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[1] TÍTULO / TITLE: - Spontaneous remission in patients with acute myeloid leukemia with
t(8;21) or cutaneous myeloid sarcoma: two case reports and a review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Zeng Q; Yuan Y; Li P; Chen T
INSTITUCIÓN / INSTITUTION: - Department of Hematology, Huashan Hospital, Fudan
University, China.
RESUMEN / SUMMARY: - Spontaneous remission (SR) in patients with acute myeloid
leukemia (AML) is rare. We herein present two such cases. The first case was of AML-
M2 accompanied by a bone marrow cytogenetic analysis revealing 46, XY,
t(8;21)(q22.q22). The second case was of isolated cutaneous myeloid sarcoma (MS)
that progressed to AML within seven months. Both of the patients had symptoms of
infection and anemia and were therefore treated with antibiotics and transfusions. The
SR lasted for two months and one month, respectively. Currently, the mechanisms
underlying SR remain ambiguous. Possible underlying mechanisms with a review of
the related literature are discussed.

[2] TÍTULO / TITLE: - Dosimetric accuracy of proton therapy for chordoma patients with
titanium implants.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Purpose: To investigate dosimetric errors in proton therapy treatment planning due to titanium implants, and to determine how these affect postoperative passively scattered proton therapy for chordoma patients with orthopedic hardware.

Methods: The presence of titanium hardware near the tumor may affect the dosimetric accuracy of proton therapy. Artifacts in the computed tomography (CT) scan can cause errors in the proton stopping powers used for dose calculation, which are derived from CT numbers. Also, clinical dose calculation algorithms may not accurately simulate proton beam transport through the implants, which have very different properties as compared to human tissue. The authors first evaluated the impact of these two main issues. Dose errors introduced by metal artifacts were studied using phantoms with and without titanium inserts, and patient scans on which a metal artifact reduction method was applied. Pencil-beam dose calculations were compared to models of nuclear interactions in titanium and Monte Carlo simulations. Then, to assess the overall impact on treatment plans for chordoma, the authors compared the original clinical treatment plans to recalculated dose distributions employing both metal artifact reduction and Monte Carlo methods.

Results: Dose recalculations of clinical proton fields showed that metal artifacts cause range errors up to 6 mm distal to regions affected by CT artifacts. Monte Carlo simulations revealed dose differences >10% in the high-dose area, and range differences up to 10 mm. Since these errors are mostly local in nature, the large number of fields limits the impact on target coverage in the chordoma treatment plans to a small decrease of dose homogeneity.

Conclusions: In the presence of titanium implants, CT metal artifacts and the approximations of pencil-beam dose calculations cause considerable errors in proton dose calculation. The spatial distribution of the errors however limits the overall impact on passively scattered proton therapy for chordoma.
the index ray was transferred to the base of the third metacarpal bone and fixed with a plate. The function of his right hand after the operation was excellent and the cosmetic appearance acceptable. There was no evidence of local recurrence or metastasis after 20 months follow up.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Froehner M; Koch R; Lossnitzer A; Schober RR; Schuler M; Wirth MP
INSTITUCIÓN / INSTITUTION: - Department of Urology, University Hospital Carl Gustav Carus, Dresden University of Technology, Fetscherstrasse 74, 01304, Dresden, Germany, Michael.Froehner@uniklinikum-dresden.de.
RESUMEN / SUMMARY: - PURPOSE: Inguinoscrotal sarcomas are exceedingly rare tumors. The aim of this study was to enable clinicians an easy and rapid access to the available information on this tumor entity. METHODS: An updated series of 21 men treated for sarcoma of the inguinoscrotal region at our institution between 1992 and 2012 was analyzed, and a systematic review of the literature with meta-analysis of outcome data was performed. The review was focused on demographic data, survival rates, prognostic factors, sites of relapse and complete remissions or successful treatments for metastatic disease. RESULTS: With only 38 %, the proportion of high-grade tumors in our sample was lower than reported in the literature and the 10-year relapse-free, disease-specific and overall survival rates were favorable with 77, 93 and 81 %. Beside our series, twelve studies including 345 patients were identified in the literature. The weighed mean 10-year relapse-free, disease-specific and overall survival rates were 63, 64 and 50 %. Only in patients with rhabdomyosarcoma, durable control of metastatic disease has been reported in more than one case (n = 4). Successful treatment in these cases consisted of a combination of complete surgical resection of metastatic lesions, subsequent chemotherapy and (optional) radiotherapy. CONCLUSIONS: Overall, about two-thirds of inguinoscrotal sarcomas may be cured. In series with a predominance of low-grade tumors, the long-term survival rates in completely excised inguinoscrotal sarcomas may be as favorable as in testicular germ cell tumors. Life-long surveillance is advisable to detect late recurrences.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm that originates from follicular dendritic cells in lymphoid follicles. FDCS has been increasingly reported in recent years. However, data on FDCS are mostly based on single case reports or case series and its natural history and standard treatment are not clear. To increase the understanding of this rare disease, we report our experience of three cases of FDCS with an analysis of the morphological and immunophenotypic characteristics, clinical course, treatment options and response to treatment. In addition, we reviewed the literature on FDCS.

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TÍTULO / TITLE: - A major response to trabectedin in metastatic malignant fibrous histiocytoma of the vertebra: a case report and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Sereno M; Merino M; Aguayo C; Hernandez S; Gutierrez-Gutierrez G; Zambrana Tevar F; Lopez-Gomez M; Gomez Raposo C; Casado-Saez E
INSTITUCIÓN / INSTITUTION: - Oncology Department, Infanta Sofia University Hospital, Madrid, España. mariasereno75@gmail.com
RESUMEN / SUMMARY: - Malignant fibrous histiocytoma is an aggressive tumor, the most common soft-tissue sarcoma of adult age. It is usually located in the extremities and retroperitoneum, and very rarely there is skeletal involvement. Surgery is the preferred treatment in early disease; in advanced disease, chemotherapy is the main therapeutic strategy. We present a 25-year-old female patient diagnosed with a vertebral mass in T5 with a severely compromised spinal cord. She underwent surgical decompression and the pathological findings were consistent with malignant fibrous histiocytoma. After several surgical treatments she had pulmonary progression and was therefore started on chemotherapy. She had a very poor response to most of the administered regimens until she initiated trabectedin 1 mg/m 2 every three weeks. She showed a significant improvement with a major response of the lung metastases. This report indicates that trabectedin is an active drug in advanced, previously treated metastatic malignant fibrous histiocytoma.

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TÍTULO / TITLE: - Uncommon synchronous association between ovarian carcinoma and gastrointestinal stromal tumor: a case study and literature review.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
 ●● Enlace al texto completo (gratuito o de pago) 1700/1283.14211
AUTORES / AUTHORS: - Favero G; Pfiffer T; Riedlinger WF; Chiantera V; Schneider A
INSTITUCIÓN / INSTITUTION: - Department of Gynecology, Charite Universitatsmedizin Berlin, Berlin, Germany. gdifavero@hotmail.com
RESUMEN / SUMMARY: - BACKGROUND: The association of gastrointestinal stromal tumors (GIST) and other cancers is well known, but its synchronous occurrence with gynecological malignancies is very uncommon. Usually, the diagnosis is accidentally established. We describe a patient with GIST and concurrent ovarian cancer and discuss the clinical implications of this finding. CASE REPORT: A 64-year-old woman with a prior diagnosis of ovarian cancer developed a second recurrence after having undergone two operations and adjuvant chemotherapy. While tumor debulking was performed, a small, nonsuspicious lesion was removed from the greater curvature of the stomach. Histology revealed a GIST. CONCLUSION: The association of GIST and ovarian cancer is a rarity and its synchronicity may alter the oncological prognosis and therapy of the patient.

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TÍTULO / TITLE: - Dense pattern of embryonal rhabdomyosarcoma, a lesion easily confused with alveolar rhabdomyosarcoma: a report from the Soft Tissue Sarcoma Committee of the Children’s Oncology Group.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
 ●● Enlace al texto completo (gratuito o de pago) 1309/AJCPA1WN7ARPCMKQ
AUTORES / AUTHORS: - Rudzinski ER; Teot LA; Anderson JR; Moore J; Bridge JA; Barr FG; Gastier-Foster JM; Skapek SX; Hawkins DS; Parham DM
INSTITUCIÓN / INSTITUTION: - Seattle Children’s Hospital, Department of Laboratories, M/S OC.8.720, 4800 Sandpoint Way NE, Seattle, WA 98015, USA. erin.rudzinski@seattlechildrens.org
RESUMEN / SUMMARY: - OBJECTIVES: To examine whether the frequency of fusion-negative alveolar rhabdomyosarcoma (ARMSn) increased coincident with changes in the definition of alveolar histology. METHODS: We re-reviewed alveolar rhabdomyosarcoma (ARMS) in the Children’s Oncology Group study D9803, comparing histopathology with fusion status. RESULTS: Our review of 255 original ARMS cases (compared with a control group of 38 embryonal rhabdomyosarcomas [ERMS] cases) revealed that many had an ARMS-like densely cellular pattern with cytologic features and myogenin expression more typical of ERMS. Following re-review, 84 (33%) cases of original ARMS were re-diagnosed as ERMS. All reclassified ERMS, including dense ERMS, were fusion negative, whereas 82% of confirmed ARMS cases were fusion positive. Total ARMS diagnoses returned to historic rates of 25% to 30% of all rhabdomyosarcomas, and ARMSn decreased from 37% to 18% of
ARMS cases. The outcome of reclassified ERMS was similar to confirmed ERMS.

