

Case Report

Adult chordoid meningioma: a case report

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ABSTRACT

Chordoid meningioma is a rare tumour accounting for less than 0.5% of all meningiomas. It is a WHO grade II tumour with an aggressive behavior. It is a rare variant characterized by cords or trabeculae of eosinophilic or vacuolated cells set in an abundant mucoid matrix. It can be associated with systemic or hematologic manifestations like Castleman disease. The tumor has a propensity for aggressive behavior and increased likelihood of recurrence. We report a case of an adult patient with chordoid meningioma who presented with headache and seizures and did not have any hematological/ systemic manifestations. She underwent total excision of the lesion and is doing well.

Keywords: Aggressive tumor, Meningioma

INTRODUCTION

Meningiomas are a group of benign slow growing neoplasms that derive from the meningothelial cells of the arachnoid layer.¹ WHO 2016 classification of tumors of the Central Nervous system describes 15 meningioma variants of which meningothelial, fibrous and transitional are common. Chordoid meningioma is a rare variant characterized by cords or trabeculae of eosinophilic or vacuolated cells set in an abundant mucoid matrix.¹ The term chordoid meningioma was first introduced by Kepes et al. in 1988 in a series of young patients with hematologic abnormalities especially Castleman's disease.² However chordoid meningiomas mostly occur without any systemic manifestations.^{3,4}

The tumour often presents as a large supratentorial mass and have very high rate of recurrence after subtotal resection.³ Aggressiveness of this grade II tumour increases if the chordoid pattern predominates histologically.

Authors report an adult case of chordoid meningioma that presented in this institute.

CASE REPORT

A 43-year-old female presented to the medicine department with a history of headache for past 3 months. She also had multiple episodes of seizure. She was referred to the neurosurgery department for evaluation. Clinical examination revealed no focal neurological deficits. Her routine hematological and biochemical investigations were within normal limits. Cranial MRI showed an extra axial lesion in the frontal region measuring 4x3.3x3.7cm. The lesion was isointense on T1, ISO to hyperintense on T2 and hyperintense on FLAIR (Figure 1).

Perilesional edema was present. MRI features were suggestive of a meningioma. Frontal craniotomy and total excision of the lesion was done, and the specimen was sent for histopathological examination.

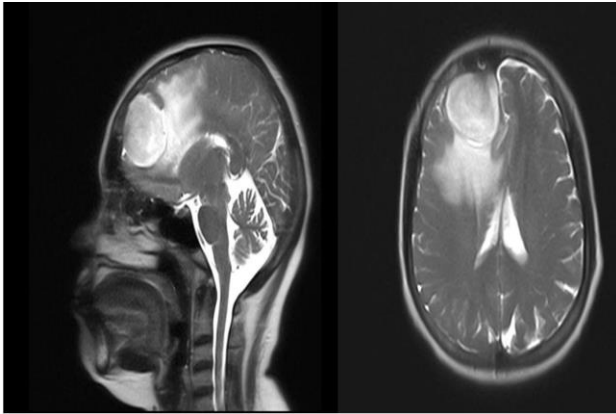


Figure 1: MRI showed contrast enhancing frontal lesion suggestive of meningioma.

In the pathology department we received multiple irregular pale white and pale brown firm tissue bits aggregate measuring 6x4x0.5cm. Hematoxylin and eosin stained sections showed a neoplasm composed of tumour cells arranged as cords and trabeculae in an abundant mucoid stroma (Figure 2A). Tumour cells had mild to moderate eosinophilic cytoplasm, round to oval nucleus with minimal nuclear atypia (Figure 2B, 2C). The tumour cells were surrounded by mucoid material. Dilated capillary sized blood vessels were seen between the cells. Focal area showed brain invasion (Figure 2D). No other typical meningioma component was identified. No necrosis/ lymphoplasmacytic infiltration was noted.

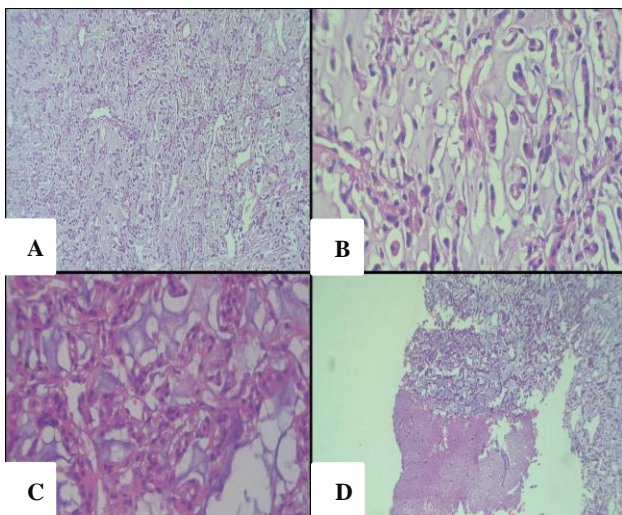


Figure 2: A) Trabeculae of tumour cells in a mucoid matrix (HandE 100x); B) Higher power of tumour cells (HandE 400x); C) Eosinophilic tumour cells (HandE 400x); D) Focus of brain invasion by the tumour.

