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Sylvian Fissure Chordoid Meningioma in an Adult Patient.

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Case Report

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ABSTRACT

Meningiomas arise from arachnoid cap cells and are usually found attached to the dura. Chordoid meningioma which comprises less than 0.5% of all meningiomas is rarely seen without dural attachment and its position in the sylvian fissure is aberrant. Our patient, a 50-yr-old female, was presented to us with headache and sensory seizures. The magnetic resonance imaging of the brain revealed left fronto temporal lesion. Tumour resection was performed through left pterional craniotomy and transsylvian approach. The tumour was seen without any dural attachment and was densely adhesive to the arachnoid layer of sylvian fissure. Histopathologic findings of resected tumour were suggestive of chordoid meningioma. Though meningiomas originate from arachnoid cap cells, it can arise at locations remote from pachymeninges. This is a rare case of adult sylvian fissure chordoid meningioma without dural attachment. The patient presented with sensory seizures, improved after surgical excision without any further recurrences.

INTRODUCTION

Meningiomas, usually are found attached to the dura and thought to arise from arachnoid cap or meningothelial cells. Meningiomas account for approximately 15–25% of all primary intracranial tumours and often seen in middle-aged and elderly patients. The vast majority of these tumours are benign; however, complete removal can be difficult and recurrence is an issue. Occasionally, meningiomas are seen developed without dural attachment in some areas, including the sylvian fissure. Chordoid meningioma without dural attachment is rare in adults and located in the sylvian fissure is even more uncommon as reported earlier by McIver et.al [1]. Here we describe the second adult case of chordoid meningioma in the sylvian fissure without dural attachment.

Case Report

A 50-yr-old woman presented to us with complaints of headache, insidious in onset, located over left frontoparietal region and focal sensory seizures involving right upper limb with slurring of speech. Neurological examination revealed normal higher mental functions. Patient had ankyloglossia with normal word output and fluency but difficulty especially in using vowels and lingual sounds. Magnetic resonance imaging scan revealed well enhanced round lesion, in left posteriofrontal perisylvian opercular region (Fig 1a,1b). Multiple flow voids with significant perilesional oedema was observed. Left pterional craniotomy, transsylvian approach was performed. Posterior sylvian fissure was opened and operculum with intervening vessels was identified. Small cortical incision was done.

The tumour was greyish in colour and firm in consistency. There was a pseudoplane of cleavage and the part of the tumour closer to the fissure was densely adhesive to the arachnoid layer of sylvian fissure, probably its site of origin. The tumour was removed in toto. The patient is under follow up of more

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than 2 years and her symptoms have improved with no seizures. Post operative MRI showed no recurrence of lesion (Fig 1c,1d).

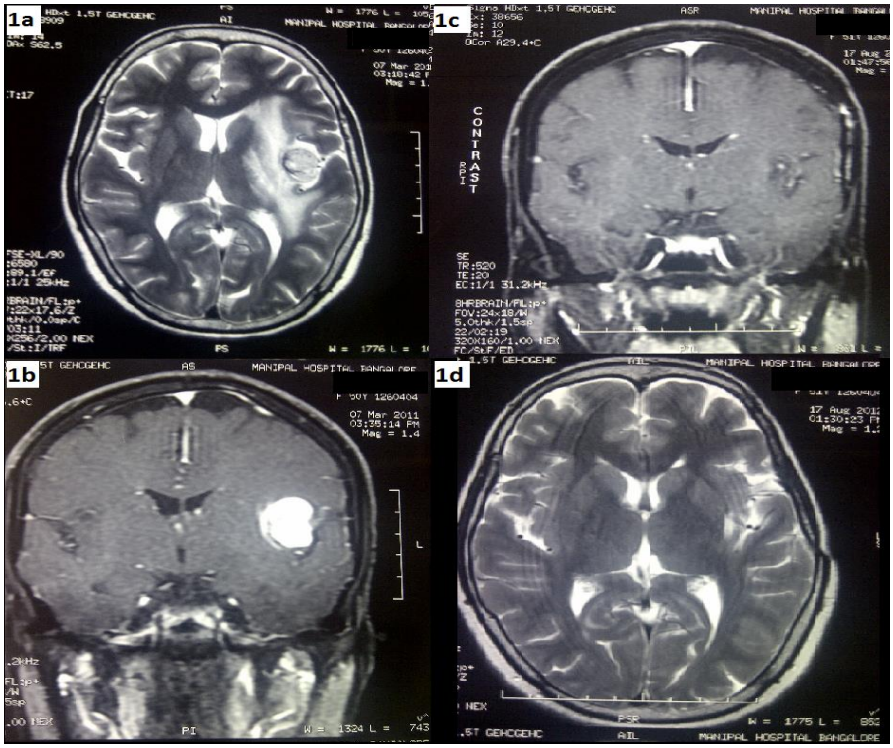

Figure 1: Preoperative (1a, 1b) and Postoperative (1c, 1d) MRI Images

DISCUSSION

Meningiomas without a dural attachment usually develop at the intraventricular region, the pineal or subcortical region and the posterior fossa[2]. Sylvian meningiomas are quite rare tumours without a visible connection to the dura mater or to the ventricular system. These tumours are more common in children than in adults, but most of them are regular histological variants.

