Benign extra-axial tumor complication – Avascular necrosis of a meningioma

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ABSTRACT

Intra-tumoral infarction is a rare complication in patients with meningioma. We report a case of a 36-year-old woman with a diagnosis of osseous cranial frontal and parietal dysplasia associated with multiple extra-axial lesions suggestive of meningiomas, that acutely developed headache, right-sided weakness, obnubilation and motor aphasia with progressive worsening. Head Computed Tomography and brain Magnetic Resonance Imaging were suggestive of intra-tumoral meningioma infarction, with a central area of necrotic-core and peripherical concentric contrast enhancement as well as signs of intracranial hypertension. The patient was submitted to surgery to remove the lesion, confirming the suspected diagnosis by histopathology analysis.

CASE REPORT

CASE REPORT

The patient was a 36-year-old woman that developed headache, right-sided weakness, obnubilation and motor aphasia with progressive worsening for the past two days. Her medical history includes a diagnosis of osseous cranial frontal and parietal dysplasia, associated with multiple extra-axial lesions suggestive of meningiomas as well as subsequent superior sagittal sinus bony obliteration.

First head-TC was unremarkable (Figure 1A,1B) despite the known pathology – with a MRI study performed two months earlier (Figure 1C,1D), but new imaging studies performed next days were suggestive of intra-tumoral meningioma infarction, with a central area of necrotic-core and peripherical concentric contrast enhancement (Figure 2), accompanied by signs of intracranial hypertension. The patient experienced clinical deterioration and the lesion was removed, allowing histopathology analysis to confirm the suspected diagnosis.

IMAGING FINDINGS

Due to clinical worsening, head CT was repeated two days after hospitalization, revealing tomodensitometric modification of one of the left frontoparietal meningioma (with 32 x 20 x 19mm of respective craniocaudal, transverse and anteroposterior diameter), with marked increase of subcortical oedema and intra-tumoral necrotic/cystic degeneration.

Later that day, she underwent brain MRI examination (Figure 3), which confirmed the signal changes of the suspected lesion with isointense T1 and FLAIR core and marked peripheral enhancement. It also showed T2 and fluid attenuated inversion recovery (FLAIR) hyperintensity on surrounding frontoparietal subcortical white matter, compatible with edematous changes (that were not present in the previous studies). The core of the lesion also demonstrated slightly b1000 hyperintensity with no ADC signal loss, which was suggestive of subacute infarction. The remaining lesions showed no changes compared with the previous imaging studies.

At that time, the main differential diagnosis was intra-tumoral infection/abscess, mainly regarding the contrast-enhancement pattern.

After MRI examination, there was clinical decline, so the patient was submitted to a resection surgery. At the time, it was performed an extemporaneous examination, which revealed necrotic changes of the left frontoparietal meningioma. The remaining specimen was sent to the Pathology department.

Management / Operative Report

Neuronavigational guided surgery was performed, namely with intraoperative ultrasound, for excision of the convexity and left parasagittal meningiomas, through parasagittal incision. After an initial careful definition of venous and bony structures, the meningioma of the lateral convexity was completely removed.

Pathology

The Pathology department received a surgical specimen representative of a meningothelial neoplasia, composed of spindle cells organized in interlacing bundles disposed in a collagenous-rich matrix (Figure 3A). Necrosis was identified (Figure 3B), and focal invasion of the trabecular bone represented was present. The mitotic index was low (<1 mitotic figures in 10 consecutive high-power fields). There was no documentation of brain tissue, and there were no histological criteria suggesting aggressive potential. Hence, a diagnosis of fibrous meningioma (grade 1) was established.

Follow-up

After the procedure the patient remained in the intensive unit care due to a pneumonia (which delayed ventilatory weaning) and acute urinary retention. The patient remained with mild motor aphasia (but capable of communication) and mild hemiparesis of the right arm and leg, with global muscular atrophy. During the hospital stay, physical therapy was initiated.

DISCUSSION

Etiology and demographics

Meningiomas are the most frequent intracranial tumor, accounting for more than a third of all primary central nervous system (CNS) neoplasms and approximately 50% of all benign brain tumors [1]. Despite having a reputation of a benign disease, patient outcome varies from benign to highly aggressive, presenting with a wide variety of non-specific, location dependent symptoms. The incidence increases with age and females are more commonly affected than males, with a ratio of approximately 2 to 1 [1-3].

