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newly diagnosed glioblastoma is based on radiotherapy combined with temozolomide. This approach doubles the 2-year survival rate to 27%, but overall prognosis remains poor. Bevacizumab is an emerging treatment alternative that deserves further study. Grade III tumors have been less well studied, and clinical trials to establish standards of care are ongoing. Patients with malignant gliomas experience frequent clinical complications, including thromboembolic events, seizures, fluctuations in neurologic symptoms, and adverse effects from corticosteroids and chemotherapies that require proper management and prophylaxis. CONCLUSIONS AND RELEVANCE: Glioblastoma remains a difficult cancer to treat, although therapeutic options have been improving. Optimal management requires a multidisciplinary approach and knowledge of potential complications from both the disease and its treatment.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Poon MT; Fung LH; Pu JK; Leung GK
INSTITUCIÓN / INSTITUTION: - Division of Neurosurgery, Department of Surgery, Li Ka Shing Faculty of Medicine, The University of Hong Kong, Queen Mary Hospital, Hong Kong, P. R. China.
RESUMEN / SUMMARY: - Background. Intracranial meningioma is a common condition in the elderly population. Surgical resection in this group of patients may be rendered more hazardous due to the patients’ ageing physiology and to multiple comorbidities. This systematic review and meta-analysis aimed to summarise outcome data of elderly patients undergoing intracranial meningioma resection. Methods. Using Ovid Medline, longitudinal studies published from 2002 to October 2012 with patients aged >/= 65 years that described outcomes after intracranial meningioma resection were reviewed. Outcome data included mortality, recurrence, complication rate and length of hospital stay (LoS). Grading score systems and covariates for predicting outcome were collected. Pooled estimates of mortality data were calculated in StatsDirect using a random effects method. I2 statistic was used to assess heterogeneity. Results. Thirteen eligible studies with a total of 7010 patients (mean age, 73.6 years) were included, in which 82% patients came from one study. The pooled estimates of 90-day and 1-year mortality from available data were 6.6% (95% confidence interval [CI], 4.6-9.1%; n = 735; I2 = 32.1) and 9.6% (95% CI, 7.0-12.6%; n = 564; I2 = 24.3), respectively. The overall complication rates ranged from 2.7% to 29.8%, and the overall incidence of complications was 20% per patient (range, 3-61%). Other outcome data were heterogeneous mainly due to incomparable study designs. Conclusions. Current evidence indicates satisfactory surgical outcomes in the elderly with intracranial meningiomas, though the risks of complications necessitate careful consideration when deciding to operate. Risk factor analysis emphasised the importance of considering pre-operative status and comorbidities during patient selection. Future research should address the causes and prevention of complications, and compare outcomes between younger and older patients using detailed stratifications of tumour characteristics.
WITHDRAWN: Comparative efficacy of epidural, subarachnoid, and intracerebroventricular opioids in patients with pain due to cancer.

BACKGROUND: Since the 1970s, when endogenous opioids and opioid receptors were first isolated in the central nervous system, attempts have been made to optimize opioid therapy by delivering the medication centrally rather than systemically. Although the vast majority of cancer patients obtain satisfactory pain relief from individualized systemic treatment, there remain the few whose pain is refractory to systemic treatments. These patients may obtain relief from neuraxial opioid therapy: intracerebroventricular, epidural or subarachnoid.

OBJECTIVES: To compare intracerebroventricular therapy with other neuraxial treatments and to determine whether intracerebroventricular (ICV) has anything to offer over epidural (EPI) and subarachnoid (SA) catheters in terms of efficacy, adverse effects, and complications.

SEARCH METHODS: A number of electronic databases were searched to retrieve information for inclusion in this review up to January 2003. Non-English language reports are awaiting assessment. Unpublished data were not sought.

SELECTION CRITERIA: Randomised studies of intracerebroventricular therapy for patients with intractable cancer pain were sought. However, this level of evidence was not available so data from uncontrolled trials, retrospective case series and uncontrolled prospective cohort studies were assessed.

DATA COLLECTION AND ANALYSIS: Our search did not retrieve any controlled trials. We therefore used data from uncontrolled studies to compare incidences of analgesic efficacy, adverse effects, and complications. We found 72 uncontrolled trials assessing ICV (13 trials, 337 participants), EPI (31 trials, 1343 participants), and SA (28 trials, 722 participants) in cancer patients. From these we extracted data on analgesic efficacy, common pharmacologic adverse effects, and complications.

MAIN RESULTS: Data from uncontrolled studies reported excellent pain relief among 73% of ICV patients compared with 72% EPI and 62% SA. Unsatisfactory pain relief was low in all treatment groups. Persistent nausea, persistent and transient urinary retention, transient pruritus, and constipation occurred more frequently with EPI and SA. Respiratory depression, sedation and confusion were most common with ICV. The incidence of major infection when pumps were used with EPI and SA was zero. There was a lower incidence of other complications with ICV therapy than with EPI or SA.

