptotic cell morphology was observed in all groups, with the exception of the control group. The MTT colorimetric analysis demonstrated that the rate of inhibition of cell proliferation in the combination group was significantly increased compared with that in the other groups (P<0.05). ERalpha and ERbeta expression was detected in the MG63 human osteosarcoma cells. TAM and ADM alone were able to inhibit cell proliferation. The combination of TAM and ADM significantly enhanced the inhibitory effect, partly through the enhanced sensitivity of the cells to ADM by TAM, which caused the inhibition of cell proliferation and apoptosis.

[786]
TÍTULO / TITLE: - Cowden syndrome complicated by a gastrointestinal stromal tumor.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Nakamura M; Hirooka Y; Yamamura T; Yamada K; Nagura A; Yoshimura T; Ohmiya N; Uehara K; Yoshioka Y; Nagino M; Goto H
INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology and Hepatology, Nagoya University Graduate School of Medicine, Nagoya, Japan.
RESUMEN / SUMMARY: - To our knowledge, this is the first report of Cowden syndrome complicated by a gastrointestinal stromal tumor (GIST) of the small bowel. A 42-year-old female patient was found to have an abdominal mass that was diagnosed as the cause of anemia and was surgically extracted. The surgical specimen was found to be a GIST. During the same period, the patient underwent an endoscopic examination of the entire gastrointestinal tract. She was also diagnosed as having Cowden syndrome based on gastrointestinal polyps and skin, thyroid and breast lesions. Cowden syndrome is associated with germline mutations in the tumor suppressor gene PTEN. PTEN expression may be essential to tumor growth and is a predictive biomarker of the prognosis of both diseases. The present report of such a case is expected to further the analysis of Cowden syndrome.

[787]
TÍTULO / TITLE: - Vascular leiomyoma of the foot: Ultrasound and histologic correlation.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Chavez-Lopez M; Reyna-Olivera G; Pedroza-Herrera G
INSTITUCIÓN / INSTITUTION: - Departamento de Reumatologia, Centenario Hospital Miguel Hidalgo, Aguascalientes, Mexico. Electronic address: drmariochavez@yahoo.com.

[788]
TÍTULO / TITLE: - Emergency excision of cardiac myxoma and endovascular coiling of intracranial aneurysm after cerebral infarction.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Cardiac myxoma is the most common primary tumor of the heart, located mainly in the left atrium. Cerebral embolization or intracranial aneurysm formation as a consequence of left atrial myxomas has been well documented, whereas myxoma embolization causing the combination of cerebral infarction and intracranial myxomatous aneurysm is rare. We report herein, a 67-year-old female with a cardiac myxoma who experienced a left hemispheric embolic ischemic stroke and in addition was found to have right internal carotid artery aneurysm. The patient underwent emergency surgical excision of left atrial myxoma 2 hours after the stroke onset and endovascular coiling of the aneurysm a week later. Although the timing of cardiac surgery is controversial in patients who have had recent ischemic stroke, we recommend immediate resection of cardiac myxoma, if feasible, and early endovascular treatment of associated intracranial myxomatous aneurysms.

[789]

Endoprosthetic replacement after extra-articular resection of bone and soft-tissue tumours around the knee.

We evaluated the clinical results and complications after extra-articular resection of the distal femur and/or proximal tibia and reconstruction with a tumour endoprosthesis (MUTARS) in 59 patients (mean age 33 years (11 to 74)) with malignant bone or soft-tissue tumours. According to a Kaplan-Meier analysis, limb survival was 76% (95% confidence interval (CI) 64.1 to 88.5) after a mean follow-up of 4.7 years (one month to 17 years). Peri-prosthetic infection was the most common indication for subsequent amputation (eight patients). Survival of the prosthesis without revision was 48% (95% CI 34.8 to 62.0) at two years and 25% (95% CI 11.1 to 39.9) at five years post-operatively. Failure of the prosthesis was due to deep infection in 22 patients (37%), aseptic loosening in ten patients (17%), and peri-prosthetic fracture in six patients (10%). Wear of the bearings made a minor revision necessary in 12 patients (20%). The mean Musculoskeletal Tumor Society score was 23 (10 to 29). An extensor lag > 10 degrees was noted in ten patients (17%). These results suggest that limb salvage after extra-articular resection with a tumour prosthesis can achieve good
functional results in most patients, although the rates of complications and subsequent amputation are higher than in patients treated with intra-articular resection.

[790]
**Título / Title:** - Avoiding unplanned resections of wrist sarcomas: an algorithm for evaluating dorsal wrist masses.

**Resumen / Summary:** - Enlace al Resumen / Link to its Summary

**Revista / Journal:** - Am J Orthop (Belle Mead NJ). 2013 Sep;42(9):401-6.

**Autores / Authors:** - Crosby SN; Alamanda VK; Weikert DR; Holt GE

**Institución / Institution:** - Orthopaedic Resident, Department of Orthopaedic Surgery, Vanderbilt University Medical Center, Nashville, Tennessee. samuel.n.crosby@vanderbilt.edu.

**Resumen / Summary:** - Ganglion cysts, soft-tissue masses that commonly occur about the wrist, are often excised without imaging or biopsy. In this article, we report a series of incompletely excised soft-tissue sarcomas about the wrist and offer an algorithm for their evaluation. We describe a series of 4 consecutive patients who each presented after incomplete resection of a soft-tissue sarcoma mistakenly diagnosed as a ganglion cyst. We also retrospectively review the cases of 7 patients with incompletely excised sarcomas of the wrist. Three of the 4 patients with sarcomas mistaken for ganglion cysts did not have prior magnetic resonance imaging (MRI), 3 of the 4 did not have an attempted aspiration, and all 4 did not have transillumination. Common atypical characteristics included ulna-based lesions (3/4), symptoms for less than 6 months (3/4), and no appreciable fluctuation in size (3/4). Functional outcomes for all patients were poor because of multiple surgical procedures, re-excisions requiring flaps, and need for additional adjuvant therapies. Dorsal wrist masses with atypical characteristics should be approached with caution. Transillumination and aspiration are 2 accessible, cost-efficient methods for evaluating these masses. If either test is abnormal, an MRI should be performed.

[791]
**Título / Title:** - Pelvic sacral and hemi lumbar spine resection of low grade pelvic chondrosarcoma: A multistage procedure involving vascular bypass, spine fixation and vascular exclusion.

**Resumen / Summary:** - Enlace al Resumen / Link to its Summary


**Autores / Authors:** - Zoccali C; Marolda G; Di Francesco A; Favale L; Salducca N; Biagini R

**Institución / Institution:** - Oncological Orthopaedics Department, Muscular-skeletal Tissue Bank, IFO-Regina Elena National Cancer Institute, Via Elio Chianesi 53, 00144 Rome, Italy; Department of Surgical Science, University of L'Aquila, L'Aquila, Italy. Electronic address: carminezoccali@libero.it.

**Resumen / Summary:** - Peripheral chondrosarcoma is a rare tumor particularly insidious when arising from the pelvis, becoming symptomatic later in time when surgery may be too difficult and dangerous due to this complex area. In the present case, the tumor arose from an exostosis located on the medial surface of the left iliac wing. Its diameter was 25cmx20cmx15cm, adhering to the last three vertebrae,
involving the left iliac vein and artery, displacing the left ureter. In a similar case, a hindquarter amputation is indicated but, if the patient refuses, a resection remains possible. In this paper, we describe a multistage technique consisting of an extra-anatomic vascular bypass, a lumbar stabilization, a neurovascular bundles anterior isolation and a posterolateral resection of this mass. After a five-year follow-up, the patient is alive and able to stand and walk with support, after undergoing twice lung metastasis removal.

[792]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace a la Editora de la Revista http://bmj.com/search.dtl
●● Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-201102
AUTORES / AUTHORS: - Khawaja A; Maheshwari PK; Haque A; Zubairi AB
INSTITUCIÓN / INSTITUTION: - Medical College, The Aga Khan University Hospital, Karachi, Sindh, Pakistan.
RESUMEN / SUMMARY: - Primary neoplasms of the respiratory tract are rarely encountered in the paediatric population. Inflammatory myofibroblastic tumour (IMT) is a rare soft tissue mesenchymal tumour but a distinct disease entity accounting for less than 1% of all primary lung tumours. We report a case of a 10-year-old boy who presented with respiratory failure and left lung collapse. On flexible fiberoptic bronchoscopy, a pedunculated mass in the lower part of the trachea originating from the left main stem bronchus was identified. The patient subsequently underwent a left-sided pneumonectomy with complete resection of the mass. The histopathological analysis was consistent with IMT. Two years of follow-up and the patient remains well.

[793]
TÍTULO / TITLE: - The natural history of uterine leiomyomas: morphometric concordance with concepts of interstitial ischemia and inanosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o de pago) 1155/2013/285103
AUTORES / AUTHORS: - Flake GP; Moore AB; Flagler N; Wicker B; Clayton N; Kissling GE; Robboy SJ; Dixon D
INSTITUCIÓN / INSTITUTION: - Cellular and Molecular Pathology Branch, National Toxicology Program (NTP), National Institute of Environmental Health Sciences (NIEHS), National Institutes of Health (NIH), Department of Health and Human Services, Research Triangle Park, NC 27709, USA.
RESUMEN / SUMMARY: - Based upon our morphologic observations, we hypothesize and also provide morphometric evidence for the occurrence of progressive developmental changes in many uterine fibroids, which can be arbitrarily divided into 4 phases. These developmental phases are related to the ongoing production of extracellular collagenous matrix, which eventually exceeds the degree of angiogenesis,
resulting in the progressive separation of myocytes from their blood supply and a condition of interstitial ischemia. The consequence of this process of slow ischemia with nutritional and oxygen deprivation is a progressive myocyte atrophy (or inanition), culminating in cell death, a process that we refer to as inanosis. The studies presented here provide quantitative and semiquantitative evidence to support the concept of the declining proliferative activity as the collagenous matrix increases and the microvascular density decreases.

[794]

**TITULO / TITLE:** - Permanent activation of HMGA2 in lipomas mimics its temporal physiological activation linked to the gain of adipose tissue.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Thies HW; Nolte I; Wenk H; Mertens F; Bullerdiek J; Markowski DN

**INSTITUCIÓN / INSTITUTION:** - Center of Human Genetics, University of Bremen, Leobener Strasse ZHG, D-28359 Bremen, Germany.

**RESUMEN / SUMMARY:** - OBJECTIVE: In this study the activation of HMGA2 and overexpression by FGF1-driven stimulation of adipose tissue derived stem cells (ADSCs) in adipose tissue tumors were analyzed. In addition, the expression of HMGA2 and PPAR-gamma mRNA were quantified in canine subcutaneous abdominal adipose tissue from normal and overweight purebred dogs. DESIGN AND METHODS: ADSCs and adipose tissue explants stimulated with FGF1 followed by gene expression analyses of HMGA2 and p14Arf using Western-blot and qRT-PCR. Furthermore, canine subcutaneous white adipose tissue (WAT) were analyzed by qRT-PCR for their expression of HMGA2 and PPAR-gamma. RESULTS: ADSCs and adipose tissue explants are able to execute a HMGA2 response upon FGF1 stimulation. FGF1 enhances proliferation of ADSCs by a HMGA2-dependent mechanism. In lipomas increase of HMGA2 is accompanied by increased expression of p14Arf. Furthermore, a significantly elevated level of HMGA2 in overweight dogs and a negative correlation between the expression of HMGA2 and PPAR-gamma in subcutaneous cWAT were noted. CONCLUSIONS: These results suggest that WAT contains cells that as essential part of adipogenesis up-regulate HMGA2 resulting from growth factor stimulation. In subgroups of lipoma, constitutive activation of HMGA2 due to rearrangements replaces the temporal response triggered by growth factors.

[795]

**TITULO / TITLE:** - Laparoscopic transgastric partial gastrectomy for a posterior fundic gastrointestinal stromal tumor.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Abdalla S; Baton O; Rouquie D; Boulanger T; Chapuis O

**INSTITUCIÓN / INSTITUTION:** - Service de chirurgie viscerale, hospital d’Instruction des Armees du Val-de-Grace, 74, boulevard de Port-Royal, 75230 Paris, France. Electronic address: solafah@hotmail.fr.
Inflammatory Myofibroblastic Tumor of the Urinary Bladder Managed by Laparoscopic Partial Cystectomy.

RESUMEN / SUMMARY: Inflammatory myofibroblastic tumor of the urinary bladder is a rare mesenchymal tumor with uncertain malignant potential. It often mimics soft tissue sarcomas both clinically and radiologically. Surgical resection in the form of partial cystectomy or transurethral resection remains the mainstay of treatment. Herein we report the case of an inflammatory myofibroblastic tumor in a young girl, which was managed by laparoscopic partial cystectomy. To the best of our knowledge, this is the first reported case of laparoscopic management of an inflammatory myofibroblastic tumor of the urinary bladder.

Maxillary bone myxoma.