Sylvian meningioma originates from the leptomeningeal foldings of the sylvian fissure and also from the arachnoid Cap cells of the pia layer of the sylvian fissure or in the Virchow–Robin space[3]. From these cap cells, meningiomas develop in locations without dural attachment like the one observed in our patient.

Chordoid meningioma is a rare variant of meningioma that bears a striking histological resemblance to chordoma and has greater likelihood of recurrence[4]. Recognition of this entity is important in cases that show similar morphologic overlap with other chondroid/myxoid neoplasm that can arise within or near the central nervous system[5]. The term ''chordoid meningiomas'' was coined by Kepes et al in 1988[4]. These tumours were composed of eosinophilic vacuolated cells disposed in chordoma-like clusters and cord in a myxoid matrix, and often featured a prominent lymphoplasmacellular infiltrate. The 1993 World Health Organization (WHO) classification of tumours of the Central Nervous System accepted chordoid meningioma as a variant of meningioma. This tumour was previously reported to be frequently associated with haematological abnormalities in paediatric population[6].

Chordoid meningioma corresponds to WHO grade II (atypical meningioma) because of its more aggressive clinical behaviour and high rate of recurrence[4].The patient described here is an adult female with symptoms of headache and seizures. Following a complete surgical excision, the patient became asymptomatic. The precise diagnosis of chordoid meningioma can only be made with an accurate histopathological analysis. Though the histopathological features are distinctive, these tumours should be distinguished from its other variants[7]. Identification of a typical feature of meningioma combined with immunostaining for EMA and vimentin in chordoid area helps establish the definitive diagnosis of chordoid meningiomas[8].

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Chordoid meningiomas can be distinguished from ependymoma based on its positive staining for EMA and negative staining for GFAP[8]. In our patient, initial possibility of ependymoma was considered and later confirmed as chordoid meningioma with immunostaining by EMA, which remains as the most effective antibody in differentiating chordoid meningioma.

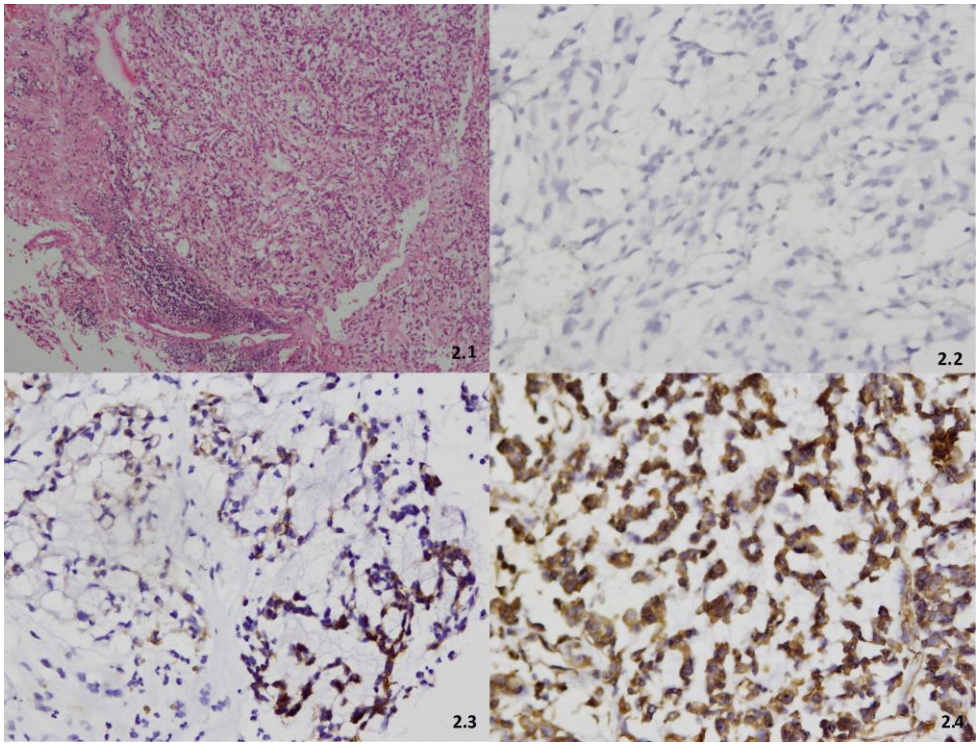

Figure 2: Immunohistochemical reactions confirming chordoid meningioma (2.1) and tumour cells showing immunoreactivity to GFAP (2.2), EMA (2.3), Vimentin (2.4)

Chordoid meningioma is recommended to be followed up at regular intervals after surgery due to its aggressive nature[9]. Its recurrence rate in 5 years is estimated as 40%. A possible explanation for the high rate of recurrence could be related to the mucoid quality of its stroma which mechanically facilitates the spread of the neoplastic cells[4]. In our patient, a follow up scan performed at 2 years after surgery showed no recurrence of tumour. She has been advised for long term follow up since her MIB-1 labelling index was high.

Chordoid meningioma without dural attachment is rare and location within the sylvian fissure is even more uncommon. This observation merits mention to stress the necessity to identify the major areas, whose covering arachnoid can be the origin of chordoid meningiomas, especially when no dural attachment is noted intra operatively.

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