AUTHORS’ CONCLUSIONS: Neuraxial opioid therapy is often effective for treating cancer pain that has not been adequately controlled by systemic treatment. However, long-term use of neuraxial therapy can be complicated by problems associated with the catheters. The data from uncontrolled studies suggests that ICV is
at least as effective against pain as other neuraxial treatments and may be a successful treatment for patients whose cancer pain is resistant to other treatments.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Rocha FC; Dos Santos Junior JG; Stefano SC; da Silveira DX
INSTITUCIÓN / INSTITUTION: - Paulista School of Medicine, Federal University of Sao Paulo, Av. Ascendino Reis, 763, Vila Clementino, Sao Paulo, SP, 04027-000, Brazil, franciscocmrocha@uol.com.br.
RESUMEN / SUMMARY: - To evaluate, through a systematic review of the literature, the antitumoral effects of cannabinoids on gliomas. Research included the following electronic databases: PUBMED, EMBASE, LILACS and The Cochrane Collaboration Controlled Trials Register. All published studies involving the antitumoral effects (cellular and molecular mechanisms) of cannabinoids were considered for this review. The bibliography search strategy included all publications of each of these databases until December 31, 2012. From 2,260 initially identified articles, 35 fulfilled the inclusion criteria for this review. All the studies included in this systematic review were experimental (in vivo and/or in vitro), except for one pilot clinical trial phase I/II involving humans. In all experimental studies included, cannabinoids exerted antitumoral activity in vitro and/or antitumoral evidence in vivo in several models of tumor cells and tumors. The antitumor activity included: antiproliferative effects (cell cycle arrest), decreased viability and cell death by toxicity, apoptosis, necrosis, autophagy, as well as antiangiogenic and antimigratory effects. Antitumoral evidence included: reduction in tumor size, antiangiogenic, and antimetastatic effects. Additionally, most of the studies described that the cannabinoids exercised selective antitumoral action in several distinct tumor models. Thereby, normal cells used as controls were not affected. The safety factor in the cannabinoids’ administration has also been demonstrated in vivo. The various cannabinoids tested in multiple tumor models showed antitumoral effects both in vitro and in vivo. These findings indicate that cannabinoids are promising compounds for the treatment of gliomas.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - van Hulsteijn LT; Niemeijer ND; Dekkers OM; Corssmit EP
INSTITUCIÓN / INSTITUTION: - Department of Endocrinology and Metabolic Diseases, Leiden University Medical Center, Leiden, The Netherlands.
RESUMEN / SUMMARY: - BACKGROUND: 131 I-MIBG therapy can be used for palliative treatment of malignant paraganglioma and pheochromocytoma. The main objective of this study was to perform a systematic review and meta-analysis assessing
the effect of 131 I-MIBG therapy on tumour volume in patients with malignant paraganglioma/pheochromocytoma. METHODS: A literature search was performed in December 2012 to identify potentially relevant studies. Main outcomes were the pooled proportions of complete response, partial response and stable disease after radionuclide therapy. A meta-analysis was performed with an exact likelihood approach using a logistic regression with a random effect at the study level. Pooled proportions with 95% confidence intervals (CI) were reported. RESULTS: Seventeen studies concerning a total of 243 patients with malignant paraganglioma/pheochromocytoma were treated with 131 I-MIBG therapy. The mean follow-up ranged from 24 to 62 months. A meta-analysis of the effect of 131 I-MIBG therapy on tumour volume showed pooled proportions of complete response, partial response and stable disease of, respectively, 0.03 (95% CI: 0.06-0.15), 0.27 (95% CI: 0.19-0.37) and 0.52 (95% CI: 0.41-0.62) and for hormonal response 0.11 (95% CI: 0.05-0.22), 0.40 (95% CI: 0.28-0.53) and 0.21 (95% CI: 0.10-0.40), respectively. Separate analyses resulted in better results in hormonal response for patients with paraganglioma than for patients with pheochromocytoma. CONCLUSIONS: Data on the effects of 131 I-MIBG therapy on malignant paraganglioma/pheochromocytoma suggest that stable disease concerning tumour volume and a partial hormonal response can be achieved in over 50% and 40% of patients, respectively, treated with 131 I-MIBG therapy. It cannot be ruled out that stable disease reflects not only the effect of MIBG therapy, but also (partly) the natural course of the disease.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Yin AA; Cai S; Dong Y; Zhang LH; Liu BL; Cheng JX; Zhang X
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Xijing Institute of Clinical Neuroscience, Xijing Hospital, Fourth Military Medical University, Changle West Road, No. 127, Xi’an, 710032, Shaanxi, People’s Republic of China.
RESUMEN / SUMMARY: - Temozolomide (TMZ) alone has been proposed as a promising alternative to radiotherapy (RT) in elderly glioblastoma (GBM) patients. We report a meta-analysis to systematically evaluate TMZ monotherapy in older GBM patients. A systematic literature search was performed using PubMed, EMBASE and the Cochrane database. Studies comparing TMZ versus RT in elderly patients (≥65 years) with newly diagnosed GBM were eligible for inclusion. Two randomized clinical trials (RCTs) and three comparative studies were included in the analyses, which revealed an overall survival (OS) advantage for TMZ compared with RT (HR [hazard ratio] 0.86, 95% CI [confidence interval] 0.74-1.00). However, a sensitivity analysis of 2 RCTs only supported its non-inferiority (HR 0.91, 95% CI 0.66-1.27). Most elderly patients tolerated TMZ despite an increased risk of grade 3-4 (G3-4) toxicities, especially hematological toxicities. The quality of life was similar between the groups. In the MGMT analysis, methylated tumors were associated with a longer OS than unmethylated tumors among elderly patients receiving TMZ monotherapy (HR 0.50, 95
% CI 0.35-0.70). Moreover, in patients with methylated tumors, TMZ was more beneficial than RT alone in improving OS (TMZ vs. RT: HR 0.66, 95 % CI 0.47-0.93) whereas the opposite was true for those with unmethylated tumors (HR 1.32, 95 % CI 1.00-1.76). Although the meta-analysis demonstrated the non-inferiority of RT in improving OS, TMZ alone was not a straightforward solution for elderly GBM patients because of an increased risk of G3-4 toxicities, especially hematological toxicities. MGMT testing might be helpful for determining individualized treatment.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Simonetti G; Trevisan E; Silvani A; Gaviani P; Botturi A; Lamperti E; Beecher D; Bertero L; Bosa C; Salmaggi A
INSTITUCIÓN / INSTITUTION: - Department of Neurooncology, Fondazione IRCCS Istituto Neurologico Carlo Besta, Milan, Italy, giorgia.simonetti@istituto-besta.it.
RESUMEN / SUMMARY: - Angiogenesis has recently become a major target for the development of new antineoplastic drugs. The most serious adverse events linked to angiogenesis inhibitors are venous or arterial thromboembolism and haemorrhage. Thus, there is need to define with more certainty the impact of these new drugs in terms of adverse effects in neurological patients. The aim of the study is to assess the risk of venous thromboembolism (VTE) and bleeding in patients with malignant gliomas treated with bevacizumab with or without concomitant anticoagulant therapy. A review of published literature was performed in Medline, from which 476 records were identified. A total of 27 full-text articles, including retrospective analyses, retrospective reviews, and open label trials, were assessed for eligibility. The investigated drugs included bevacizumab alone, bevacizumab plus chemotherapy with/without concomitant radiation therapy; only two articles dealt with bevacizumab in association with anticoagulant treatment. A total of 2,208 patients with malignant gliomas, were identified and included in the analysis. From data it appears that patients receiving bevacizumab had a major risk of developing VTE that increased when bevacizumab is associated with radio-chemotherapy (4.27 vs 7.46 %). Regarding bleeding, data showed that patients treated with anticoagulant had a significantly increased risk of severe central nervous system (CNS) bleeding compared to patients not receiving anticoagulant therapy (0.6 vs 8.2 %). The use of bevacizumab combined with chemoradiotherapy seems to be associated with a higher risk for VTE compared to patients receiving antiangiogenic therapy alone. The associated use of anticoagulants and bevacizumab far increases the risk of developing CNS and non-CNS bleeding higher than grade 3, compared to patients receiving bevacizumab alone.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
While the ERCC1 C8092A and ERCC2 K751Q polymorphisms have received much attention for their potential associations with adult glioma risk, inferences from such studies are hindered by their limited statistical power and conflicting results. The aim of this meta-analysis is to provide a relatively comprehensive account of the association between these two polymorphisms and adult glioma risk. A literature search for eligible studies published before September 1, 2013 was conducted in PubMed, Embase, Web of Science, Cochrane Library, and CNKI databases. Pooled odds ratios (ORs) with their corresponding 95% confidence intervals (95% CIs) were used to evaluate the strength of the association under a fixed or random effect model according to heterogeneity test results. All analyses were performed using STATA software, version 12.0. Ten case-control studies were included in this meta-analysis, with a total of 5,843 adult glioma patients and 8,139 healthy controls. For ERCC1 C8092A (dbSNP: rs3212986, C>A), the combined results show that carriers of the AA genotype may be associated with a higher risk of adult glioma than carriers of the CA and CC genotypes. Stratified analyses show that the magnitude of the effect was especially significant among Asians, indicating ethnicity differences in adult glioma susceptibility. For ERCC2 K751Q (dbSNP: rs13181, A>C), the pooled ORs were not significant in the overall population, although all of the ORs were greater than 1. However, Asians seem to be significantly more susceptible to adult glioma than Caucasians. The results of this meta-analysis indicate that the AA genotype of ERCC1 C8092A may be associated with a higher risk of adult glioma than the CA and CC genotypes and that the risk allele of ERCC2 K751Q confers a significant susceptibility to adult glioma, especially in Asian populations. These polymorphisms may be used along with other genetic markers to identify individuals at high risk for adult glioma.
BACKGROUND: Tuberous sclerosis complex is an autosomal dominant disorder predisposing to the development of benign lesions in different body organs, mainly in the brain, kidney, liver, skin, heart, and lung. Subependymal giant cell astrocytomas are characteristic brain tumors that occur in 10% to 20% of tuberous sclerosis complex patients and are almost exclusively related to tuberous sclerosis complex. Subependymal giant cell astrocytomas usually grow slowly, but their progression ultimately leads to the occlusion of the foramen of Monro, with subsequent increased intracranial pressure and hydrocephalus, thus necessitating intervention. During recent years, secondary to improved understanding in the biological and genetic basis of tuberous sclerosis complex, mammalian target of rapamycin inhibitors have been shown to be effective in the treatment of subependymal giant cell astrocytomas, becoming an alternative therapeutic option to surgery.

METHODS: In June 2012, an International Tuberous Sclerosis Complex Consensus Conference was convened, during which an expert panel revised the diagnostic criteria and considered treatment options for subependymal giant cell astrocytomas. This article summarizes the subpanel's recommendations regarding subependymal giant cell astrocytomas. CONCLUSIONS: Mammalian target of rapamycin inhibitors have been shown to be an effective treatment of various aspects of tuberous sclerosis complex, including subependymal giant cell astrocytomas. Both mammalian target of rapamycin inhibitors and surgery have a role in the treatment of subependymal giant cell astrocytomas. Various subependymal giant cell astrocytoma-related conditions favor a certain treatment.

[10]

TÍTULO / TITLE: Children are not just little adults: recent advances in understanding of diffuse intrinsic pontine glioma biology.

RESUMEN / SUMMARY: Diffuse intrinsic pontine glioma (DIPG) is a high-grade glioma that originates in the pons and is seen exclusively in children. Despite numerous efforts to improve treatment DIPG remains incurable with 90% of children dying within two years of diagnosis, making it one of the leading causes of death in children with brain tumors. With the advent of new genomic tools the genetic landscape of DIPG is slowly being unraveled. The most common genetic alterations include a K27M mutation in H3.3 or H3.1, which are found in up to 78% of DIPGs while p53 mutations are found in up to 77%. Other recently discovered alterations include amplification of components of the Receptor Tyrosine Kinase-Ras-PI3K signaling pathway, particularly PDGFR-A. Recapitulating such alterations, genetically engineered DIPG preclinical models have been developed, and DIPG xenograft models have also been established. Both models have strengths and weaknesses but can help with the prioritization of novel agents for clinical trials for children with DIPG. As we move forward, it is important that we continue to study the complex and unique biology of DIPG and develop improved preclinical models to increase our understanding of DIPG pathogenesis, allowing

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - di Russo P; Perrini P; Pasqualetti F; Meola A; Vannozzi R
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Azienda Ospedaliero Universitaria Pisana (AOPU), Via Paradisa 2, 56100, Pisa, Italy.
RESUMEN / SUMMARY: - BACKGROUND: Multicentric malignant gliomas are well-separated tumours in different lobes or hemispheres, without anatomical continuity between lesions. The purpose of this study was to explore the clinical features, the pathology and the outcome according to the management strategies in a consecutive series of patients treated at a single institution. In addition, an analysis of the existing literature is presented. METHODS: For the institutional analysis, a retrospective review of all patients who underwent treatment for multicentric gliomas in the last 7 years was performed. For the analysis of the literature, a MEDLINE search with no date limitations was accomplished for surgical treatment of multicentric malignant gliomas. RESULTS: Two hundred and thirty-nine patients with glioma were treated in our department. Eighteen patients (7.5%) with a mean age of 64 years (age range, 37-78 years) presented multicentric malignant gliomas. Thirteen patients (72%) underwent surgical resection of at least one lesion that was followed by adjuvant treatment in all but one case. Five patients (28%) underwent stereotactic biopsy and thereafter received chemotherapy. A survival advantage was associated with resection of at least one lesion followed by adjuvant treatment (median overall survival 12 months) compared with 4 months for stereotactic biopsy followed by chemotherapy. Similar results were obtained from the review of the literature. CONCLUSIONS: Resection of at least one lesion seems to play a significant role in the management of selected patients with multicentric malignant gliomas. Multi-institutional studies on larger series are warranted to define how aggressively the patients with malignant multicentric gliomas should be treated.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Mendelson ZS; Husain Q; Elmoursi S; Svider PF; Eloy JA; Liu JK