RESUMEN / SUMMARY: INTRODUCTION: Maxillary bone myxoma is a rare benign mesenchymal tumor, slow-growing but locally aggressive. Pathogenesis remains disputed. OBJECTIVE: To study the clinical, radiological and histological features and treatment of maxillary myxoma, based on a pediatric case report. CASE REPORT: An infant of two and a half months presented with endonasal tumor extending to ethmoid. Surgical excision was performed on an endonasal approach. Myxoma was diagnosed by histologic examination of the surgical specimen, whereas initial biopsy had suggested fibrous dysplasia. No recurrence was observed after two and a half years’ surveillance. CONCLUSION: Positive diagnosis of maxillary myxoma is histological. Treatment is primarily surgical. Strict long-term surveillance is required because of the high risk of recurrence.

Central odontogenic fibroma of the maxilla.

RESUMEN / SUMMARY: INTRODUCTION: Maxillary bone myxoma is a rare benign mesenchymal tumor, slow-growing but locally aggressive. Pathogenesis remains disputed. OBJECTIVE: To study the clinical, radiological and histological features and treatment of maxillary myxoma, based on a pediatric case report. CASE REPORT: An infant of two and a half months presented with endonasal tumor extending to ethmoid. Surgical excision was performed on an endonasal approach. Myxoma was diagnosed by histologic examination of the surgical specimen, whereas initial biopsy had suggested fibrous dysplasia. No recurrence was observed after two and a half years’ surveillance. CONCLUSION: Positive diagnosis of maxillary myxoma is histological. Treatment is primarily surgical. Strict long-term surveillance is required because of the high risk of recurrence.
The central odontogenic fibroma (COF) is a rare benign odontogenic mesenchymal tumor of jaw bones. The World Health Organization (WHO) recognizes two variants of COF namely: 1) Epithelial-rich type (WHO) and 2) epithelial-poor type (simple type). Rare variants like ossifying COF, COF associated with giant cell lesions, and amyloid have been documented. This article presents a case of an epithelial-rich variant of COF in a 24-year-old female. It presented as a bony swelling of the maxilla and appeared as a mixed lesion in radiographs. Histopathology showed a highly cellular fibrous connective tissue stroma with plump fibroblasts and long strands of odontogenic epithelium exhibiting mild eosinophilic to clear cytoplasm. Numerous cementum-like hematoxyphilic calcifications of various sizes akin to dentin or acellular cementum were observed. We believe that clinical and radiographic features of this case may add valuable knowledge to the already existing literature.

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Primary malignant fibrous histiocytoma involving the left pulmonary vein presenting as a left atrial tumor.

A 35-year-old woman presented with 4 months history of progressively increasing intermittent dyspnea and hemoptysis. Transthoracic echocardiography revealed a loculated mass in the left atrium (LA). A provisional diagnosis of LA myxoma was made. Intraoperatively the tumor was found extending into and closely adherent to the left pulmonary vein and could not be completely cleared off from the pulmonary venous wall. The histopathological examination of the tumor revealed it to be a myxoid malignant fibrous histiocytoma.

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Gastrointestinal stromal tumors and second primary malignancies before and after the introduction of imatinib mesylate.

A very late recurrence of a formerly misdiagnosed low grade endometrial stromal sarcoma metastasized to the colon.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Bakker IS; Hoven-Gondrie ML; Moll FC; de Haan HH
INSTITUCIÓN / INSTITUTION: Department of Surgery, University Medical Center Groningen, University of Groningen, Groningen, The Netherlands. Electronic address: i.s.bakker@umcg.nl.
RESUMEN / SUMMARY: INTRODUCTION: Endometrial stromal sarcomas are rare mesenchymal neoplasms of the uterus with an indolent clinical course but a high risk of recurrence. PRESENTATION OF CASE: We report a case of a 78 year old woman who presented with rectal bleeding and recurrent urinary tract infections, caused by a very late recurrence of a formerly misdiagnosed low grade endometrial stromal sarcoma, metastasized to the colon. DISCUSSION: Endometrial stromal sarcomas are difficult to diagnose, both due to the rarity of the tumor and because of the close resemblance of the tumor to normal stromal tissue. These tumors are known for a high tendency of recurrence, therefore long term follow up is required in patients with endometrial stromal sarcoma. CONCLUSION: In patients with a history known for endometrial stromal sarcoma recurrence should always be considered.

TÍTULO / TITLE: GSTT1 copy number gain and ZNF overexpression are predictors of poor response to imatinib in gastrointestinal stromal tumors.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Lee EJ; Kang G; Kang SW; Jang KT; Lee J; Park JO; Park CK; Sohn TS; Kim S; Kim KM
INSTITUCIÓN / INSTITUTION: Department of Pathology, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea.
RESUMEN / SUMMARY: Oncogenic mutations in gastrointestinal stromal tumors (GISTs) predict prognosis and therapeutic responses to imatinib. In wild-type GISTs, the tumor-initiating events are still unknown, and wild-type GISTs are resistant to imatinib therapy. We performed an association study between copy number alterations (CNAs) identified from array CGH and gene expression analyses results for four wild-type GISTs and an imatinib-resistant PDGFRA D842V mutant GIST, and compared the results to those obtained from 27 GISTs with KIT mutations. All wild-type GISTs had multiple CNAs, and CNAs in 1p and 22q that harbor the SDHB and GSTT1 genes, respectively, correlated well with expression levels of these genes. mRNA expression levels of all SDH gene subunits were significantly lower (P<=0.041), whereas mRNA expression levels of VEGF (P=0.025), IGF1R (P=0.026), and ZNFs (P<0.05) were significantly higher in GISTs with wild-type/PDGFRA D842V mutations than GISTs with KIT mutations. qRT-PCR validation of the GSTT1 results in this cohort and 11 additional malignant GISTs showed a significant increase in the frequency of GSTT1 CN gain and increased mRNA expression of GSTT1 in wild-type/PDGFRA D842V GISTs than KIT-mutant GISTs (P=0.033). Surprisingly, all four malignant GISTs with KIT exon 11 deletion mutations with primary resistance to imatinib had an increased
GSTT1 CN and mRNA expression level of GSTT1. Increased mRNA expression of GSTT1 and ZNF could be predictors of a poor response to imatinib. Our integrative approach reveals that for patients with wild-type (or imatinib-resistant) GISTs, attempts to target VEGFRs and IGF1R may be reasonable options.

[803] TÍTULO / TITLE: - Giant cell tumor arising from anterior arc of the rib.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Heo W; Kang do K; Min HK; Jun HJ; Hwang YH
INSTITUCIÓN / INSTITUTION: - Department of Thoracic and Cardiovascular Surgery, Inje University Haeundae Paik Hospital, Inje University College of Medicine, Korea.
RESUMEN / SUMMARY: - A primary giant cell tumor of the rib is very rare. The most common site of a giant cell tumor arising from the rib is the posterior arc. A giant cell tumor arising from the anterior arc of the rib is extremely rare. The treatment of a giant cell tumor of the rib is not well defined. Generally, a complete surgical resection is performed in a patient with a primary giant cell tumor of the rib. We report a case of a giant cell tumor arising from the anterior arc of the rib that was treated with a wide excision and chest wall reconstruction.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Seethala S
INSTITUCIÓN / INSTITUTION: - Srikanth Seethala, Department of Internal Medicine, University of New Mexico, Albuquerque, NM 87131, United States.
RESUMEN / SUMMARY: - Left ventricular myxomas account for 2.5% of all cardiac myxoma cases. There are very few case reports on left ventricular myxoma (LVM) presented after complete surgical resection of left atrial myxoma. Here we report a case of a 58-year-old male presented to the hospital for transient limb weakness, numbness and dysarthria. Magnetic resonance image of the brain revealed multiple thromboembolic cerebrovascular accidents. Transthoracic echocardiogram (TTE) revealed a left atrial myxoma. It was resected completely with good surgical margins. After one and half year he started having dizziness, and transient right sided weakness. Computer tomography scan of the head revealed a progression of thromboembolic disease. TTE revealed a LVM that was confirmed by transesophageal echocardiogram. It was resected with good surgical margins 3 wk after recurrent cerebrovascular accident.

[805] TÍTULO / TITLE: - Left atrial myxoma presenting with unusual cystic form.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
**Cardiac myxomas are the most common primary benign tumors of uncertain etiology. They usually present as polypoid or oval-shaped masses projecting into a heart chamber from the interatrial septum and have a soft, gelatinous consistency without a cystic structure. We report a case of left atrial myxoma with a single cystic form.**

[806]

**Título / Title:** Recurrent and self-remitting sixth cranial nerve palsy: pathophysiological insight from skull base chondrosarcoma.

**Resumen / Summary:** Palsy of the abducens nerve is a neurological sign that has a wide range of causes due to the nerve's extreme vulnerability. Need of immediate neuroimaging is a matter of debate in the literature, despite the risks of delaying the diagnosis of a skull base tumor. The authors present 2 cases of skull base tumors in which the patients presented with recurrent and self-remitting episodes of sixth cranial nerve palsy (SCNP). In both cases the clinical history exceeded 1 year. In a 17-year-old boy the diagnosis was made because of the onset of headache when the tumor reached a very large size. In a 12-year-old boy the tumor was incidentally diagnosed when it was still small. In both patients surgery was performed and the postoperative course was uneventful. Pathological diagnosis of the tumor was consistent with that of a chondrosarcoma in both cases. Recurrent self-remitting episodes of SCNP, resembling transitory ischemic attacks, may be the presenting sign of a skull base tumor due to the anatomical relationships of these lesions with the petroclival segment of the sixth cranial nerve. Physicians should promptly recommend neuroimaging studies if SCNP presents with this peculiar course.

[807]

**Título / Title:** Bimaxillary unilateral gingival fibromatosis with localized aggressive periodontitis (eating the tooth at the same table).

**Resumen / Summary:**

**INSTITUCIÓN / INSTITUTION:** - Department of Oral Surgery ACPM Dental Collage and Hospital, Dhule, Maharashtra, India.

**RESUMEN / SUMMARY:** - This case reports a unique presentation of two different clinical entities amidst few similarities and differences. Usually, aggressive periodontitis and gingival fibromatosis occur independently. Their simultaneous occurrence is rarely found. This report deals with the clinical features and management aspect of such a case.

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**TÍTULO / TITLE:** - Malignant inflammatory myofibroblastic tumor of the maxillary sinus.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


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**TÍTULO / TITLE:** - Lingual neurofibroma causing dysaesthesia of the tongue.

**RESUMEN / SUMMARY:** - British Medical J (BMJ). Acceso gratuito al texto completo.

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**TÍTULO / TITLE:** - Neurofibroma is a benign tumour derived from the neural sheaths of peripheral nerves and composed of Schwann cells, fibroblast-like cells and intermediate cells. The usual clinical presentation of an oral neurofibroma is swelling. A 62-year-old woman was referred to our department of otorhinolaryngology with irritation.
and dysaesthesia of the lateral aspect of the tongue. The only finding was a slightly red
area from which a biopsy was taken. The macroscopic findings observed by the
surgeon were consistent with normal tongue tissue. The histopathological examination
showed a small, rounded tumour closely approximated to an invagination of the surface
epithelium and with a small lymphatic infiltrate. The tumour was a neurofibroma. A
Schwannoma type B was considered but the presence of small nerves and positive
neurofilament reaction favoured a neurofibroma. The patient had no other
neurofibromas or skin lesions. At a 3-week follow-up visit all symptoms had resolved.

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**TÍTULO / TITLE:** An unusual cause of spontaneous hemothorax: cardiac
angiosarcoma.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** Turk Kardiyol Dern Ars. 2013 Sep;41(6):526-8. doi:

**AUTORES / AUTHORS:** Lafci G; Cagli K; Tok D; Yalcinkaya A

**INSTITUCIÓN / INSTITUTION:** Department of Cardiovascular Surgery, Turkiye Yuksek
Ihtisas Hospital, Ankara, Turkey.

**RESUMEN / SUMMARY:** Angiosarcoma, the most common primary malignant
neoplasm of the heart in adults, usually presents as pericardial effusion or right-sided
heart failure. Rupture of an angiosarcoma-infiltrated cardiac chamber as a cause of
hemothorax is very rare in the literature. In this report, we describe a 34-year-old male
patient, who presented to emergency service with sudden chest pain and dyspnea. The
diagnostic work-up revealed spontaneous right-sided hemothorax and a large right
atrial (RA) mass with suspicious atrial perforation. An urgent surgery showed a
vascularized irregular RA mass invading the parietal pericardium and pleura and a
perforation of the RA free wall. Histopathologic examination confirmed the diagnosis of
angiosarcoma, and the patient was subsequently referred for radiotherapy and
chemotherapy.

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**TÍTULO / TITLE:** Primary cardiac angiofibroma.

**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** Korean Circ J. 2013 Sep;43(9):636-9. doi:

**AUTORES / AUTHORS:** Kim YJ; Kim YJ; Kim SH; Youn YN; Park S

**INSTITUCIÓN / INSTITUTION:** Cardiology Division, Severance Cardiovascular
Hospital, Yonsei University College of Medicine, Seoul, Korea.

