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Case Report

Pathology Section

Intraparenchymal Angiomatous Meningioma: A Diagnostic Dilemma

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ABSTRACT

Meningioma arises from the arachnoid cap cells of the cerebrum. Intraparenchymal meningiomas or meningiomas without dural attachment are rare. We report a case of 40-year-old male who presented with a history of headache, dizziness and gradual loss of vision since one year. Clinicoradiological diagnosis of a high grade glioma was considered. Tumour was excised and haematoxylin and eosin stained sections revealed a tumour comprised predominantly of variable sized blood vessels showing hyalinization in a background of plump spindle cells with oval vesicular nuclei. In view of these features angiomatous meningioma was suspected. However, to confirm the diagnosis, a panel of immunohistochemical markers including vimentin, EMA and GFAP was done and a final diagnosis of angiomatous meningioma was offered. Angiomatous meningioma is a rare variant of meningioma and even much rarer in the intraparenchymal location. Angiomatous meningioma should be considered in the differential diagnosis of highly vascular intraparenchymal brain tumours.

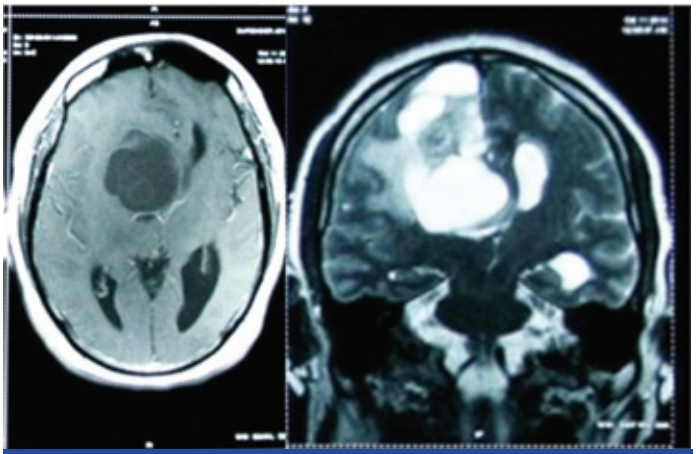
Keywords: Brain tumour, Meningiomas without dural attachment, Vascular

CASE REPORT

A 40-year-old male presented with history of headache, dizziness and gradual loss of vision since 1 year. He also complained of increasing unsteadiness in gait, along with weakness in left upper and lower limb since 5 months. Magnetic Resonance Imaging (MRI) scan revealed a large lobulated intraaxial subcortical mass lesion involving the right parietal lobe. The mass appeared hypointense on T1W and heterogenously hyperintense on T2W images with some cystic component. Surrounding white matter oedema and mass effect was also present [Table/Fig-1].

A standard right fronto-parietal-temporal flap craniotomy was done in view of gross mass effect and significant midline shift. Abnormal grayish tissue was found in subcortical region which was firm, non-suckable, and moderately vascular. No clear tumour-brain margin could be identified. Internal decompression followed by gross total tumour removal was done with minimal retraction of surrounding neural and vascular structures. On clinicoradiological correlation, a diagnosis of high grade glioma was offered. The postoperative period was uneventful.

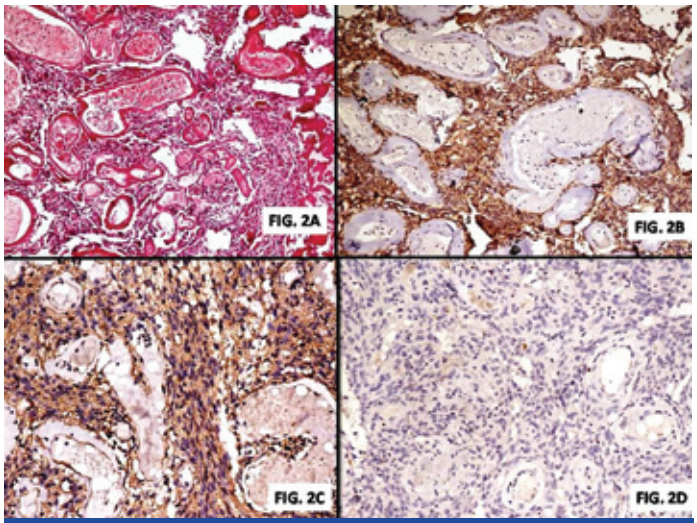
Tissue was submitted for histopathological examination and comprised of multiple grey to reddish brown soft tissue pieces, firm in consistency, together measuring 4.5x2.5x2 cm. Entire tissue was processed. Sections revealed a highly vascular tumour tissue comprised predominantly of variable sized hyalinized blood vessels, and intervening areas showed tumour cells in a syncytial pattern with indistinct cytoplasm and oval vesicular nucleus suggestive of angiomatous meningioma [Table/Fig-2a]. Further a battery of immunohistochemical (IHC) markers was done including vimentin, epithelial membrane antigen (EMA) and glial fibrillary acidic protein (GFAP) to confirm the diagnosis. Vimentin showed cytoplasmic positivity and EMA revealed membranous positivity in the tumour cells while GFAP was negative in the tumour cells [Table/Fig-2b-d]. A final diagnosis of angiomatous meningioma was made. Patient is on follow up since 10 months and is asymptomatic with unremarkable repeat MRI.