Enlace al texto completo (gratuito o de pago) 1016/j.jocn.2013.07.008
RESUMEN / SUMMARY: - Rathke’s cleft cysts (RCC) arise from the development of the Rathke’s cleft pouch. These commonly occurring cysts are typically asymptomatic, but sometimes present with headaches, endocrine dysfunction, and visual loss. Recurrence is common after either drainage or surgical removal. The purpose of this study was to review published outcomes for RCC management, and determine whether specific factors, including patient demographics, cyst pathology, radiologic parameters, or surgical techniques predispose to their recurrence. A systematic review of studies for RCC from 1990 to 2012 was conducted. Patients were identified using a Medline/PubMed search, and from the bibliographies of relevant articles obtained from the primary search. Relevant studies reporting recurrence rate were identified, and data were extracted regarding patient demographics, presenting symptoms, cyst characteristics, surgical treatment, and outcomes. A meta-analysis for recurrence rates was also performed. Twenty-eight journal articles comprising a total of 1151 RCC revealed an average follow-up of 38 months (range 16-79 months). In the studies reviewed, there was a relatively equal distribution of treatment approaches, with 35% subtotal resection, 33% gross total resection, and 32% complete drainage with wall biopsy. The microsurgical transsphenoidal approach was found to have a higher recurrence rate (14% versus 8%) and new endocrine dysfunction rate (25% versus 10%) compared to the endoscopic approach. The data demonstrates a notable overall recurrence rate for RCC (12.5%). However, there appears to be no conclusive evidence that more aggressive resection of the cyst wall results in lower rates of recurrence.

[13]
TÍTULO / TITLE: - Association between glutathione S-transferase T1 null genotype and glioma susceptibility: a meta-analysis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Zhang B; Wang J; Niu H; Li Y; Yuan F; Tian Y; Zhou F; Hao Z; Zheng Y; Li Q; Wang W
INSTITUCIÓN / INSTITUTION: - Department of Neurology, The 2nd Affiliated Hospital, Harbin Medical University, Harbin, 150086, China.
RESUMEN / SUMMARY: - The relationship between genetic polymorphisms of glutathione S-transferase (GST) and the development of glioma has been investigated in several epidemiologic studies. However, these studies report inconsistent results. In order to get this precise result, a meta-analysis was conducted by calculating the pooled odds ratios (OR) and the 95% confidence intervals (95% CI). Eleven case-control research studies with a total of 2,416 glioma cases and 4,850 controls were included into this meta-analysis. The combined results based on all studies showed that there was no significant association between the GSTT1 null allele and glioma risk (OR = 1.188, 95% CI = 0.929-1.520, P heterogeneity = 0.003, P = 0.170). In the subgroup analysis, the same results were found in our work. There was no risk of
publication bias in this meta-analysis. Our results suggest that GSTT1 null genotype was not associated with the increased risk of glioma.

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


**AUTORES / AUTHORS:** - Sano K; Kitai R; Yoshimura H; Ohba S

**INSTITUCIÓN / INSTITUTION:** - Professor, Division of Dentistry and Oral Surgery, Department of Sensory and Locomotor Medicine, Faculty of Medical Sciences, University of Fukui, Fukui, Japan. Electronic address: sano@u-fukui.ac.jp.

**RESUMEN / SUMMARY:** - Recent advances in diagnostic tools, such as computed tomography and magnetic resonance imaging (MRI), have provided clinicians with the opportunity to detect asymptomatic meningiomas. This report describes a case of frontal convexity meningioma detected incidentally at MRI during the preoperative assessment of tongue cancer. To the best of the authors’ knowledge, this case report is the first regarding the successful treatment of tongue cancer in a patient with incidental meningioma. The incidence, perioperative management, and various imaging tests to detect meningiomas are discussed, with a review of the literature.

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


**AUTORES / AUTHORS:** - Zheng J; Geng M; Shi Y; Jiang B; Tai Y; Jing H

**INSTITUCIÓN / INSTITUTION:** - Department of Pathology, The General Hospital, Jinan Military Command, Shifan Road 25, Jinan 250031, Shandong Province, China.

**RESUMEN / SUMMARY:** - Oncocytic meningioma is an uncommon variant of meningioma, with only 20 reported cases to date, that is histologically characterized by the presence of neoplastic cells with granular eosinophilic cytoplasm rich in mitochondria. We present the clinicopathological features of a case of oncocytic meningioma in a 49-year-old Chinese female, along with a literature review. Brain computed tomography and magnetic resonance imaging demonstrated a slightly hyperintense mass located in the right frontal region and attached to the dura. In addition, it was homogeneously enhanced following contrast administration. She underwent gross total surgical resection of the tumor and adjacent dura. Grossly, the well-demarcated, nonencapsulated mass had a solid and tan-white appearance with soft and rubbery consistency. The lesions were composed primarily of sheets, nests, and cords of large polygonal bland cells with finely granular eosinophilic cytoplasm rich in mitochondria. Mitotic figures were rare, and necrosis was absent. There was no infiltration of the dura or brain cortex. Immunohistochemical staining revealed that the
neoplastic cells were positive for vimentin, epithelial membrane antigen, antimitochondrial antibody, and progesterone receptor, whereas MIB-1 stained only approximately 1% of the tumor cells. This is the first known report of an oncocytic meningioma arising in a Chinese patient. The patient was followed for 19 months without any evidence of metastasis or recurrence.

[16] TÍTULO / TITLE: - Plurihormonal pituitary adenoma with concomitant adrenocorticotropic hormone (ACTH) and growth hormone (GH) secretion: a report of two cases and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Rasul FT; Jaunmuktane Z; Khan AA; Phadke R; Powell M
INSTITUCIÓN / INSTITUTION: - Victor Horsley Department of Neurosurgery, National Hospital for Neurology and Neurosurgery, Queen Square, London, WC1N 3BG, UK, fahidrasul@doctors.org.uk.
RESUMEN / SUMMARY: - Plurihormonal pituitary adenomas are tumours that show immunoreactivity for more than one hormone that cannot be explained by normal adenohypophysial cytodifferentiation. The most common combinations in these adenomas include growth hormone (GH), prolactin (PRL) and one or more glycoprotein hormone sub-units (beta-TSH, beta-FSH, beta-LH and alphaSU). The authors report two cases of a plurihormonal pituitary adenoma expressing the rare combination of ACTH and GH. They both underwent successful transphenoidal hypophysectomy (TSH). Long-term post-operative follow-up revealed no evidence of tumour recurrence. Due to the multiple secretions and plurihormonal characteristics clinical diagnosis of composite pituitary adenomas can be difficult. The authors discuss the diagnosis and management of composite pituitary adenomas and review the literature regarding this rare phenomenon.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Tarantino R; Marruzzo D; Colistra D; Mancarella C; Delfini R
INSTITUCIÓN / INSTITUTION: - Department of Neurology and Psychiatry, Division of Neurosurgery, University of Rome “Sapienza”, Rome, Italy.
RESUMEN / SUMMARY: - There are only three cases of arachnoid cysts inducing twelfth nerve paresis described in English medical literature. We herein report one more instance. Six weeks after surgery, the patient has almost fully recovered. This case underlines the importance of considering the arachnoid cyst as a possible cause of twelfth nerve paresis.
TÍTULO / TITLE: Familial syndromes associated with intracranial tumours: a review.
RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Ranger AM; Patel YK; Chaudhary N; Anantha RV
INSTITUCIÓN / INSTITUTION: Department of Clinical Neurological Sciences, Western University, 800 Commissioners Road East, London, ON, N6A 5W9, Canada, adrianna.ranger@gmail.com.
RESUMEN / SUMMARY: BACKGROUND: Most cancers of the central nervous system (CNS) occur sporadically in the absence of any known underlying familial disorder or multi-systemic syndrome. Several syndromes are associated with CNS malignancies, however, and their recognition has significant implications for patient management and prognosis. Patients with syndrome-associated CNS malignancies often have multiple tumours (either confined to one region or distributed throughout the body), with similar or different histology. OBJECTIVE: This review examines syndromes that are strongly associated with CNS cancers: the phakomatosis syndromes, familial syndromes such as Li-Fraumeni and familial polyposis syndromes and dyschondroplasia.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Figueiredo N; Brooks N; Resnick DK
INSTITUCIÓN / INSTITUTION: UW-Madison - Department of Neurological Surgery University of Wisconsin School of Medicine and Public Health, Madison, WI, USA - n.brooks@neurosurgery.wisc.edu.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Aoki H; Ogura R; Tsukamoto Y; Okada M; Natsumeda M; Isogawa M; Yoshida S; Fujii Y
INSTITUCIÓN / INSTITUTION: Department of Neurosurgery, Brain Research Institute, Niigata University.
RESUMEN / SUMMARY: The efficacy and toxicity of high-dose methotrexate (HD-MTX)-based chemotherapy were retrospectively reviewed in patients with primary central nervous system lymphoma (PCNSL). All immunocompetent patients with histologically or radiographically diagnosed PCNSL treated between 2006 and 2012 at Niigata University Hospital were enrolled. Thirty-eight patients with a diagnosis of PCNSL were treated with one of two regimens during different time periods. During the
first period, from 2006 to 2009, three 3-week cycles of MPV (MTX + procarbazine + vincristine) were administered (MPV3 group). In the second period, from 2010 to 2012, five 2-week cycles of MTX were administered (MTX5 group). High-dose cytarabine was used in both groups following HD-MTX-based chemotherapy. Whole-brain radiotherapy was used for patients who did not attain a complete response (CR) based on magnetic resonance images. In the MPV3 group, 20 out of 23 patients (87%) completed the planned treatment. The CR rate after chemotherapy was 30%, and 57% after radiation therapy. Thirteen out of 15 patients (87%) in the MTX5 group completed the planned treatment. The CR rates after chemotherapy and radiation therapy were 53% and 93%, respectively. Renal dysfunction was assessed by measuring creatinine clearance rates, which were very similar in both groups. In terms of hematologic toxicity and other adverse reactions, there was no significant difference between the two groups. In conclusion, dose-dense MTX chemotherapy improved outcome with acceptable toxicity compared with the treatment schedule for three cycles of MPV treatment.