**RESUMEN / SUMMARY:** Cardiac Angiofibroma is an uncommon intracardiac tumor.
Thus far, only 4 cases of the rare intracardiac tumor have been reported. The present
case-report describes an intracardiac angiofibroma in a 57-year-old healthy female.
The patient was incidentally diagnosed with a left ventricle mass during
echocardiography. We performed cardiac imaging, surgical excision and histological
evaluation of the mass. The angiofibroma demonstrated features different from the
relatively common cardiac tumors such as fibroma, myxoma and angiosarcoma. The
cardiac MRI showed slightly high signal intensity on both T1 and T2, with the central
core of lower signal intensity. The resected tumor was a whitish and rubbery mass.
Histologically, the tumor showed the benign vascular proliferations associated with the surrounding collagen deposition.

[812] TÍTULO / TITLE: Giant cell angiofibroma of the scalp: A benign rare neoplasm with bone destruction.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Arifin MZ; Tjahjono FP; Faried A; Gill AS; Cahyadi A; Hernowo BS
INSTITUCIÓN / INSTITUTION: Department of Neurosurgery, Universitas Padjadjaran-Dr. Hasan Sadikin General Hospital, Bandung, Indonesia; Oncology Working Group, Health Research Unit, Universitas Padjadjaran-Dr. Hasan Sadikin General Hospital, Bandung, Indonesia.
RESUMEN / SUMMARY: BACKGROUND: The incidence of extraorbital giant cell angiofibroma (GCA) is rare, with only one case located in the scalp reported in the literature. The morphological hallmark is histopathological examination showing richly vascularized pattern-less spindle cell proliferation containing pseudovascular spaces and floret-like multinucleate giant cells. CASE DESCRIPTION: We report a case of a 30-year-old female with a primary complaint of a painless solitary nodule arising on the left parietal region of the scalp. Complete tumor removal through surgical intervention was achieved, and the postoperative period was uneventful. CONCLUSION: Diagnosing a highly vascularized tumor in the head and neck is challenging. Our case is unique in that it is presented as a GCA of the scalp, which is an extremely rare clinical entity, and also demonstrated bone destruction.

[813] TÍTULO / TITLE: A rare presentation of an intraosseous lipoma in the proximal femur.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Rabbani SA; Ilyas I; Alrumaih H
INSTITUCIÓN / INSTITUTION: King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia.
RESUMEN / SUMMARY: Patient: Female, 32 Final Diagnosis: Intraosseous lipoma
Symptoms: Swelling of the thigh Medication: - Clinical Procedure: Excision of the mass Specialty: Surgery. OBJECTIVE: Rare disease. BACKGROUND: Intraosseous lipomas happen to be one of rarest benign soft tissue tumors with only a little known about its etiology and pathophysiology. A pubmed search using key word “Intraosseous lipoma”. Came back with 165 results. All the sizes and presentations were reviewed and compared with the case we are presenting. We are reporting a 15x20 cm intraosseous lipoma. CASE REPORT: 32-year-old lady with a large intraosseous lipoma in the proximal thigh. The largest reported in English literature. CONCLUSIONS: A intraosseous lesion with this size could still be benign even if its lager than 5 cm. Keeping in mind that with a lesion this size malignancy should always be ruled out.
TÍTULO / TITLE: - Undifferentiated uterine sarcoma metastatic to the brain.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Stofko DL
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Philadelphia College of Osteopathic Medicine, Philadelphia, PA 19131, USA.
RESUMEN / SUMMARY: - BACKGROUND: Undifferentiated uterine sarcoma (UUS) is a rare tumor with an aggressive growth pattern. They occur in women from 40 to 60 years and are generally characterized by poor prognosis, a high rate of local recurrence, and distant metastases. UUS accounts for 0.2% of all gynecological malignancies. Possible treatments include surgery, radiotherapy, and chemotherapy.
CASE DESCRIPTION: A 65-year-old female with postmenopausal bleeding was found to have a uterine mass for which she underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy, and omentectomy. The pathologic evaluation was consistent with undifferentiated endometrial sarcoma. She began experiencing headaches with associated visual disturbances. Magnetic resonance imaging (MRI) of the brain showed a homogenous enhancing occipital dural-based mass measuring 1.6 x 1.8 x 1.7 cm. Due to the rarity of metastatic uterine sarcoma to the brain, this was believed to represent a meningioma and subsequently observed. Interval MRI scan revealed a significant increase in size of the right occipital mass to 2.3 cm with increased edema and mass effect. She underwent right occipital image guided craniotomy for resection of the mass. Histopathology confirmed UUS metastases.
CONCLUSION: Randomized trials analyzing these treatment options are limited due to the rarity of this disease; therefore, a standard therapy is not established. Based on a review of the literature, this is only the fourth case reported of UUS metastatic to the brain.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Guttikonda V; Taneeru S; Gaddipati R; Madala J
INSTITUCIÓN / INSTITUTION: - Department of Oral Pathology and Microbiology, Mamata Dental College, Khammam, Andhra Pradesh, India.
RESUMEN / SUMMARY: - Juvenile ossifying fibroma (JOF) is an uncommon, benign, bone-forming neoplasm with an aggressive local growth that is distinguished from other fibro-osseous lesions primarily by its age of onset, clinical presentation and aggressive behaviour. JOF is considered as a variant of the ossifying fibroma (OF) and the former includes psammomatoid JOF (PsJOF) and Trabecular JOF (TrJOF). Both variants involve the craniofacial bones with the trabecular variant being more common in the jaws and the psammomatoid variant being more common in the craniofacial skeleton.
PsJOF is an unique variant of JOF that has a predilection for the sinonasal tract and the orbit particularly centered on the periorbital, frontal, and ethmoid bones. We report a rare case of massive PsJOF involving the maxillary sinus in a 20-year-old female.

[816]  
**TÍTULO / TITLE:** Chordoma of skull base presenting as nasopharyngeal mass.  
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary  
**AUTORES / AUTHORS:** Kataria SP; Batra A; Singh G; Kumar S; Sen R  
**INSTITUCIÓN / INSTITUTION:** Department of Pathology, Pt B.D. Sharma PGIMS, Rohtak, Haryana, India.  
**RESUMEN / SUMMARY:** While the nasopharynx is most commonly regarded by the otolaryngologist as a primary site of neoplastic involvement, it is also an avenue of spread of base-of-the-skull tumors presenting as bulging nasopharyngeal masses. Chordoma is a relatively rare tumor of the skull base and sacrum thought to originate from embryonic remnants of the notochord. Chordomas arising from the skull base/clivus are typically locally aggressive with lytic bone destruction. The optimal treatment may be photon/proton radiotherapy alone or combined with a gross total resection, when feasible. We report a case of intracranial chordoma presenting as nasopharyngeal mass.

[817]  
**TÍTULO / TITLE:** Large atrial myxoma mimicking severe mitral stenosis associated with right heart enlargement and severe pulmonary hypertension.  
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary  
**AUTORES / AUTHORS:** Leo S; Yang K; Weng C; Liang Z  
**INSTITUCIÓN / INSTITUTION:** Department of Cardiology, Third Xiangya Hospital, Central South University, Changsha, Hunan 410013, P.R. China.

[818]  
**TÍTULO / TITLE:** Moritz (Maurice) Kaposi: Kaposi’s sarcoma.  
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary  
**REVISTA / JOURNAL:** J Perioper Pract. 2013 Sep;23(9):208.  
**AUTORES / AUTHORS:** Ellis H  
**INSTITUCIÓN / INSTITUTION:** Department of Anatomy, University of London, Guy’s Campus, London, SE1 1UL.

[819]  
**TÍTULO / TITLE:** Peripheral ossifying fibroma.  
**RESUMEN / SUMMARY:** Enlace al Resumen / Link to its Summary  
**ENLACE AL TEXTO COMPLETO (GRATUITO O DE PAGO):** 4103/0972-124X.118325
AUTORES / AUTHORS: - Mohiuddin K; Priya NS; Ravindra S; Murthy S
INSTITUCIÓN / INSTITUTION: - Department of Periodontics, Al-Ameen Dental College, Bijapur, India.
RESUMEN / SUMMARY: - Peripheral ossifying fibroma (POF) is one of the inflammatory reactive hyperplasia of gingiva. It represents a separate clinical entity rather than a transitional form of pyogenic granuloma and shares unique clinical characteristics and diverse histopathological features. We present a case of POF in a 65-year-old male patient in the posterior maxillary gingiva, the clinical presentation of which differs from the usual presentation. Differential diagnosis and some interesting facts of POF are discussed.

[820]
TÍTULO / TITLE: - INCIDENCE OF SOFT TISSUE SARCOMAS IN AN ITALIAN AREA AFFECTED BY ILLEGAL WASTE DUMPING SITES.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Benedetti M; Fazzo L; Buzzoni C; Comba P; Magnani C; Fusco M
INSTITUCIÓN / INSTITUTION: - a Unit of Environmental Epidemiology, Department of Environment and Primary Prevention, National Institute of Health, Rome, Italy.
RESUMEN / SUMMARY: - Abstract Aim of the present study was to investigate the possible association between occurrence of soft tissue sarcomas (STS) and residence in an Italian area affected by illegal practices of dumping and setting fire to both hazardous and solid urban waste. Standardized Incidence Ratios (SIR) were computed separately for STS and some specific STS subtypes. The analysis was performed for the total population and for specific age-groups, namely children, adolescents and adults. In adults, no significant increase in STS was found other than for gastrointestinal stromal tumours in males. A non-significant increase in incidence of STS was observed for male children and female adolescents. The results of the present study do not allow conclusions for a causal association. In the absence of previous epidemiological studies on this issue further investigation are needed.

[821]
TÍTULO / TITLE: - Duodenal metastasis from subcutaneous angiosarcoma of the head: Rare cause of obscure gastrointestinal bleeding.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Katsurahara M; Horiki N; Takei Y
INSTITUCIÓN / INSTITUTION: - Department of Endoscopic Medicine, Mie University Graduate School of Medicine, Tsu, Japan.

[822]
TÍTULO / TITLE: - Probable initial pulmonary lymphangioleiomyomatosis and mediastinal lymphangioleiomyoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Pontes M; Barbosa C; Coelho ML; Carvalho L

INSTITUCIÓN / INSTITUTION: - Servico de Anatomia Patologica, Centro Hospitalar Universitario de Coimbra, Hospitales da Universidade de Coimbra, Coimbra, Portugal. Electronic address: manuelpontes1@sapo.pt.

RESUMEN / SUMMARY: - A 68 year old woman was submitted to a mediastinal lymphangioleiomyoma resection found in a follow-up study of lower left lung resection due to bronchiectasis complicated by chylothorax. This led to a revaluation of the pulmonary specimen that revealed, in addition to inflammatory bronchiecstasy, small spindle cell nodules in the lung parenchyma, similar to minute pulmonary meningotheial-like nodules, but with smooth muscle actin immunohistochemical positivity. The possibility of initial pulmonary development of lymphangioleiomyomatosis is discussed.

[823]

TÍTULO / TITLE: - Incidental finding of isolated colonic neurofibroma.

AUTORES / AUTHORS: - Chelimilla H; Chandrala CK; Niazi M; Kumbum K

INSTITUCIÓN / INSTITUTION: - Division of Gastroenterology, Department of Medicine, Bronx Lebanon Hospital Center, Affiliated with Albert Einstein College of Medicine, Bronx, N.Y., USA.

RESUMEN / SUMMARY: - Neurofibromatosis is a genetic disorder manifested by characteristic cutaneous lesions called neurofibromas. There are two distinct neurocutaneous syndromes named neurofibromatosis type 1 (also called von Recklinghausen disease or NF1) and neurofibromatosis type 2 (NF2). NF1 is by far the most common presentation and is caused by an autosomal dominant mutation in the NF1 gene mapped to chromosome 17q11.2. The literature shows that gastrointestinal involvement is noted in systemic neurofibromatosis in up to 25% of patients, but isolated intestinal neurofibromatosis is a very rare manifestation. We herein present the case of a 70-year-old woman who was diagnosed with an isolated colonic neurofibroma without any systemic signs of neurofibromatosis; only a few case reports of this condition have been published to date.

[824]


AUTORES / AUTHORS: - Zhang Z; Gu B; Zhu W; Zhu L

INSTITUCIÓN / INSTITUTION: - Orthopedic Department, The Affiliated Taizhou People’s Hospital of Nantong University, Taizhou, Jiangsu 225300, P.R. China.
RESUMEN / SUMMARY: This study was conducted to evaluate the bioactivity of manganese-incorporated TiO2 (Mn-TiO2) coating prepared on titanium (Ti) plate by plasma electrolytic oxidation (PEO) technique in Ca-, P- and Mn-containing electrolytes. The surface topography, phase and element compositions of the coatings were investigated using scanning electron microscopy (SEM), X-ray diffraction (XRD) and energy dispersive spectrometry (EDS), respectively. The adhesion of osteoblast-like MG63 cells onto Ti, TiO2 and Mn-TiO2 surfaces was evaluated, and the signal transduction pathway involved was confirmed by the sequential expression of the genes for integrins beta1, beta3, alpha1 and alpha3, focal adhesion kinase (FAK), and the extracellular regulated kinases (ERKs), including ERK1 and ERK2. The results obtained indicated that Mn was successfully incorporated into the porous nanostructured TiO2 coating, and did not alter the surface topography or the phase composition of the coating. The adhesion of the MG63 cells onto the Mn-incorporated TiO2 coating was significantly enhanced compared with that on the Mn-free TiO2 coating and the pure Ti plates. In addition, the enhanced cell adhesion on the Mn-TiO2 coatings may have been mediated by the binding of the integrin subunits, beta1 and alpha1, and the subsequent signal transduction pathway, involving FAK and ERK2. The study indicated that the novel Mn-TiO2 coating has potential for orthopedic implant applications, and that further investigations are required.