CONCLUSIONS: To address the role of fusion status in risk stratification, pathologists should include both a histologic diagnosis and an evaluation of fusion status for all new ARMS diagnoses.

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

AUTORES / AUTHORS: - Drolet BA; Trenor CC 3rd; Brandao LR; Chiu YE; Chun RH; Dasgupta R; Garzon MC; Hammill AM; Johnson CM; Tlougan B; Blei F; David M; Elluru R; Frieden JI; Friedlander SF; Iacobas I; Jensen JN; King DM; Lee MT; Nelson S; Patel M; Pope E; Powell J; Seefeldt M; Siegel DH; Kelly M; Adams DM

INSTITUCIÓN / INSTITUTION: - Departments of Pediatrics and Dermatology, Medical College of Wisconsin, Milwaukee, WI. Electronic address: bdrolet@mcw.edu.

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

AUTORES / AUTHORS: - Jiao G; Ren T; Lu Q; Sun Y; Lou Z; Peng X; Liang W; Guo W

INSTITUCIÓN / INSTITUTION: - Musculoskeletal Tumor Center, Peking University People’s Hospital, 100044, Beijing, People’s Republic of China.

RESUMEN / SUMMARY: - Published studies researching the prognostic significance of cyclooxygenase-2 (COX-2) expression in patients with osteosarcoma are inconclusive and heterogeneous. We conducted a meta-analysis to assess its prognostic value more precisely. The pooled odds ratios (ORs) or hazard ratios (HRs) with corresponding 95 % confidence intervals (CIs) were calculated to evaluate the effects. Fourteen studies with 735 osteosarcoma patients were included to estimate the relationship between COX-2 and metastasis of tumor, clinical stage, and 3-year overall survival. High expressions of COX-2 predicted neoplasm metastasis (OR = 1.891, 95 % CI 1.276-2.803, P = 0.002), advanced clinical stage (OR = 1.801, 95 % CI 1.257-2.581, P = 0.001). In addition, high COX-2 expression tended to be associated with a poor 3-year survival (HR = 1.741, 95 % CI 0.762-3.979, P = 0.188), but the difference was not significant. Therefore, this meta-analysis demonstrated that high COX-2 expression might be an unfavorable prognostic effect in osteosarcoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
  ●● Enlace al texto completo (gratuito o de pago) 1016/j.jpeds.2013.05.024
AUTORES / AUTHORS: - Fisher PG

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
  ●● Enlace al texto completo (gratuito o de pago) 1016/j.ejca.2013.06.036
AUTORES / AUTHORS: - Furtwangler R; Gooskens SL; van Tinteren H; de Kraker J; Schleiermacher G; Bergeron C; de Camargo B; Acha T; Godzinski J; Sandstedt B; Leuschner I; Vujanic GM; Pieters R; Graf N; van den Heuvel-Eibrink MM
INSTITUCIÓN / INSTITUTION: - Department of Pediatric Hematology/Oncology, University Hospital for Children, Homburg, Germany.
RESUMEN / SUMMARY: - PURPOSE: Clear Cell Sarcoma of the Kidney (CCSK) is a rare childhood renal tumour. Only a few homogeneously treated CCSK cohorts have been reported. This study aims to describe clinical characteristics and survival of CCSK patients treated according to recent International Society of Pediatric Oncology (SIOP) protocols. PATIENTS AND METHODS: We analysed the prospectively collected data of patients with a histologically verified CCSK, entered onto SIOP 93-01/2001 trials. RESULTS: A total of 191 CCSK patients (64% male) were analysed, with a median age at diagnosis of 2.6 years. Stage distribution for stages I, II, III and IV was 42%, 23%, 28% and 7%, respectively. Pre-operative chemotherapy was administered to 169/191 patients. All patients underwent total nephrectomy and 189/191 patients received post-operative chemotherapy. Radiotherapy was applied in 2/80 stage I, 33/44 stage II, 44/54 stage III and 6/13 stage IV patients. Five year event-free survival (EFS) and overall survival (OS) were 79% (95% confidence interval (CI): 73-85%) and 86% (95% CI: 80-92%) respectively. Stage IV disease and young age were significant adverse prognostic factors for event-free survival. Factors such as gender, tumour volume and type of initial treatment were not found to be prognostic for EFS and OS. CONCLUSION: In this largest SIOP cohort described so far, overall outcome of CCSK is reasonable, although treatment of young and advanced-stage disease patients is challenging. As further intensification of treatment is hampered by direct and late toxicity, future directions should include the development of targeted therapy based on specific molecular aberrations of CCSK.
TÍTULO / TITLE: - Toward Better Soft Tissue Sarcoma Staging: Building on American Joint Committee on Cancer Staging Systems Versions 6 and 7.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Maki RG; Moraco N; Antonescu CR; Hameed M; Pinkhasik A; Singer S; Brennan MF
INSTITUCIÓN / INSTITUTION: - Tisch Cancer Institute, Departments of Medicine, Pediatrics and Orthopaedics, Mount Sinai School of Medicine, New York, NY, USA, bobmakimd@gmail.com.

RESUMEN / SUMMARY: - BACKGROUND: Based on review of patient data in case conferences over time, we hypothesized that clinically relevant data are omitted in routine soft tissue sarcoma staging. METHODS: We examined subsets of a prospectively collected single institution soft tissue sarcoma database with respect to criteria of the AJCC versions 6 (2002) and 7 (2010) staging systems and examined their clinical outcomes. RESULTS: Relapse-free survival decreases with increasing primary tumor size in four categories, versus two categories used in AJCC 6 and 7 staging. Disease-specific survival decreases over three categories. Conversely, omission of tumor depth as a prognostic factor in version 7 appears supported, since tumor depth is not an independent risk factor for disease-specific survival by multivariate analysis. Patients with nodal disease and no other metastases fare better than patients with other metastases, but have inferior outcomes compared with patients with large high-grade tumors without nodal metastasis. Multivariate analysis identified size, site, grade, age, nodal metastatic disease, and other metastatic disease as independent risk factors for disease-specific survival. Versions 6 and 7 criteria are tacit regarding anatomic site and histology for tumors with identical FNCLCC grade. CONCLUSIONS: Improved patient risk assessment may be achieved by staging using a larger number of size categories. Staging system refinements come at the cost of a larger number of staging categories. Histology or site-specific staging systems, nomograms or Bayesian belief networks may provide more accurate means to assess clinical outcomes.

[13]
TÍTULO / TITLE: - Primary sarcomas of the salivary glands: Case series and literature review.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cockerill CC; Daram S; El-Naggar AK; Hanna EY; Weber RS; Kupferman ME
BACKGROUND: Mesenchymal malignancies of salivary origin are rare and are histologically diverse. We reviewed our experience with these tumors, as well as the published literature, with an emphasis on treatment modalities and prognosis. METHODS: We identified 17 patients treated for malignant mesenchymal cell tumors at The University of Texas MD Anderson Cancer Center between 1990 and 2007. We compared our results to the literature from January 1990 to July 2010. RESULTS: Tumors were located primarily in the parotid gland and were primarily T1 tumors (<5 cm). All patients were treated with surgical resection, and 13 patients were given adjuvant therapy. Seven patients (41%) had recurrence, and 4 developed distant metastases. The overall 5-year and 10-year survival rates were 42% and 20%, respectively. CONCLUSIONS: Our case series and literature review show that sarcomas of the salivary glands have a high rate of recurrence and are associated with a poor prognosis. © 2013 Wiley Periodicals, Inc. Head Neck, 2013.

[14]

TÍTULO / TITLE: - Desmoplastic fibroma of the pediatric cranium: case report and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cho BH; Tye GW; Fuller CE; Rhodes JL
INSTITUCIÓN / INSTITUTION: - Virginia Commonwealth University School of Medicine, Richmond, VA, 23298, USA, chobh@vcu.edu.
RESUMEN / SUMMARY: - PURPOSE: Desmoplastic fibromas are primary bone tumors that seldom occur in the cranial bones. Furthermore, reports of desmoplastic fibromas of the skull in children are exceedingly rare. Although desmoplastic fibromas are histologically benign, they are locally aggressive and have a propensity to reoccur. Their radiographic appearance may mimic other more common central nervous system and bone neoplasms. There are only 19 reported cases of desmoplastic fibroma of the cranium in the literature, and only seven occurred in the pediatric age group. We present a case report of an 11-year-old female patient with a desmoplastic fibroma of the parieto-occipital region and review the literature. CASE REPORT: An 11-year-old female presented to the craniofacial clinic complaining of intermittent pain and a soft mass in the occipital region. There was a distant history of trauma to the region that did not require medical intervention. Computed tomography imaging revealed a lytic bone lesion overlying the sagittal sinus in the parieto-occipital region. Surgical resection with wide margins and immediate autologous reconstruction was performed. Pathological analysis revealed a desmoplastic fibroma. At 4 months of follow-up, no recurrence has been noted. CONCLUSION: Desmoplastic fibroma of the cranium is rare. Complete surgical resection with careful follow-up is the treatment of choice.
TÍTULO / TITLE: - Epstein-Barr virus negative primary hepatic leiomyoma: Case report and literature review.

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


AUTORES / AUTHORS: - Luo XZ; Ming CS; Chen XP; Gong NQ

INSTITUCIÓN / INSTITUTION: - Xian-Zhang Luo, Chang-Sheng Ming, Xiao-Ping Chen, Nian-Qiao Gong, Institute of Organ Transplantation, Department of Surgery, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430030, Hubei Province, China.

