ACTA RADIOLÓGICA PORTUGUESA
September-December 2018 | Vol 30 nº 3 | 19-22

Radiological Case Report / Caso Clínico

Unusual Skull Base Lesion: Hemangiopericytoma of the Middle Cranial Fossa

Lesão Incomum da Base do Crânio: Hemangiopericitoma da Fossa Craniana Média

Carlos Bilreiro[1], Domingos Coiteiro[2], Manuela Mafra[3], Alexandra Borges[1]

[1]Instituto Português de Oncologia Francisco Gentil, Serviço de Radiologia. Lisboa, Portugal
[2]Centro Hospitalar Lisboa Norte, Serviço de Neurocirurgia. Lisboa, Portugal
[3]Centro Hospitalar Lisboa Central, Serviço de Anatomia Patológica. Lisboa, Portugal

Address

Carlos Bilreiro
Rua Giacomino Ferrari,
lote 56, -2B
8500-782 Portimão, Portugal
email: carlosbilreiro@hotmail.com

Abstract

Meningioma is the most common intracranial tumor affecting the skull base and the diagnosis is usually straightforward on imaging studies. However, other lesions may present similar findings.
A 60 year-old-man presented with memory loss and behavioral changes. Computed Tomography (CT) and Magnetic Resonance (MR) imaging disclosed an extra-axial, dural-based lesion of the middle cranial fossa. The lesion was strongly enhanced and of hypervascular nature, peripheral vessels and vasogenic edema of the adjacent brain parenchyma.
Although the most common dural-based lesion of the central nervous system (CNS) is the meningioma, radiologists should be aware of other differential diagnoses, particularly when facing atypical imaging features. These comprise metastases, lymphoma and leukemia, histiocytic lesions, sarcoidosis and hemangiopericytoma. Based on imaging findings we were able to suggest the hemangiopericytoma diagnosis and the patient was subject to tumoral resection surgery and adjuvant radiotherapy. A small tumoral residue remains stable on follow-up studies.

Keywords

Hemangiopericytoma; Meningioma; Solitary fibrous tumor; Skull base; Computed tomography; Magnetic resonance.

Resumo

O meningioma é o tumor intracraniano mais comum na base do crânio e o seu diagnóstico imagiológico é habitualmente claro, mas outras lesões podem apresentar-se com achados semelhantes.
Um homem de 60 anos apresentou-se com perda de memória e alterações comportamentais. Efectuou Tomografia Computorizada (TC) e Ressonância Magnética (RM) de crânio que demonstraram uma lesão extra-axial de base dural na fossa média. A lesão possuía forte realce com natureza hipervascular, vasos periféricos e edema vasogénico do parênquima cerebral adjacente.
Embora a lesão de base dural mais comum do sistema nervoso central (SNC) seja o meningioma, devem-se considerar outros diagnósticos diferenciais, particularmente na presença de características de imagem atípicas. Nestes incluem-se metástases, linfoma e leucemia, lesões histiocíticas, sarcoidose e hemangiopericitoma. Com base nos achados de imagem, sugerimos o diagnóstico de hemangiopericitoma e o doente foi sujeito a cirurgia de ressecção tumoral e radioterapia adjuvante, encontrando-se um pequeno resíduo tumoral estável nos estudos de seguimento.

Palavras-chave

Hemangiopericitoma; Meningioma; Tumor fibroso solitário; Base do crânio; Tomografia computorizada; Ressonância magnética.

Introduction

Meningioma, a benign tumor originating from arachnoid cap cells, is the most common intracranial tumor representing 33.8% of all primary brain tumors diagnosed in the United States.[1,2] Often incidentally discovered when investigating non-specific neurological symptoms, diagnosis is usually performed on cross-sectional imaging (CT and MR). However, other rarer meningeal-related lesions may be easily overlooked as they mimic meningioma features on imaging studies. Hemangiopericytoma is one such lesion, representing less than 0.4% of CNS tumors.[3]
This rare tumor has recently been reclassified as being the same entity as solitary fibrous tumor of the dura.[4,5] Usually, being a more aggressive lesion than a meningioma, with a high rate of local recurrence and possible distant metastases, it is crucial that the diagnosis is made as early as possible and that a close and prolonged follow-up is performed.[6]