[825]

TÍTULO / TITLE: Odontogenic myxoma.
RESUMEN / SUMMARY: Odontogenic myxoma is a rare intraosseous neoplasm, which is benign but locally aggressive. It rarely appears in any bone other than the jaws. It is considered to be derived from the mesenchymal portion of the tooth germ. Clinically, it is a slow-growing, expansile, painless, non-metastasizing, central tumor of jaws, chiefly the mandible. Here we report the case of a typical odontogenic myxoma in a 26-year-old female patient, which had acquired large dimensions and involved the entire left half of the mandible including the ramus, resulting in a gross facial deformity, within a span of one and a half years.

[826]

TÍTULO / TITLE: Myeloid Sarcoma of the Nasopharynx Mimicking an Aggressive Lymphoma.
RESUMEN / SUMMARY: Myeloid Sarcoma of the Nasopharynx Mimicking an Aggressive Lymphoma.
AUTORES / AUTHORS: Raphael J; Val lent A; Hanna C; Auger N; Casiraghi O; Ribrag V; De Botton S; Saada V
INSTITUCIÓN / INSTITUTION: - Clinical Haematology Department, Institut Gustave Roussy (IGR), 114 rue Edouard Vaillant, 94805, Villejuif, France, Raphaeljack13@hotmail.com.

RESUMEN / SUMMARY: - BACKGROUND: Myeloid sarcoma (MS) is a rare extra-medullary tumour of the myeloid lineage, which can be a difficult diagnosis to make. CASE PRESENTATION: We report the case of a 73-year-old male with a right-sided nasopharyngeal mass revealed on CT scan and MRI. RESULTS: An initial cytological and histological examination suggested a high-grade lymphoma. Nevertheless, the final diagnosis was a MS with an unusual involvement of the nasopharynx that was treated with a conventional induction leukemia therapy. Eight months later, the patient had persistent thrombocytopenia and a bone marrow aspiration showed the dysplasia of a high grade myelodysplastic syndrome and cytogenetics detected t(3;21). The patient was treated with a 5-Azacitidine (Vidaza) protocol until overt progression and disease evolution. CONCLUSION: In conclusion few cases of MS involving the nasopharynx have been reported. Its diagnosis is often difficult and should be considered especially when a high index of suspicion is present and the immunophenotype of the malignant haematological cells is not clearly in favour of a lymphoma.

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TÍTULO / TITLE: - The roles of hyaluronan/RHAMM/CD44 and their respective interactions along the insidious pathways of fibrosarcoma progression.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Nikitovic D; Kouvidi K; Karamanos NK; Tzanakakis GN

INSTITUCIÓN / INSTITUTION: - Department of Histology-Embryology, School of Medicine, University of Crete, 71003 Heraklion, Greece.

RESUMEN / SUMMARY: - Fibrosarcomas are rare malignant mesenchymal tumors originating from fibroblasts. Importantly, fibrosarcoma cells were shown to have a high content and turnover of extracellular matrix (ECM) components including hyaluronan (HA), proteoglycans, collagens, fibronectin, and laminin. ECMs are complicated structures that surround and support cells within tissues. During cancer progression, significant changes can be observed in the structural and mechanical properties of the ECM components. Importantly, hyaluronan deposition is usually higher in malignant tumors as compared to benign tissues, predicting tumor progression in some tumor types. Furthermore, activated stromal cells are able to produce tissue structure rich in hyaluronan in order to promote tumor growth. Key biological roles of HA result from its interactions with its specific CD44 and RHAMM (receptor for HA-mediated motility) cell-surface receptors. HA-receptor downstream signaling pathways regulate in turn cellular processes implicated in tumorigenesis. Growth factors, including PDGF-BB, TGFbeta2, and FGF-2, enhanced hyaluronan deposition to ECM and modulated HA-receptor expression in fibrosarcoma cells. Indeed, FGF-2 through upregulation of specific HAS isoforms and hyaluronan synthesis regulated secretion and net hyaluronan deposition to the fibrosarcoma pericellular matrix modulating these cells’ migration capability. In this paper we discuss the involvement of
hyaluronan/RHAMM/CD44 mediated signaling in the insidious pathways of fibrosarcoma progression.

[828]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 4103/0019-5049.118543
AUTORES / AUTHORS: - Koul A; Sood J
INSTITUCIÓN / INSTITUTION: - Department of Anaesthesiology, Pain and Perioperative Medicine, Sir Gangaram Hospital, New Delhi, India.

[829]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 4103/2229-516X.117093
AUTORES / AUTHORS: - Momin YA; Kulkarni MP; Pandav AB; Sulhyan KR
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Government Medical College, Miraj, Maharashtra, India.
RESUMEN / SUMMARY: - Sex cord stromal tumor with annular tubules (SCTAT) is a distinctive, rare subtype of sex cord stromal tumor of the ovary, predominant component of which has morphological features intermediate between that of granulosa cell and sertoli cell. The majority of ovarian SCTAT are benign. So far, malignant behavior in SCTAT has been reported only in sporadic cases. We have presented a case of malignant SCTAT in a 35-year-old lady with no associated Peutz-Jegher (P-J) syndrome.

[830]
TÍTULO / TITLE: - eComment. Soft-tissue sarcomas in a 'nutshell'.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 1093/icvts/ivt446
AUTORES / AUTHORS: - Challoumas D; Dimitrakakis G
INSTITUCIÓN / INSTITUTION: - Department of Cardiothoracic Surgery, University Hospital of Wales, Cardiff, UK.

[831]
TÍTULO / TITLE: - A massive posterior neck mass: lipoma or something more sinister?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 1155/2013/205936
AUTORES / AUTHORS: - Ryan MF; Allen B
INSTITUCIÓN / INSTITUTION: - Department of Emergency Medicine, University of Florida, 1329 SW 16th Street, P.O. Box 1000186, Gainesville, FL 32610-0186, USA.
RESUMEN / SUMMARY: - Lipomas are slow-growing benign soft-tissue tumors which are typically asymptomatic and occur in approximately 1% of the population. A lipoma is considered to be of excessive size when it is greater than 10 cm in length (in any dimension) or weighs over 1000 g (Kransdorf (1995)). We describe a case of a man presenting with a giant posterior neck mass which greatly reduced the sagittal range of cervical spine. A discussion of the pathophysiology of lipomas and a literature review regarding giant lipomas versus malignancy follows.

[832]
TÍTULO / TITLE: - Intraneural angioleiomyoma of the median nerve at the wrist.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Jing S; Giesen T

[833]
TÍTULO / TITLE: - Penile subcutaneous fibrolipoma postaugmentative phalloplasty.
RESUMEN / SUMMARY: - Fibrolipomas are a rare subtype of lipomas. We describe a case of a man suffering from subcutaneous penile fibrolipoma, who three months earlier has been submitted to an augmentative phalloplasty due to aesthetic dysmorphophobia. After six months from the excision of the mass, the penile elongation and penile enlargement were stable, and the patient was satisfied with his sexual intercourse and sexual life. To our knowledge, this is the first reported penile subcutaneous fibrolipoma case in the literature. The diagnostics and surgical features of this case are discussed.

[834]
TÍTULO / TITLE: - Bilateral primary angiosarcoma of the breast.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Keshav P; Hegde SS

INSTITUCIÓN / INSTITUTION: - Associate Professor, Department of Surgery, KMC, Mangalore, Manipal University, Karnataka 575003, India.
RESUMEN / SUMMARY: - Primary breast sarcomas are very rare entities, accounting for 0.04% of all malignant neoplasms. Angiosarcoma of breast is infrequent and is an endothelial malignant tumor with bad prognosis because of the frequency of metastasis and recurrence. We present a case of a 30-year-old female who presented with an ulcerated left breast lesion which on further workup revealed to be a primary angiosarcoma of breast with metastasis to right breast.

[835]
TÍTULO / TITLE: - Mining Sarcomas by Proteomics Approaches: Ewing Sarcoma on the Spotlight.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Mackintosh C; Madoz-Gurpide J
INSTITUCIÓN / INSTITUTION: - Molecular Pathology Program, Centro de Investigacion del Cancer-IBMCC, Universidad de Salamanca-CSIC, Salamanca, 37007, España. JMadoz@fjd.es.
RESUMEN / SUMMARY: - Sarcomas are a class of tumors defined by their mesenchymal origin that comprise very different neoplasms. Although some sarcomas harbor pathogenomic molecular alterations (i.e. specific balanced translocations and their associated chimeric fusion genes), others still lack an ultimate diagnostic tool, which could be of great interest as in some cases different sarcomas share a similar clinical manifestation. High throughput tools are contributing new ways to molecularly delinew.hupoeate the boundaries of each sarcoma subtype. Moreover, they are also shedding light into other research subjects of immediate concern: (i) the elucidation of the molecular targets of chimeric fusion proteins and their interactome; (ii) the discovery of new biomarkers and therapeutic targets; and (iii) the delineation of the response to therapeutic agents. Here we review the application of proteomics approaches to sarcomas, with special emphasis in Ewing sarcoma. Proteomics strategies offer the focus, the analytical potential, and the high throughput capabilities to decipher the hidden agenda of the biology of sarcomas, a knowledge that will surely be the subject of future patents intended to develop new diagnostic and therapeutic tools.

[836]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Puri A; Jaffe N; Gelderblom H
INSTITUCIÓN / INSTITUTION: - Department of Surgical Oncology, Tata Memorial Hospital, Mumbai 400012, India.

[837]
TÍTULO / TITLE: - Solitary fibrous tumor of neck mimicking cold thyroid nodule in 99m tc thyroid scintigraphy.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
A 68-year-old man had a rapidly growing, painless neck mass, thought to be nodular goiter. Ultrasonography showed a giant, heterogeneous mass occupying the middle and superior poles and protruding outside of the left thyroid lobe. The results of the thyroid function tests were normal. Thyroid scintigraphy revealed a large hypoactive nodule in the left thyroid lobe. Complete surgical removal of tumor was performed and macroscopically demonstrated a well-demarcated lesion outside the thyroid gland. Microscopically, the lesion was composed of fibroblast-like spindle cells in a patternless architecture and extensive stromal hyalinization. Immunohistochemistry showed positive reaction for CD34 in spindle cells and diffuse bcl-2 staining. The pathology was confirmed as solitary fibrous tumor. In the follow-up period after surgery, thyroid scintigraphy showed normal left thyroid lobe. Solitary fibrous tumor originated from or associated with thyroid gland is extremely rare. According to our knowledge, this is the first reported solitary fibrous tumor presenting like a cold thyroid nodule. This pathology must be considered for differential diagnosis of neck masses in the thyroid region.
AUTORES / AUTHORS: - Jolepalem P; Yeow RY; Cosner D; Seitz JP
INSTITUCIÓN / INSTITUTION: - Department of Diagnostic Radiology and Molecular Imaging, Oakland University William Beaumont School of Medicine and Health System, Royal Oak, MI, USA.
RESUMEN / SUMMARY: - We present a case of a 70-year-old male who was referred for a technetium-99m methylene-diphosphonate bone scan for mild left hip pain and an elevated alkaline phosphatase level of 770 units/L. No additional information was provided and the patient's history was limited due to a language barrier. We were able to ascertain that the patient had a remote history of prostate cancer, which had been treated with radiation. Originally, we felt the bone scan was compatible with Paget's disease; however, further work-up revealed the presence of osteosarcoma, which was potentially radiation-induced.

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TÍTULO / TITLE: - Intracaval and intracardiac leiomyomatosis of uterine origin.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
  ● Enlace a la Editora de la Revista http://bmj.com/search.dtl
  ● Enlace al texto completo (gratuito o de pago) 1136/bcr-2012-008368

AUTORES / AUTHORS: - Stoleriu C; Rizas K; Gawaz M; Geisler T
INSTITUCIÓN / INSTITUTION: - Department of Cardiology, Uniklinik Tuebingen, Tuebingen, Germany.
RESUMEN / SUMMARY: - A 37-year-old woman, gravida 2, para 1 presented in the outpatient ward with dyspnoea and tachycardia of unknown origin. The physical examination was unremarkable. Echocardiography revealed an intracardiac mass protruding through the tricuspid orifice into the right ventricle during diastole. The patient was admitted to the intensive care unit with the suspicion of vena cava thrombosis with intracardiac expansion. An abdominal sonography showed a mass in the uterus, presumed to be a benign tumour, with extension into the vena cava inferior. Owing to the extent of the mass in the right atrium and the risk for pulmonary embolism, after interdisciplinary discussion, a decision to remove the atrial mass was made. The case was managed by a two-stage procedure. Pathological examination of the intracardiac portions of the tumour revealed a benign tumour that consisted of proliferating smooth muscle fibres without abnormal mitotic activity.