RESUMEN / SUMMARY: - Primary hepatic leiomyoma is a neoplasm of mesenchymal origin and occurs only rarely. Secondary to benign smooth muscle proliferation, it is usually found in adult women and is associated with Epstein-Barr virus (EBV) infection. Here, we report the 29(th) case of primary hepatic leiomyoma with its unique features related to diagnosis, treatment and developmental biology. A 48-year-old man, with an immunocompromised status, complained of pain in the upper quadrant of the abdomen. Serological analysis indicated no presence of hepatitis virus, no human immunodeficiency virus, and no EBV infection. The levels of alpha-fetoprotein and carcinoembryonic antigen were normal. A mass was detected in segment III of the hepatic lobe by ultrasonography and an abdominal computed tomography scan. Endoscopy had negative findings. Exploratory laparotomy found no existing extrahepatic tumor and left lateral lobectomy was performed. Pathological examination showed the mass to be a typical leiomyoma. The cells were positive for alpha-smooth muscle actin and desmin, and negative for the makers of gastrointestinal stromal tumor (GIST), including CD117, CD34 and DOG1 (discovered on GIST1). In situ hybridization revealed negative status for EBV-encoded small RNA. After left lateral lobectomy, the patient was not given chemotherapy or radiotherapy. During a 2-year follow-up, no sign of local recurrence or distant metastasis was observed. In conclusion, we report a rare case of primary hepatic leiomyoma in a male patient without EBV infection. Hepatic resection was curative. This case presents data to expand our knowledge concerning the complex and heterogeneous nature of primary liver leiomyoma, indicating that EBV infection is important but neither necessary nor sufficient for the development of primary liver leiomyoma.

TÍTULO / TITLE: - A case of lipoma of lateral anterior neck treated with surgical enucleation.

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

Lipoma arise in almost 50% of all soft tumours. The neck lipomas are rare tumours that may present as painless masses with slow growth, in the lateral portions of the neck. Some lipomas, such as the one studied in our case, grow deep in the subcutaneous tissue, in close contact with muscles. Here, we report a case of lipoma extending from pre-tragal region up to the ascending branch of the mandible in a 62 year old man, treated with enucleation. The inferior margin of lipoma involved the pharyngeal and the superior margin was achieved by the top of the skull base. The mass of lipoma caused breathing difficulties in the patient, preventing regular sleep. No complication was recorded in the post-operative period and no further surgery was performed. The complete resolution after one year's follow-up, together with the rarity of the anatomical site, makes this case worthy of description. A correct diagnosis facilitated removal of this lesion with a surgical method.
our patient received rapamycin therapy with about six-fold reduction of the tumor growth rate upon 10-month follow-up neuroimaging. This is the first case of chordoma in whom chemotherapy was tailored on the basis of the sensitivity of patient-derived tumor cells.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Bhadage CJ; Vaishampayan S; Kolhe S; Umarji H
INSTITUCIÓN / INSTITUTION: Department of Oral Medicine and Radiology, MGV’s KBH Dental College and Hospital, Nashik, India.
RESUMEN / SUMMARY: Inflammatory lesions, like periapical/odontogenic abscesses, are by far the most common pathologic condition of the jaws. Radiographically, these lesions commonly manifest as widening of periodontal ligament space, discontinuity of lamina dura and ill-defined periapical radiolucency. There are some rare disorders which could cause similar radiographic changes in the jaw bone. With careful scrutiny of periapical radiolucency, regular periodic follow-up radiographs and histopathologic examination, the periapical abscess or infection can be differentiated from rare fatal disorders. CLINICAL RELEVANCE: This paper highlights the need for vigilant examination of even the commonest, innocuous-appearing periapical changes which sometimes are produced by some rare fatal disorders.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Fraioli MF; Lecce M; Fraioli C; Paolo C
INSTITUCIÓN / INSTITUTION: University of Rome Tor Vergata, Rome, Italy.
RESUMEN / SUMMARY: BACKGROUND: Patients affected by tuberous sclerosis (TS) have a greater incidence of tumors than the healthy population. Spinal tumours in TS are reported very rarely and consist mainly of sacrococcygeal and cervical chordomas. METHOD: Case report. FINDINGS: A 21-year-old man, affected by TS, presented a spinal dorsal T2 tumor that caused medullary compression. He underwent decompressive laminectomy and microsurgical excision of a giant cell tumor and an associated aneurysmal bone cyst. Postoperative hypofractionated radiotherapy was
performed on the surgical field. At 2.4 years of follow-up the patient reported total recovery of neurological deficits and was free from tumor recurrence. CONCLUSION: Considering this association, which is the first reported in the literature, spinal magnetic resonance imaging with gadolinium should be performed at the onset of spinal pain in patients affected by TS.

[19]
TÍTULO / TITLE: - Primary inflammatory malignant fibrous histiocytoma of the breast: A case report of an unusual variant and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Tashjian R; Gilani SM; Falk J; Kelly MJ; Ockner D; Danforth R
INSTITUCIÓN / INSTITUTION: - Department of Pathology, St. John Hospital & Medical Center, Detroit, MI, USA. Electronic address: randy.tashjian.md@gmail.com.
RESUMEN / SUMMARY: - Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma of adults, but its presence in the breast is rare. We report a case of primary inflammatory MFH in a 72-year-old Caucasian female with no previous medical history and no prior radiation exposure. She presented with a palpable mass that was suspicious for malignancy on mammography. Histologic evaluation of the core needle biopsy revealed sheets of large, pleomorphic neoplastic cells within a dense background of acute and chronic inflammatory cells. The neoplastic cells exhibited a moderate to abundant amount of finely vacuolated cytoplasm and atypical nuclei with vesicular nuclear chromatin and prominent nucleoli. Mitotic activity was readily identified, and foci of necrosis were noted. The neoplastic cells were immunoreactive with CD68, alpha 1-antitrypsin, alpha 1-antichymotrypsin, and vimentin. The diagnosis of MFH was rendered after thorough microscopic examination of the entire mass following mastectomy. MFH of the breast is a diagnosis of exclusion. The definitive treatment of MFH is surgical, either with wide local excision or total mastectomy. The roles of sentinel lymph node biopsy, axillary lymph node dissection, chemotherapy, and radiation have yet to be definitively clarified. The prognosis of MFH of the breast is generally poor.

[20]
TÍTULO / TITLE: - Prognosis analysis of sarcomatous intrahepatic cholangiocarcinoma from a review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Watanabe G; Uchinami H; Yoshioka M; Nanjo H; Yamamoto Y
RESUMEN / SUMMARY: - BACKGROUND: Sarcomatous intrahepatic cholangiocarcinoma (ICC) is a rare histological variant of ICC. The prognosis of sarcomatous ICC is poorly understood. METHODS: We analyzed the prognosis of sarcomatous ICC by reviewing the previous reports and our own case. RESULTS: Only 15 cases of sarcomatous ICC have been reported in the English-language literature so far. Median survival time of patients with sarcomatous ICC with and without surgery was 11 and 3 months, respectively. Survival rate of patients operated on for sarcomatous ICC was similar to that of patients with ordinary ICC without surgery in the early postoperative period. In the long-term view, however, the prognosis for the patients with sarcomatous ICC receiving surgery was better than that for the patients with ordinary ICC without surgery. CONCLUSION: Although the prognosis for the patients with sarcomatous ICC was poor even after curative resection, surgery would be justified as the primary treatment for sarcomatous ICC.


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


AUTORES / AUTHORS: - Zuo D; Zheng L; Sun W; Fu D; Hua Y; Cai Z

INSTITUCIÓN / INSTITUTION: - Department of Musculoskeletal Oncology, Shanghai Tenth People’s Hospital, Tongji University School of Medicine, Shanghai 200072, China. yingqi.hua@yahoo.com.

RESUMEN / SUMMARY: - BACKGROUND: Reports of recurrence following restructuring of primary giant cell tumor (GCT) defects using polymethyl methacrylate (PMMA) bone cementation or allogeneic bone graft with and without adjuvants for intralesional curettage vary widely. Systematic review and meta-analysis were conducted to investigate efficacy of PMMA bone cementation and allogeneic bone grafting following intralesional curettage for GCT. METHODS: Medline, EMBASE, Google Scholar, and Cochrane databases were searched for studies reporting GCT of bone treatment with PMMA cementation and/or bone grafting with or without adjuvant therapy following intralesional curettage of primary GCTs. Pooled risk ratios and 95% confidence intervals (CIs) for local recurrence risks were calculated by fixed-effects methods. RESULTS: Of 1,690 relevant titles, 6 eligible studies (1,293 patients) spanning March 2008 to December 2011 were identified in published data. Treatment outcomes of PMMA-only (n = 374), bone graft-only (n = 436), PMMA with or without adjuvant (PMMA + adjuvant; n = 594), and bone graft filling with or without adjuvant (bone graft
+ adjuvant; n = 699) were compared. Bone graft-only patients exhibited higher recurrence rates than PMMA-treated patients (RR 2.09, 95% CI (1.64, 2.66), Overall effect: Z = 6.00; P <0.001), and bone graft + adjuvant patients exhibited higher recurrence rates than PMMA + adjuvant patients (RR 1.66, 95% CI (1.21, 2.28), Overall effect: Z = 3.15, P = 0.002). CONCLUSIONS: Local recurrence was minimal in PMMA cementation patients, suggesting that PMMA is preferable for routine clinical restructuring in eligible GCT patients. Relationships between tumor characteristics, other modern adjuvants, and recurrence require further exploration.

