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ANGIOMATOUS MENINGIOMA WITH CLASSIC HISTOMORPHOLOGICAL FEATURES

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ABSTRACT
Meningioma comprises majority of the intracranial tumors. They arise from the meningothelial cells (MECs) and are usually slow growing and benign in nature. Few meningiomas have a complicated histology and occur in compromising locations are very difficult to treat. Such cases have poor prognostic outcomes. This necessitates prompt diagnosis of the tumor and a proper understanding of its severity that can help in better management of the patients in future.
Keywords: Meningioma, histomorphology**,** intracranial tumors

Introduction

Meningioma is one of the most common CNS tumours which consists of about 36.4% of all intracranial neoplasms.[1][2] The different types of meningiomas according to WHO classification are meningothelial, fibrous, transitional, psammomatous, microcystic, secretory, angiomatous, lymphoplasmacytic- rich, metaplastic, chordoid, clear cell, atypical, papillary, rhabdoid and anaplastic(malignant). It is believed that about 1% to 3% of meningiomas can turn into malignant forms with a 5-year survival of 32% to 64%.[3] Despite the benign nature of disease, these dural based tumors can become a cause of death when present in unsual locations with non-specific symptoms.[4]

Case Report

A 43-year-old female presented with complaints of headache which progressed to severe intensity along with multiple episodes of projectile vomiting and drowsiness. The patient also had complaints of left upper limb and left lower limb weakness for a month. She had no other comorbidities. On examination, the pulse rate was 82/minute and BP was 150/90 mm Hg. Computed tomography (CT) brain demonstrated an Ill- defined hypodense lesion in the right fronto- parieto- occipital region with edema, mass effect with 10mm midline shift to left side, mostly suggestive of a neoplastic etiology (Figure 1A). Magnetic resonance imaging (MRI) brain showed a well-defined smooth marginated, extra-axial mass lesion measuring 3.2 x 3.1 x 2.5 cm, arising from the falx cerebri in the right para- falacine area of the fronto-parietal region. A midline shifts of 7 mm and sub-falacine herniation was noted towards the left side.

The features were most likely suggestive of meningioma (Figure1B). The Glasgow coma scale of the patient was 4/15 and the patient was intubated and put on a ventilator in view of neurological deterioration. Thin mucoid endotracheal tube secretion was noted. Cough reflex and gag reflex were present. The patient was planned for craniotomy and tumour excision. The specimen was sent for the histopathological examination. The specimen was received in multiple grey white soft tissue bits aggregating to 1.5x1x0.8 cms. Haematoxylin and eosin-stained sections revealed a tumour composed of spindle shaped cells having round to oval nuclei and eosinophilic cytoplasm with distinct border arranged in whorled pattern. Many well-formed and thick hyalinised stromal vasculature were seen comprising >50% of tumour (Figure 2A, 2B). The sections also showed microcystic change along with foamy cells which are related to leakage of plasma lipids across thin vessel walls. Immunohistochemistry showed strong EMA positivity of tumour cells (Figure 2C, 2D). The final diagnosis of Angiomatous meningioma was made. Post operatively, the patient became unconscious and was on ventilator and had multiple episodes of supraventricular tachycardia with heart rate reaching 300bpm followed by an episode of hypotension for which injection Adenosine and tablet Ivabrad was given. The patient became unresponsive with bleeding or oozing from the surgical site. The complete blood count and biochemical values were deranged. Bilateral pupils were fixed and dilated, non-reactive to light and the Doll's eye reflex, corneal reflex, gag and cough reflexes were absent. The caloric test was negative. Hence, an apnoea test was done which came positive twice over an interval of 6 hours. The patient was finally declared brain dead.

Discussion

Angiomatous meningioma is a rarely encountered subtype, comprising 2.1% of all meningiomas.[1][2][5][6][7][8] The male to female ratio for meningioma is 1:2 generally but for angiomatous meningioma it seems to be higher.[1][2][5] There are two histological subtypes-macrovascular (diameter of >50% vessels larger than 30 um and microvascular (diameter of >50% vessels smaller than 30 um).[2][5] Unlike other meningiomas, this subtype has numerous chromosomal polysomies (7,20,13,12 and 5) which suggests that the signals for vascular differentiation and proliferation outweighs the inhibitory growth signals.[1][5] The vascular endothelial growth factor (VEGF) pathway and the VEGF gene(chromosome 5)also plays an important role in the pathogenesis of peritumoral brain edema which is frequently present in this subtype.[1][5][6][8] On T2 weighted MRI, the width of peritumoral edema at the largest tumour level is classified as mild(</= 2cm), moderate (>2cm or </= ½ hemisphere) and severe (>/= ½ hemisphere).[8] The differential diagnosis includes vascular tumours like hemangiopericytoma and capillary hemangioblastoma and non-neoplastic vascular lesions like vascular malformations. These can be differentiated from Angiomatous meningioma by the use of immunohistochemical stains.[2][5][8] Hemangiopericytoma and capillary hemangioblastoma stain negative with EMA but angiomatous meningioma is EMA immunoreactive.[2][3][8] This variant has low MIB/Ki67 indices and thus low probability of recurrence.[2][3][4] These tumours belong to the WHO grade 1 and patients have a very favourable prognosis, despite the aneuploidy state.[1][2][5][6][7][8] Metastasis is quite rare (only 0.1%) but is more frequent for meningiomas invading dural sinuses.[2] Craniotomy with total tumour resection is the treatment of choice and in cases of incomplete removal, gamma knife is recommended for small residual tumour.[2][8]

Conclusion

There are many subtypes of meningiomas with varied histomorphological features that need to be differentiated from other resembling CNS tumours. Angiomatous meningioma is a rare subtype with favourable prognosis. Hence, early detection and management can prevent mortality.

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LEGENDS OF FIGURES: -

Fig. 1A: CT brain showing an ill-defined hypodense lesion with edema and 10mm shift to left.
Fig. 1B: MRI brain showing well-defined and smoothly marginated mass lesion.

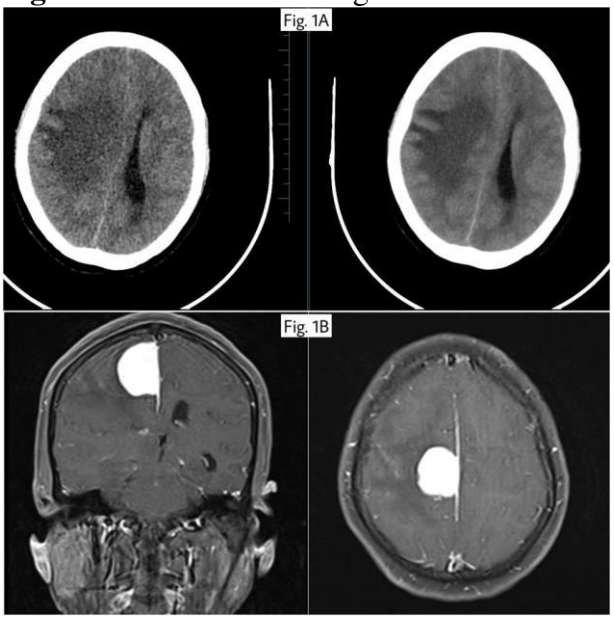

Fig. 2A &2 B: Meningothelial cells arranged in short fascicles and syncytial pattern with multiple hyalinized thick-walled vessels comprising >50% or tumour. (H & E; 100X)
Fig. 2C & 2D: EMA immunostaining highlighting the meningothelial cells around the vasculature. (IHC; 100X)