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Case Report

Intraparenchymal meningioma in the parieto-occipital region: A case report of a diagnostically challenging tumor

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ABSTRACT

Background: Intraparenchymal meningioma is a rare entity of one of the most common brain tumors. It is challenging to diagnose preoperatively due to the vague clinical presentation and absence of stereotypical radiological features. These atypical features might mislead the differential to favor high-grade gliomas or brain metastasis.

Case Description: We describe a case of a 46-year-old male who presented with vertigo, right-sided sensorineural hearing loss, and bilateral blurred vision. Contrast-enhanced magnetic resonance imaging of the brain revealed a large parieto-occipital contrast-enhanced mass with a multi-loculated cystic component and diffusion restriction but without dural attachment. A gross total reaction was achieved, and the histopathological results yielded a World Health Organization Grade I meningioma diagnosis. The patient exhibited no signs of recurrence after 2 years of follow-up.

Conclusion: Intraparenchymal meningiomas are difficult to identify without histopathological assessment. We emphasize the importance of considering this diagnosis when outlining an initial differential as it may direct management planning. Total surgical resection is the best treatment modality for such cases; however, radiotherapy is a valuable option. The prognosis of intraparenchymal meningiomas is generally favorable.

Keywords: Differential diagnosis, Intraparenchymal, Meningioma, Subcortical meningioma, Transitional meningioma

INTRODUCTION

Meningiomas are the most common nonmalignant central nervous system (CNS) tumors representing 53.9% of all non-malignant cases and 37.9% of all CNS tumors.[11] Comprising 26.99% of all adult primary CNS tumors, meningiomas are the most common single tumor entity in adults from Saudi Arabia.[1] Classically, meningiomas are extra-axial tumors originating from the dura mater and displaying dural attachment on radiographic imaging.[9] In 1938, Cushing and Eisenhardt classified meningiomas without dural attachment into plexus choroidal tumors, telae choroidal tumors, deep sylvian psammomeningiomas, and extra-cerebellar psammomeningiomas.[3] Later, this classification was revised, and among other changes, intraparenchymal and subcortical meningiomas were included as part of the classification.[22]

Brain tissue surrounding a meningioma is the defining feature of intraparenchymal meningiomas. Hence, they lack the classical signs of dural attachment on imaging.[7] The classical appearance of meningiomas on imaging is imperative in making the preoperative diagnosis of meningiomas. However, intraparenchymal meningiomas display unusual features that can mimic other types of tumors, completely changing the differential diagnosis of the lesion.[5] Intraparenchymal meningiomas are a rare entity with an unclear etiology. Here, we report the first case of a primary intraparenchymal meningioma in the middle east, describing clinical features, preoperative and postoperative imaging, and histopathologic features.

CASE REPORT

Presentation

A 46-year-old male presented to the otolaryngology clinic complaining of vertigo, right-sided hearing loss, and ear discharge for 5 months. Contrast-enhanced computed tomography brain showed a parieto-occipital lesion with a well-demarcated enhanced capsule consisting of mixed solid and cystic parts [Figure 1]. The patient was planned for further investigations. However, 3 weeks later, he presented to the emergency department with blurred vision for a few hours. His medical history is remarkable for lower limb paraplegia due to a road traffic accident 20 years before presentation and a mild, dull, on-and-off headache for years. His examination was remarkable for the right-sided sensorineural hearing loss and bilateral blurred vision. The patient was admitted for further investigation.

Imaging

Contrast-enhanced magnetic resonance imaging (MRI) brain showed a large, complex solid, and cystic intra-axial mass centered in the right parietal and occipital lobes. Diffusion restriction was noted in the solid component and the walls of the cystic part. A large solid enhancing component was seen in the inferior and posterior margins of the mass. The cystic component had multiple loculations with a thick and irregular wall enhancement. The mass exerted a local mass effect with effacement of the cortical sulci and surrounding vasogenic edema, as well as compression on the posterior horn of the right lateral ventricle with a subtle midline shift to the left side [Figure 2]. These findings were suggestive of high-grade glioma. The differential diagnosis included glioblastoma multiforme, anaplastic oligodendroglioma, and gliosarcoma. Hence, the patient was planned for a craniotomy and resection of the tumor.

Surgical resection

A right parietal-occipital craniotomy was performed, exposing the dura overlying the tumor, which appeared unremarkable. The lesion started to bulge out upon opening of the dura. Multiple specimens were taken for permanent histopathology. Internal debulking of the lesion started using suction and bipolar. The lesion was soft and greyish and highly vascular. However, there was a decent plane between the lesion and the surrounding parenchyma with some areas of invasion. There was no attachment to the adjacent falx cerebri, and no dural blood vessels supplying the tumor were identified intraoperatively. The lesion was then dissected circumferentially in the brain tissue following the plane, and a gross total resection was achieved.