Case report

A 60-year-old man presented with short and long-term memory loss and behavioral changes which had been slowly progressing in the last 2 years. Also, discreet personality changes were recently observed by close relatives. Family history was irrelevant. The remainder neurological and physical examination were only remarkable for slight hypoesthesia of the left face.
A non-contrast-enhanced CT of the brain revealed a large, spontaneously hyperdense extra-axial mass, in the left middle cranial fossa, with lobulated contours (Fig. 1), extending into the infratemporal fossa through the greater

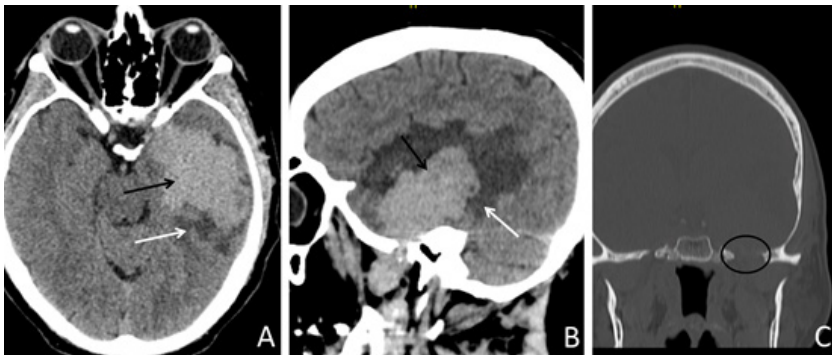

Figure 1 - Non-contrast-enhanced head CT scan, with multiplanar reformats on the axial (A), sagittal (B) and coronal (C) planes. A and B shows soft-tissue window and C shows bone window. There is a large apparently extra-axial mass in the left middle cranial fossa (black arrows), spontaneously hyperdense with a lobulated contour. The mass extends through the left central skull base, causing destruction of the greater sphenoid wing (C, black circle). Note the hypodensity of the surrounding brain parenchyma (white arrows), due to vasogenic brain edema.