[21]

TÍTULO / TITLE: - English consensus protocol evaluating candidacy for auditory brainstem and cochlear implantation in neurofibromatosis type 2.

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


AUTORES / AUTHORS: - Tysome JR; Axon PR; Donnelly NP; Evans DG; Ferner RE; O' Connor AF; Freeman SR; Gleeson M; Halliday D; Harris F; Jiang D; Kerr R; King A; Knight RD; Lloyd SK; Macfarlane R; Mannion R; Mawman D; O'Driscoll M; Parry A; Ramsden J; Ramsden R; Rutherford SA; Saeed SR; Thomas N; Vanat ZH

INSTITUCIÓN / INSTITUTION: - *Department of Otolaryngology, Cambridge University Hospitals NHS Foundation Trust, Cambridge; daggerDepartment of Genetics, Manchester Royal Infirmary, Manchester; double daggerDepartment of Neurology, and section signDepartment of Otolaryngology, Guy’s and St. Thomas' NHS Foundation Trust, London; parallelDepartment of Otolaryngology, Manchester Royal Infirmary, Manchester; paragraph signDepartment of Otolaryngology, National Hospital for Neurology and Neurosurgery, London; #Department of Neurology, John Radcliffe Hospital, Oxford; ** Emmeline Centre for Hearing Implants, Cambridge University Hospitals NHS Foundation Trust, Cambridge; daggerdaggerDepartment of Neurosurgery, John Radcliffe Hospital, Oxford; double daggerdouble daggerDepartment of Neurosurgery, Manchester Royal Infirmary, Manchester; section sign section signDepartment of Audiology, and parallel parallelDepartment of Neurosurgery, Cambridge University Hospitals NHS Foundation Trust, Cambridge; paragraph sign paragraph signAudiology, Manchester Royal Infirmary, Manchester; ##Department of Otolaryngology, John Radcliffe Hospital, Oxford; ***Department of Otolaryngology, Royal National Throat Nose and Ear Hospital; and daggerdaggerdaggerDepartment of Neurosurgery, King’s College Hospital NHS Trust, London, U.K.

RESUMEN / SUMMARY: - OBJECTIVE: Hearing loss resulting from bilateral vestibular schwannomas (VSs) has a significant effect on the quality of life of patients with
neurofibromatosis Type 2 (NF2). A national consensus protocol was produced in England as a guide for cochlear implantation (CI) and auditory brainstem implantation (ABI) in these patients. STUDY DESIGN: Consensus statement. SETTING: English NF2 Service. PARTICIPANTS: Clinicians from all 4 lead NF2 units in England. MAIN OUTCOME MEASURES: A protocol for the assessment, insertion and rehabilitation of CI and ABI in NF2 patients. RESULTS: Patients should undergo more detailed hearing assessment once their maximum aided speech discrimination score falls below 50% in the better hearing ear. Bamford-Kowal-Bench sentence testing scores below 50% should trigger assessment for auditory implantation, as recommended by the National Institute for Clinical Excellence guidelines on CI. Where this occurs in patients with bilateral stable VS or a unilateral stable VS where the contralateral cochlear nerve was lost at previous surgery, CI should be considered. Where VS surgery is planned, CI should be considered where cochlear nerve preservation is thought possible, otherwise an ABI should be considered. Intraoperative testing using electrically evoked auditory brainstem responses or cochlear nerve action potentials may be used to determine whether a CI or ABI is inserted. CONCLUSION: The NF2 centers in England agreed on this protocol. Multisite, prospective assessments of standardized protocols for auditory implantation in NF2 provide an essential model for evaluating candidacy and outcomes in this challenging patient population.
a T11-T12 laminectomy with fenestration of the cyst. RESULTS: She experienced an immediate relief of pain symptoms and by the 7th postoperative day she was able to stand without help and walk a few meters with assistance. By the 6th postoperative month, the patient resulted significantly improved, having gained the ability to walk alone without assistance with complete resolution of the bladder dysfunctions, with no cyst recurrence after about 2 years’ follow-up. CONCLUSIONS: Intramedullary arachnoid cysts should be considered in the differential diagnosis for intramedullary cystic lesions. A particular consideration deserves their occurrence in the asymptomatic patients, who should be adequately informed on the possible natural evolution: when symptomatic, surgical therapy should be promptly offered, considering that a postoperative complete recovery is usually observed, regardless the surgical technique.

[23]
**TITULO / TITLE:** - Current concepts and future strategies in the management of intracranial germinoma.

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


**AUTORES / AUTHORS:** - Kortmann RD

**INSTITUCIÓN / INSTITUTION:** - Department of Radiation Therapy, University of Leipzig, Stephanstr. 9a, 04103 Leipzig, Germany.

Radiation therapy is the backbone in the management of intracranial germinoma. In localized disease chemotherapy followed by whole brain irradiation is the present standard providing cure rates in excess of 90%. Craniospinal irradiation alone in metastatic disease provides equally excellent outcome. Chemotherapy is able to convert macroscopic to microscopic disease permitting a dose reduction to the tumor site and possibly the ventricular system and is investigated in prospective trials. Chemotherapy alone cannot replace radiotherapy as sole treatment. Whole-ventricular radiotherapy followed by a boost to tumor site without chemotherapy might be feasible. New treatment technologies such as intensity-modulated radiotherapy or proton therapy permit a dose reduction to non-target brain. Data on functional outcome are conflicting and based on small heterogeneous series only mandating prospective investigations.

[24]
**TITULO / TITLE:** - Prevalence of neurobehavioral, social, and emotional dysfunction in patients treated for childhood craniopharyngioma: a systematic literature review.

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


**AUTORES / AUTHORS:** - Zada G; Kintz N; Pulido M; Amezcua L

**INSTITUCIÓN / INSTITUTION:** - Department of Neurosurgery, Keck School of Medicine of USC, Los Angeles, California, United States of America.
RESUMEN / SUMMARY: - BACKGROUND: Craniopharyngiomas (CP) are locally invasive and frequently recurring neoplasms often resulting in neurological and endocrinological dysfunction in children. In addition, social-behavioral impairment is commonly reported following treatment for childhood CP, yet remains to be fully understood. The authors aimed to further characterize the prevalence of neurobehavioral, social, and emotional dysfunction in survivors of childhood craniopharyngiomas. MATERIALS AND METHODS: A systematic literature review was conducted in PubMed to identify studies formally assessing neurobehavioral, social, and emotional outcomes in patients treated for CP prior to 18 years of age. Studies published between the years 1990-2012 that reported the primary outcome (prevalence of neurobehavioral, social, emotional/affective dysfunction, and/or impaired quality of life (QoL)) in >/=10 patients were included. RESULTS: Of the 471 studies screened, 11 met inclusion criteria. Overall neurobehavioral dysfunction was reported in 51 of 90 patients (57%) with available data. Social impairment (i.e. withdrawal, internalizing behavior) was reported in 91 of 222 cases (41%). School dysfunction was reported in 48 of 136 patients (35%). Emotional/affective dysfunction was reported in 58 of 146 patients (40%), primarily consisting of depressive symptoms. Health related quality of life was affected in 49 of 95 patients (52%). Common descriptors of behavior in affected children included irritability, impulsivity, aggressiveness, and emotional outbursts. CONCLUSIONS: Neurobehavioral, social, and emotional impairment is highly prevalent in survivors of childhood craniopharyngioma, and often affects quality of life. Thorough neurobehavioral/emotional screening and appropriate counseling is recommended in this population. Additional research is warranted to identify risk factors and treatment strategies for these disorders.

[25]

TÍTULO / TITLE: - The Role of Levetiracetam in Treatment of Seizures in Brain Tumor Patients.

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


AUTORES / AUTHORS: - Fonkem E; Bricker P; Mungall D; Aceves J; Ebwe E; Tang W; Kirmani B

INSTITUCIÓN / INSTITUTION: - The Brain Tumor Center, Scott & White Healthcare, Temple, TX, USA; Texas A&M Health Science Center College of Medicine, Temple, TX, USA.

RESUMEN / SUMMARY: - Levetiracetam, trade name Keppra, is a new second generation antiepileptic drug that is being increasingly used in brain tumor patients. In patients suffering with brain tumors, seizures are one of the leading neurologic complications being seen in more than 30% of patients. Unlike other antiepileptic drugs, levetiracetam is proposed to bind to a synaptic vesicle protein inhibiting calcium release. Brain tumor patients are frequently on chemotherapy or other drugs that induce cytochrome P450, causing significant drug interactions. However, levetiracetam does not induce the P450 system and does not exhibit any relevant drug interactions. Intravenous delivery is as bioavailable as the oral medication allowing it to be used in emergency situations. Levetiracetam is an attractive option for brain tumor patients suffering from seizures, but also can be used prophylactically in patients with brain tumors, or patients undergoing neurological surgery. Emerging studies have also
demonstrated that levetiracetam can increase the sensitivity of Glioblastoma tumors to the chemotherapy drug temozolomide. Levetiracetam is a safe alternative to conventional antiepileptic drugs and an emerging tool for brain tumor patients combating seizures.