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TÍTULO / TITLE: - Sarcomas of the uterine cervix: a united and multidisciplinary approach is required.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
  ● Enlace al texto completo (gratuito o de pago) 2217/whe.13.59

AUTORES / AUTHORS: - Khosla D; Patel FD; Kumar R
INSTITUCIÓN / INSTITUTION: - Department of Radiotherapy & Oncology, Regional Cancer Centre, Postgraduate Institute of Medical Education & Research, Sector 12, Chandigarh 160012, India. dr_divya_khosla@yahoo.com.
[842]
**TÍTULO / TITLE:** - Vascular leiomyosarcoma of thigh - A rare tumour at an unusual site.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](https://doi.org/10.4066/AMJ.2013.1874)
**AUTORES / AUTHORS:** - Roy AD; Deka M; Dutta UC
**INSTITUCIÓN / INSTITUTION:** - Gauhati Medical College, Guwahati, Assam, India.
**RESUMEN / SUMMARY:** - Leiomyosarcomas of vascular or origin are rare. They originate from the smooth muscles of tunica media of major blood vessels. The majority of such tumours arising in the extremities affect the femoral vascular bundle. There is limited knowledge and experience of the clinical presentation, pathological reports and results of treatment of this type of tumour. A case of primary leiomyosarcoma of femoral vein is being reported from a subtropical region of India that developed over the right thigh of a 35-year-old male farmer and was clinically diagnosed as benign soft tissue tumour. The diagnosis was confirmed by histopathology and immunohistochemistry.

[843]
**TÍTULO / TITLE:** - Pleomorphic undifferentiated sarcoma of urinary bladder with calcified pulmonary metastasis: A rare entity.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](https://doi.org/10.4103/0970-1591.117263)
**AUTORES / AUTHORS:** - Mylarappa P; Prathvi; Javali T; Ramesh D; Prabhu V; Rau AR
**INSTITUCIÓN / INSTITUTION:** - Department of Urology, M. S. Ramaiah Medical College and Hospital, Bangalore, Karnataka, India.
**RESUMEN / SUMMARY:** - We report the case of a 29-year-old male who presented to us with hematuria, dysuria and bilateral flank pain. On evaluation, the patient was found to have primary pleomorphic undifferentiated sarcoma of bladder with calcified pulmonary metastasis, confirmed with computerized tomography scan and immunohistochemistry.

[844]
**TÍTULO / TITLE:** - Peripheral ossifying fibroma secondary to pulpo-periodontal irritation.
**RESUMEN / SUMMARY:** - [Enlace al Resumen / Link to its Summary](https://doi.org/10.7860/JCDR/2013/6009.3410)
**AUTORES / AUTHORS:** - Akkara F; Chalakkal P; Boyapati CM; Pavaskar R
**INSTITUCIÓN / INSTITUTION:** - Assistant Professor, Department of Oral and Maxillofacial Surgery, Goa Dental College and Hospital, Bambolim-403202, Goa, India.
**RESUMEN / SUMMARY:** - This report has discussed the endodontic and surgical management of a peripheral ossifying fibroma that had occurred in the anterior maxillary anterior region, between a central and a lateral incisor in a 12-year-old child. The lesion was diagnosed to have occurred secondary to a pulpo-periodontal irritation.
TÍTULO / TITLE: - Clear Cell Adenocarcinoma Arising from Clear Cell Adenofibroma of the Ovary: Value of DWI and DCE-MRI.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Takeuchi M; Matsuzaki K; Uehara H; Furumoto H; Harada M
INSTITUCIÓN / INSTITUTION: - Department of Radiology, University of Tokushima.
RESUMEN / SUMMARY: - Clear cell adenofibroma (CCAF) is a rare surface epithelial-stromal tumor of the ovary and recently considered another precursor of clear cell adenocarcinoma (CCA) other than endometrioma. We report magnetic resonance (MR) findings of a borderline CCAF that contained a small CCA focus. The tumor manifested a characteristic "black sponge" appearance. The CCA focus showed high signal intensity on diffusion-weighted imaging (DWI) and early enhancement on dynamic contrast-enhanced (DCE) MR imaging (DCE-MRI), and the CCAF components showed low signal intensity on DWI and gradually increasing contrast enhancement on DCE-MRI.

TÍTULO / TITLE: - Metastatic transitional cell carcinoma of the tibia radiologically mimicking osteosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cunningham LP; O'Neill BJ; Quinlan JF
INSTITUCIÓN / INSTITUTION: - The Adelaide & Meath Hospital Incorporating The National Children's Hospital, Tallaght, Dublin, Ireland.
RESUMEN / SUMMARY: - We report a case of a 73-year-old lady with transitional cell carcinoma and no evidence of metastatic disease presenting with gradual weight loss, pretibial swelling and painful weightbearing. Investigations revealed a lesion of the right tibial diaphysis. The radiological and clinical appearance was that of primary osteosarcoma. Biopsy results revealed metastatic transitional cell carcinoma of the tibia. Intramedullary nailing was performed which relieved pain on weightbearing. The patient declined radiotherapy and was started on a palliative care regimen. This case illustrates the importance of histological diagnosis in the treatment of diaphyseal lesions.

TÍTULO / TITLE: - Vulvar fibroadenoma with lactational changes in ectopic breast tissue.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Lev-Cohain N; Kapur P; Pedrosa I
Ectopic breast tissue represents any type of breast tissue found outside its normal location in the pectoral region. The second most common location for ectopic breast tissue after axilla is the vulvar region. We present a case of a healthy 20-year-old female, G1P1, who presented to the Emergency Department with a sudden increase in size of a painful mass located in her vulva, which started 4 days after a spontaneous vaginal delivery and 3 days after initiation of breast-feeding of her newborn. She reported a stable, smaller, painless mass in the same location for almost 2 years prior to this episode. After surgical excision, a fibroadenoma with lactation changes within ectopic breast tissue was confirmed.

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**TÍTULO / TITLE:** - Production of Bioactive Compounds with Antitumor Activity Against Sarcoma 180 by Pleurotus sajor-caju.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Assis IS; Chaves MB; Silveira ML; Gern RM; Wisbeck E; Junior AF; Furlan SA

**INSTITUCIÓN / INSTITUTION:** - 1 Master’s Program in Health and Environment, University of the Joinville Region (Univille), Joinville, Santa Catarina, Brazil.

**RESUMEN / SUMMARY:** - Abstract This work studied the influence of culture medium composition and pH on exopolysaccharide (EPS) production by Pleurotus sajor-caju and validates the antitumor activity of the produced EPSs and of the mycelial biomass (intracellular polysaccharides [IPS]) against Sarcoma 180 (S180) cells. The effect of the initial concentrations of (NH4)2SO4, yeast extract and soy peptone on EPS production by P. sajor-caju was studied in shake flasks. A bioreactor was used to evaluate the pH values and the initial CaCO3 and glucose concentrations. Extracts of EPSs (PE1) and IPSs obtained through two different separation processes (PM1 and PM2) were tested on mice inoculated with S180 cells. A medium containing 2.5, 1.0, and 1.0 g/L of (NH4)2SO4, yeast extract and soy peptone, respectively, provided the highest EPS concentration (0.6 g/L). The use of pH 4.0, 1.0 g/L CaCO3 and 20 g/L initial glucose concentration enhanced EPS productivity (3.84 g/L per hour). The PE1 extract promoted the highest reduction of S180 growth (86%), followed by the PM2 extract (80%); growth reduction was dose-independent for both substances. This work provides information about culture medium and conditions that enhanced the production of extracellular polysaccharides by P. sajor-caju. The results can contribute to the search for new bioactive products bringing novel aspects to the medical and pharmaceutical areas.

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**TÍTULO / TITLE:** - Benign metastasizing leiomyoma of the lung.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Ki EY; Hwang SJ; Lee KH; Park JS; Hur SY
RESUMEN / SUMMARY: - Benign leiomyomas of the uterus are uncommonly found in association with benign smooth muscle tumors beyond the confines of the uterus. Benign metastasizing leiomyoma (BML) is a rare disease in which the lung is described to be the most afflicted extrauterine organ. We present a brief review of the literature, along with case reports for four patients who were followed up after resection of a pulmonary lesion or after pathological confirmation by biopsy. The clinical course of BML varies from chronic asymptomatic appearance to rapid progression, leading to respiratory failure and death. Our BML patients did not complain of pulmonary symptoms, such as cough, dyspnea, or chest tightness. Pathology revealed benign leiomyomas with no atypia and mitotic activity <5 per 10 high-power field. Immunohistochemical staining was positive for actin and desmin. A standard treatment for BML has not yet been established. Because of the hormone-sensitive characteristics of BML, treatments are based on hormonal manipulation along with either surgical or medical oophorectomy. Benign metastasizing leiomyoma can be observed in postmenopausal women. We observed four patients who did not receive adjuvant hormonal therapy because they were postmenopausal or perimenopausal. All patients are still healthy and show no evidence of recurrence or progression of the disease.

TÍTULO / TITLE: - Conjunctival Kaposi’s sarcoma as the initial manifestation of acquired immunodeficiency syndrome.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Izquierdo Rodriguez C; Cordova JM

RESUMEN / SUMMARY: - Abstract Introduction: Kaposi’s sarcoma is a common neoplasm in patients with acquired immunodeficiency syndrome (AIDS). Its presentation as an initial manifestation of AIDS is very rare. Objective: To report a rare case with Kaposi’s sarcoma as an initial manifestation of AIDS. Case: We report the case of a 37-year-old man who was a parenteral drug addict, HIV seropositive and was not under any treatment with a conjunctival lesion which was diagnosed as Kaposi’s sarcoma after surgical resection. Conjunctival Kaposi’s sarcoma is present frequently in HIV patients and lesions may be mistaken with other conjunctival lesions.

TÍTULO / TITLE: - Synovial sarcoma of the neck masquerading as a malignant second branchial cleft cyst.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary


AUTORES / AUTHORS: - Teng YS; Lin ZH; Li Y; Cao XL; Lin FC; Xiang JJ

INSTITUCIÓN / INSTITUTION: - Department of Otorhinolaryngology, Hangzhou First People's Hospital Hangzhou, 310006, P.R. China.

RESUMEN / SUMMARY: - Synovial sarcoma is an uncommon, aggressive malignant tumor of the soft tissues primarily involving the extremities of young adults. Head and neck synovial sarcoma is rare, and its diagnosis and therapy are still challenging. We report a case of a young patient with synovial sarcoma, clinically masquerading as
cystic mass of the neck and malignant second branchial cleft cyst. The pathological diagnosis of the sarcoma was confirmed by a multimodality diagnostic protocol, including histological, immunohistochemical and molecular genetic analysis. The patient underwent complete surgical excision followed by postoperative radiotherapy and recovered well.

[852]
**Título / Title:** Low-grade central osteosarcoma of distal femur, resembling fibrous dysplasia.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


**Autores / Authors:** Vasiliadis HS; Arnaoutoglou C; Plakoutsis S; Doukas M; Batistatou A; Xenakis TA

**Institución / Institution:** Haris S Vasiliadis, Christina Arnaoutoglou, Sotiris Plakoutsis, Theodoros A Xenakis, Department of Orthopaedics, University Hospital of Ioannina, 45500 Ioannina, Greece.

**Resumen / Summary:** We report a case of a 32 year-old male, admitted for a lytic lesion of the distal femur. One month after the first X-ray, clinical and imaging deterioration was evident. Open biopsy revealed fibrous dysplasia. Three months later, the lytic lesion had spread to the whole distal third of the femur reaching the articular cartilage. The malignant clinical and imaging features necessitated excision of the lesion and reconstruction with a custom-made total knee arthroplasty. Intra-operatively, no obvious soft tissue infiltration was evident. Nevertheless, an excision of the distal 15.5 cm of the femur including 3.0 cm of the surrounding muscles was finally performed. The histological examination of the excised specimen revealed central low-grade osteosarcoma. Based on the morphological features of the excised tumor, allied to the clinical findings, the diagnosis of low-grade central osteosarcoma was finally made although characters of a fibrous dysplasia were apparent. Central low-grade osteosarcoma is a rare, well-differentiated sub-type of osteosarcoma, with clinical, imaging, and histological features similar to benign tumours. Thus, initial misdiagnosis is usual with the condition commonly mistaken for fibrous dysplasia. Central low-grade osteosarcoma is usually treated with surgery alone, with rare cases of distal metastases. However, regional recurrence is quite frequent after close margin excision.

[853]
**Título / Title:** Primary bone carcinosarcoma of the fibula with chondrosarcoma and squamous cell carcinoma components.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


**Autores / Authors:** Ishida M; Kodama N; Takemura Y; Iwai M; Yoshida K; Kagotani A; Matsusue Y; Okabe H

**Institución / Institution:** Department of Clinical Laboratory Medicine and Division of Diagnostic Pathology, Shiga University of Medical Science Shiga, Japan.