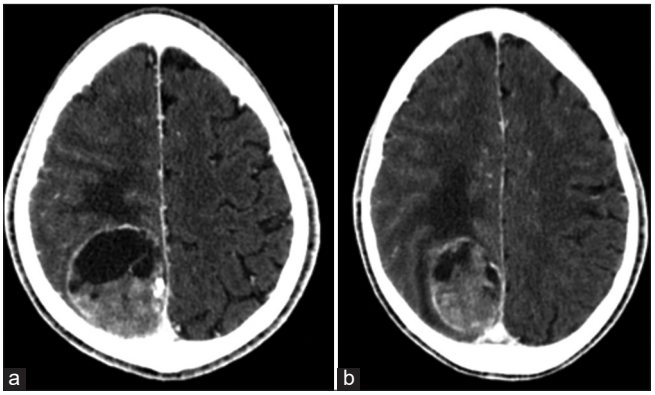

Figure 1: A contrast-enhanced axial brain computed tomography image (a and b) showing a right parieto-occipital lesion, well-demarcated, and enhanced capsule consisting of solid and cystic parts with perifocal edema and subtle midline shift to the left.

Histopathology

Microscopic examination of hematoxylin and eosin stained sections shows a proliferation of neoplastic cells, arranged in whorls and fascicles, with prominent vascular spaces and microcystic pattern. Psammoma bodies were identified, and no atypical features were seen. Fragments of brain parenchyma were identified without evidence of invasion by tumor cells; however, infiltration of the dura was seen. In immunohistochemistry, epithelial membrane antigen and progesterone receptor were positive. Glial fibrillary acidic protein was negative with Ki67 <2% proliferative index [Figure 3]. Thus, the diagnosis of the WHO Grade I transitional meningioma with no invasion of the surrounding brain structures was made.

Follow up

The patient's symptoms started to improve by the 2nd day after the surgery and his headache fully resolved. Aside from the above-mentioned right-sided sensorineural hearing loss, his postoperative examination was only remarkable. MRI brain with and without contrast showed no evidence of residual tumor. On discharge, the patient had a modified Rankin scale score of one. Brain MRI with and without contrast was