Immunohistochemistry was done. The tumour cells showed diffuse strong immunoreactivity for vimentin highlighting the ribbon like architecture of the tumour (Figure 3A, 3B). Ki67 revealed a high mitotic index of

11% (Figure 3C, 3D). With these the diagnosis of Chordoid meningioma WHO grade II was given.

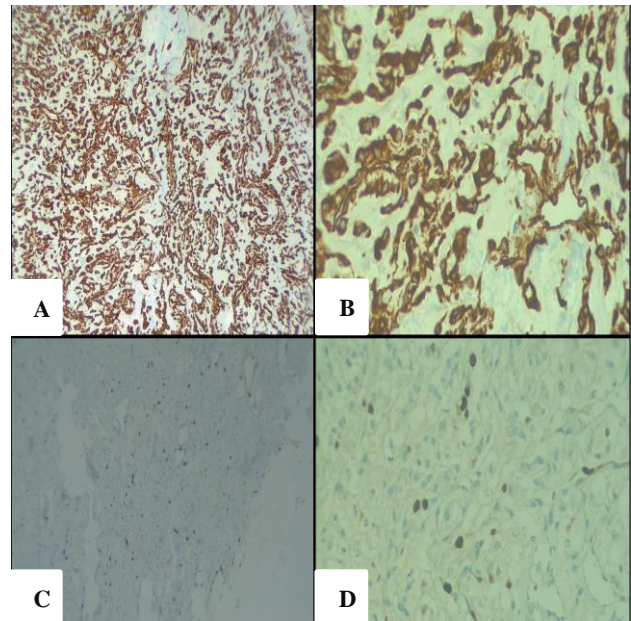


Figure 3: A) Tumour cells showed diffuse strong positivity for vimentin highlighting ribbon like architecture (IHC 100x); B) Tumour cells with cytoplasmic positivity for vimentin- high power (IHC 400x); C) Ki67 showed proliferation index of 11% (IHC 100x); D) Ki67 nuclear staining of tumour cells in high power (IHC 400x).

DISCUSSION

Chordoid meningioma is an uncommon variant of meningioma with a propensity for aggressive behavior and increased likelihood of recurrence.² Kepes et al, in 1988 first described the entity of chordoid meningioma which earlier used to be known as myxoid/myxomatous variant of meningioma.^{2,5} Chordoid meningioma was also thought to be associated with many systemic features like Castleman disease and anemia. Some later studies that followed Kepes disproved the same. A larger series by Couce et al, failed to identify an association of chordoid meningioma with systemic manifestations; however, the aggressive nature of this neoplasm was confirmed, with 42% of cases showing 1 or more recurrences, ranging from 1.8 years to as long as 16 years postoperatively.^{3,6} Recurrences are more common with incomplete excision of the tumour.

Chordoid meningioma often presents as large supratentorial masses and one of the commonest presenting complaints is headache. The incidence ranges from 0.5% to 1% of all meningiomas and there is no age or sex predilection.⁴

Histologically the tumour shows cords and trabeculae of eosinophilic /vacuolated cells in a mucoid matrix.¹

Chordoid areas may be interspersed with varying amount of typical meningioma cells.

If the histology or imaging studies are not typical chordoid meningioma can pose a diagnostic challenge. Chordoma, chordoid glioma, myxoid chondrosarcoma (skeletal and extraskel et al), low-grade chondrosarcoma, myxopapillary ependymoma, and mucinous metastatic carcinomas enter in the differential diagnosis at times.^{6,7} A wide panel of IHC markers can be used to differentiate all these tumours which include GFAP, EMA, Vimentin, S100 and Cytokeratin.^{6,8}

A unique unbalanced translocation involving chromosomes 1 and 3 identified in 3 cases of chordoid meningioma and not observed in other meningioma subtypes suggests that cytogenetic studies and potentially fluorescence in-situ hybridization methodologies could be of diagnostic utility, although further investigation in this area is needed.^{6,9}

Our patient was an adult female who did not have any hematological abnormalities or features suggestive of Castleman disease. The tumour was completely excised.

CONCLUSION

Chordoid meningioma is a rare and aggressive neoplasm. Aggressive behavior increases with the predominance of chordoid pattern within the tumor. Recurrence rates are higher with incomplete excision of the tumour.

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Ethical approval: Not required

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