[22]
TÍTULO / TITLE: - Giant cell tumor of tendon sheath: study of 64 cases and review of literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Di Grazia S; Succi G; Fragetta F; Perrotta RE
RESUMEN / SUMMARY: - The giant cell tumor of tendon sheath (GCTTS) is the most common benign neoplasm in the hand after the ganglion cyst. Several hypotheses were formulated about the etiological factors of these tumors, but still there is not a common opinion on etiology, prognostic factors and recurrence rate. This article presents a review of literature of the last 15 years about GCTTS to assess the demographic, clinical and histological profile. We compared the information obtained from literature with our experience of 64 cases between 2000 and 2012. Our study showed similar results to those reported in literature, except for the recurrence rate: only 3 cases (4.7%) of 64 patients reported recurrence (versus about 15% on average in literature). Among the various possible factors that predispose to recurrence, it is necessary that the surgeon ensures complete excision of the tumor and removal of any residual satellite nodules. Although the marginal excision is the treatment of choice, it is often difficult to perform due to for the location and the strict adherence of the tumor to the tendon or neurovascular bundles. We used in all cases a magnifying loupe to help a careful research of satellite lesions and to respect surrounding structures.

[23]
TÍTULO / TITLE: - Giant retroperitoneal leiomyoma A case report and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Fama F; Patti R; Linard C; Saint Marc O; Piquard A; Gioffre Florio M
RESUMEN / SUMMARY: - Retroperitoneal leiomyomata are infrequent, and their prevalence among primary retroperitoneal tumours has been estimated as 0.5-1.2%. The authors report a case of symptomatic retroperitoneal leiomyoma with a favourable prognosis. A 53-year-old woman presented for abdominal pain associated to an
inflammatory syndrome. A contrast-enhanced computed tomography revealed a large abdominopelvic mass and patient underwent open surgical excision. Definitive diagnosis was done after immunohistochemical assessment. Immunoreactivity was strong for smooth muscular actin. Presence of oestrogen and progesterone receptor proteins was also detected. Prognosis of these well-differentiated smooth muscle tumours is generally favourable but a postoperative surveillance is always recommended. KEY WORDS: Leiomyoma, Retroperitoneal space, Surgery.

[24]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Wang Z; He ML; Zhao JM; Qing HH; Wu Y
INSTITUCIÓN / INSTITUTION: - Graduate School of Guangxi Medical University, Nanning, Guangxi, China E-mail: zhaojinmin@hotmail.com.
RESUMEN / SUMMARY: - Various studies examining the relationship between Ezrin overexpression and response to chemotherapy and clinical outcome in patients with osteosarcoma have yielded inconclusive results. We accordingly conducted a meta-analysis of 7 studies (n = 318 patients) that evaluated the correlation between Ezrin and histologic response to chemotherapy and clinical prognosis (death). Data were synthesized in receiver operating characteristic curves and with fixed-effects and random-effects likelihood ratios and risk ratios. Quantitative synthesis showed that Ezrin is not a prognostic factor for the response to chemotherapy. The positive likelihood ratio was 0.538 (95% confidence interval [95% CI], 0.296-0.979; random-effects calculation), and the negative likelihood ratio was 2.151 (95% CI, 0.905-5.114; random-effects calculations). There was some between-study heterogeneity, but no study showed strong discriminating ability. Conversely, Ezrin positive status tended to be associated with a lower 2-year survival (risk ratio, 2.45; 95% CI, 1.26-4.76; random-effects calculation) with some between-study heterogeneity that disappeared when only studies that employed immunohistochemistry were considered (risk ratio, 2.97; 95% CI, 2.01-4.40; fixed-effects calculation). To conclude, Ezrin is not associated with the histologic response to chemotherapy in patients with osteosarcoma, whereas Ezrin positivity was associated with a lower 2-year survival rate regardless of risk of death at 2 years. Expression change of Ezrin is an independent prognostic factor in patients with osteosarcoma.

[25]
TÍTULO / TITLE: - Combination therapy for scalp angiosarcoma using bevacizumab and chemotherapy: a case report and review of literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Yang P; Zhu Q; Jiang F

INSTITUCIÓN / INSTITUTION: - Department of Oncology, PLA Navy General Hospital, Beijing 100048, China.

RESUMEN / SUMMARY: - Bevacizumab, an angiogenesis inhibitor, is a recombinant humanized monoclonal antibody against vascular endothelial growth factor and a promising therapeutic option for angiosarcoma management. This is a case report and review of the literature using bevacizumab and combination chemotherapy for angiosarcoma. The understanding of the effectiveness of combined therapy of bevacizumab and chemotherapy agents is still limited. The benefits of bevacizumab treatment for angiosarcoma will need to be weighed against the risks of venous thromboembolism in this population.

[26]

TÍTULO / TITLE: - Isolated plexiform neurofibroma over left palm: a case report and review of literature.

AUTORES / AUTHORS: - Kudur MH; Hulmani M

INSTITUCIÓN / INSTITUTION: - Departments of Dermatology and Venereology, Srinivas Institute of Medical Sciences and Research Centre, Mukka, Surathkal, Mangalore, India.

RESUMEN / SUMMARY: - Plexiform neurofibroma is common over the branches of trigeminal and cervical nerves over the face. Plexiform neurofibroma over palm is rare and affects the individuals with fine motor functions of hand. Here, we report a case of isolated plexiform neurofibroma over the palm with review of literature.

[27]

AUTORES / AUTHORS: - Sampson VB; Gorlick R; Kamara D; Anders Kolb E

INSTITUCIÓN / INSTITUTION: - Nemours Center for Childhood Cancer and Blood Disorders, Alfred I. duPont Hospital for Children, Wilmington, DE, USA.
RESUMEN / SUMMARY: Osteosarcoma, the most common malignant bone tumor of childhood, is a high-grade primary bone sarcoma that occurs mostly in adolescence. Standard treatment consists of surgery in combination with multi-agent chemotherapy regimens. The development and approval of imatinib for Philadelphia chromosome-positive acute lymphoblastic leukemia in children and the fully human monoclonal antibody, anti-GD2, as part of an immune therapy for high-risk neuroblastoma patients have established the precedent for use of targeted inhibitors along with standard chemotherapy backbones. However, few targeted agents tested have achieved traditional clinical endpoints for osteosarcoma. Many biological agents demonstrating anti-tumor responses in preclinical and early-phase clinical testing have failed to reach response thresholds to justify randomized trials with large numbers of patients. The development of targeted therapies for pediatric cancer remains a significant challenge. To aid in the prioritization of new agents for clinical testing, the Pediatric Preclinical Testing Program (PPTP) has developed reliable and robust preclinical pediatric cancer models to rapidly screen agents for activity in multiple childhood cancers and establish pharmacological parameters and effective drug concentrations for clinical trials. In this article, we examine a range of standard and novel agents that have been evaluated by the PPTP, and we discuss the preclinical and clinical development of these for the treatment of osteosarcoma. We further demonstrate that committed resources for hypothesis-driven drug discovery and development are needed to yield clinical successes in the search for new therapies for this pediatric disease.

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TÍTULO / TITLE: Converging paths to progress for skull base chordoma: Review of current therapy and future molecular targets.

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


AUTORES / AUTHORS: Di Maio S; Kong E; Yip S; Rostomily R

INSTITUCIÓN / INSTITUTION: Division of Neurosurgery, McGill University, Jewish General Hospital, Montreal, QC, Canada.

RESUMEN / SUMMARY: BACKGROUND: Chordomas of the skull base are rare locally aggressive neoplasms with a predilection for encapsulating critical neurovascular structures, bony destruction and irregular growth patterns, and from which patients succumb to recurrence and treatment failures. METHODS: A review of the medical literature is performed, using standard search engines and identifying articles related to skull base chordomas, surgery, radiation therapy, chemotherapy, molecular genetics, and prospective trials. RESULTS: A synthesis of the literature is presented, including sections on pathology, treatment, molecular genetics, challenges, and future directions. CONCLUSION: Beyond an understanding of the current treatment paradigms for skull base chordomas, the reader gains insight into the collaborative approach applied to orphan diseases, of which chordomas is a prime exemplar.

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RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary  
●● Enlace al texto completo (gratuito o de pago) 1007/s12262-012-0587-4  
AUTORES / AUTHORS: - Deo SV; Manjunath NM; Shukla NK  
INSTITUCIÓN / INSTITUTION: - Department of Surgical Oncology, Institute Rotary cancer Hospital, All India Institute of Medical Sciences, Ansari Nagar, New Delhi, 110029 India.  
RESUMEN / SUMMARY: - Soft tissue sarcomas (STS) constitute a rare and challenging group of solid tumor in the field of oncology. Unlike other malignancies STS can affect a wide variety of anatomical regions in the body with varied histo-pathological variants and clinical outcomes. There are controversies in the diagnosis and management of STS due to rarity and heterogeneity of the disease entity. Due to dedicated research and advances made in the field of imaging, pathology, surgery, radiotherapy and chemotherapy certain controversies were laid to rest and treatment approach to STS could be standardized to a large extent in the recent past. A review of controversies related to STS was performed in this article and an attempt was made to present a balanced view pertaining to these issues.  
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RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary  
●● Enlace al texto completo (gratuito o de pago) 3109/2000656X.2013.812969  
AUTORES / AUTHORS: - Akram J; Wooler G; Lock-Andersen J  
INSTITUCIÓN / INSTITUTION: - Department of Plastic Surgery.  
RESUMEN / SUMMARY: - Abstract Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous sarcoma that frequently recurs locally, but rarely metastasizes. The purpose of this work is to present a clinical series of DFSP patients and national Danish incidence data in the period 2000-2012. Furthermore, the aim is to present guidelines on the management based on a review of the literature. Medical records of 26 consecutively treated patients at the Department of Plastic Surgery in Health Care Region Zealand were reviewed and histological specimens were reassessed. To investigate national Danish incidence in the period 2000-2012, data were extracted from the national pathology registry. Finally, a literature search was performed in Pubmed and Cochrane, and 23 major publications were reviewed. Studies on Mohs Micrographic surgery were excluded. All patients were treated with wide local excision (WLE) with a median margin of 2.8 cem and a median follow-up time of 36 months. We found a local recurrence rate of 4%. Our national incidence data were based on 374 patients. The overall incidence was 0.53 per 100,000 persons. The prevalence of
DFSP in the age group 20-50 years was significantly higher than the group below 20 years (p < 0.0001). Surgery is the treatment of choice for primary DFSP, local recurrences, and metastases. If clear margins cannot be obtained by WLE or surgery is not an option because of unacceptable functional or cosmetic outcome, adjuvant radiotherapy or imatinib can be considered. Chemotherapy can be a final option if other treatments fail.