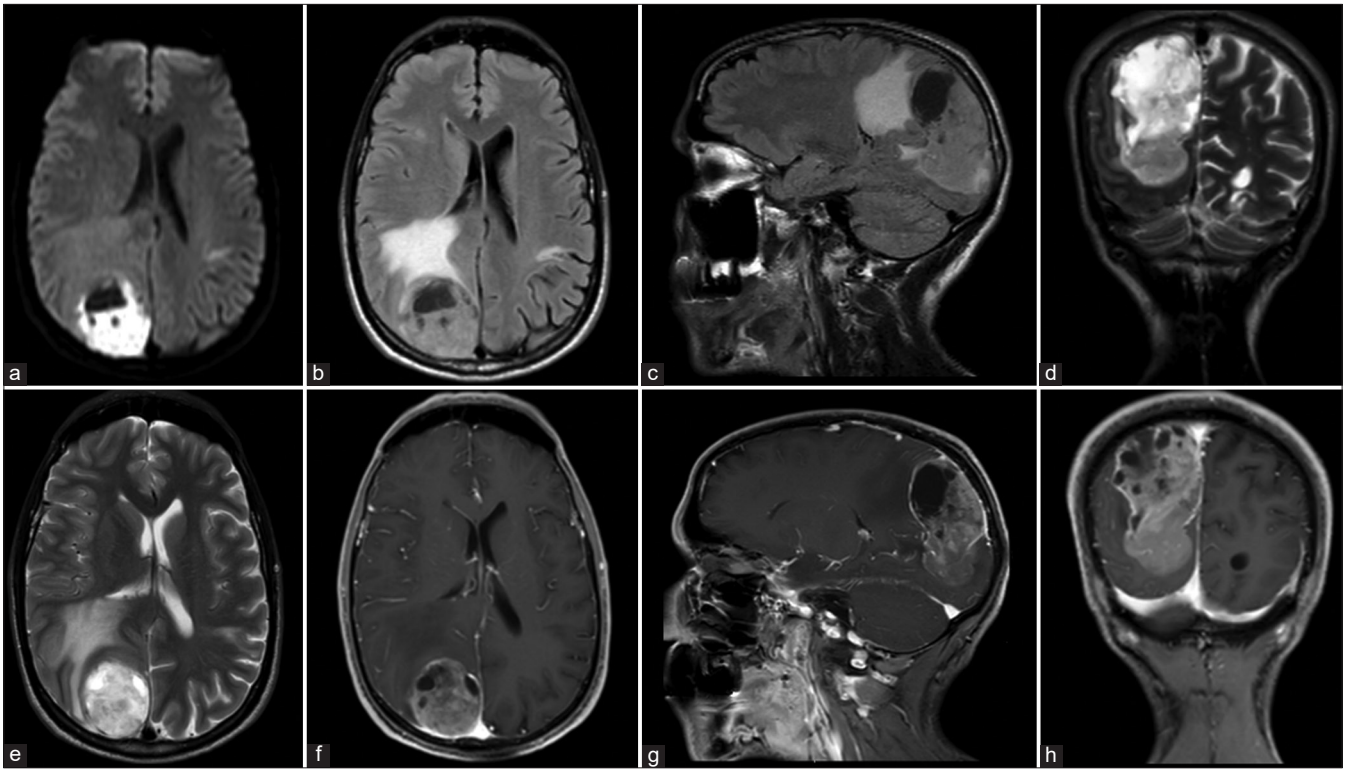

Figure 2: (A-H) Brain magnetic resonance imaging was done preoperatively. (a) axial diffusion-weighted image showing diffusion restriction in the solid part of the lesion. Fluid attenuated inversion recovery sequence images in (b) axial and (c) coronal planes, in addition to (d) coronal and (e) axial T2-weighted images showing a large solid and cystic intra-axial mass in the right parieto-occipital region with surrounding vasogenic edema. (f) axial, (g) sagittal, and (h) coronal gadolinium-enhanced T1-weighted image demonstrating an enhanced lesion and multiple loculations in the cystic component with a thick irregular enhancement of the wall.

repeated 2–3 months later and showed no evidence of residual disease or tumor recurrence [Figure 4]. On his most recent visit 2 years after surgery, the patient remains well and has no progression of his symptoms, unremarkable neurological examination, with no signs of recurrence or complications.

DISCUSSION

Meningiomas are the most common primary tumors in the CNS comprising over half of the benign brain tumors in adults.[14] They are slow-growing tumors originating from the arachnoid cap cells in the dura matter, which is recognized by the cytoarchitectural similarities with the meningioma.[10] Intraparenchymal meningioma is a rare entity identified as a meningioma that arises within the brain parenchyma without attachment to the dura.[22] Cushing and Eisenhardt first described this phenomenon as a meningioma lacking dural attachment, yet it can extend to the brain surface.[3] The terms intraparenchymal meningioma and subcortical meningioma are often used interchangeably by many due to shared characteristics between the two types.[7,17,22] A consensus over the etiology behind intraparenchymal meningiomas has yet to be reached and, to this day, remains unclear. However, it has been hypothesized that arachnoid cap cells of the pia matter migrate to the parenchyma through penetrating blood vessels during development. Another theory suggests that abnormal levels of growth factors or a neurofibromatosis 2 mutation might result in the proliferation of the meningothelial cells encircling the vessels.[12]

Unlike typical meningioma, intraparenchymal meningioma does not have a gender preference favoring females. Surprisingly, they occur more in males than females (M: F ratio, 1.9:1), while most meningiomas commonly occur more in females, with a male-to-female ratio of 1:2.5.[15,18] They also present at an unusually early age, with symptoms and signs of increased intracranial pressure, seizures, and focal neurological signs.[8] Intraparenchymal meningiomas most commonly involve the cerebrum, with few reported cases involving the cerebellum and brainstem. More than half of intraparenchymal meningiomas are in the frontal and temporal lobes. Lesions arising in the parieto-occipital region are rare,[13,19] and here, we report the second case of an intraparenchymal meningioma in this region. It is challenging to diagnose intraparenchymal meningioma before obtaining histopathological results, as neither clinical symptoms nor radiological features can reliably

aid in the diagnosis, and lack of any clinical feature or imaging finding that seems pathognomonic for such a tumor.[13]