The 2021 WHO Classification recognizes fifteen phenotypes: nine benign (grade 1), three intermediate-grade (grade 2) and three malignant (grade 3), based on histopathology [4,5]. These dural-based tumors are usually benign, slow growing tumors that are considered to arise from meningothelial (arachnoid) cells [1,2]. WHO CNS endorses the use of molecular biomarkers for classification and malignancy grading3, since pathology is a strong prognostic factor, critical both for therapeutic strategies and follow-up plans [2,3,5]. However, it is not required for the diagnosis, if definitive histopathology is present. Finally, the proliferation rate varies with the morphology grading, and it is typically low for grade I tumors (<4%), with significant prognostic value [6].

Grade I meningiomas are more likely to be found at the skull base. Convexity meningiomas comprise fibroblastic and transitional subtypes, most commonly CNS WHO grade 2 and 3. Hyperostosis associated with intracranial meningiomas is a well-described entity, but there is still some disagreement related to the cause effect of hyperostotic changes (secondary to the tumor formation versus bone invasion) [7,8].

Clinical and imaging findings

While most often benign and with typical imaging appearance of unilobed well demarcated extra-axial masses (with iso to hypointense signal on T1 and iso to hyperintense signal on T2), meningiomas may also show an heterogenous pattern in approximately 15% of cases. [1,8,9] Atypical imaging features include cyst formation, intramural hemorrhage or necrosis, which may be associated with more aggressive behavior of the tumor (namely higher rate of recurrence and worse prognosis).

Their presentation is often non-specific, with insidious unset and features of raised intracranial pressure (ICP), seizure and focal neurologic deficits (including cranial nerve deficits), owing to the location and compression of adjacent parenchyma and vascular structures [1]. Although uncommon, acute presentation is also possible: occasionally with intratumoral bleed or exceedingly rare, as an acute ischemic stroke, mainly due to vessel compression, hypoperfusion and/or thromboembolism [1,10-12].

Despite of being well known the association between meningiomas, necrosis and, on the other hand, acute ischemic stroke, there are few case reports of pathology-proven spontaneous necrosis with focal neurological deficits, compatible with an intralesional stroke without large vessel involvement. Moreover, the prominent ipsilateral hyperostotic changes in a young patient provide a more unique character to the clinical case.

Treatment and prognosis

The management of a patient with an asymptomatic meningioma usually involves a conservative approach [13]. If the patient is symptomatic, for example in a case of an infarcted meningioma, the initial management includes stabilization; steroids are usually not effective in reducing peritumoral edema, comparing to malignant tumors but can still be employed. The final step includes surgical resection. Due to the known diagnosis of hyperostosis, although firstly steroids were administered, due to clinical worsening (and keeping in mind that our patient was particularly susceptible to intracranial hypertension due to the hyperostotic changes), surgical approach was considered in an early phase.

Differential diagnosis

This atypical presentation may also be misinterpreted as a stroke mimic like a brain abscess or a malignant tumor, enhancing the importance of an accurate differential diagnosis. With only 15 cases described in the literature, intra-tumoral abscess was the main concern, namely due to peripheral ring-enhancement pattern. Keeping in mind some microenvironmental predisposing factors, like venous congestion (that was present in this case), a known cause of bacteriemia is almost always identified, but it was not the case in this patient. Additionally, MRI diffusion restriction was different from expected in relation to an abscess (which was expected to document low ADC values).

Intra-tumoral infarction may differ from a parenchymal ischemic lesion due to necrosis progression following infarction (that do not occur in parenchymal lesions), which can explain the differences on diffusion restriction within the tumor, which was less conspicuous than normally seen and may actually be absent. Besides, infarction may also justify an incremental evolution of vasogenic edema (although the cause of this process is not well-kown), contributing to the clinical symptoms at presentation, as it was seen in our case.

TEACHING POINT

We report and discuss thereby the work-up using neuroimaging techniques, resective surgery and histopathology. The purpose of the present article is to highlight the imaging features of meningioma intra-tumoral infarction, which is by itself a rare entity, as well as emphasize the role of magnetic resonance imaging (namely DWI) to establish the correct diagnosis and exclude other differentials, namely intra-tumoral abscess.

Authors' contributions

Inês Prisco and Sofia Vedor wrote all the manuscript sections except the Pathology description. Ana Rita Coelho and Roberto Silva provided pathology report and sample description. Gonçalo Alves reviewed the whole manuscript.