[26] **TÍTULO / TITLE:** - Prospective trial of a short hospital stay protocol after endoscopic endonasal pituitary adenoma surgery.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Thomas JG; Gadgil N; Samson S; Takashima M; Yoshor D
**INSTITUCIÓN / INSTITUTION:** - The Pituitary Center at Baylor College of Medicine and St. Luke’s Episcopal Hospital, Houston, TX 77030, USA; Department of Neurosurgery, Baylor College of Medicine, Houston TX 77030, USA.

**RESUMEN / SUMMARY:** - OBJECT: Patients typically remain hospitalized for several days after transsphenoidal surgery for pituitary adenoma resection for reasons including pain control, serial neurological assessments, surveillance for CSF leak, and management of endocrine issues. We sought to determine if an evidence-based perioperative care protocol combined with an endoscopic approach could lead to routine and safe discharge on post-operative day 1 (POD1). METHODS: Our multidisciplinary pituitary group prospectively implemented a perioperative care protocol that emphasizes patient education, early mobilization, and scheduled inpatient and outpatient endocrine assessments on 50 consecutive patients who underwent surgical resection of a pituitary adenoma (82% macroadenomas, 2.1+/−0.8 cm, max 4.5 cm, 18% microadenomas). Endoscopic endonasal surgery characterized by aggressive tumor resection and avoidance of nasal packing and lumbar drains was employed in all cases. Lengths-of-stay, readmissions, and postoperative outcomes were analyzed.
**RESULTS:** Using the short-stay-protocol, 92% (46/50) of patients were successfully discharged on POD1. The average length-of-stay for all patients was 1.16 +/-0.55 days (range 1-4). Postoperative diabetes insipidus (DI) occurred in 16% of patients (8/50), was effectively managed on an outpatient basis did not delay discharge. Readmission was required in 2 patients, in both cases for delayed presentation of a CSF leak.
**CONCLUSION:** A short-stay protocol allows for an overnight hospital stay for patients following pituitary surgery with a low rate of complications or readmission. This study offers evidence-based guidelines that may be used to avoid complications and facilitate early discharge after transsphenoidal surgery.

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[27] **TÍTULO / TITLE:** - Successful treatment of paraganglioma with sorafenib: a case report and brief review of the literature.
**RESUMEN / SUMMARY:** - Enlace al Resumen / Link to its Summary

**AUTORES / AUTHORS:** - Lin Y; Li Q; Huang W; Jia X; Jiang H; Gao Y; Li Q
INTRODUCTION: To date, no effective systemic therapies have been made available for paraganglioma. However, multiple mutations in susceptibility genes have been identified that are potential targets for sorafenib, an oral multitargeted tyrosine-kinase inhibitor. CASE PRESENTATION: We report the case of a 69-year-old Chinese man with mediastinal paraganglioma that had metastasized to the bone. The paraganglioma responded to sorafenib, a novel multitargeted tyrosine kinase inhibitor that targets angiogenesis, the Raf-kinase pathway, the platelet-derived growth factor Ret, and c-Kit. The patient was diagnosed as having paraganglioma after biopsy of the mediastinal mass. We first treated the patient with radiotherapy. Then he tolerated an etoposide-and-cisplatin chemotherapy regimen. Subsequently, he received 6 months of maintenance treatment with sorafenib (400 mg twice daily). A dramatic reduction in tumor volume was observed. At present, the patient has achieved a partial response, and his clinical status remains unchanged.

CONCLUSION: We suggest that sorafenib should be further investigated in the management of patients with paraganglioma.
**ENLACE AL TEXTO COMPLETO (GRATUITO O DE PAGO)** 2147/OTT.S50208

AUTORES / AUTHORS: - Thon N; Kreth S; Kreth FW

INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Hospital of the University of Munich, Campus Grosshadern, Munich, Germany.

RESUMEN / SUMMARY: - The identification of molecular genetic biomarkers considerably increased our current understanding of glioma genesis, prognostic evaluation, and treatment planning. In glioblastoma, the most malignant intrinsic brain tumor entity in adults, the promoter methylation status of the gene encoding for the repair enzyme O6-methylguanine-DNA methyltransferase (MGMT) indicates increased efficacy of current standard of care, which is concomitant and adjuvant chemoradiotherapy with the alkylating agent temozolomide. In the elderly, MGMT promoter methylation status has recently been introduced to be a predictive biomarker that can be used for stratification of treatment regimes. This review gives a short summy of epidemiological, clinical, diagnostic, and treatment aspects of patients who are currently diagnosed with glioblastoma. The most important molecular genetic markers and epigenetic alterations in glioblastoma are summarized. Special focus is given to the physiological function of DNA methylation-in particular, of the MGMT gene promoter, its clinical relevance, technical aspects of status assessment, its correlation with MGMT mRNA and protein expressions, and its place within the management cascade of glioblastoma patients.

[30]

TÍTULO / TITLE: - Everolimus in the treatment of subependymal giant cell astrocytomas, angiomyolipomas, and pulmonary and skin lesions associated with tuberous sclerosis complex.

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


AUTORES / AUTHORS: - Franz DN

INSTITUCIÓN / INSTITUTION: - Department of Pediatrics, Tuberous Sclerosis Clinic, Cincinnati Children’s Hospital Medical Center, Cincinnati, OH, USA.

RESUMEN / SUMMARY: - Tuberous sclerosis complex (TSC) is an autosomal dominant genetic disorder caused by inactivating mutations in either the TSC1 or TSC2 genes. It is characterized by the development of multiple, benign tumors in several organs throughout the body. Lesions occur in the brain, kidneys, heart, liver, lungs, and skin and result in seizures and epilepsy, mental retardation, autism, and renal and pulmonary organ system dysfunction, as well as other complications. Elucidation of the molecular pathways and etiological factors responsible for causing TSC has led to a paradigm shift in the management and treatment of the disease. TSC1 or TSC2 mutations lead to constitutive upregulation of the mammalian target of rapamycin pathway, which affects many cellular processes involved in tumor growth. By targeting mammalian target of rapamycin with everolimus, an orally active rapamycin derivative, clinically meaningful and statistically significant reductions in tumor burden have been achieved for the main brain (subependymal giant cell astrocytoma) and renal manifestations (angiomyolipoma) associated with TSC. This review provides an overview of TSC, everolimus, and the clinical trials that led to its approval for the treatment of TSC-associated subependymal giant cell astrocytoma and renal angiomyolipoma.
Adoptive Cell Therapies for Glioblastoma.

Título / Title: Adoptive Cell Therapies for Glioblastoma.
Resumen / Summary: Enlace al Resumen / Link to its Summary

Resumen / Summary: Glioblastoma (GBM) is the most common and most aggressive primary brain malignancy and, as it stands, is virtually incurable. With the current standard of care, maximum feasible surgical resection followed by radical radiotherapy and adjuvant temozolomide, survival rates are at a median of 14.6 months from diagnosis in molecularly unselected patients (1). Collectively, the current knowledge suggests that the continued tumor growth and survival is in part due to failure to mount an effective immune response. While this tolerance is subtended by the tumor being utterly "self," it is to a great extent due to local and systemic immune compromise mediated by the tumor. Different cell modalities including lymphokine-activated killer cells, natural killer cells, cytotoxic T lymphocytes, and transgenic chimeric antigen receptor or alphabeta T cell receptor grafted T cells are being explored to recover and or redirect the specificity of the cellular arm of the immune system toward the tumor complex. Promising phase I/II trials of such modalities have shown early indications of potential efficacy while maintaining a favorable toxicity profile. Efficacy will need to be formally tested in phase II/III clinical trials. Given the high morbidity and mortality of GBM, it is imperative to further investigate and possibly integrate such novel cell-based therapies into the current standards-of-care and herein we collectively assess and critique the state-of-the-knowledge pertaining to these efforts.

Antiangiogenic therapy for high-grade gliomas: current concepts and limitations.

Título / Title: Antangiogenic therapy for high-grade gliomas: current concepts and limitations.
Resumen / Summary: Enlace al Resumen / Link to its Summary

Resumen / Summary: Glioblastoma (GBM) is associated with a high degree of angiogenesis. Therefore, antiangiogenic therapy could have a role in the treatment of this tumor. The currently available treatment approaches acting against angiogenesis are mainly directed toward three pathways: VEGF pathway, VEGF-independent pathways and inhibition of vascular endothelial cell migration. It has been demonstrated that antiangiogenic therapy can produce a rapid radiological response and a decrease of brain edema, without significantly influencing survival. Future studies...
should consider that: animal models are inadequate and cells used for animal models
(mainly U87) are deeply different from patient GBM cells; GBM cells may become
resistant to antiangiogenic therapy and some cells may be resistant to antiangiogenic
therapy ab initio; and angiogenesis in the peritumor tissue has been poorly
investigated. Therefore, the ideal target of angiogenesis is probably yet to be identified.