[31] TÍTULO / TITLE: - Surgical margins and handling of soft-tissue sarcoma in extremities: a clinical practice guideline.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ● Enlace al texto completo (gratuito o de pago) 3747/co.20.1308
AUTORES / AUTHORS: - Kandel R; Coakley N; Werier J; Engel J; Ghert M; Verma S
INSTITUCIÓN / INSTITUTION: - Department of Pathology and Laboratory Medicine, Mount Sinai Hospital, Toronto, ON.
RESUMEN / SUMMARY: - QUESTIONS: In limb salvage surgery for extremity soft-tissue sarcoma (sts), what is an adequate surgical margin? What is the appropriate number of samples to take from the margins of a surgical resection specimen? What is the appropriate handling of surgical resection specimens?
BACKGROUND: Surgery is the primary treatment for extremity sts. The combination of radiotherapy with surgery allows for limb salvage by using radiation to biologically “sterilize” microscopic extensions of tumour and to spare neurovascular and osseous structures. Adjuvant chemotherapy in sts-except for rhabdomyosarcoma and Ewing sarcoma-continues to be controversial.
METHODS: The medline and embase databases (1975 to June 2011) and the Cochrane Library were searched for pertinent studies. The Web sites of the main guideline organizations and the American Society of Clinical Oncology conference proceedings (2007-2010) were also searched.
RESULTS AND CONCLUSIONS: Thirty-three papers, including four guidelines, one protocol, and one abstract, were eligible for inclusion. The data suggest that patients with clear margins have a better prognosis, but no prospective studies have indicated how wide margins should be. In limb-salvage surgery for extremity sts, the procedure should be planned to achieve a clear margin. However, to preserve functionality, surgery may result in a very close (<1 cm) or even microscopically positive margin. In this circumstance, the use of preoperative or postoperative radiation should be considered. No studies described the optimal number of tissue sections required to assess adequacy of excision nor the appropriate handling of surgical resection specimens. The Sarcoma Disease Site Group made its recommendations based on expert opinion and consensus.

[32] TÍTULO / TITLE: - Postradiation osteosarcoma in an older prostate cancer survivor: case study and literature review with emphasis on geriatric principles.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
La población envejecida y el aumento del número de supervivientes de cáncer probablemente estarán asociados con más malignidades secundarias debido a tratamiento previo de cáncer. Dado que la incidencia de la mayoría de los cánceres aumenta con la edad, estos trastornos asociados con el tratamiento maligno probablemente afectarán de manera desproporcionada a los adultos mayores. Aquí, presentamos el caso de un hombre de 78 años con historia de cáncer prostático localizado tratado con radioterapia externa hace 11 años, quien desarrolló osteosarcoma de la ilíaca. Sheet oro de salud mostró un hombre mayor de buena forma con pocos comorbilidades, independencia funcional y no otro síndrome geriátrico. Dado que el paciente prefirió una operación que preservara la extremidad, se llevó a cabo un tratamiento neoadyusante. Con la escasez de datos clínicos en osteosarcoma en adultos mayores, el paciente se sometió a un régimen de carboplatin (sustituido para cisplatino), doxorubicina y metotrexato. Desafortunadamente, desarrolló insuficiencia renal aguda inducida por metotrexato. El tratamiento quimioterápico fue discontinuado y se realizó hemipelvectomía. La evolución postoperatoria estuvo marcada por numerosas complicaciones, incluyendo delirio, depresión y rehospitalizaciones recurrentes. Finalmente se desarrolló un recidiva local y el paciente eligió cuidados paliativos.

Este caso subraya los desafíos de manejar a adultos mayores con malignidades asociadas con el tratamiento. Los clínicos se enfrentan a la falta de datos clínicos de los que extrapolar las limitaciones de las opciones terapéuticas debido a tratamiento previo y una limitada capacidad para predecir con precisión cuáles abuelos experimentarán resultados adversos. Se necesitan mejores enfoques para ayudar a los pacientes mayores a tomar decisiones que cumplan con sus objetivos de cuidado y mejorar el cuidado de adultos mayores con malignidades asociadas con el tratamiento.
the literature from 1975 to 2011. In total, 45 patients with giant osteomas arising from the ethmoid and frontal sinuses (including the present cases) have been reported in 41 articles. Headache and ocular signs are the most common symptoms. This disease often leads to intracranial or intraorbital complications. The main treatment for giant osteoma is surgery via an external approach. The outcome of surgery for giant osteoma is good, with rare recurrence, no malignant transformation and few persistent symptoms.

[34]

TÍTULO / TITLE: - Unusual presentation of giant cell tumor originating from a facet joint of the thoracic spine in a child: a case report and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Siribumrungwong K; Tangtrakulwanich B; Nitiruangjaras A
INSTITUCIÓN / INSTITUTION: - Department of Orthopedic Surgery, Faculty of Medicine, Prince of Songkla University, Hat Yai, Songkla 90110, Thailand.
koopongs@hotmail.com.

RESUMEN / SUMMARY: - INTRODUCTION: Giant cell tumor of the synovium is a common benign lesion that frequently occurs at the tendon sheaths in the hand; it is usually found in adults over 30 years old. It is related to pigmented villonodular synovitis. Giant cell tumor of the synovium or pigmented villonodular synovitis has been described rarely in the axial skeleton especially in the thoracic vertebrae of a child. CASE PRESENTATION: A previously healthy 7-year-old Thai girl presented with back pain and progressive paraparesis and was unable to walk for 1 month. She had weakness and hyperreflexia of both lower extremities. Magnetic resonance imaging showed a well-defined homogeneously and intensely enhanced extradural mass with cord compression at T4 to T7 levels. The patient underwent laminectomy at T4 through to T7 and total tumor removal. Permanent histopathologic sections and immunostains revealed a giant cell tumor of the synovium. Postoperative neurological status recovered to grade V. Magnetic resonance imaging at the 1-year follow-up showed no recurrence and there was no clinical recurrence at the 2-year follow-up.
CONCLUSION: We report an extremely rare case of giant cell tumor in the epidural space that extended from a thoracic facet joint. The tumor was removed successfully through laminectomies. Although giant cell tumor of a facet joint of the thoracic spine is very rare, it must be considered in the differential diagnosis for masses occurring in the epidural space in a child. Total tumor removal is the best treatment. Careful monitoring of recurrence can achieve a good clinical outcome.

[35]

TÍTULO / TITLE: - Primary pulmonary rhabdomyosarcoma in children: Report of three cases with review of literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Enlace al texto completo: 4103/0971-5851.113423

AUTORES / AUTHORS: Lokesh KN; Premalata CS; Aruna Kumari BS; Appaji L

INSTITUCIÓN / INSTITUTION: Department of Medical Oncology, Kidwai Memorial Institute of Oncology, Bangalore, Karnataka, India.

RESUMEN / SUMMARY: Primary pulmonary rhabdomyosarcoma in children is rare. Three children aged three, nine and three years were evaluated for abnormal shadows on radiological examination with pneumothorax in two cases. Resection and histopathological examination revealed embryonal rhabdomyosarcoma in all and cystic malformation in first case. All the three children were treated with surgery and first two received adjuvant chemotherapy. The disease free duration was 160 months, 19 months and seven months respectively. The literature on primary pulmonary rhabdomyosarcoma in children was reviewed.

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


AUTORES / AUTHORS: Liu JJ; Liu S; Wang JG; Zhu W; Hua YQ; Sun W; Cai ZD

INSTITUCIÓN / INSTITUTION: Department of Orthopedic Surgery, Shanghai Tenth People’s Hospital, Tongji University School of Medicine, Shanghai, People’s Republic of China.

RESUMEN / SUMMARY: Telangiectatic osteosarcoma is a rare variant of osteosarcoma and hence its occurrence, presentation, and prognosis are poorly understood. With advancements in technology and available treatment options, the scenario of its diagnosis, management, and outcome has changed. Chemotherapy with surgery was challenged previously, but has now been proved to be beneficial. We reviewed the available literature and compared results to define the characteristics of the disease, its presentation, radiographic and pathologic features, optimal treatment, and prognosis.

Enlace al texto completo: 2147/OTT.S41351

AUTORES / AUTHORS: Cetin M; Cakici M; Ercisli M; Polat M

RESUMEN / SUMMARY: Unusually located left ventricular outflow myxoma: a brief review of the literature.

Among all myxomas, left ventricular outflow tract (LVOT) myxomas are very rare. This article reports an LVOT myxoma in a 67-year-old woman presenting with palpitations and weight loss. Surgical excision of the LVOT myxoma was performed.