QUESTIONS

QUESTION 1: Which of the following is false regarding meningiomas?

1. They are more common in females with a 2:1 ratio.
2. It is the most common intracranial tumor.
3. They are dural-based tumors arising from meningothelial cells.
4. Since they are typically benign, pathology correlation is usually not necessary for therapeutic strategies. (applies)
5. Atypical imaging features are present in about 15% of cases.

Explanation for question 1:

5. [WHO CNS endorses the use of molecular biomarkers for classification and malignancy grading3, since pathology is a strong prognostic factor, critical both for therapeutic strategies and follow-up plans]

QUESTION 2: What is the main differential diagnosis of a smooth ring enhancement lesion within a known benign tumor?

1. Malignant degeneration
2. Venous sinus thrombosis
3. Intra-tumoral abscess (applies)
4. Arterial ischemic stroke
5. Incremental size of the lesion

Explanation for question 2: [At that time, the main differential diagnosis was intra-tumoral infection/abscess, mainly regarding the contrast-enhancement pattern.]

QUESTION 3: Intra-tumoral abscess formation is usually associated with:

1. Arterial ischemic stroke
2. A known cause of bacteriemia.
3. Systemic hypoperfusion
4. Intracranial venous hyperperfusion
5. Trauma

Explanation for question 3:

2. [Keeping in mind some microenvironmental predisposing factors, like venous congestion (that was present in this case), a known cause of bacteriemia is almost always identified]

QUESTION 4: Regarding intra-tumoral infarction management, which of the following is false:

1. Patients usually develop new symptoms due to edematous changes related to the infarction.
2. In asymptomatic patients, conservative approach is the first approach.
3. Steroids are very effective in reducing peritumoral edema just like in malignant lesions.
4. Surgical approach is the last resource of management.
5. Pathology is mandatory to confirm the suspected diagnosis.

Explanation for question 4:

2. [steroids are usually not effective in reducing peritumoral edema, comparing to malignant tumors but can still be employed]

QUESTION 5: What is the most sensible MRI sequence to differentiate intra-tumoral infarction from abscess?

T1-weighted imaging.
FLAIR weighted imaging.
T2*/SWI-weighted imaging.
DWI and ADC map
T2/DP-weighted imaging.

Explanation for question 5:

2. [Additionally, MRI diffusion restriction was different from expected in relation to an abscess (which was expected to document low ADC values).]

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FIGURES

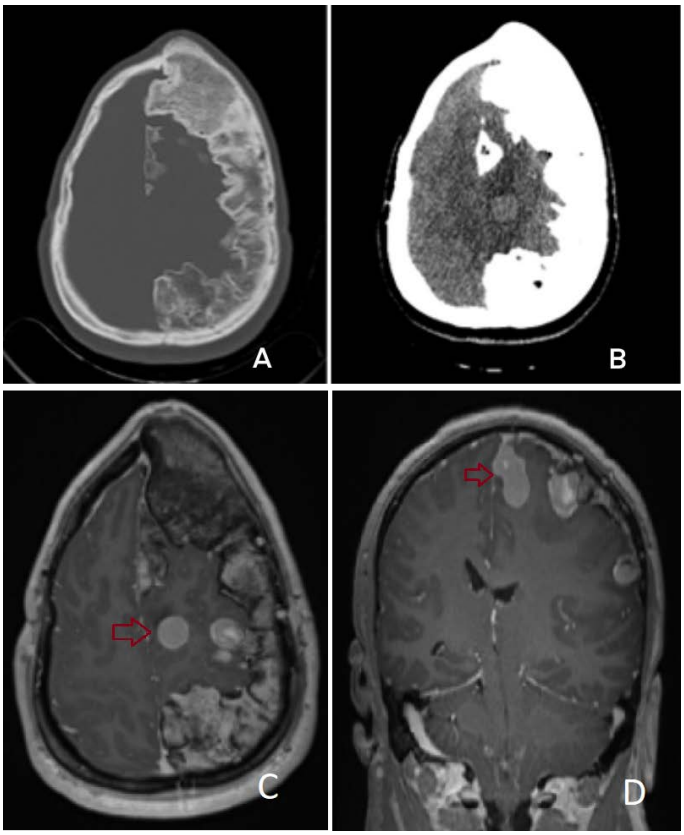

Figure 1: Female, 36 years-old, Basal Head CT showing known diffuse left convexity hyperostosis (A) with a spontaneously hyperdense left parasagittal meningioma (B) with homogeneous enhancement on MRI. (C, D). MRI (1.5 Tesla) images (axial T1-weighted images) also documents other known extra-axial homogenously enhancing lesions, compatible with meningiomas.