[33] TÍTULO / TITLE: - Optic Neuritis as Isolated Manifestation of Leptomeningeal
Carcinomatosis: A Case Report and Systematic Review of Ocular Manifestations of
Neoplastic Meningitis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Lanfranconi S; Basilico P; Trezzi I; Borellini L; Franco G;
Civelli V; Pallotti F; Bresolin N; Baron P
INSTITUCIÓN / INSTITUTION: - Department of Neurological Sciences, Fondazione
I.R.C.C.S. Ca’ Granda Ospedale Maggiore Policlinico, Dino Ferrari Center, 20122
Milan, Italy.
RESUMEN / SUMMARY: - Introduction. Leptomeningeal carcinomatosis occurs in about
5% of cancer patients. Ocular involvement is a common clinical manifestation and often
the presenting clinical feature. Materials and Methods. We report the case of a 52-year
old lady with optic neuritis as isolated manifestation of neoplastic meningitis and a
review of ocular involvement in neoplastic meningitis. Ocular symptoms were the
presenting clinical feature in 34 patients (83%) out of 41 included in our review, the
unique manifestation of meningeal carcinomatosis in 3 patients (7%). Visual loss was
the presenting clinical manifestation in 17 patients (50%) and was the most common
ocular symptom (70%). Other ocular signs were diplopia, ptosis, papilledema,
anisocoria, exophthalmos, orbital pain, scotomas, hemianopsia, and nystagmus.
Associated clinical symptoms were headache, altered consciousness, meningism, limb
weakness, ataxia, dizziness, seizures, and other cranial nerves involvement. All
patients except five underwent CSF examination which was normal in 1 patient,
pleocytosis was found in 11 patients, increased protein levels were observed in 16
patients, and decreased glucose levels were found in 8 patients. Cytology was positive
in 29 patients (76%). Conclusion. Meningeal carcinomatosis should be considered in
patients with ocular symptoms even in the absence of other suggestive clinical
symptoms.

[34] TÍTULO / TITLE: - Gelastic epilepsy in combination with hypothalamic hamartoma and
partial agenesis of the corpus callosum: A case report and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Cheng B; Sun C; Li S; Gong Q; Lui S
INSTITUCIÓN / INSTITUTION: - Huaxi MR Research Center (HMRRC), Department of
Radiology, West China Hospital of Sichuan University, Chengdu, Sichuan 610041, P.R.
China.
RESUMEN / SUMMARY: Gelastic epilepsy has been reported to originate from various conditions, particularly from hypothalamic hamartoma (HH). In the present study, we report a patient with gelastic seizures (GSs), followed by complex partial and tonic-clonic seizures. Magnetic resonance imaging (MRI) revealed a rare combination of HH and partial agenesis of the corpus callosum (ACC). Following resectioning of the HH, the seizures were reduced, but not fully controlled, with medication by the one year follow-up. HH and partial ACC patients may experience seizures; the seizures in the case presented in this study may have originated from HH, partial ACC or both. Considering the fact that seizure frequency reduced following surgery, they may have mainly occurred from HH. Additionally it was considered to be likely that the seizures following surgery were due to secondary epileptogenesis, partial ACC, or both.

RESUMEN / SUMMARY: Extraventricular neurocytomas (EVNs) are rare neuronal tumors included in the definition of neoplasms in the 2007 World Health Organization classification of tumors of the central nervous system. Although a small case series of EVNs in adults has been previously reported, EVNs in pediatric populations are extremely rare. The current case report presents the clinicopathological features of an EVN in a 2-year-old female who presented with nausea and vomiting that had lasted for five days. In addition, an analysis of the imaging features, histology, treatment and prognosis of these reported rare lesions is presented. Immunohistochemically, EVNs are characterized by the robust expression of synaptophysin, but with a lack of oligodendrocyte transcription factor 2, isocitrate dehydrogenase enzyme isoform 1 (IDH1) R132/IDH2 R172 mutations and p53 immunoexpression. The treatment for EVNs in pediatric and adult populations is gross total resection, with post-operative radiation reserved for subtotal resection or recurrent disease. In addition, drop metastasis must be carefully avoided.

[36] TÍTULO / TITLE: Radiotherapy plus concurrent or sequential temozolomide for glioblastoma in the elderly: a meta-analysis.
RESUMEN / SUMMARY: Radiotherapy plus concurrent or sequential temozolomide for glioblastoma in the elderly: a meta-analysis.
AUTORES / AUTHORS: Yin AA; Zhang LH; Cheng JX; Dong Y; Liu BL; Han N; Zhang X
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Xijing Institute of Clinical Neuroscience, Xijing Hospital, Fourth Military Medical University, Xi'an, Shaanxi Province, PR China.

RESUMEN / SUMMARY: - BACKGROUND: Many physicians are reluctant to treat elderly glioblastoma (GBM) patients as aggressively as younger patients, which is not evidence based due to the absence of validated data from primary studies. We conducted a meta-analysis to provide valid evidence for the use of the aggressive combination of radiotherapy (RT) and temozolomide (TMZ) in elderly GBM patients.

METHODS: A systematic literature search was conducted using the PubMed, EMBASE and Cochrane databases. Studies comparing combined RT/TMZ with RT alone in elderly patients (≥65 years) with newly diagnosed GBM were eligible for inclusion. RESULTS: No eligible randomized trials were identified. Alternatively, a meta-analysis of nonrandomized studies (NRSs) was performed, with 16 studies eligible for overall survival (OS) analysis and nine for progression-free survival (PFS) analysis. Combined RT/TMZ was shown to reduce the risk of death and progression in elderly GBM patients compared with RT alone (OS hazard ratio [HR] 0.59, 95% confidence interval [CI] 0.48-0.72; PFS: HR 0.58, 95% CI 0.41-0.84). Evaluable patients were reported to tolerate combined treatment but certain toxicities, and especially hematological toxicities, were more frequently observed. Limited data on O6-methylguanine-DNA methyltransferase (MGMT) promoter status and quality of life were reported. CONCLUSION: The meta-analysis of NRSs provided level 2a evidence (Oxford Centre for Evidence-Based Medicine) that combined RT/TMZ conferred a clear survival benefit on a selection of elderly GBM patients who had a favorable prognosis (e.g., extensive resection, favorable KPS). Toxicities were more frequent but acceptable. Future randomized trials are warranted to justify a definitive conclusion.

TÍTULO / TITLE: - Early outcome in endoscopic extended endonasal approach for removal of supradiaphragmatic craniopharyngiomas: a case series and a comprehensive review.

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


AUTORES / AUTHORS: - Bosnjak R; Benedicic M; Vittori A

INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, University Medical Centre, 1000 Ljubljana, Slovenia.

RESUMEN / SUMMARY: - BACKGROUND: The choice of endoscopic expanded endonasal approach introduces the possibility of improved gross total resection of craniopharyngioma while minimizing surgical morbidity in a significant subset of patients. METHODS: From our trans-sphenoidal surgical series of 331 cases, we retrospectively reviewed visual, endocrine and neuro-cognitive outcomes in the first consecutive eight patients (median age 63 years; range 47-73 years) with newly diagnosed supradiaphragmatic craniopharyngioma (median tumour height 23 mm; range 15-34 mm), removed by expanded endonasal approach (median follow-up 27 months; range 10-69 months). Gross total resection was attempted in all patients. RESULTS: Gross total resection was achieved in 6 of 8 patients. Visual improvement was present in 6 of 8 patients of patients or in 14 of 16 eyes. New endocrinopathy,
including diabetes insipidus, appeared in 5 of 8 patients. Stalk was preserved in 4 patients. Cognitive decline was present in 2 cases. Five of 8 patients retained previous quality of life. CONCLUSIONS: Our early outcome results are comparable to the recent few expanded endonasal approach series, except for the incidence of new endocrinopathy and cerebrospinal fluid leak rate. This was influenced by higher number of transinfundibular tumours in our series, where stalk preservation is less likely, and not using nasoseptal flap or gasket closure in the first half of cases. Including data from the literature and ours, expanded endonasal approach shows a trend for improved gross total resection rate with less morbidity, more obviously for visual outcome and quality of life than for endocrine outcome. However, validity of expanded endonasal approach should be confirmed in a larger number of patients with a longer follow-up period.

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[38]

**TÍTULO / TITLE:** Intracranial Hemangiopericytoma, our experience in 30 years: a series of 43 cases. A case series and review of the literature.

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


**AUTORES / AUTHORS:** Melone AG; D’Elia A; Santoro F; Salvati M; Delfini R; Cantore G; Santoro A

**INSTITUCIÓN / INSTITUTION:** Department of Neurological Sciences-Neurosurgery, University of Rome “Sapienza”, Italy. Electronic address: melgra80@yahoo.it.

**RESUMEN / SUMMARY:** INTRODUCTION: Meningeal hemangiopericytoma (HPC) is a rare, aggressive CNS tumor that tends to invade locally, metastasize, and has a high rate of recurrence. METHODS: The authors present a retrospective review of patients managed for intracranial HPC at Rome University Hospital. RESULTS: A total of 43 patients with intracranial HPC were treated from 1980 to 2010. Treatment and follow-up information was available for analysis on 36 patients. The median survival for all patients was 83.5 months after date of diagnosis, with 1-year, 5-year, and 10-year survival rates of 100%, 94.4%, and 72.2%, respectively. Eighteen patients (41.86%) had HPC recurrence. The median time until recurrence was 72.24 months, with 1-year, 5-year, and 10-year progression-free survival rates of 98%, 51%, and 29%, respectively. Five patients (11.62%) developed extracranial metastasis. Patients undergoing any form of adjuvant radiation treatment, including EBRT, Gamma Knife radiosurgery, and/or proton beam therapy, had no longer median OS (178 vs 154 months, respectively; p = 0.2); but did have a significantly improved recurrence-free interval (108 vs 64 months; p = 0.04) compared with patients who did not undergo radiation treatment. Tumor characteristics associated with earlier recurrence included size >/= 7 cm (log-rank, p < .05) and sinus invasion (log-rank, p < .05). CONCLUSIONS: Strategies combining adjuvant radiation with tumor resection appeared to hinder tumor progression, but had no effect on overall survival or the development of metastases. Greater extent of resection was associated with increased overall survival (log-rank, p < .05). Anaplastic HPC was associated with reduced overall survival and with reduced recurrence interval (log-rank, p < .05).

