Abstract:
Meningioma is the most common extraaxial neoplasm of adults. Among its histological variant, Chordoid meningioma is uncommon. We report a 19 years young girl who presented with simple partial seizure, headache for 3 months. The neurological examination was normal. The imaging of the patient showed a lobulated lesion with solid and cystic component located in the left occipital region. We proceeded with left occipital craniotomy and total excision of the lesion. The histopathological report was chordoid meningioma. The post-operative period was uneventful.

Keyword:
Chordoid meningioma, chordoma.

Introduction:
Meningioma is the most common extra–axial neoplasm of adults accounting for about 20% of primary intracranial neoplasm. Meningioma exhibits a wide variety of histological diversity. Chordoid meningioma is a metastatic meningioma with aggressive behaviour (WHO Gr II). They are locally invasive and the recurrence rate is high after subtotal excision. Kepes et al coined the term chordoid meningioma, as its histological features resemble chordoma.

Case report:
A 19 years young girl, presented with complaints of seizure-3 months headache for 2 months. The seizure starts with perioral twitching followed by tonic clonic movements of the right upper and lower limbs, lasting for about 2-3 minutes. The consciousness was preserved during the
attack and there was no post ictal confusion or weakness. She had about 2 such episodes per month. She had headache in the occipital region lasting for about 1 hour initially, which became holocranial later. There was no diurnal or postural variation. There were no aggravating or relieving factors. It was not associated with vomiting or blurring of vision. There was no history of trauma, loss of consciousness, fever, and loss of weight, loss of appetite or any chronic cough. No history suggestive of cranial nerve disturbance, motor/sensory deficit. Bladder and bowel habits were normal.

**Examination:**
The general examination was normal with stable vitals. On neurological examination, her higher mental functions were normal. The cranial nerves, motor/sensory system, cerebellum, spine, cranium, autonomic system did not reveal any abnormality.

**Clinical and Radiological investigation:**
The routine investigations, complete haemogram, blood sugar, urea, creatinine were within normal limits. X-rays chest was normal and mantoux was negative. Ophthalmic examination did not show any abnormality.

![Fig 1](image1.png) Pre op CT scan Brain shows solid, contrast enhancing lesion with cystic component in the left occipital region

![Fig 2](image2.png) MRI Brain T1 images show hypo-intense and T2 hyper intense lesion
**Fig:3** MRI Brain T 1 contrast images show enhancement of the solid component.

CT scan brain of the patient shows an enhancing lesion in the left occipital region. MRI brain shows T1 iso-hypo intense lesion, T2 hyper intense, with cystic component not suppressed in flair images. Solid component shows contrast enhancement in the left occipital region with attachment to the posterior portion of falx.

**Intra operative photos:**

**Fig:4** Intra operative images

**Procedure:**

Left occipital craniotomy was done, and through trans sulcal approach, the cystic component was aspirated and the solid component was removed in Toto. The size was about 4x4 cm. The specimen was subjected to histopathological examination.
The post op CT brain shows complete excision of the tumour. The post operative period was uneventful. She recovered well without any neurological deficit and she was discharged with advice to come for regular follow up.

Fig: 6 Visual charting was Normal complete removal.

Fig: 7 HPE shows myxoid matrix and lymphocytic infiltrates

Fig: 8 Meningothelial cells in whorls
The tumour contains a bluish myxoid matrix mimicking a chordoma. Focal areas of meningiothelial cells in whorls and lymphoplasmacytic infiltrates. The histopathological report was Chordoid meningioma.
Discussion:
Meningiomas are the most common primary intracranial tumours, and the most remain asymptomatic throughout the patient's lifetime. They arise from arachnoid cap cells and not from Dura. Incidence peaks at 45 years of age. Female: Male ratio is 1.8:1. Most commonly located along falx, convexity, or sphenoid bone.

WHO classified meningioma based on the histopathology as:
- **Grade I**
  - Meningothelial, fibrous, transitional, psammomatous, angiomatous, microcystic, secretory, metaplastic
- **Grade II**
  - Chordoid, clear cell, atypical
- **Grade III**
  - Papillary, rhabdoid, anaplastic

Chordoid meningioma is an uncommon variant of meningioma, classified under WHO Grade II. Histologically, it has the characteristic features of meningioma and chordoma. The tumour contains trabeculae of vacuolated or eosinophilic cells in a bluish matrix mimicking a chordoma. Focal areas of meningothelial and transitional features and lymphoplasmacytic infiltrates are common. The immunohistochemistry shows scant positivity for epithelial membrane antigen and S-100. Differential diagnosis includes chordoid chordoma, myxoid chondrosarcoma and chordoid glioma.

Total surgical excision is the treatment of choice. Post-operative radiotherapy is less useful when the residual or recurrent tumour size is large. They are aggressive, locally invasive. The chance of recurrence is high when it is not removed totally. In the literature, less than 75 cases have been reported. The largest case report was 42 cases (1975 to 1997) by Couce et al in the American Journal of Surgical Pathology. We report this case because; it is an uncommon variant of a common extra-axial neoplasm.

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