[38] TÍTULO / TITLE: A large cellular angiofibroma of the male pelvis presenting with obstructive voiding: A case report and review of the literature.
RESUMEN / SUMMARY: Cellular angiofibromas (CAF) are rare, benign soft-tissue tumours. The diagnosis of CAF is important given the heavy resemblance to other tumours. Herein, we describe a case of a rapidly growing, very large (13.5 cm) CAF located in the deep pelvis of a middle-aged male who presented with difficulty voiding.

[39] TÍTULO / TITLE: Multifocal adult rhabdomyoma of the head and neck manifestation in 7 locations and review of the literature.
RESUMEN / SUMMARY: Background. Adult rhabdomyoma is a rare benign tumour with the differentiation of striated muscle tissue, which mainly occurs in the head and neck region. Twenty-six cases of multifocal adult rhabdomyoma are documented in the literature. Method. We report a 55-year-old male with simultaneous diagnosis of 7 adult rhabdomyomas and review the literature of multifocal adult rhabdomyoma. Result. Review of the literature revealed 26 cases of multifocal adult rhabdomyoma, of which only 7 presented with more than 2 lesions. Mean age at diagnosis was 65 years with a male to female ratio of 5.5 : 1. Common localizations were the parapharyngeal space (36%), larynx (15%), submandibular (14%), paratracheal region (12%), tongue (11%), and floor of mouth (9%). Besides the known radiological features of adult rhabdomyoma, our case showed FDG-uptake in (18) F-FDG PET/CT. Conclusion.
This is the first case of multifocal adult rhabdomyoma published, with as many as 7 simultaneous adult rhabdomyomas of the head and neck.

[40]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
   ●● Enlace al texto completo (gratuito o de pago) 12659/AJCR.883928
AUTORES / AUTHORS: - Tsalis K; Antoniou N; Kalfadis S; Dimoulas A; Dagdilelis AK; Lazaridis C
INSTITUCIÓN / INSTITUTION: - D’ Surgical Department “G Papanikolaou” Hospital, Aristotle University of Thessaloniki, Thessaloniki, Greece.
RESUMEN / SUMMARY: - Patient: Female, 40 Final Diagnosis: Esophageal lipoma
Symptoms: - Medication: - Clinical Procedure: Laparoscopic enucleation Specialty: Surgery Objective: Rare disease. BACKGROUND: Benign tumors of the esophagus are very rare, constituting only 0.5% to 0.8% of all esophageal neoplasms. Approximately 60% of benign esophageal neoplasms are leiomyomas, 20% are cysts, 5% are polyps, and less than 1% are lipomas. CASE REPORT: A 40-year-old woman was referred to our department with dysphagia that had progressively worsened during the previous 2 years. Physical examination on admission produced normal findings. Upper gastrointestinal endoscopy revealed a submucosal space-occupying mass in the posterior wall of the lower esophagus, with normal mucosa. The mass was yellowish and soft. A computed tomography (CT) of the chest revealed a submucosal esophageal lesion in the posterior wall, with luminal narrowing of the distal esophagus. Thus, a submucosal tumor was identified in this region and esophageal submucosal lipoma was considered the most likely diagnosis. A laparoscopic operation was performed. The tumor was completely enucleated, and measured 10x7x2.5 cm. The pathology showed lipoma. The postoperative course was uneventful, and the patient was discharged 4 days after the operation. CONCLUSIONS: Benign tumors of the esophagus are very rare. Laparoscopic transhiatal enucleation of lower esophageal lipomas and other benign tumors is a safe and effective operation.

[41]
TÍTULO / TITLE: - Liposarcoma of the retropharyngeal space with rapidly worsening dyspnea: 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.2013.1310
AUTORES / AUTHORS: - He JG; Jiang H; Yang BB; Lin PF
INSTITUCIÓN / INSTITUTION: - Department of Otolaryngology, Second Affiliated Hospital, School of Medicine, Zhejiang University, Hangzhou, Zhejiang 310009, P.R. China.
RESUMEN / SUMMARY: - Liposarcomas represent a significant proportion of soft-tissue sarcomas. However, their occurrence in the head and neck is infrequent and they are
exceedingly rare in the retropharyngeal space. The present study reports the case of a 58-year-old patient with retropharyngeal liposarcoma. Uniquely, the patient presented with rapidly worsening dyspnea. The diagnosis of liposarcoma was established following retropharyngeal tumor excision, although biopsies were performed twice. Adjuvant radiotherapy was refused by the patient. However, during the post-operative follow-up period, no sign of either local tumor recurrence or distant metastasis was observed. Previously reported cases were also reviewed to analyze the diagnosis, treatment and prognosis of this disease.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Xiang L; Zhou J; Gu W; Yang B
INSTITUCIÓN / INSTITUTION: Department of Hematology, The First People’s Hospital of Changzhou, Third Affiliated Hospital of Suzhou University, Changzhou, Jiangsu 213003;
RESUMEN / SUMMARY: Angiomatoid fibrous histiocytoma (AFH) is a rare, low-grade malignant potential soft tissue tumor which occurs most commonly in children and young adults. Only a few case reports have been described that typically occur in the extremities of the deep dermis and subcutaneous tissue, followed by the trunk, as well as the head and neck. A case report of retroperitoneal AFH is described. This presentation for patients with AFH has not yet been reported. AFH may occur in the retroperitoneum, in the future patients with retroperitoneal tumor should be considered the possibility of having AFH.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Chavez C; Hoffman MA
INSTITUCIÓN / INSTITUTION: Division of Hematology-Oncology, Department of Medicine, Long-Island Jewish Medical Center, North Shore-LIJ Health System, New Hyde Park, NY 11040, USA.
RESUMEN / SUMMARY: We report a case in which a 52-year-old female developed a multifocal inflammatory myofibroblastic tumor (IMT) of the lung. The tumor did not overexpress the anaplastic lymphoma kinase (ALK) protein, indicating a lack of ALK rearrangement. The patient required two wedge resections in 15 months due to recurrent disease. Recurrence after the second surgery was treated with
corticosteroids, which only led to a transient response (6 months). Introduction of celecoxib, a cyclooxygenase-2 inhibitor, induced a complete remission in the patient. Maintenance on celecoxib further led to a progression-free survival of 34 months. A literature review retrieved a total of eight case reports, comprising ten patients, of IMT of various anatomical sites successfully treated with non-steroidal anti-inflammatory agent (NSAID) therapy. Nine of the ten patients achieved durable complete remission. Remission occurred rapidly and persisted even after termination of NSAID therapy. Although such a successful outcome may only be achieved rarely, a trial of an NSAID should be considered in any patient in whom complete resection is not an option. Our case also demonstrates that NSAID therapy may be successful in a non-ALK rearranged tumor in which ALK inhibition is not an option.

[44]
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Ohta K; Taki M; Ogawa I; Ono S; Mizuta K; Fujimoto S; Takata T; Kamata N
INSTITUCIÓN / INSTITUTION: - Department of Oral and Maxillofacial Surgery, Institute of Biomedical and Health Sciences, 1-2-3 Kasumi, Minami-Ku, Hiroshima 734-8553, Japan. otkouji@hiroshima-u.ac.jp
RESUMEN / SUMMARY: - Ossifying fibromyxoid tumor (OFMT) is a rare mesenchymal neoplasm that arises in subcutaneous tissue, with that in the oral cavity extremely rare. We present a case of malignant OFMT in the tongue. A 26-year-old male noticed a painless mass in the tongue, which was extracted at a general hospital. Four years later, the tumor recurred and was resected at our department. Histologically, the recurrent tumor was composed of the closely packed cells positive for vimentin and S-100 proliferating in a nodular fashion. It showed high cellularity and mitotic activity. In the primary tumor, some tumor cells were arranged in a diffuse or cord-like manner within an abundant fibromyxoid matrix, along with a small amount of metaplastic ossification, corresponding with the histopathological characteristic of OFMT. Accordingly, a diagnosis of malignant OFMT arising in typical OFMT was established. This is the first reported case of malignant OFMT in the tongue. Long-term follow-up is needed for confirmation of prognosis and biological behavior.

[45]
TÍTULO / TITLE: - Intrascrotal lipoblastoma in a ten year old: case report and review of literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
Lipoblastoma is a rare benign soft tissue tumor encountered almost exclusively in infancy and early childhood. The location of tumors varies, but most occur in the extremities, trunk, head and neck. Less frequently, lipoblastomas have been reported in the mediastinum, the retroperitoneum and the inguinal region. Only 7 cases of lipoblastoma in the scrotum have been reported so far in the English literature, with none of the patients older than 8. We report an intrascrotal lipoblastoma in a 10 year-old boy. The differential diagnosis is discussed with reference to the literature.

[46]

Angiosarcoma of the spleen presenting as spontaneous splenic rupture: A rare case report and review of the literature.

INTRODUCTION: Angiosarcoma of the spleen is a rare malignancy of vascular origin with a high rate of metastasis and poor prognosis. We report one such rare case of spontaneous splenic rupture, along with a review of current literature. PRESENTATION OF CASE: A 30 year old man presented to our emergency services with severe abdominal pain, distension, hypotension and splenomegaly. Investigations revealed a marked anaemia, coagulopathy, severe lactic acidosis, and acute kidney injury. Imaging demonstrated splenomegaly with acute haemorrhage and lymphadenopathy. Laparotomy and splenectomy revealed piecemeal spleen and nodular omentum. The patient suffered an intra-operative cardiorespiratory arrest, and despite successful resuscitation, fatally arrested postoperatively in ICU. Histology revealed a primary splenic angiosarcoma with omental metastases. DISCUSSION: Primary splenic angiosarcoma was first reported in 1879, with only 200 cases reported to date, largely as isolated case reports, with an annual incidence of 0.14-0.25 per million. With variable symptomatology and a potential to present with life-threatening complications, early diagnosis is paramount. CT scanning shows distinctive changes and is invaluable in disease assessment. Tissue diagnosis is often possible only after splenectomy. Spontaneous rupture carries the worst prognosis. CONCLUSION: Primary splenic angiosarcoma is a rare and aggressive malignancy that often presents with metastatic disease, and largely carries
a dismal prognosis. Definitive diagnosis is challenging, but imaging with CT scanning can show characteristic changes and establish any metastatic disease. With no established adjuvant therapy long term outlook remains poor even if treated successfully by surgery.