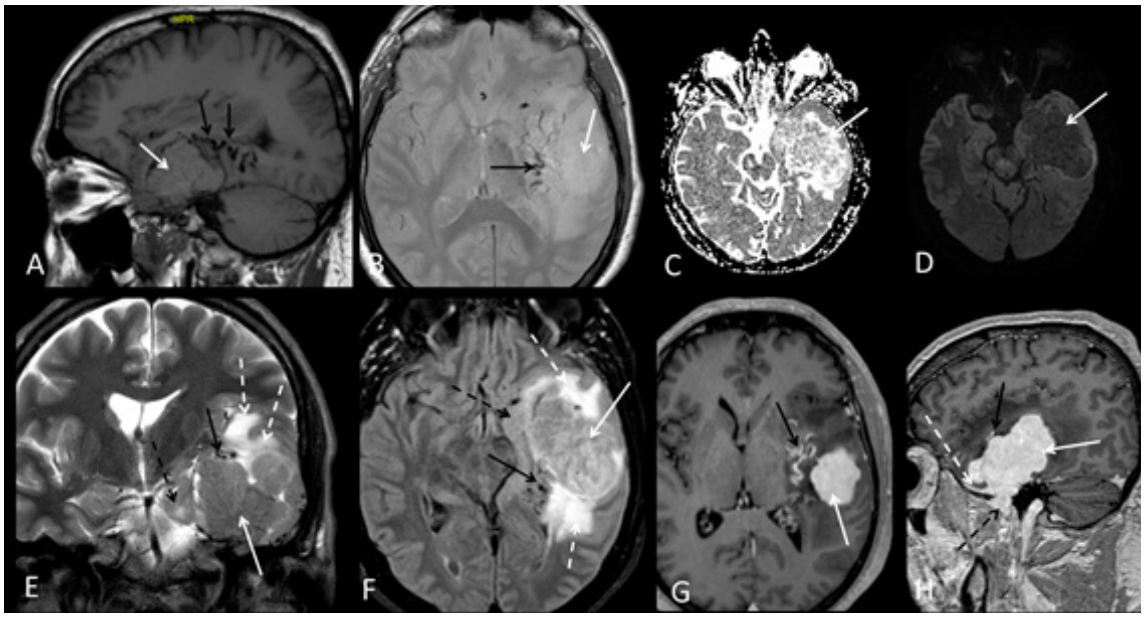

Figure 2 – Contrast-enhanced MR study. The lesion (white arrows) is discreetly hyperintense on sagittal T1-weighted images (A) and isointense to gray matter on axial Proton-density-weighted: PDW (B) images. On coronal T2-weighted images (E) and axial Fluid-attenuated inversion recovery: FLAIR (F) images, the lesion (white arrows) is isointense to gray matter. Note the extensive vascular flow voids surrounding the lesion (black arrows). There is mass effect with a rightward tilt of the mesencephalon, effacement of the ambiens cistern and inferior uncal herniation (E and F, black dashed arrows). The surrounding brain parenchyma reveals high signal intensity (E and F, white dashed arrows) due to vasogenic edema. On Diffusion-weighted imaging: DWI the lesion shows high ADC values (C, white arrow) and low signal intensity on b1000 (D, white arrow). Axial (G) and coronal (H) contrast-enhanced fat-suppressed T1-weighted images reveal intense and homogeneous enhancement (white arrows). The inferior extension to the infratemporal fossa and the surrounding neovascularization is clearly depicted (H, black dashed arrow). Also note the linear enhancement or the surrounding dura - "dural tail" sign (H, white dashed arrow).

sphenoid wing and foramen ovale. The lesion showed mass-effect and vasogenic edema of the surrounding temporal lobe.

On MR, the lesion was discreetly hyperintense on T1-weighted and isointense to gray matter on T2-weighted images (Fig. 2). Multiple vascular flow-voids were seen both at the periphery and central core of the lesion, representing prominent branches from the middle and posterior left cerebral arteries. The surrounding brain parenchyma showed vasogenic edema. After gadolinium administration, strong homogeneous enhancement and a "dural tail" sign were seen. There was no restricted diffusion. Additional MR spectroscopy (MRS) showed a myoinositol peak and no alanine peak. With a presumed diagnosis of hemangiopericytoma, the patient underwent a chest CT and a Positron emission tomography (PET-CT), which were both negative.

Surgical resection of the tumor was performed through a left temporal craniotomy. On histopathology the tumor was composed of polygonal cells arranged in a solid pattern (Fig. 3A). Focally, some collagen fibers were seen (Fig. 3B). The tumor was highly vascularized with numerous capillary blood vessels, often with 'staghorn' configuration (Fig. 3C). The neoplastic cells had indistinct cytoplasmic contours and round to oval nuclei. Three mitotic figures in 10 high-power fields were counted (Fig. 3D). Immunohistochemistry revealed reactivity with CD34 (Fig. 3E), BCL2 (Fig. 3F) and vimentin and no expression of EMA, S100 protein or progesterone receptor. Diagnosis of hemangiopericytoma (WHO Grade II) was made.

Post-surgical MR disclosed complete resection of the intracranial component and a 12-mm tumor residue adjacent to foramen ovale and upper masticator space. The patient received adjuvant radiotherapy (SBRT), in a total dose of 50Gy (2,5Gy/session).

Upon treatment completion, the patient maintained short memory deficit and hypoesthesia of the left face. After a two-year follow-up the small tumor residue remained unchanged with no signs of local recurrence. Also, no distant metastases were noted on serial PET-CT and chest CT.