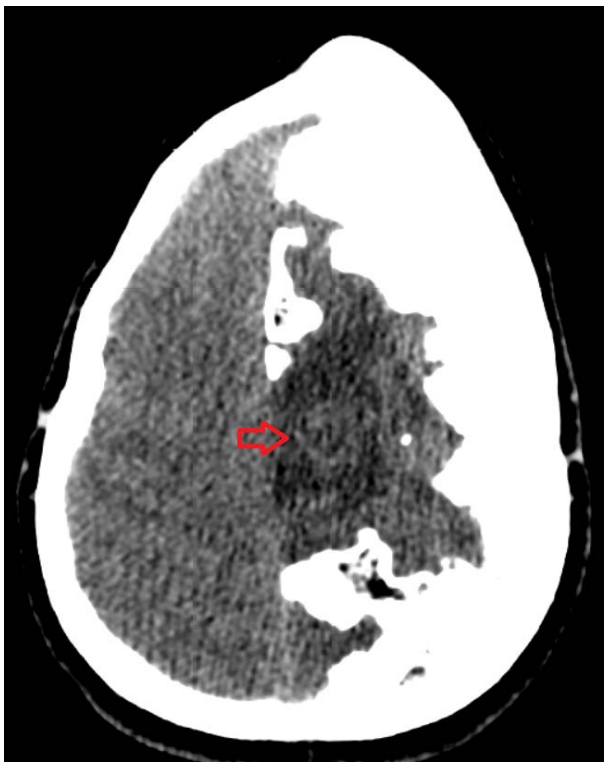

Figure 2: Female, 36 years old. Head CT (day 2 after hospitalization): necrotic/cystic degeneration of the meningioma core, with oedema on the frontoparietal subcortical white matter and higher mass effect, with attenuation of the adjacent cortical sulcus.