**Resumen / Summary:** Carcinosarcoma is defined as a malignant neoplasm that is composed of both carcinomatous and sarcomatous components. The occurrence of carcinosarcoma in the bone is extremely rare. In this report, we describe the third
documented de novo case of carcinosarcoma of the bone. A 59-year-old Japanese female presented with a painful tumor in her right lower leg. Plane radiography revealed an osteolytic destructive lesion with periosteal reaction and mineralization in the right fibula. Resection of the fibula tumor was performed under a clinical diagnosis of chondrosarcoma. Histopathological study revealed that the tumor was comprised of three components. The main component was proliferation of small round to short spindle cells (approximately 50%), and the remaining components were chondrosarcoma (30%) and squamous cell carcinoma (20%). Immunohistochemically, SOX9 was expressed in the small round to spindle cells and chondrosarcoma component, and p63 and p40 were expressed in all three components. Accordingly, an ultimate diagnosis of carcinosarcoma of the bone was made. The clinicopathological analysis of carcinosarcoma of the bone revealed that this type of tumor affects the middle-aged to elderly persons and occurs in the long bone. All three de novo cases had chondrosarcoma and squamous cell carcinoma components. One of the 3 patients died of the disease. The histogenesis of carcinosarcoma of the bone remains a matter of controversy, although a multipotential stem cell theory has been proposed. Additional studies are required to clarify the clinical behavior and histogenesis of carcinosarcoma of the bone.

[854]

TÍTULO / TITLE: Duodenal subepithelial hyperechoic lesions of the third layer: Not always a lipoma.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary

TÍTULO / TITLE: MiR-320a downregulation is associated with imatinib resistance in gastrointestinal stromal tumors.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary

Gastrointestinal stromal tumor (GIST) is one of the most common mesenchymal tumors of the gastrointestinal tract. Though imatinib improves the outcome, drug resistance remains the major problem for extending patient survival. Genetic mutation of the drug targets is the known mechanism for imatinib resistance. However, it cannot explain all of the phenomena of imatinib resistance, and numerous additional mechanisms have been proposed to account for imatinib resistance in various model systems. In this study, we applied the SYBR green quantitative polymerase chain reaction-based array approach to screen the differentially expressed miRNAs between primary GIST patients and imatinib-resistant patients. The selected candidate miRNAs were validated in a cohort of 12 GIST patients. We found that low expression of miR-320a was correlated with short time to imatinib resistance, and proposed the potential mechanism of miR-320a for imatinib resistance.

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The neurology of rhizomelic chondrodysplasia punctata.

BACKGROUND: To describe the neurologic profiles of Rhizomelic chondrodysplasia punctata (RCDP); a peroxisomal disorder clinically characterized by skeletal abnormalities, congenital cataracts, severe growth and developmental impairments and immobility of joints. Defective plasmalogen biosynthesis is the main biochemical feature. METHODS: Observational study including review of clinical and biochemical abnormalities, genotype, presence of seizures and neurophysiological studies of a cohort of 16 patients with RCDP. RESULTS: Patients with the severe phenotype nearly failed to achieve any motor or cognitive skills, whereas patients with the milder phenotype had profound intellectual disability but were able to walk and had verbal communication skills. Eighty-eight percent of patients developed epileptic seizures. The age of onset paralleled the severity of the clinical and biochemical phenotype. Myoclonic jerks, followed by atypical absences were most frequently observed. All patients with clinical seizures had interictal encephalographic evidence of epilepsy. Visual evoked (VEP) and brain auditory potential (BAEP) studies showed initial normal latency times in 93% of patients. Deterioration of VEP occurred in a minority in both the severe and the milder phenotype. BAEP andsomatosensory evoked potentials (SSEP) were more likely to become abnormal in the severe phenotype. Plasmalogens were deficient in all patients. In the milder phenotype levels of plasmalogens were significantly higher in erythrocytes than in the severe phenotype. Phytanic acid levels ranged from normal to severely
increased, but had no relation with the neurological phenotype. CONCLUSION: Neurodevelopmental deficits and age-related occurrence of seizures are characteristic of RCDP and are related to the rest-activity in plasmalogen biosynthesis. Evoked potential studies are more likely to become abnormal in the severe phenotype, but are of no predictive value in single cases of RCDP.

[857]
**TITULO / TITLE:** A giant cystic leiomyoma mimicking an ovarian malignancy.
**RESUMEN / SUMMARY:** [Enlace al Resumen / Link to its Summary](#)
**AUTORES / AUTHORS:** Aydin C; Eris S; Yalcin Y; Sen Selim H
**INSTITUCIÓN / INSTITUTION:** Gynecology and Obstetrics Department of Ataturk Training and Research Hospital, Basin Sitesi, Yesilyurt, 35360 Izmir, Turkey. Electronic address: cetinaydin2005@mynet.com.
**RESUMEN / SUMMARY:** INTRODUCTION: Leiomyoma of the uterus is the most common type of tumor affecting the female pelvis and arises from uterine smooth muscle. The size of leiomyomas varies from microscopic to giant; giant myomas are exceedingly rare. We report an unusual case of a large, cystic, pedunculated uterine leiomyoma mimicking a primary malignant ovarian tumor on sonography and CT. PRESENTATION OF CASE: A 58-year-old postmenopausal nulliparous woman presented with a history of lower abdominal pain and distension for a period of approximately 12 months. The patient's personal history revealed difficulty in walking, tiredness and recent weight gain of approximately 25kg. Sonography and CT examination showed a large mass that filled the abdomen. A preoperative diagnosis of a primary malignant ovarian tumor was made. The patient underwent laparotomy, total hysterectomy and bilateral salpingo-oophorectomy. The histology revealed a leiomyoma with extensive cystic degeneration. DISCUSSION: The current established management of uterine fibroids may involve expectant, surgical, or medical management or uterine artery embolization or a combination of these treatments. A surgical approach is preferred for management of giant leiomyomas. CONCLUSION: Pedunculated leiomyomas should be considered in the differential diagnosis of a multilocular and predominantly cystic adnexal mass.

[858]
**TITULO / TITLE:** Multiple liver abscess formation and primary gastrointestinal stromal tumor.
**RESUMEN / SUMMARY:** [Enlace al Resumen / Link to its Summary](#)
**AUTORES / AUTHORS:** E Chang A; N Mann G; Hoch B; T Loggers E; M Pollack S; Kolokythas O; L Jones R
**INSTITUCIÓN / INSTITUTION:** University of Washington Medical Center, Fred Hutchinson Cancer Research Center, Seattle, WA, USA.
**RESUMEN / SUMMARY:** Gastrointestinal stromal tumors are the most common mesenchymal tumors of the gastrointestinal tract. The introduction of a number of small molecule tyrosine kinase inhibitors has revolutionized the management of metastatic
disease. Surgery is the mainstay of management for localized disease. Patients with high risk tumors are treated with adjuvant imatinib. We report the rare presentation of a localized primary small bowel gastrointestinal stromal tumor in association with multiple liver abscesses. Cystic liver lesions should be fully evaluated in gastrointestinal tumor patients to exclude an infective cause. Treatment with intravenous antibiotics resulted in clinical and radiological improvement of the liver abscesses. The small bowel tumor was treated with surgical resection.

[859] TÍTULO / TITLE: - Osteolitic and osteoblastic lesions of the skull.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace a la Editora de la Revista http://bmj.com/search.dtl
   ●● Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-201228
AUTORES / AUTHORS: - Fernandes AM; Pedreira DG; Lopes P; Fera M
INSTITUCIÓN / INSTITUTION: - Servico de Medicina Interna, Hospital S Bernardo, Centro Hospitalar de Setubal, EPE, Portugal.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1093/icvts/ivt444
AUTORES / AUTHORS: - Lazopoulos A; Gogakos A; Rallis T; Barbetakis N
INSTITUCIÓN / INSTITUTION: - Department of Thoracic Surgery, Theagenio Cancer Hospital, Thessaloniki, Greece.

[861] TÍTULO / TITLE: - Tricuspid valve obstruction and right heart failure due to a giant right atrial myxoma arising from the superior vena cava.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 1186/1749-8090-8-200
AUTORES / AUTHORS: - Xiao ZH; Hu J; Zhu D; Shi YK; Zhang EY
RESUMEN / SUMMARY: - Myxomas are the most common primary cardiac tumors. The cardiac myxomas are mostly diagnosed within the atria, and only a few such tumors are reported to have arisen from atrioventricular valves or pulmonary vessels. The authors here present a case of 59-year-old Chinese woman who was hospitalized for exacerbating symptoms of tricuspid stenosis and right heart failure. Echocardiography revealed a giant right atrial myxoma arising from an extremely rare site, the anterior wall of the superior vena cava. With the aid of transesophageal echocardiography, the surgical resection was performed successfully with the patient achieving complete recovery.

[862]
TÍTULO / TITLE: - Secondary aneurysmal bone cyst following chondroblastoma of the patella.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Tomoyuki K; Susa M; Nakayama R; Watanabe I; Horiuchi K; Toyama Y; Morioka H
INSTITUCIÓN / INSTITUTION: - Department of Orthopedics Surgery, Keio University School of Medicine , Tokyo, Japan.
RESUMEN / SUMMARY: - Aneurysmal bone cyst (ABC) is a rare benign cystic lesion of the bone that composes 1-2% of the entire bone tumors. Some are idiopathic, and some occur secondary to other tumors such as giant cell tumor and chondroblastoma. In this article, we report the clinical, radiographic, and histological findings of a secondary ABC following chondroblastoma of the patella with a review of the literature.

[863]
TÍTULO / TITLE: - Renal leiomyosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Beardo P; Jose Ledo M; Jose Luis RC
INSTITUCIÓN / INSTITUTION: - Department of Urology, Hospital de Jerez, Cadiz University.
RESUMEN / SUMMARY: - Leiomyosarcoma (LMS) is a rare malignant tumor of smooth muscle origin that generally stems from soft tissues and uterine tissue. Although, a small percentage of these may originate from the smooth muscle or vessel walls, most of which are of venous origin. Renal leiomyosarcomas may arise from the smooth muscle fibers of renal pelvis, renal capsule or renal vessels, last one is the most frequent. We report a case of renal LMS that could be originated in the renal capsule.

[864]
TÍTULO / TITLE: - Salinomycin increases chemosensitivity to the effects of doxorubicin in soft tissue sarcomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Liffers ST; Tilkorn DJ; Stricker I; Junge CG; Al-Benna S; Vogt M; Verdoort B; Steinau HU; Tannapfel A; Tischoff I; Mirmohammadsadegh A
RESUMEN / SUMMARY: - BACKGROUND: Chemotherapy for soft tissue sarcomas remains unsatisfactory due to their low chemosensitivity. Even the first line chemotherapeutic agent doxorubicin only yields a response rate of 18-29%. The antibiotic salinomycin, a potassium ionophore, has recently been shown to be a potent compound to deplete chemoresistant cells like cancer stem like cells (CSC) in adenocarcinomas. Here, we evaluated the effect of salinomycin on sarcoma cell lines, whereby salinomycin mono- and combination treatment with doxorubicin regimens were analyzed. METHODS: To evaluate the effect of salinomycin on fibrosarcoma, rhabdomyosarcoma and liposarcoma cell lines, cells were drug exposed in single and combined treatments, respectively. The effects of the corresponding treatments were
monitored by cell viability assays, cell cycle analysis, caspase 3/7 and 9 activity assays. Further we analyzed NF-kappaB activity; p53, p21 and PUMA transcription levels, together with p53 expression and serine 15 phosphorylation. RESULTS: The combination of salinomycin with doxorubicin enhanced caspase activation and increased the sub-G1 fraction. The combined treatment yielded higher NF-kappaB activity, and p53, p21 and PUMA transcription, whereas the salinomycin monotreatment did not cause any significant changes. CONCLUSIONS: Salinomycin increases the chemosensitivity of sarcoma cell lines - even at sub-lethal concentrations - to the cytostatic drug doxorubicin. These findings support a strategy to decrease the doxorubicin concentration in combination with salinomycin in order to reduce toxic side effects.

[865]
TITULO / TITLE: - Epithelioid angiosarcoma of the septum pellucidum.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 1155/2013/603671
AUTORES / AUTHORS: - Baldovini C; Martinoni M; Marucci G
INSTITUCIÓN / INSTITUTION: - Section of Pathology M. Malpighi at Bellaria Hospital, Department of Biomedical and Neuro Motor Sciences, University of Bologna, Via Altura 3, 40139 Bologna, Italy.
RESUMEN / SUMMARY: - Primary cerebral intra-axial epithelioid angiosarcoma is an extremely rare malignancy. To the best of our knowledge we describe the first case of epithelioid angiosarcoma arisen in the septum pellucidum of a 54-years-old man. Albeit extremely rare, this neoplasia is a potential source of misdiagnosis for other aggressive malignant tumors, and it should be taken into consideration.