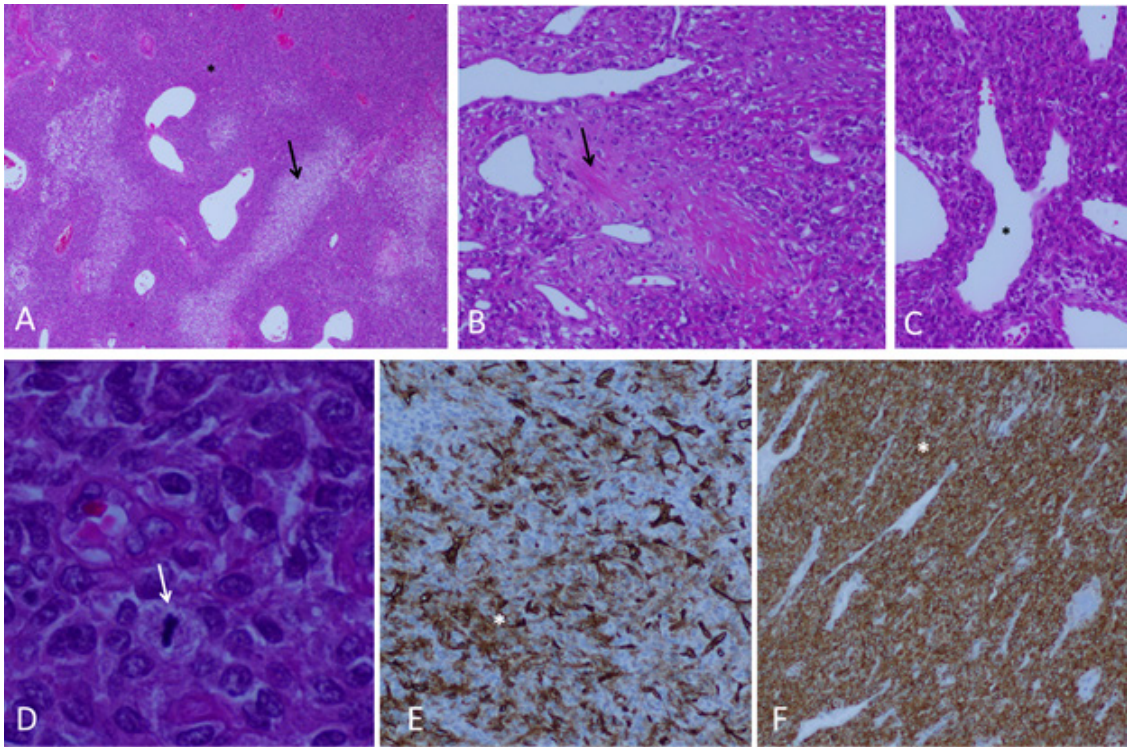

Figure 3 – Lesion histology, Hematoxylin and eosin stain: H&E (A 20x, B and C 200x). There is compact tumor, alternating hypo (arrow) and hypercellular (asterisk) areas (A). Note focal collagen deposition (B, arrow) and the "staghorn" vessel (C, asterisk). D (H&E 400x) shows a typical mitotic figure (arrow). CD34 immunohistochemistry (E, 100x) shows immunoreactive neoplastic cells in some areas (asterisk), with a rich vascular network. There is also BCL2 immunoreactivity in neoplastic cells (F, 100x, asterisk).

Discussion

The imaging diagnosis of dural-based lesions is perceived as straightforward, although this is not always the case.

Typical imaging features of meningioma: "dural tail sign", regular contours and adjacent bone hyperostosis, signal intensity similar to gray matter on T1-weighted and T2-weighted MR images along with vivid contrast enhancement, should be enough to confidently diagnose a meningioma in an otherwise "healthy" patient. However, in a patient suffering from other co-morbidities, especially a malignancy, a high index of suspicion for dural metastasis must be maintained, even if the lesion looks "typical".

In the case reported, working diagnoses were atypical meningioma and hemangiopericytoma. Both lesions appear on imaging studies as extra-axial masses with a broad dural base and "dural tail" sign, with signal intensity similar to gray matter, often absent restriction on diffusion-weighted images and homogeneous contrast enhancement.

Imaging features favoring hemangiopericytoma included the lobulated contours, peri-lesional neovascularization, lytic bone destruction of the central skull base and a high myoinositol peak without alanine peak, on MRS.[6,7]

Other lesions which can present themselves as broad dural-based masses are summarized on table 1.

Hemangiopericytoma is a rare neoplasm thought to arise from the Zimmermann pericytes surrounding capillaries and postcapillary venules that is more common in musculoskeletal system and skin and represents less than 0.4% of CNS tumors.[6] Its typical age of appearance is 38-42 years, while meningiomas occur frequently in slightly older patients (50 year-olds).[6,8] These are highly cellular tumors, composed of closely packed cells with scant cytoplasm and interrupted by gaping capillary caliber vessels with a branching "staghorn" pattern, as shown in the present case (Fig. 3A-D). Hemangiopericytomas can show malignant behavior, with metastases occurring in 20% of the cases, usually in the liver, lung and bone.[6,9] Recurrence rates are high and there is a known long-term risk for metastatic disease.[4] Nowadays, it is considered part of a spectrum of lesions, as described in The 2016 World Health Organization Classification of Tumors of the Central Nervous System, which attributed 3 grades for the combined entity hemangiopericytoma/solitary fibrous tumor: grade I (solitary fibrous tumor); grade II (hemangiopericytoma) and grade III (anaplastic hemangiopericytoma).[4] Currently, immunophenotypic analysis, assessing for STAT6 expression and/or NAB2-STAT6 fusion, further confirms the diagnosis.

Recommended therapy is surgical resection and preoperative embolization may be considered, due to the high vascularization of the tumor. These are followed by adjuvant radiotherapy in all WHO grade III and incompletely resected WHO grade II tumors to prevent recurrence (such as in the present case report). Long-term follow-up is recommended for early detection of local recurrence and/or distant metastases.