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[39]
**Título / Title:** Xanthomatous meningioma: a case report with review of the literature.

**Resumen / Summary:**

Enlace al Resumen / Link to its Summary


**Autores / Authors:** Ishida M; Fukami T; Nitta N; Iwai M; Yoshida K; Kagotani A; Nozaki K; Okabe H

**Institución / Institution:** Department of Clinical Laboratory Medicine and Division of Diagnostic Pathology, Shiga University of Medical Science Shiga, Japan.

**Resumen / Summary:**

Xanthomatous meningioma is an extremely rare variant of meningioma that is characterized histopathologically by the presence of tumor cells with lipid-filled vacuolated cytoplasm. In this report, we describe the fifth documented case of xanthomatous meningioma and review its clinicopathological features. A 76-year-old Japanese male presented with dizziness. Magnetic resonance imaging demonstrated a well-circumscribed tumor in the left parasagittal to frontal region with attachment of the dura mater. Histopathological examination of the resected specimen revealed proliferation of polygonal to spindle cells with eosinophilic cytoplasm and bland round to oval nuclei. Whorl formation and psammomas were scattered, and mitotic figures were rarely seen. A peculiar finding was the presence of extensive xanthomatous change continuing to the above-mentioned typical meningothelial meningioma. These tumor cells had clear vacuolated cytoplasm and bland round to oval nuclei. Immunohistochemically, xanthomatous cells were positive for epithelial membrane antigen. Accordingly, an ultimate diagnosis of xanthomatous meningioma was made. Our clinicopathological analysis revealed that xanthomatous meningioma affects children to young persons or the elderly, and four of five cases were located in the supratentorial region. Although the detailed mechanism underlying the xanthomatous change has not been clarified, this change is thought to result from a metabolic abnormality of the neoplastic meningothelial cells. Further, xanthomatous change has also been reported in atypical and anaplastic meningiomas. Therefore, it is important to recognize that xanthomatous change can occur in meningiomas, and to avoid misidentifying these cells as macrophages.

**Metastasis of lung adenosquamous carcinoma to meningioma: case report with literature review.**

**Resumen / Summary:**

Enlace al Resumen / Link to its Summary


**Autores / Authors:** Glass R; Hukku SR; Gershenhorn B; Alzate J; Tan B

**Institución / Institution:** Department of Radiation Biology, Memorial Sloan Kettering Cancer Center New York, NY.

**Resumen / Summary:**

The occurrence of metastasis of a systemic neoplasm to an intracranial tumor is a rare phenomenon. Meningiomas have been reported as the most common intracranial tumor to harbor a systemic metastasis, with breast and lung carcinomas being the most common sites of origination. Here, we report a case of an adenocarcinoma metastasis of an adenosquamous lung carcinoma found within a meningioma, resulting in the patient’s first clinical manifestations. We also review the literature for other cases of adenocarcinoma metastatic to a meningioma and suggest mechanisms that make meningiomas likely to harbor systemic metastases including
increased vascularity, slow growth rate, increased hyaline content and expression of cell-cell adhesion molecules.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Spitler K; Drazin D; Hanna G; Patel A; Chu R
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Cedars-Sinai Medical Center, Los Angeles, CA 90048, USA.
RESUMEN / SUMMARY: - Asymptomatic intracranial abnormalities are increasingly becoming a focus of attention with the utilization of high-resolution imaging. The concurrence of tumor and aneurysm has been described, largely, by case reports and single-surgeon experiences. Recent papers have outlined the ethics of incidental findings and possible treatment algorithms. Incidental finding of an aneurysm occurs most commonly in patients with meningiomas, pituitary adenomas, and gliomas. Such an association may explain the mechanisms of aneurysm formation, growth, and rupture in acromegalic patients; however, insufficient data are available to link aneurysm with either glioma or meningioma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Krishnan P; Kartikueyan R
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, National Neurosciences Centre, Peerless Hospital Complex, Kolkata, India.

[43] TITULO / TITLE: - Implications of Rho GTPase Signaling in Glioma Cell Invasion and Tumor Progression.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Fortin Ensign SP; Mathews IT; Symons MH; Berens ME; Tran NL
INSTITUCIÓN / INSTITUTION: - Cancer and Cell Biology Division, Translational Genomics Research Institute, Phoenix, AZ, USA; Cancer Biology Graduate Interdisciplinary Program, University of Arizona, Tucson, AZ, USA.
RESUMEN / SUMMARY: - Glioblastoma (GB) is the most malignant of primary adult brain tumors, characterized by a highly locally invasive cell population, as well as abundant proliferative cells, neoangiogenesis, and necrosis. Clinical intervention with chemotherapy or radiation may either promote or establish an environment for
manifestation of invasive behavior. Understanding the molecular drivers of invasion in the context of glioma progression may be insightful in directing new treatments for patients with GB. Here, we review current knowledge on Rho family GTPases, their aberrant regulation in GB, and their effect on GB cell invasion and tumor progression. Rho GTPases are modulators of cell migration through effects on actin cytoskeleton rearrangement; in non-neoplastic tissue, expression and activation of Rho GTPases are normally under tight regulation. In GB, Rho GTPases are deregulated, often via hyperactivity or overexpression of their activators, Rho GEFs. Downstream effectors of Rho GTPases have been shown to promote invasiveness and, importantly, glioma cell survival. The study of aberrant Rho GTPase signaling in GB is thus an important investigation of cell invasion as well as treatment resistance and disease progression.

[44]
TÍTULO / TITLE: - Malignant intracerebral nerve sheath tumors: a case report with review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Shweikeh F; Drazin D; Bannykh SI
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Cedars-Sinai Medical Center, Maxine Dunitz Neurosurgical Institute, Los Angeles, CA 90048, USA.
RESUMEN / SUMMARY: - The occurrence of benign nerve sheath tumors within the neuroaxis is uncommon. Even rarer is the finding within brain parenchyma, termed malignant intracerebral nerve sheath tumors (MINST). We present a case of MINST which occurred in the frontal lobe of an 18-year-old male that recurred almost 4 years later. Imaging demonstrated a 4.0 cm lesion with an associated mass effect. He underwent a right frontoparietal craniotomy for gross total resection. Pathology was inconclusive with a Glioblastoma Multiforme (GBM) as the most likely diagnosis, though gliosarcoma and MINST were also highly considered. Postoperatively, he was treated with chemotherapy and radiation and followed for almost 4 years, when an MRI indicated a recurrence. Resection of the recurrence was highly suggestive of MINST. Surgery was followed by radiation and chemotherapy, but, less than 7 months later, he was readmitted for a surgical-site infection, and, after multiple surgeries, and his family terminated care. Recognizing this unusual tumor in the differential diagnosis of a heterogeneously enhancing intracerebral mass can help surgeons diagnose and treat it. This report also exhaustively reviews the literature and presents diagnostic and treatment strategies.

[45]
TÍTULO / TITLE: - Malignant extra-adrenal pancreatic paraganglioma: case report and literature review.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Al-Jiffry BO; Alnemary Y; Khayat SH; Haiba M; Hatem M
RESUMEN / SUMMARY: - BACKGROUND: Pancreatic paragangliomas are rare tumors, with only 16 reported cases to date. One of these cases demonstrates
metastasis to lymph node, while another case was functional, however, none of these cases showed malignant and large, pancreatic paraganglioma with marked invasion. Also another unique feature was the age of our patient compared to the average reported ages in published literature (42--85 years). **CASE PRESENTATION:** A 19-year-old woman presented with a one-year history of intermittent abdominal pain. Physical examination showed a palpable mass in the right upper abdomen, but initial laboratory results were within normal ranges; tumor markers (CEA, AFP, and CA19-9) were negative. An abdominal and pelvic computed tomography (CT) scan showed a well-defined retroperitoneal para-aortic mass. The CT scan revealed that the surrounding lymph nodes were not enlarged, but the liver showed evidence of parenchymal infiltration. Intraoperatively, a large, firm tumor originating from the head of pancreas was found pushing on the caudate hepatic lobe and the inferior vena cava (IVC). The tumor was resected through a pancreaticoduodenectomy, involving segment VI of the liver and a small segment of the IVC. The blood pressure spiked (>220 mm Hg) when the tumor was manipulated during the operation. The final pathology report showed a 9-cm tumor with lymphovascular invasions; immunohistochemistry was positive for synaptophysin and chromogranin. All resection margins were negative and 1/15 lymph nodes was positive for metastasis. Postoperative recovery was unremarkable. One month after discharge, the patient was re-admitted with abdominal pain and found to have an abdominal collection at the resection site, which was drained under CT guidance. She received a therapeutic dose of I131-metaiodobenzylguanidine (MIBG). Follow-ups showed the absence of recurrence, and she has remained disease free. **CONCLUSION:** This patient was an extraordinary example of a rare tumor. Even more remarkable was that the tumor was malignant with lymph node invasion. To our knowledge, a case similar to that presented here has not been previously reported in the literature.

[46]

**TÍTULO / TITLE:** - Primary medulla oblongata germinomas: two case reports and review of the literature.

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


- Enlace al texto completo (gratuito o de pago) [1186/1477-7819-11-274]

**AUTORES / AUTHORS:** - Hao S; Li D; Feng J; Wang L; Wu Z; Zhang J; Zhang L

**RESUMEN / SUMMARY:** - An intracranial germinoma is a tumor that is sensitive to radiotherapy. As medulla oblongata germinomas are extremely rare, determining an accurate preoperative diagnosis is challenging. Two cases of medulla oblongata lesions were surgically treated, and a postoperative diagnosis of germinoma was determined in both of the cases. The tumor in one patient completely resolved after a treatment course consisting of surgical intervention, radiotherapy and chemotherapy; the other patient, who did not receive any type of adjuvant treatment after surgery, suffered from tumor relapse and died from pneumonia 8 months following surgery. A preoperative diagnosis of medulla oblongata germinoma is difficult because of the lack of specific clinical signs and symptoms. If the correct diagnosis is reached, patients can have a favorable prognosis with proper evaluation and treatment. An invasive operation can potentially lesion and impair the function of the medulla oblongata, which is fatal to the patient.