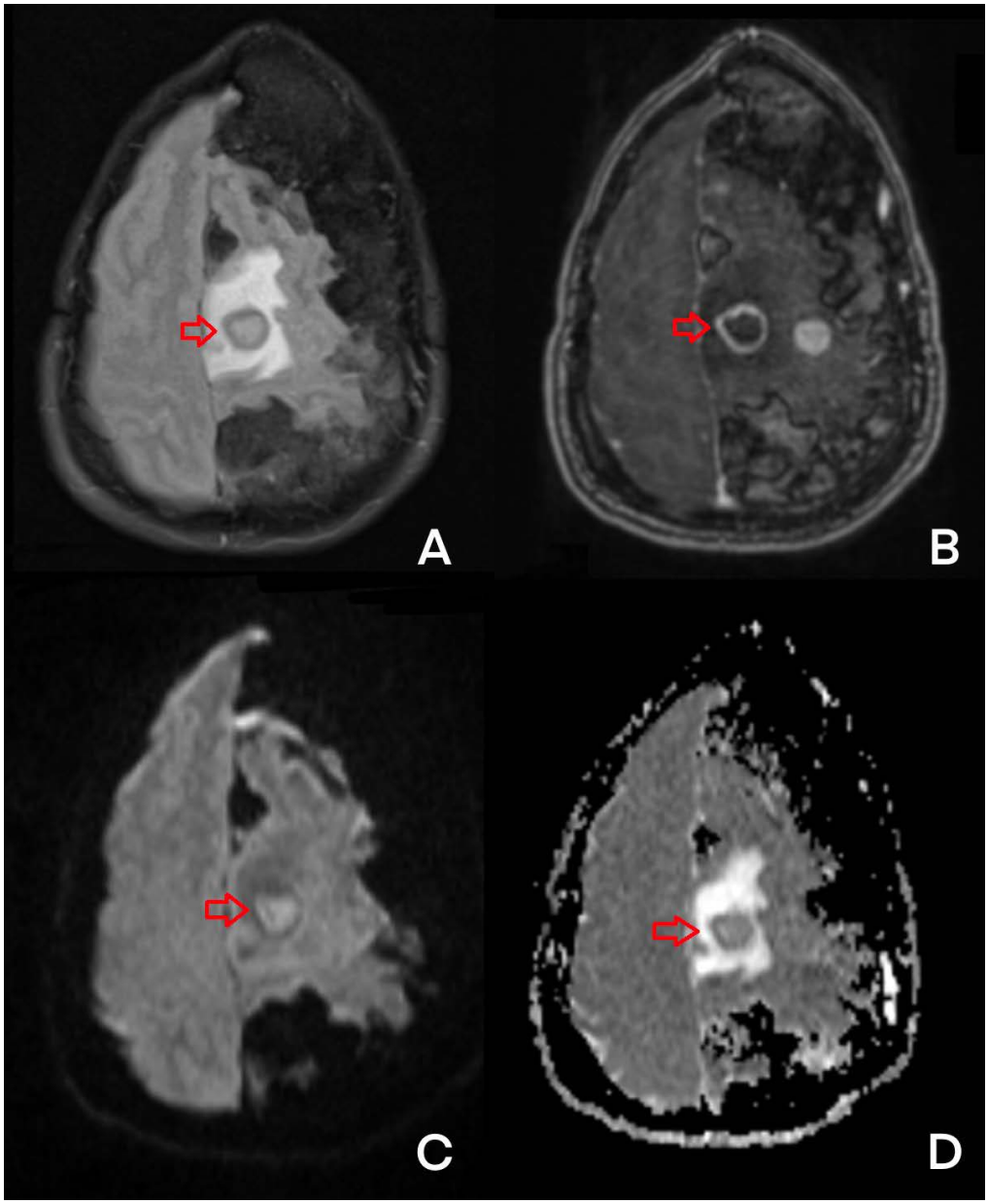

Figure 3: Female, 36 years old. Multiple MRI (1.5 Tesla) axial images show loss of signal of meningioma on T2-weighted FLAIR imaging (A). The lesion demonstrates peripheral ring enhancement (with an additional known left frontoparietal meningioma) (B) with slightly central b1000 hyperintensity (C) with no ADC signal loss (D).

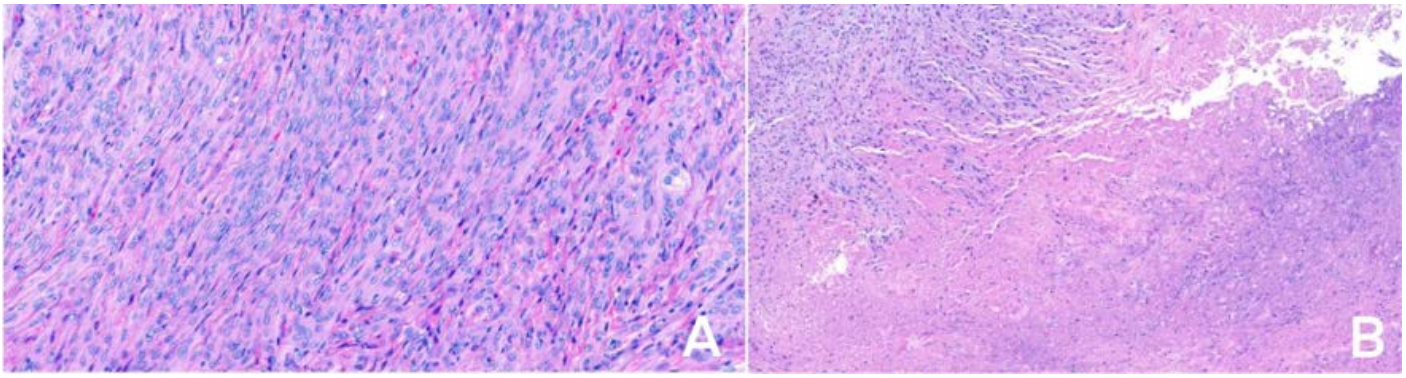

Figure 4: Female, 36 years old. H&E sections of the resected specimen illustrate spindle cells organized in interlacing bundles disposed in a collagenous-rich matrix (A), as well as necrosis signs (B).

KEYWORDS

Meningioma; Infarction; MRI; Diffusion; Tumor oesophagus

ABBREVIATIONS

H&E: HEMATOXYLIN AND EOSIN
CT: COMPUTED TOMOGRAPHY
MRI: MAGNETIC RESONANCE IMAGING