Conclusion

Meningeal hemangiopericytomas are dural-based lesions with malignant behavior that should not be overlooked on imaging studies. A high degree of suspicion along with the clinical history may help the diagnosis. Imaging features mimic those of the much more common meningioma but the presence of lobulated contours, vasogenic brain edema, strong arterial-like enhancement, presence of multiple flow voids, adjacent bone erosion and a high myoinositol peak on MRS allow a pre-operative diagnosis and adequate patient management. Therapy consists of surgical resection and adjuvant radiotherapy.

Table 1 – Differential diagnosis of mass lesions with a broad dural base

Mass lesions with broad dural base	Distinctive features
Meningioma	- hemispheric, well-defined borders - "dural tail" sign - signal intensity of gray matter on all pulse sequences - presence of calcifications - adjacent bone hyperostosis - homogeneous enhancement - alanine peak on MRS
Hemangiopericytoma/ solitary fibrous tumor	- lobulated contours - neovascularization (flow voids) - no calcifications - local aggressiveness (vasogenic edema of adjacent brain) - lytic erosion of adjacent bone - vivid, homogeneous, arterial-like enhancement (on DCE-PWI) - myoinositol peak on MRS
Dural metastasis	- irregular margins - high T2W signal intensity - restricted diffusion - usually multiple - most frequently from breast, lung and prostate cancer
Primary dural lymphoma	- indistinct brain-tumor interface - low T2W signal intensity - restricted diffusion (mADC< 1x10-3mm2/s)
Rosai-Dorfman disease	- iso/hypo T2W signal intensity (often with central hypointensity) - adjacent brain edema - cervical lymphadenopathy (common) - fever, night sweats and weight loss
Erdheim-Chester disease	- iso/hypo on T2W signal intensity - associated involvement of facial bones and orbits (common)
Melanocytic neoplasms	- high T1W signal intensity - iso/hypo T2W signal intensity - susceptibility artifact on T2*/ SWI
Neurosarcoidosis	- iso/hypo T2W signal intensity - diffuse meningeal thickening or a focal mass - often associated to systemic sarcoidosis

Abbreviations: MRS: magnetic resonance spectroscopy; DCE-PWI: dynamic contrast-enhanced perfusion weighted imaging; T2W: T2 weighted; T1W: T1 weighted; mADC: mean apparent diffusion coefficient; SWI: susceptibility weighted imaging.

Received / Recebido 19/07/2018
Acceptance / Aceite 01/09/2018

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.
Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.
Financing Support: This work has not received any contribution, grant or scholarship.
Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.
Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.
Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.
Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).
Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

References

1 - Claus EB, Bondy ML, Schildkraut JM, et al. Epidemiology of intracranial meningioma. Neurosurgery. 2005;57:1088-95.
2 - Wiemels J, Wrensch M, Claus EB. Epidemiology and etiology of meningioma. J Neurooncol. 2010;99:307-14.
3 - Giannini C, Rushing EJ, Hainfellner JA. Haemangiopericytoma. In: WHO classification of tumours of the central nervous system. Lyon, France:IARC,2007;178-180.
4 - Louis DN, Perry A, Reifenberger G, et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. Acta Neuropathol. 2016;131: 803-20.
5 - Schweizer L, Koelsche C, Sahm F, et al. Meningeal hemangiopericytoma and solitary fibrous tumors carry the NAB2-STAT6 fusion and can be diagnosed by nuclear expression of STAT6 protein. Acta Neuropathol. 2013;125:651-658.
6 - Smith AB, Horkanyne-Szakaly I, Schroeder JW, et al. From the radiologic pathology archives: mass lesions of the dura: beyond meningioma-radiologic-pathologic correlation. Radiographics. 2014;34:295-312.
7 - Barba I, Moreno A, Martinez-Pérez I, et al. Magnetic resonance spectroscopy of brain hemangiopericytomas: high myoinositol concentrations and discrimination from meningiomas. J Neurosurg. 2001;94:55-60.
8 - Liu G, Chen ZY, Ma L, et al. Intracranial hemangiopericytoma: MR imaging findings and diagnostic usefulness of minimum ADC values. J Magn Reson Imaging 2013;38(5): 1146–1151.
9 - Chiechi MV, Smirniotopoulos JG, Mena H. Intracranial hemangiopericytomas: MR and CT features. AJNR Am J Neuroradiol. 1996;17:1365-71.