[866]
TITULO / TITLE: - Testicular fibroma of gonadal stromal origin with minor sex cord elements, presenting with hydrocele.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 4081/rt.2013.e34
AUTORES / AUTHORS: - Datta S; Dey S; Mukherjee S; Chandra Paul P; Bhattacharyya A; Biswas S; Tudu B
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Nilratan Sircar Medical College, Kolkata, India.
RESUMEN / SUMMARY: - Testicular fibroma of gonadal stromal origin is a rare benign tumor of testis which usually presents as a slow growing testicular mass. Only 25 cases of testicular fibroma have been reported in the literature. Presence of minor sex cord elements in this tumor is even rarer. We report a case of testicular fibroma with minor sex cord elements that involved almost the entire testis and tunica vaginalis. The patient presented with hydrocele, a rare presentation for this entity. The rarity of the diagnosis and the clinical presentation prompted this case report.

[867]
TÍTULO / TITLE: - Real-time image-guided recontouring in the management of craniofacial fibrous dysplasia.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Gui H; Zhang S; Shen SG; Wang X; Bautista JS; Voss PJ
INSTITUCIÓN / INSTITUTION: - Associate Professor, Department of Oral and Maxillofacial Surgery, Ninth People's Hospital, affiliated to Shanghai Jiao Tong University School of Medicine, Shanghai, China.
RESUMEN / SUMMARY: - OBJECTIVE: This report presents our experience using computer-aided recontouring in the surgical management of complex craniofacial fibrous dysplasia with the use of a navigation system developed by the authors.
STUDY DESIGN: A total of 21 patients (9 men and 12 women) with craniofacial fibrous dysplasia and mean age of 23 years (range, 17-32 years) were included in this study from 2007 to 2012. By creating a mirror image of the unaffected side on the affected side as a virtual treatment template, we completed the recontouring procedures in real time with the aid of the navigation system that we developed (Accu-Navi). The surgical outcome was assessed by superimposing the postoperative computed tomography (CT) images onto the preoperative CT images. RESULTS: The precise preoperative simulation and intraoperative navigation enabled the surgeon to complete the recontouring procedure visually. Postoperative CT was compared with the preoperative plan, yielding an average discrepancy of <1.0 mm. Postoperative follow-up found that both facial aesthetics and patient satisfaction improved remarkably. CONCLUSIONS: Navigation-guided recontouring shows benefits in improving accuracy and safety for this complicated procedure.

[868]

TÍTULO / TITLE: - Primary intracranial leiomyosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Alijani B; Yousefzade S; Aramnia A; Mesbah A
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Poursina Hospital, Guilan University of Medical Sciences, Rasht, Iran.drbabakali@gmail.com.
RESUMEN / SUMMARY: - Primary intracranial leiomyosarcomas are rare tumors that arise from the mesenchymal cells of the dura mater or cerebral blood vessels Here we report the case of an extra axial leiomyosarcoma in the right parieto-occipital region of a 19-year-old male who had normal clinical and laboratory findings. Diagnostic imaging showed bony destruction, dural involvement and no parenchymal invasion. No primary site was found after metastasis work up. Specific serology tests were negative. The patient underwent a craniotomy, total tumor resection, duraplasty and skull reconstruction. He received radiotherapy and after 18 months of follow-up, no clinical and radiological signs of recurrence have been found.

[869]

TÍTULO / TITLE: - Schistocytes, echinocytes, iron deficiency anemia, and thrombocytopenia - hematologic manifestations of splenic angiosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Splenic angiosarcoma is a rare and aggressive malignancy with an incidence of less than one per million and a fatality rate over 90%. Early diagnosis is of great importance for optimal management. Here, we report the case of a patient with splenic angiosarcoma who presented with prominent schistocytes, echinocytes, thrombocytopenia, and iron deficiency anemia, which in combination with radiographic evidence of a splenic mass, raised the suspicion for angiosarcoma and resulted in a prompt surgical intervention with curative intent. Resolution of the hematologic findings following splenectomy suggests that patients with this malignancy should be monitored for recurrent hematologic abnormalities as they may herald recurrence of the disease. We present a literature review on the hematologic manifestations that is associated with this malignant disease.

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**Título / Title:** Subconjunctival hibernoma in a dog.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


**Autores / Authors:** Stuckey JA; Rankin AJ; Romkes G; Slack J; Kiupel M; Dubielzig RR

**Institución / Institution:** Veterinary Health Center, Kansas State University, 106 Mosier Hall, Manhattan, KS, 66506, USA.

**Resumen / Summary:** A 10-year-old, castrated male, German Shepherd mixed-breed dog was presented to Kansas State University Veterinary Health Center for evaluation of a subconjunctival swelling in the ventral fornix of the left orbit. The owner elected to pursue excision of the mass 2 years after initial consultation following a sudden change in the size and color of the lesion. An excisional biopsy was performed, and the mass along with its associated capsule were submitted to the Comparative Ocular Pathology Laboratory of Wisconsin for histopathologic evaluation, which confirmed the diagnosis of a hibernoma. Fourteen months following excision, the patient showed no evidence of tumor regrowth.

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**Título / Title:** Inactivated autograft-prosthesis composite have a role for grade III giant cell tumor of bone around the knee.

**Resumen / Summary:** Enlace al Resumen / Link to its Summary


**Autores / Authors:** Xu S; Yu X; Xu M; Fu Z

**Institución / Institution:** Department of Orthopaedics, General Hospital of Ji’Nan Military Region, Ji’Nan 250031, China. yxch48@vip.sina.com
**RESUMEN / SUMMARY:** - BACKGROUND: Giant cell tumors (GCT) around the knee are common and pose a special problem of reconstruction after tumor excision, especially for grade III GCT. We questioned whether en bloc resection and reconstruction with alcohol inactivated autograft-prosthesis composite would provide (1) local control and long-term survival and (2) useful limb function in patients who had grade III GCT around the knee. METHODS: We retrospectively reviewed eight patients (5 males and 3 females) treated with this procedure with mean age of 31 years (range 20 to 43 years) from Jan 2007 to Oct 2008. 5 lesions were located in distal femur and 3 in proximal tibia. 4 patients were with primary tumor and the other 4 with recurrence. 2 patients showed pathological fracture. RESULTS: Mean Follow-up is 54 months ranging from 38 to 47 months. No recurrence, metastasis, prosthesis loosening were found. The mean healing time between autograft and host bone was 5.5 months. The mean MSTS score was 26.3 (88%) ranging from 25 to 29. The mean ISOLS composite graft score was 32.8 (88.5%) ranging from 28 to 35. Creeping substitution is possibly the main way in bony junction. The healing time in femoral lesion is faster than that in tibial lesion. CONCLUSIONS: The technique of alcohol inactivated autograft-prosthesis composite could be able to achieve satisfactory oncological and functional outcomes in Grade III GCT.

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**TÍTULO / TITLE:** - Primary Extraskeletal Mesenchymal Chondrosarcoma of the Anterior Mediastinum.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Jeong SS; Choi PJ; Kim DW; Son C; Roh MS

**INSTITUCIÓN / INSTITUTION:** - Department of Thoracic and Cardiovascular Surgery, Dong-A University College of Medicine, Busan, Korea.

[872]

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**TÍTULO / TITLE:** - Malignant Solitary Fibrous Tumor of Tandem Lesions in the Skull and Spine.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary


**AUTORES / AUTHORS:** - Son S; Lee SG; Jeong DH; Yoo CJ

**INSTITUCIÓN / INSTITUTION:** - Department of Neurosurgery, Gachon University, Gil Medical Center, Incheon, Korea.

**RESUMEN / SUMMARY:** - A Solitary Fibrous Tumor (Sft) Is A Rare Neoplasm Originated From The Pleura, But They Can Occur In A Variety Of Extrathoracic Regions. Although Many Cases Of Primary Sft Have Been Reported, There Are Extremely Rare Repots To Date Of A Malignant Sft In The Spine Or Skull. A 54-year-old Woman Visited Our Hospital Due To Low Back Pain And Both Leg Radiating Pain. Several Imaging Studies Including Magnetic Resonance Imaging And Computed Tomography Revealed Expansive Enhanced Lesions In The Occipital Bone, T8, S1-2, And Ilium, With Neural Tissue Compression. We Performed Surgical Resection Of The...
Tumor In Each Site, And Postoperative Radiosurgery And Chemotherapy Were Performed. However, After Six Months, Tumors Were Recurred And Metastasized In Multiple Regions Including Whole Spine And Lung. The Authors Report Here The First Case Of Patient With Malignant Sft Of Tandem Lesions In The Various Bony Structures, Including Skull, Thoracic Spine, And Sacral Spine, With A Rapid Recurrence And Metastasis. Although Malignant Sft Is Extremely Rare, It Should Be Considered In The Differential Diagnosis And Careful Follow-up Is Needed.

[874]
**TÍTULO / TITLE:** - Cavernous haemangioma of small intestine mimicking gastrointestinal stromal tumour.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Ersoy O; Akin E; Demirezer A; Koseoglu H; Balci S; Kiyak G
**INSTITUCIÓN / INSTITUTION:** - Yildirim Beyazit University, Faculty of Medicine, Department of Gastroenterology, Ankara, Turkey. Electronic address: oersoy@yahoo.com.tr.

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[875]
**TÍTULO / TITLE:** - Phyllodes Tumor of Anogenital Mammary-like Glands with Diffuse Pseudoangiomatous Stromal Hyperplasia.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Eliyatkin N; Top OE; Yalcin E; Zengel B; Ozgur H; Aykas A; Vardar E
**INSTITUCIÓN / INSTITUTION:** - Department of Pathology, Izmir Education and Research Hospital, IZMIR, TURKEY.
**RESUMEN / SUMMARY:** - Anogenital mammary-like glands may give rise to various pathologic lesions identical to those known in mammary pathology. Tumor occurring in the anogenital region is extremely rare. The histogenetic origin of this tumor is controversial as it is being debated whether such lesions evolve from ectopic breast tissue and most recently, anogenital mammary-like gland. We report a 28-year-old girl who presented with a painless mass in the anogenital region, which was subsequently excised. Microscopic examination revealed morphologic pattern characteristic of benign phyllodes tumor with pseudoangiomatous stromal hyperplasia. We present this case to emphasize the importance of recognizing this uncommon lesion occurring at an extremely unusual site. We also discuss the histogenesis of phyllodes tumor and related lesions occurring in the anogenital region in light of the current literature along with a brief review of the previously reported cases of anogenital mammary-like glands.

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[876]
**TÍTULO / TITLE:** - Functioning adrenal myelolipoma: A rare cause of hypertension.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary
Co-occurrence of adrenal incidentaloma with hypertension calls for evaluation of endocrine causes including pheochromocytoma, Cushing’s disease, and primary aldosteronism. We are reporting a 40-year-old man who presented with hypertension and an adrenal mass. He had elevated metanephrines, histology of resected adrenal mass revealed adrenal myelolipoma, and immuno-histochemistry was positive for chromogranin A. Both his blood pressure and urinary metanephrines returned to normal after surgery. The association of hypertension and adrenal myelolipoma may not be entirely coincidental, as it may be associated with secreting catecholamine. Literature on such an uncommon association is reviewed briefly as well.

Characterization of a Novel Polyomavirus Isolated from a Fibroma on the Trunk of an African Elephant (Loxodonta africana).

Viruses of the family Polyomaviridae infect a wide variety of avian and mammalian hosts with a broad spectrum of outcomes including asymptomatic infection, acute systemic disease, and tumor induction. In this study a novel polyomavirus, the African elephant polyomavirus 1 (AelPyV-1) found in a protruding hyperplastic fibrous lesion on the trunk of an African elephant (Loxodonta africana) was characterized. The AelPyV-1 genome is 5722 bp in size and is one of the largest polyomaviruses characterized to date. Analysis of the AelPyV-1 genome reveals five putative open-reading frames coding for the classic small and large T antigens in the early region, and the VP1, VP2 and VP3 capsid proteins in the late region. In the area preceding the VP2 start codon three putative open-reading frames, possibly coding for an agnoprotein, could be localized. A regulatory, non-coding region separates the 2 coding regions. Unique for polyomaviruses is the presence of a second 854 bp long non-coding region between the end of the early region and the end of the late region. Based on maximum likelihood phylogenetic analyses of the large T antigen of the AelPyV-1 and 61 other polyomavirus sequences, AelPyV-1 clusters within a heterogeneous group of polyomaviruses that have been isolated from bats, new world primates and rodents.

Cardiac rhabdomyoma in familial tuberous sclerosis.
RESUMEN / SUMMARY: Cardiac rhabdomyomas are often associated with tuberous sclerosis in infants. We report a 5 month old child presented with a tumor in the right ventricle and echocardiography features of rhabdomyoma. Both the child and her father had cutaneous features of tuberous sclerosis. In the absence of features of congestive heart failure, surgery is rarely required.