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Hypertension secondary to a periprostatic paraganglioma: case report and review of the literature.

BACKGROUND: Around 10 per cent of catecholamine-secreting tumours can be found outside the adrenal medulla (paraganglioma). We report a case of a functional sporadic paraganglioma that was localized later to the prostate without causing lower urinary tract symptoms. CASE PRESENTATION: A 76-year old male with an extensive history of cardiovascular disease suffered from hypertension and an unexplained hypochromic microcytic anaemia for years before the coincidental discovery of a 2.5 x 3.5 cm periprostatic mass upon abdominal contrast-enhanced CT scanning. Transrectal biopsies revealed a paraganglioma. The urinary levels of the catecholamine metabolites were found increased. The paraganglioma showed uptake of iodine-123-metaiodobenzylguanidine by SPECT scanning, indicating a solitary lesion. Successful preperitoneal endoscopic resection of the tumour was performed, which resulted in a decrease in blood pressure and a normalization of the urinary catecholamine metabolites. None of the to date known genetic mutations that have been shown to relate to the existence of paragangliomas were identified in the current case. CONCLUSION: An intra- or periprostatic localization of a paraganglioma is very rare. We reviewed the literature and found 6 other cases. Three of the described cases presented with lower urinary tract symptoms. In these three patients, the tumour had a size of 4 cm or larger and in 67 per cent of these cases the paragangliomas were situated within the prostate. The periprostatic region might be considered as a possible location for paragangliomas, especially in the presence of lower urinary tract symptoms even though they were absent in the current case.

Gastric diffuse large B cell lymphoma presenting as paraneoplastic cerebellar degeneration: Case report and review of literature.

Paraneoplastic cerebellar degeneration (PCD) is a type of paraneoplastic neurological disorder (PND) that is associated with many solid tumors, Hodgkin’s lymphoma (HL) and very rarely with non-Hodgkin’s lymphoma (NHL). We report a case of PCD associated with gastric diffuse large B-cell lymphoma (DLBCL) in a patient who presented with acute onset of giddiness and double vision and had complete remission of the gastric lesion and marked improvement of cerebellar syndrome with rituximab-based combination chemotherapy. A brief review of the literature is also presented.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Ramirez C; Hernandez-Ramirez LC; Espinosa-de-Los-Monteros AL; Franco JM; Guinto G; Mercado M
INSTITUCIÓN / INSTITUTION: - Endocrinology Service and Experimental Endocrinology Unit, Hospital de Especialidades Siglo XXI, Instituto Mexicano del Seguro Social, Aristoteles 68, Col, Polanco, 11560 Mexico City, Mexico. moises.mercado@endocrinologia.org.mx.

RESUMEN / SUMMARY: - BACKGROUND: In more than 98% of cases, acromegaly is due to a GH-secreting pituitary adenoma. The term “ectopic acromegaly” includes neuroendocrine tumors secreting GH releasing hormone (GHRH), usually located in the lungs, thymus and endocrine pancreas. Considerably less frequent are cases of ectopic acromegaly due to GH-secreting tumors located out of the pituitary fossa; except for one isolated case of a well-documented GH-secreting lymphoma, the majority of these lesions are located in the sphenoid sinus. CASE PRESENTATION: We present the case of a 45 year old woman with acromegaly whose MRI showed an empty sella without evidence of a pituitary adenoma but revealed a large mass within the sphenoid sinus. She underwent transsphenoidal surgery and the excised sphenoid sinus mass, proved to be a GH-secreting adenoma; the sellar floor was intact and no other lesions were found in the pituitary fossa. She required postoperative treatment with somatostatin analogs and cabergoline for clinical and biochemical control.
CONCLUSIONS: This case highlights the importance of carefully evaluating the structures surrounding the sellar area when a pituitary adenoma is not found with currently available imaging techniques. The finding of an intact sellar floor and duramater lead us to conclude that the patient’s tumor originated de novo from embryological pituitary remnants. Upon a careful review of the literature and a critical evaluation of our case we found neither clinical nor biochemical features that would distinguish an ectopic from the more common eutopically located somatotrophinoma.

TÍTULO / TITLE: - Subcutaneous metastasis in medulloblastoma: A case report and review of literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

AUTORES / AUTHORS: - Maiti T; Sabharwal P; Pandey P; Devi BI
INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, National Institute of Mental Health and Neuro Sciences, Bangalore, Karnataka, India.
RESUMEN / SUMMARY: - Extraneural metastasis (ENM) in patients with medulloblastoma is a rare but a well-described phenomenon, both in children and adults. Most of the ENM involve bone and bone marrow and rarely involve other solid
organs. Subcutaneous and muscular metastasis is an extremely rare event, more so in children, with only two cases documented in the pediatric population. We describe a case of medulloblastoma with ENM in right masseter and subcutaneous plane along with concomitant central nervous system relapse, 4 years after the primary diagnosis, with a brief review of the literature.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Arabi M; Ibrahim M; Camelo-Piragua S; Shah G
INSTITUCIÓN / INSTITUTION: Department of Radiology, University of Michigan Health System, Ann Arbor, MI, USA.
RESUMEN / SUMMARY: Neurenteric (NE) cysts are uncommon congenital cysts of endodermal origin. These cysts are commonly encountered in the posterior fossa surrounding the brain stem structures. We present a case of pathologically proven supratentorial NE cyst that mimicked a hydatid cyst in its clinical presentation and imaging appearance. Including this pathology in the differential diagnosis of supratentorial cystic lesions is important due to the differences in medical and surgical management.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Liu YQ; Yue JQ
INSTITUCIÓN / INSTITUTION: Department of Pathology, People’s Hospital of Yingcheng Yingcheng, Hubei, China.
RESUMEN / SUMMARY: Paraganglioma is a neuroendocrine neoplasm, which is extremely rare in the vulva and only one case has been reported. Here we present a case of vulvar paraganglioma in a 48-year-old woman and a literature review. The patient found a lump located in the genitals below the symphysis pubis 3 months before presentation when she complained that the lump was increasing in size. A 3.2 cm x 2.3 cm x 1.5 cm nodule was excised from subcutaneous soft tissue in the vulva. Microscopy showed a diversity of cell morphologies and structures in the rich vascular network of the tumor separated the chief cells into round cell nests (Zellballen pattern). Some areas of the tumor presented epithelioid and spindle-shaped cells with increased cell density and indistinct structural characteristics. Hyaline degeneration of collagen fibers or mucoid degeneration was found in tumor interstitium. Immunohistochemical staining showed diffused expression of synaptophysin in the chief cells, focal expression of S-100 protein in the sustentacular cells and high expression of CD34 in the vascular components. Based on morphological and immunohistochemical results, a rare paraganglioma of the vulva was diagnosed.
Involvement of tumor acidification in brain cancer pathophysiology.

Gliomas, primary brain cancers, are characterized by remarkable invasiveness and fast growth. While they share many qualities with other solid tumors, gliomas have developed special mechanisms to convert the cramped brain space and other limitations afforded by the privileged central nervous system into pathophysiological advantages. In this review we discuss gliomas and other primary brain cancers in the context of acid-base regulation and interstitial acidification; namely, how the altered proton (H+) content surrounding these brain tumors influences tumor development in both autocrine and paracrine manners. As proton movement is directly coupled to movement of other ions, pH serves as both a regulator of cell activity as well as an indirect readout of other cellular functions. In the case of brain tumors, these processes result in pathophysiology unique to the central nervous system. We will highlight what is known about pH-sensitive processes in brain tumors in addition to gleaning insight from other solid tumors.

Neurological manifestations of oculodentodigital dysplasia: a Cx43 channelopathy of the central nervous system?

GJs are formed by connexins of which Cx43 is most widespread in the human body. In the brain, Cx43 GJs are mostly found in astroglia where they coordinate the propagation of Ca2+ waves, spatial K+ buffering, and distribution of glucose. Beyond its role in direct intercellular communication, Cx43 also forms unapposed, non-junctional hemichannels in the plasma membrane of glial cells. These allow the passage of several neuro- and gliotransmitters that may, combined with downstream paracrine signaling, complement direct GJ communication among glial cells and sustain glial-neuronal signaling. Mutations in the GJA1 gene encoding Cx43 have been identified in a rare, mostly autosomal dominant syndrome called oculodentodigital dysplasia (ODDD). ODDD patients display a pleiotropic phenotype reflected by eye, hand, teeth, and foot abnormalities, as well as craniofacial and bone malformations. Remarkably, neurological symptoms such as dysarthria, neurogenic bladder (manifested as urinary incontinence), spasticity or muscle weakness, ataxia, and epilepsy are other prominent features observed in ODDD patients. Over 10 mutations detected in patients diagnosed with neurological disorders are associated with altered
functionality of Cx43 GJs/hemichannels, but the link between ODDD-related abnormal channel activities and neurologic phenotype is still elusive. Here, we present an overview on the nature of the mutants conveying structural and functional changes of Cx43 channels and discuss available evidence for aberrant Cx43 GJ and hemichannel function. In a final step, we examine the possibilities of how channel dysfunction may lead to some of the neurological manifestations of ODDD.