[47]
TÍTULO / TITLE: - Ewing’s sarcoma/primitive neuroectodermal tumour of the prostate: A case report and literature review.
RESUMEN / SUMMARY: -  
AUTORES / AUTHORS: - Wu T; Jin T; Luo D; Chen L; Li X
INSTITUCIÓN / INSTITUTION: - Department of Urology, West China Hospital, Sichuan University, Chengdu, China.
RESUMEN / SUMMARY: - We present a case of Ewing’s sarcoma and primitive neuroectodermal tumour (PNET) of the prostate. A 29-year-old male presented with difficult defecation and anus distention; on magnetic resonance imaging scan of the pelvis, we found a prostate tumour. A transrectal ultrasound-guided needle biopsy confirmed the diagnosis. The patient underwent cystoprostatectomy and replacement ileocystoplasty and was followed by multi-agent chemotherapy. PNET/ Ewing’s sarcoma of the prostate is extremely rare. The prognosis is very poor, so we should pay enough attention to the differential diagnosis and treatment.

[48]
TÍTULO / TITLE: - Solitary myofibroma of the sigmoid colon: case report and review of the literature.
RESUMEN / SUMMARY: - 
AUTORES / AUTHORS: - Kim MJ; Lee SH; Youk EG; Lee S; Choi JH; Cho KJ
INSTITUCIÓN / INSTITUTION: - Departments of Pathology, Daehang Hospital, Seoul, Korea.
RESUMEN / SUMMARY: - A 58-year-old woman presented with a solitary myofibroma that arose in the sigmoid colon. Computed tomography revealed a highly enhanced intramural mass (1.3-cm maximum diameter) in the proximal sigmoid colon. Histologically, the tumor exhibited a biphasic growth pattern, which comprised haphazardly arranged, interwoven fascicles of plump, myoid-appearing spindle cells with elongated nuclei and abundant eosinophilic cytoplasm, and more cellular areas of primitive-appearing polygonal cells that were arranged in a hemangiopericytomatous pattern. The tumor cells were positive for smooth muscle actin (SMA), and negative for desmin, h-caldesmon, CD34, cytokeratin, S100 protein, and CD117. The Ki-67 labeling index was not high (up to 7%). Based on these histologic and immunohistochemical features, our patient was diagnosed with a myofibroma of the sigmoid colon. The
presence of solitary myofibroma in the intestine of an adult requires attention to avoid misdiagnosis as a more aggressive mesenchymal tumor. VIRTUAL SLIDES: The virtual slide(s) for this article can be found here: http://www.diagnosticpathology.diagnomx.eu/vs/2096403796957687.

[49] TÍTULO / TITLE: - Primary renal angiosarcoma: Case report and literature review.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Chaabouni A; Rebai N; Chabchoub K; Fourati M; Bouacida M; Slimen MH; Bahloul A; Mhiri MN
INSTITUCIÓN / INSTITUTION: - Department of Urology, Habib Bourguiba University Hospital, Tunisia.
RESUMEN / SUMMARY: - Angiosarcoma is a rare malignant tumour occurring in less than 2% of soft tissue sarcomas. Angiosarcoma involving the kidney usually represents metastasis from skin or visceral primary lesions, while angiosarcoma primarily occurring in the kidney is a very rare neoplasm. We report a case of angiosarcoma of the right kidney in a 59-year-old male. The computed tomography scan showed a solid tumour with a low increased density after administration of contrast medium. Histological examination of the piece of nephrectomy confirmed the diagnosis.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - An SY; Shin HI; Choi KS; Park JW; Kim YG; Benavides E; Kim JW; An CH
INSTITUCIÓN / INSTITUTION: - Assistant Professor, Department of Oral and Maxillofacial Radiology, School of Dentistry, Kyungpook National University, Daegu, South Korea.
RESUMEN / SUMMARY: - A 10-year-old boy presented with a swelling on the right side of the jaw. He had undergone excision of the lesion about 10 months ago at a private dental clinic and the swelling began to regrow 4 months after surgery. A panoramic radiograph revealed 4 sclerotic round masses with radiolucent rims surrounded by sclerosis of the right posterior mandible. Computed tomography scan showed 4 round bony masses centered on the buccal cortex and bone marrow space, sclerosis of the adjacent bone and periosteal reaction. He underwent a marginal resection under general anesthesia and the final histopathological report confirmed the diagnosis of osteoid osteoma. Postoperative course was uneventful, and there was no evidence of
recurrence at the 5.5-year follow-up. In the review of the literature, 20 osteoid ostemas were found in the jaw and to the best of our knowledge, the present case is the only one showing multifocal nidi.

[51]
TÍTULO / TITLE: - Low-grade central osteosarcoma of the rib: a case report and brief review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o pago) 1155/2013/798435
AUTORES / AUTHORS: - Moghadamfalahi M; Alatassi H
INSTITUCIÓN / INSTITUTION: - Department of Pathology and Laboratory Medicine, 503 South Jackson Street, Louisville, KY 40202, USA.
RESUMEN / SUMMARY: - Low-grade central osteosarcoma is a rare variant of osteosarcoma which comprises less than 1-2% of all osteosarcomas. Most low-grade osteosarcomas involve long bones, most commonly distal femur, and proximal tibia. Histologically this tumor is difficult to diagnose, and an unusual location makes this diagnosis even more challenging. Here we report a case of low-grade osteosarcoma presenting as a chest wall mass involving the left 6th-8th ribs. This unusual site of presentation significantly added to the diagnostic difficulties of this rare tumor with challenging histologic features. To the best of our knowledge, only six cases of low-grade central osteosarcoma of the ribs have been reported in the English literature.

[52]
TÍTULO / TITLE: - Fibrosarcomatous pigmented dermatofibrosarcoma protuberans: A case report with review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
●● Enlace al texto completo (gratuito o pago) 3892/ol.2012.765
AUTORES / AUTHORS: - Ishida M; Okabe H
INSTITUCIÓN / INSTITUTION: - Department of Clinical Laboratory Medicine and Division of Diagnostic Pathology, Shiga University of Medical Science, Otsu, Shiga 520-2192, Japan.
RESUMEN / SUMMARY: - Dermatofibrosarcoma protuberans (DFSP) is a relatively rare low-grade sarcoma that occasionally exhibits melanin-containing spindle cells within the tumor. Fibrosarcomatous DFSP (FS-DFSP) is a variant that is characterized by areas which are histopathologically indistinguishable from fibrosarcoma. In the present study, we describe a case of fibrosarcomatous pigmented DFSP and review the clinicopathological features of this extremely rare lesion. A 51-year-old male presented with a slow-growing nodular cutaneous mass in his left upper arm. Histopathologically, the resected tumor was comprised of pigmented DFSP in approximately 20% of the tumor, with the remaining area further covered by a fibrosarcomatous component. A review of the clinicopathological features of the five previously reported cases as well
as the present case indicated that this lesion mainly affects middle-aged males and occurs mostly in the extremities. Melanin-containing spindle cells are present only in the conventional DFSP component. The prognosis appears to be poor; in the six cases reviewed, four demonstrated multiple metastases and three succumbed to the disease. Our analyses revealed that the presence of a fibrosarcomatous component in pigmented DFSP is associated with aggressive behavior; therefore, careful assessment for the presence of a fibrosarcomatous component is necessary in the diagnosis of this disease.

[53]


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


AUTORES / AUTHORS: - Xiao J; Selvaggi SM; Leith CP; Fitzgerald SA; Stewart J 3rd

INSTITUCIÓN / INSTITUTION: - Department of Pathology and Laboratory Medicine, University of Wisconsin School of Medicine and Public Health, Madison, Wisconsin.

RESUMEN / SUMMARY: - BACKGROUND: Primary effusion lymphoma (PEL) is a rare subtype of large B-cell lymphoma that arises in body cavities without detectable tumor masses. PEL is universally associated with Kaposi sarcoma herpesvirus (KSHV)/human herpesvirus-8 (HHV8). Despite overlapping features, KSHV/HHV8-negative effusion-based lymphoma is a distinct entity from PEL. To date, 52 cases have been reported. The authors report 3 additional cases received in their laboratory from 2007 to 2012. METHODS: Clinical data, cytomorphologic features, and immunophenotypic features of the 3 cases were described and compared with those reported in the literature. RESULTS: The cells in HHV8-negative effusion lymphoma commonly revealed large cell, immunoblastic morphology and B-cell immunophenotype. The 3 cases demonstrated cytomorphologic and immunophenotypic variability. Cytomorphologically, 1 case contained large, highly atypical cells with a moderate amount of cytoplasm, round nucleus, coarsely granular chromatin, and a single macronucleolus. The other 2 cases had medium to large atypical cells with high nuclear-to-cytoplasmic ratios, slightly irregular to cleaved nuclei, and multiple conspicuous nucleoli. One case had a null phenotype with aberrant cytokeratin expression. B-cell phenotype was established by clonal immunoglobulin heavy-chain rearrangement using polymerase chain reaction, whereas the other 2 cases demonstrated a B-cell phenotype by flow cytometry and immunohistochemical staining. All 3 cases were negative for both HHV8 and Epstein-Barr virus. CONCLUSIONS: HHV8-negative effusion lymphoma exhibits clinical, cytomorphologic, and immunophenotypic variability. Cases with a null-phenotype can be particularly challenging. When effusion lymphoma is suspected, ancillary tests are helpful. Moreover, HHV8 detection is critical in differentiating PEL and HHV8-negative effusion lymphoma, because they have overlapping features yet different prognoses. Cancer (Cancer Cytopathol) 2013. © 2013 American Cancer Society.
TÍTULO / TITLE: - Vulvar “proximal-type” epithelioid sarcoma: report of a case and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Patrizi L; Corrado G; Saltari M; Perracchio L; Scelzo C; Piccione E; Vizza E

RESUMEN / SUMMARY: - BACKGROUND: The “proximal-type” epithelioid sarcoma is a very rare kind of mesenchimal tumor characterized by the difficulty in histological diagnosis and the very aggressive biological behavior. Case We report of a case of a 63 years old woman with a vulvar “proximal-type” epithelioid sarcoma that underwent a radical surgical staging followed by an adjuvant radiotherapy. She is on follow-up care for 14 months and there is no clinical evidence of disease. CONCLUSION: Even if quite rare the proximal type epithelioid sarcoma should be regarded as a separate entity of particularly aggressive biologic behaviour. Its diagnosis attracts controversies and criticism related to the surgical approach and the choice of an adjuvant therapy.