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TÍTULO / TITLE: Cytotoxic Effects of Fucoidan Nanoparticles against Osteosarcoma.
RESUMEN / SUMMARY: In this study, we analyzed the size-dependent bioactivities of fucoidan by comparing the cytotoxic effects of native fucoidan and fucoidan lipid nanoparticles on osteosarcoma in vitro and in vivo. In vitro experiments indicated that nanoparticle fucoidan induced apoptosis of an osteosarcoma cell line more efficiently than native fucoidan. The more potent effects of nanoparticle fucoidan, relative to native fucoidan, were confirmed in vivo using a xenograft osteosarcoma model. Caco-2 cell transport studies showed that permeation of nanoparticle fucoidan was higher than native fucoidan. The higher bioactivity and superior bioavailability of nanoparticle fucoidan could potentially be utilized to develop novel therapies for osteosarcoma.

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TÍTULO / TITLE: Management of Cervical Fibroid during the Reproductive Period.
RESUMEN / SUMMARY: This is a case report of a 29-year-old lady who presented with excessive vaginal discharge and sessile cervical fibroid arising from the vaginal portion of the cervix. She was not suitable for uterine artery embolization as she has never previously been pregnant before. She was encouraged to get pregnant and to avoid surgical excision which can lead to hysterectomy. Shortly after, she became...
pregnant. She had many admissions during pregnancy due to bleeding from the fibroid, and in one occasion she had blood transfusions. The fibroid increased in size to become larger than the head of the baby. An emergency caesarean section was performed at 37 weeks when she attended in labour before the date of her elective caesarean section. She was managed conservatively following delivery in the hope that the fibroid becomes smaller making surgery easier. The fibroid degenerated and reduced in size. Vaginal myomectomy was carried out. The patient is now pregnant for the second time and had a cervical suture at 20 weeks gestation. In this educational case report we discuss the different management options of cervical fibroids and review the literature of other similar cases and their outcome.

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[881] TÍTULO / TITLE: - A large left atrial myxoma causing multiple cerebral infarcts.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace a la Editora de la Revista http://bmj.com/search.dtl
●● Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-010007
AUTORES / AUTHORS: - Kebede S; Edmunds E; Raybould A
INSTITUCIÓN / INSTITUTION: - Department of Medicine, Hywel Dda Health Board, Carmarthen, UK.
RESUMEN / SUMMARY: - A 52-year-old man presented with a history of sudden onset diplopia. On neurological examination, the only abnormality was a right-sided oculomotor (third nerve) palsy. A brain CT was performed and reported as showing no abnormality. He was discharged to be investigated as an outpatient. He presented 1 month later with a new expressive dysphasia and confusional state. MRI was performed which revealed multiple cerebral infarcts. He was discharged on secondary stroke prevention medication. Six months elapsed, before a transthoracic echocardiogram was performed. This showed a large left atrial myxoma. The patient underwent an emergency resection and made a good postoperative recovery. This case report showed the importance of considering a cardiogenic source of emboli in patients who present with cerebral infarcts. Performing echocardiography early will help to detect treatable conditions such as atrial myxoma, and prevent further complications.
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RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o de pago) 3390/cancers5030890
AUTORES / AUTHORS: - Tejani MA; Galloway TJ; Lango M; Ridge JA; von Mehren M
INSTITUCIÓN / INSTITUTION: - Division of Hematology/Oncology, University of Rochester Medical Center, Rochester, NY 14642, USA. margaret.vonmehren@fccc.edu.
RESUMEN / SUMMARY: - Head/neck sarcomas are rare, accounting for about 1% of head/neck malignancies and 5% of sarcomas. Outcomes have historically been worse
in this group, due to anatomic constraints leading to difficulty in completely excising tumors, with high rates of local recurrence. We retrospectively analyzed cases of head/neck soft tissue sarcomas (STS) and osteogenic sarcomas managed in a multi-disciplinary setting at Fox Chase Cancer Center from 1999-2009 to describe clinicopathologic characteristics, treatment, outcomes, and prognostic factors for disease control and survival. Thirty patients with STS and seven patients with osteogenic sarcoma were identified. Most STS were high grade (23) and almost all were localized at presentation (28). Common histologies were synovial cell (6), rhabdomyosarcoma (5), angiosarcoma (4), liposarcoma (4) and leiomyosarcoma (3). The type of primary therapy and disease outcomes were analyzed. Cox proportional hazards regression analysis was performed to identify predictors of disease-free survival (DFS) and overall survival (OS). The HR and 95% CI for Cox model and median DFS/OS analyzed by Kaplan-Meier curves were calculated.

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**TÍTULO / TITLE:** Difficulty in complete transarterial embolization for pulmonary benign metastasizing leiomyoma with massive hemoptysis.

**RESUMEN / SUMMARY:**

Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** Jpn J Radiol. 2013 Nov 25.

- Enlace al texto completo (gratuito o de pago) 1007/s11604-013-0266-9

**AUTORES / AUTHORS:**

Miyazaki M; Nakayama A; Noda D; Maehara Y; Tsushima Y

**INSTITUCIÓN / INSTITUTION:** Department of Diagnostic and Interventional Radiology, Gunma University Hospital, 3-39-15 Showa-machi, Maebashi, Gunma, 371-8511, Japan, mmiyazak@gunma-u.ac.jp.

**RESUMEN / SUMMARY:**

A 43-year-old woman suffering from massive hemoptysis from pulmonary benign metastasizing leiomyoma (BML) at the left lung hilum underwent bronchial and nonbronchial transarterial embolization (TAE) using gelatin sponge particles and n-butyl 2-cyanoacrylate during three interventional procedures. However, since complete embolization of the tumor was difficult despite decreased tumor size, the tumor was surgically resected 3 months after the last interventional procedure. This case demonstrates the difficulty of complete TAE for pulmonary BML because of its hypervascularity and the rich communications between bronchial and nonbronchial anastomotic arteries.

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**TÍTULO / TITLE:** Rb1 loss modifies but does not initiate alveolar rhabdomyosarcoma.

**RESUMEN / SUMMARY:**

Enlace al Resumen / Link to its Summary


- Enlace al texto completo (gratuito o de pago) 1186/2044-5040-3-27

**AUTORES / AUTHORS:**

Kikuchi K; Taniguchi E; Chen HI; Svalina MN; Abraham J; Huang ET; Nishijo K; Davis S; Louden C; Zarzabal LA; Recht O; Bajwa A; Berlow N; Suelves M; Perkins SL; Meltzer PS; Mansoor A; Michalek JE; Chen Y; Rubin BP; Keller C

**RESUMEN / SUMMARY:**

BACKGROUND: Alveolar rhabdomyosarcoma (aRMS) is a myogenic childhood sarcoma frequently associated with a translocation-mediated fusion gene, Pax3:Foxo1a. METHODS: We investigated the complementary role of Rb1 loss in aRMS tumor initiation and progression using conditional mouse models. RESULTS: Rb1 loss was not a necessary and sufficient mutational event for
rhabdomyosarcomagenesis, nor a strong cooperative initiating mutation. Instead, Rb1 loss was a modifier of progression and increased anaplasia and pleomorphism. Whereas Pax3:Foxo1a expression was unaltered, biomarkers of aRMS versus embryonal rhabdomyosarcoma were both increased, questioning whether these diagnostic markers are reliable in the context of Rb1 loss. Genome-wide gene expression in Pax3:Foxo1a,Rb1 tumors more closely approximated aRMS than embryonal rhabdomyosarcoma. Intrinsic loss of pRb function in aRMS was evidenced by insensitivity to a Cdk4/6 inhibitor regardless of whether Rb1 was intact or null. This loss of function could be attributed to low baseline Rb1, pRb and phospho-pRb expression in aRMS tumors for which the Rb1 locus was intact. Pax3:Foxo1a RNA interference did not increase pRb or improve Cdk inhibitor sensitivity. Human aRMS shared the feature of low and/or heterogeneous tumor cell pRb expression. CONCLUSIONS: Rb1 loss from an already low pRb baseline is a significant disease modifier, raising the possibility that some cases of pleomorphic rhabdomyosarcoma may in fact be Pax3:Foxo1a-expressing aRMS with Rb1 or pRb loss of function.

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[885]

TÍTULO / TITLE: - Leiomyosarcoma of the splenic vein.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Aguilar C; Socola F; Donet JA; Gallastegui N; Hernandez GA
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Edgardo Rebagliati Hospital, Lima, Peru.
RESUMEN / SUMMARY: - Leiomyosarcomas arising from the wall of blood vessels are rare and aggressive neoplasm. We report a case of a previously healthy 66-year-old woman who presented with intermittent abdominal pain, progressive constipation, and weight loss. Abdominal computed tomography showed a 12 cm solid heterogeneous tumor in the tail of the pancreas. The patient subsequently underwent surgical resection of the pancreatic mass. Surprisingly, histological and immunohistochemical analyses revealed leiomyosarcoma arising from the smooth muscle of the splenic vein. After surgery, she received adjuvant chemotherapy. One year later, there was no evidence of local recurrence. In this paper, we discuss the available information about leiomyosarcomas of splenic vein and its management.

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[886]

TÍTULO / TITLE: - Primary fibrosarcoma of the heart.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Kabashi S; Hoxha N; Gashi S; Ahmegjekaj I; Bejta I; Sadiku M; Ymeri H; Kabashi A; Bicaj X; Mucaj S
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Diagnostic Centre, UCCK, Pristine, Kosovo; Faculty of Medicine, Pristine University, Pristine, Kosovo.
RESUMEN / SUMMARY: - Primary malignant heart tumors represent rare entities where fibrosarcoma represents about 3% of all. Introducing the patient: A 15 years old patient
with cardiac insufficiency (heart failure) symptoms, such as weakness, cyanosis, palpitations and breathing difficulties; enlargement of upper mediastinum and pleural effusion. Through echocardiography a pericardial effusion and intracavitary thrombus in atrium was diagnosed. With computed tomography is diagnosed a tumoral mass in right atrium which is also spread in the right ventricle of the heart. Tumor is completely removed; pat histology result showed primary fibro sarcoma of the heart. At that time no metastasis was found. Conclusion. Primary malignant heart tumors may manifest like cardiac insufficiency or like systemic diseases. Fibrosarcomas are rare and have bad prognosis. On average patients can live around six months after initial symptoms appeared and diagnosis of the tumor was done. In the case of cardiac insufficiency with differential diagnosis we should also think of heart tumors, which could certainly be proved for or eliminated by echocardiography.

[887]

**TITULO / TITLE:** - Giant leiomyoma of the oesophagus with eosinophilic infiltration.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - British Medical J (BMJ). Acceso gratuito al texto completo.

- Enlace a la Editora de la Revista [http://bmj.com/search.dtl](http://bmj.com/search.dtl)


- Enlace al texto completo (gratuito o de pago) [1136/bcr-2013-201343](http://bmj.com/search.dtl)

**AUTORES / AUTHORS:** - Mutairi H; Al-Akkad M; Afzal M; Chaudhry I

**INSTITUCIÓN / INSTITUTION:** - Department of Thoracic Surgery, King Fahad Specialist Hospital, Dammam, Saudi Arabia.

**RESUMEN / SUMMARY:** - The most common benign tumour of the oesophagus is leiomyoma. Haemopoietic elements rarely infiltrate oesophageal leiomyoma. We report the case of a 24-year-old man with a long history of intermittent dysphagia. Endoscopy revealed external compression with normal oesophageal mucosa. A barium swallow study showed a defect in the oesophageal wall and a narrow oesophageal lumen. A CT scan of the chest confirmed a mass in the mid-oesophagus. A tumour was excised and the histology report revealed leiomyoma infiltrated with eosinophils, which is a rare variant.

[888]

**TITULO / TITLE:** - Lipoma of the sinus tarsi.

**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**REVISTA / JOURNAL:** - British Medical J (BMJ). Acceso gratuito al texto completo.

- Enlace a la Editora de la Revista [http://bmj.com/search.dtl](http://bmj.com/search.dtl)


- Enlace al texto completo (gratuito o de pago) [1136/bcr-2013-200904](http://bmj.com/search.dtl)

**AUTORES / AUTHORS:** - Fan KY; Lui TH

**INSTITUCIÓN / INSTITUTION:** - Department of Orthopaedics and Traumatology, North District Hospital, Hong Kong.

**RESUMEN / SUMMARY:** - Lipoma is rarely found in the foot. It is usually asymptomatic although sometimes can present with pain. We report a case of lipoma of the sinus tarsi presenting with functional instability.
Multifocal thoracic chordoma mimicking a paraganglioma.

Chordoma of thoracic vertebras is a very rare locally invasive neoplasm with low grade malignancy arising from embryonic notochordal remnants. Radical surgery remains the cornerstone of the treatment. We describe a case of multifocal T1-T2 chordoma, without bone and disc involvement, incidentally misdiagnosed as a paraganglioma, occurring in a 47-year-old male asymptomatic patient. Neoplasm was radically removed by an endocrine surgeon through a right extended cervicotomy. A preoperative reliable diagnosis of chordoma, as in the reported case, is often difficult. Radical surgery can provide a favorable outcome but, given the high rates of local recurrence of this neoplasm, a strict and careful follow-up is recommended. Although very rare, chordoma should be suggested in the differential diagnosis of the paravertebral cervical masses of unknown origin. Spine surgeon consultation and a FNB should be routinely included in the multidisciplinary preoperative work-up of these neoplasms.