[Table/Fig-1]: Axial T1W and coronal T2W images showing intraparenchymal mass lesion in right parietal lobe; hypointense on T1W and heterogeneously hyperintense on T2W images

DISCUSSION

Meningioma accounts for 36.1% of the primary intracranial tumours with an age predilection towards middle aged and elderly patients [1]. Meningiomas arise from arachnoid cap cells that are present on the surface of pachinonian granulations, stroma of choroid plexus, tela choroidea and pia mater [2-5]. Location like tela choroidea and choroid plexus accounts for 1-2% of intraventricular meningiomas



[Table/Fig-2]: (a) Highly vascular tumour consisting predominantly of variable sized blood vessels showing hyalinization with intervening areas showing tumour cells (H & E X 100) (b) Tumour cells showing membranous positivity for EMA (DAB chromogen X 100) (c) Tumour cells showing cytoplasmic positivity for Vimentin (DAB chromogen X 100) (d) Tumour cells negative for GFAP (DAB chromogen X 100)

[6]. Primary occurrence of meningiomas without dural attachment is rare [7]. Few authors suggest that meningioma without dural attachment arises from the arachnoid cap cells in Virchow Robin spaces along the cerebral vasculature or from the pia mater within the brain sulcus while others suggest that the arachnoid cap cells rest during the migration process [7,8].

Meningiomas without dural attachment are divided into supratentorial and posterior cranial fossa tumours on the basis of location. Supratentorial non dural based meningiomas are further classified in five varieties: 1) intraventricular [9], 2) pineal region [9], 3) deep sylvian [10], 4) intraparenchymal/subcortical [11], and 5) others. Abraham and Chandy classified meningiomas of the posterior cranial fossa without dural attachment in five varieties: 1) meningioma arising from the choroid of the fourth ventricle and lying wholly within it, 2) meningiomas arising from the inferior tela and lying partially in the fourth ventricle and partially in the cerebellar hemisphere; and 3) meningiomas lying in the cisterna magna, 4) meningiomas arising from the choroid plexus and lying in the lateral cerebellomedullary cistern, 5) intraparenchymal meningioma [12].

A total of 30 cases of intraparenchymal meningioma have been reported in the English literature till date[11,13,14]. Incidence of paediatric intraparenchymal meningiomas was higher than adults and showed a male predominance. The most common symptom of intraparenchymal meningiomas was seizures followed by headache and hemiparesis, based on tumour location. On MR imaging none of the case was diagnosed as meningioma, preoperatively. Frontal lobe was the most common location followed by parietal and temporal lobes. Amongst the intraparenchymal meningiomas, fibroblastic variant was the most common subtype as compared to the meningothelial variant in dural based meningiomas. No case of angiomatous meningioma in intraparenchymal location has been reported till date.

Angiomatous meningioma is a rare and distinct variant of meningioma and accounts for 2.1% of meningiomas [15]. Mostly, angiomatous meningiomas affect cerebral convexity, and its occurrence in subcortical location without dural attachment is unusual and has not been found on literature search. It shares the histological and clinical features of benign meningioma but with a predominance of vascular component. The distribution of age, sex and tumour localization of angiomatous meningioma usually resembles typical meningiomas in most of the cases [15]. However, on MR imaging some specific feature like perilesional oedema due to increased vascularity is usually seen but is not a sign of atypia or malignancy. In angiomatous meningioma, the blood vessel component exceeds 50% of the total tumour area. Hasselblatt et al., described two histological patterns based on vessel diameter i.e. macrovascular with >50% of vessels having diameter larger than 30μm and microvascular subtype in which > 50% of vessels having diameter smaller than 30μm in their study but found no clinical importance of these subtypes [15].

The differentials of angiomatous meningioma like haemangioblastoma and haemangiopericytoma were ruled out in the present case on the basis of morphology and results of immunohistochemistry. Haemangioblastoma shows two components on histomorphology: vacuolated clear cells and a network of thin-walled blood vessels while haemangiopericytoma reveals irregular cells with oval nuclei, ill defined cytoplasm and staghorn vascular pattern. Haemangiopericytoma is vimentin, CD34 positive but EMA negative.

Gross total resection is the treatment of choice for angiomatous meningioma. Postoperative radiotherapy may help the patients with residual tumour. Angiomatous meningiomas belong to WHO grade I and has a favourable prognosis. The prognosis of meningiomas without dural attachment is similar to dural based meningiomas [11]. However, the completeness of surgical resection is the single most important factor in deciding prognosis.

The present case describes the unusual location of angiomatous meningioma and highlights the fact that making a diagnosis of intraparenchymal meningioma preoperatively is challenging and difficult. MRI findings of meningiomas are nonspecific in intraparenchymal location and thus, can be misleading too. Therefore, correlation of clinical, radiological, histopathological and immunohistochemical findings is always required to reach a particular diagnosis.

CONCLUSION

Although reports on angiomatous meningiomas are not uncommon but its intraparenchymal location has not been described before. Angiomatous meningioma should be considered in the differential diagnosis of a highly vascular intraparenchymal brain tumour.

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