Virtual slides The virtual slides for this article can be found here: http://www.diagnosticpathology.diagnomx.eu/vs/1508554852942125.

TÍTULO / TITLE: - Solitary intestinal neurofibroma with no associated systemic syndromes causing intussusception: Case report and literature review.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Al-Harake A; Chour M; Al Beteddini OS
INSTITUCIÓN / INSTITUTION: - Department of Surgery, Al Rassoul Al Aazam Hospital, Beirut, Lebanon.

RESUMEN / SUMMARY: - INTRODUCTION: The isolated presence of neurofibromatous lesions in the gastrointestinal tract, with no associated systemic syndromes, is a rarely reported clinical entity. PRESENTATION OF CASE: A 48-year-old lady, with no history of neurofibromatosis or other systemic disease, presented with small bowel obstruction secondary to an ileo-ileal intussusception induced by an isolated ileal neurofibromatous mass. The patient underwent a segmental enterectomy and after a smooth recovery, she was put on a long-term follow-up schedule. DISCUSSION: This article presents a review of the literature of this area clinical entity. Very few reports of gastrointestinal isolated neurofibromas could be found. Similarly, extra-digestive isolated lesions have been rarely reported. CONCLUSION: Isolated ileal neurofibroma is a rare pathological entity. The clinical significance of such a diagnosis lies mainly in the need of further follow up of these patients as the bowel involvement could be the first manifestation of neurofibromatosis type 1 or multiple endocrine neoplasia type 2b.
[56] Título / Title: Primary Ewing sarcoma of the kidney: a symptomatic presentation and review of the literature.
Resumen / Summary: Enlace al Resumen / Link to its Summary

- Enlace al texto completo (gratuito o de pago) 1756287212471095 [pii]
- Enlace al texto completo (gratuito o de pago) 1756287212471095

Autores / Authors: Hakky TS; Gonzalvo AA; Lockhart JL; Rodriguez AR
Institución / Institution: Department of Urology, University of South Florida, Tampa, FL, USA.

Resumen / Summary: The objective of this review is to discuss the unique nature of primary renal Ewing sarcoma, including incidence, presentation and management. We also report on a common pattern of presentation, consisting of acute flank pain mimicking a renal stone colic, with or without hydronephrosis, and a renal mass discovered during imaging studies of renal Ewing sarcoma. We present our case of renal Ewing sarcoma along with imaging and pathological analysis. We also performed a retrospective review of all cases of renal Ewing sarcoma using PubMed. A total of 48 cases of renal EWS sarcoma have been reported and analyzed in this review. A mean age of 30.4 years was found along with a 61% male predominance. The mean survival was 26.14 months with a lower median survival in patients with advanced metastatic disease. Primary Ewing sarcoma of the kidney is rare. The diagnosis of primary renal EWS can be difficult and is based on a combination of electron microscopy, immunohistochemistry, chromosomal analysis, fluorescence in situ hybridization (FISH) and light microscopy.

[57] Título / Title: Brief S2k guidelines—Dermatofibrosarcoma protuberans.
Resumen / Summary: Enlace al Resumen / Link to its Summary

- Enlace al texto completo (gratuito o de pago) 1111/ddg.12015_4

Autores / Authors: Ugurel S; Kortmann RD; Mohr P; Mentzel T; Garbe C; Breuninger H
Institución / Institution: Department of Dermatology, Graz University Hospital, Graz, Austria. selma.ugurel@medunigraz.at

[58] Título / Title: Brief S1 guidelines—Cutaneous angiosarcoma and Kaposi sarcoma.
Resumen / Summary: Enlace al Resumen / Link to its Summary

- Enlace al texto completo (gratuito o de pago) 1111/ddg.12015_2
**AUTORES / AUTHORS:** - Vogt T; Brockmeyer N; Kutzner H; Schofer H

**INSTITUCIÓN / INSTITUTION:** - Department of Dermatology, Saarland University Hospital, Homburg/Saar, Germany. thomas.vogt@uks.eu

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**TÍTULO / TITLE:** - Bilateral ewing sarcoma/primitive neuroectodermal tumor of the breast: a very rare entity and review of the literature.

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


**AUTORES / AUTHORS:** - Majid N; Amrani M; Ghissassi I; El Cadi M; El Bouzidi M; El Kabous M; Kherbach A; Errihani H

**INSTITUCIÓN / INSTITUTION:** - Department of Medical Oncology, National Institute of Oncology, Rabat, Morocco.

**RESUMEN / SUMMARY:** - Peripheral primitive neuroectodermal tumors (PNET) are rare malignant tumors, affecting mostly children and adolescents and have been described in breast in eight case reports only. In this paper, we present a case of bilateral mammary ES/PNET where distinction between primary and metastatic diseases was discussed through a literature review. The aim of this work is to demonstrate that although rare, the possibility of PNET should be kept in mind while evaluating a palpable breast abnormality in a young female.

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**TÍTULO / TITLE:** - A gist of gastrointestinal stromal tumors: A review.

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


**AUTORES / AUTHORS:** - Rammohan A; Sathyanesan J; Rajendran K; Pitchaimuthu A; Perumal SK; Srinivasan U; Ramasamy R; Palaniappan R; Govindan M

**INSTITUCIÓN / INSTITUTION:** - Ashwin Rammohan, Jeswanth Sathyanesan, Kamalakannan Rajendran, Anbalagan Pitchaimuthu, Senthil-Kumar Perumal, UP Srinivasan, Ravi Ramasamy, Ravichandran Palaniappan, Manoharan Govindan, The Institute of Surgical Gastroenterology and Liver Transplantation, Centre for GI Bleed, Division of HPB Diseases, Stanley Medical College Hospital, Chennai 600001, India.

**RESUMEN / SUMMARY:** - Gastrointestinal stromal tumors (GISTs) have been recognized as a biologically distinctive tumor type, different from smooth muscle and neural tumors of the gastrointestinal tract (GIT). They constitute the majority of gastrointestinal mesenchymal tumors of the GIT and are known to be refractory to conventional chemotherapy or radiation. They are defined and diagnosed by the expression of a proto-oncogene protein detected by immunohistochemistry which serves as a crucial diagnostic and therapeutic target. The identification of these mutations has resulted in a better understanding of their oncogenic mechanisms. The
remarkable antitumor effects of the molecular inhibitor imatinib have necessitated accurate diagnosis of GIST and their distinction from other gastrointestinal mesenchymal tumors. Both traditional and minimally invasive surgery are used to remove these tumors with minimal morbidity and excellent perioperative outcomes. The revolutionary use of specific, molecularly-targeted therapies, such as imatinib mesylate, reduces the frequency of disease recurrence when used as an adjuvant following complete resection. Neoadjuvant treatment with these agents appears to stabilize disease in the majority of patients and may reduce the extent of surgical resection required for subsequent complete tumor removal. The important interplay between the molecular genetics of GIST and responses to targeted therapeutics serves as a model for the study of targeted therapies in other solid tumors. This review summarizes our current knowledge and recent advances regarding the histogenesis, pathology, molecular biology, the basis for the novel targeted cancer therapy and current evidence based management of these unique tumors.

[61]

TÍTULO / TITLE: - Low-grade myofibroblastic sarcoma of the larynx: a rare entity with review of literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Khosla D; Yadav BS; Kumar R; Ghoshal S; Vaiphei K; Verma R; Sharma SC
INSTITUCIÓN / INSTITUTION: - Department of Radiotherapy and Oncology, Regional Cancer Centre, Chandigarh, India.
RESUMEN / SUMMARY: - Mesenchymal neoplasms of the larynx are rare and make up approximately 0.3% to 1.0% of all malignancies at this location. Low grade myofibroblastic sarcoma (LGMS) of larynx is a rare entity. We describe a rare case of LGMS of larynx who presented with complaint of hoarseness of voice. The patient was treated with total laryngectomy plus partial pharyngectomy followed by post-operative radiotherapy. Histopathologically, the lesion was composed of spindle cells that manifested variable cellular anaplasia and expressed smooth muscle actin (SMA) and focally S-100. One of the resection limits was involved so patient was given post-operative radiotherapy. The patient is alive and disease free 14 months after surgery. The characteristic clinical, histopathological features and treatment of this case are described with a literature review.

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