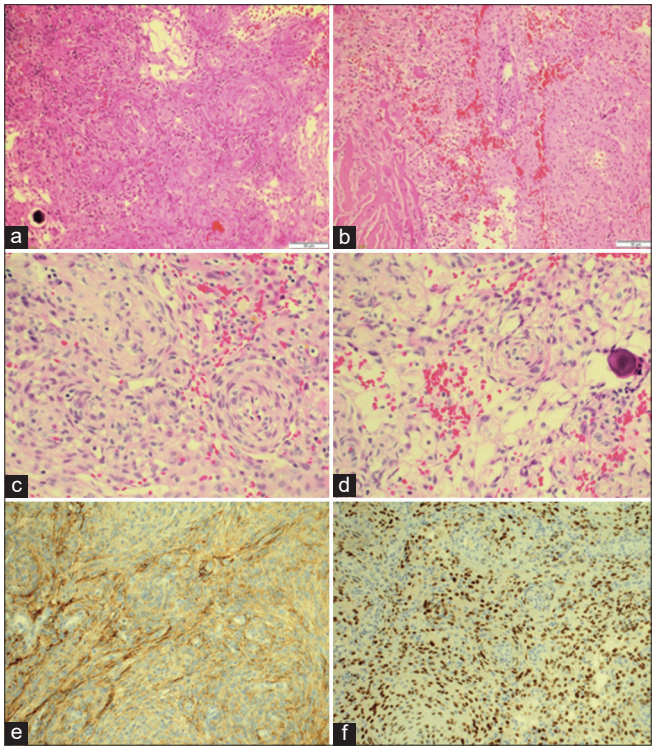

Figure 3: Microscopic examination of hematoxylin and eosin (H and E) stained sections. (a and b) (H and E- ⟡10) Proliferation of neoplastic meningothelial cells, infiltrating the dura. (c and d) (H and E- ⟡20) Tumor cells are arranged in whorls, fascicles, and microcystic pattern. Psammoma bodies seen. (e) epithelial membrane antigen immunohistochemistry patchy positive staining and (f) Progesterone receptor immunohistochemistry positive nuclear staining.

Typical radiological features seen with a meningioma include a "dural tail" sign, hyperostosis, calcifications, homogeneous enhancement with contrast, and a clear demarcation between the tumor and the normal brain tissue.[2,20,21] These characteristic features, however, tend not to be found in intraparenchymal meningiomas; instead, they are usually part cystic with contrast-enhanced solid parts and exhibit significant vasogenic edema.[6,7] Such findings make it difficult to distinguish this tumor from intra-axial lesions with cystic and solid components. Nonetheless, it has been found that peritumoral edema on brain MRI developing from one part of the tumor surface could be a feature that favors subcortical meningiomas over high-grade gliomas or brain metastases, where the latter tends to develop edema around the whole tumor surface.[19] In the present case, the radiological features seen on the brain MRI included mass effect, irregular enhancement with contrast, diffuse restriction, and mixed solid and cystic components, which were not indicative of a benign tumor, let alone a meningioma. Therefore, based on the MRI features and the patient's presentation. The preoperative differential diagnosis of our case was a glioma, oligodendroglioma, and metastatic tumor.

Surgical resection remains the gold standard of management for meningiomas. A gross total resection is preferable to decrease the rate of recurrence.[4] Moreover, surgery with maximal safe resection debulks the tumor, improves symptoms, allows for better outcomes with chemoradiotherapy, and, most importantly, improves survival.[16] While the standard treatment for meningiomas is surgical excision, observation or stereotactic radiosurgery is plausible alternatives.[13] Intraparenchymal meningiomas generally have a better prognosis in adults than in pediatric patients, with a lower incidence of recurrence and mortality.

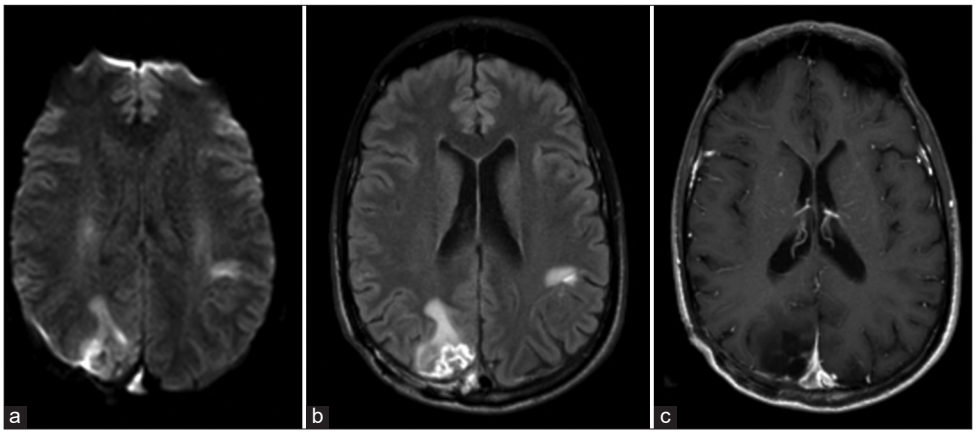

Figure 4: A brain magnetic resonance imaging was done 2-month postoperatively. Axial (a) Diffusion-weighted image, (b) FLAIR, and (c) contrast-enhanced T1-weighted images showing postoperative changes noted in the right parietal and occipital lobes with areas of gliosis and focal volume loss. Nodular areas of enhancement are seen at the surgical site, consistent with scarring-no evidence of residual disease or tumor recurrence.

Still, the prognosis largely depends on the pathological grade of the lesion, the extent of excision, and the tumor's location.[8,13]

CONCLUSION

The radiological features of intraparenchymal meningiomas are largely atypical of this tumor type and tend to mislead the diagnosis and steer it toward more malignant lesions. Preoperative diagnosis of this rare entity is challenging. However, it should be part of the differential diagnosis as it might direct the course of treatment and foreseen outcomes.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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