Research Paper

Volume : 4 | Issue : 2 | February 2015 • ISSN No 2277 - 8179

Chordoid Meningioma with CastlemanSyndrome-A rare case report

Medical Science

KEYWORDS : Chordoid Meningioma, Lymphoplasmacytic infiltration, Castleman Syndrome.

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Introduction:

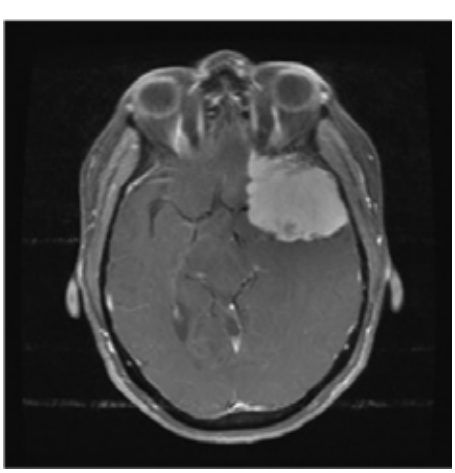
Meningiomas are generally slowly growing benign tumors derived from leptomeningeal arachnoid cells and account for approximately 15% of all primary tumors of central nervous system (1,2). Meningiomas have a wide range of histopathological appearances due to the capacity of arachnoid cells to exhibit divergent differentiation. The latest World Health Organization (WHO) classification of tumours of the central nervous system lists 15 histopathological variants of meningioma(3).

In contrast to the most common types of meningiomas, such as meningothelial, fibrous and transitional, the chordoidmeningiomas are relatively rare variantoften associated with peritumorallymphoplasmacytic infiltration causing Castleman syndrome. The majority of these reports suggest that chordoidmeningiomas are frequently associated with hematological abnormalities or systemic manifestations of Castleman syndrome (2,5,8,9). However, recent reports of chordoid meningioma do not show any association with hematological abnormalities or systemic manifestations ofCastleman syndrome (4,6,7).We present herein a case of chordoid meningioma with Castleman syndrome that occurred in a 20-year old female.

Case History:

A 20 years old female presented with complaints of sudden onset of convulsions associated with low grade fever. On investigation her Hb was 9.8gm/dl (Normal range of 12-14gm/dL), MCV 70 fl (Normal range of 80-98fl), ESR 98 mm in 1 hour. On MRI there was a space occupying lesion in the fronto-temporal region which was given differential diagnosis of either Tuberculoma or Meningioma.(Figure 1.1) So the patient was started with AKT empirically. She again had an episode of convulsion with severe headache even after 4 months of AKT. She was having low grade fever also since last 4 months. In spite of AKT,lesion didn't improved on follow up MRI. Then she was operated twice with initial diagnosis of Inflammatory Pseudotumor on Frozen section with limited resection.

Figure 1.1:Axial T1 MRI with contrast showing left frontotemporal region meningioma.



Later on, complete resection of the tumor was done.

On gross examination they were greyish multiple tissue bits measuring 4 cm in aggregate.

On microscopic examination:

Low power view of tumor resembled myxoid or mucoid tumor with dispersed tumor cell cords & clusters.(Figure 1.2) There were also well defined inflammatory foci.

In High power view the tumor cells resembledmeningothelial cells having round to oval bland looking nuclei & moderate eosinophilic cytoplasm.(Figure 1.3)Meningothelial whorls were also seen at places.

Inflammatory foci consist of lymphoplasmacytic infiltrate with occasional lymphoid follicle formation.(Figure 1.4)

Figure 1.2:

(10x view)Tumor with loosely arranged tu mor cells forming fascicles & whorls.

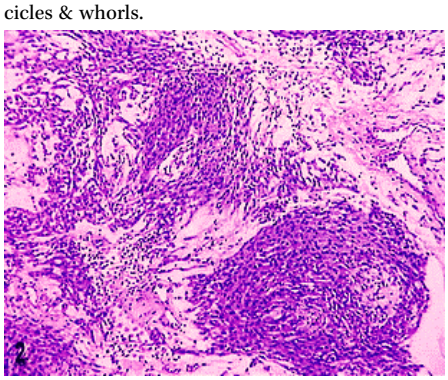


Figure 1.3:

(20x view) Tumor cells are arranged in small clusters & cords in mucoid or myxoid background.

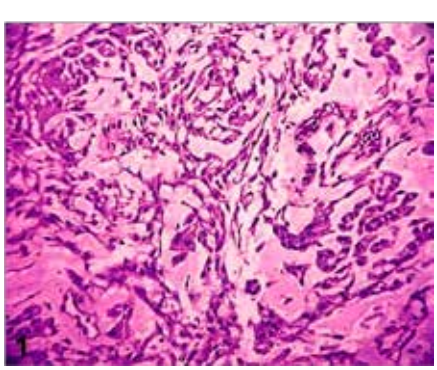
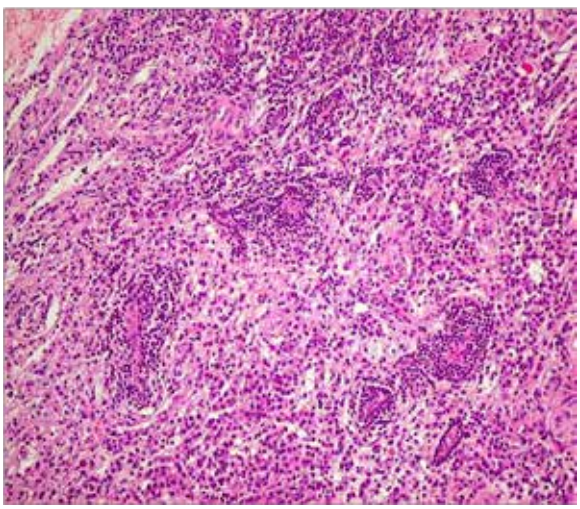
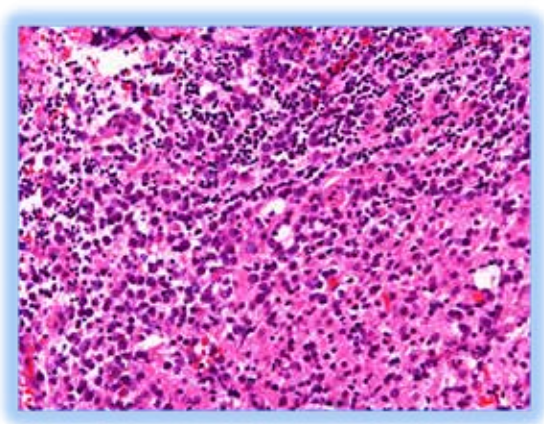


Figure 1.4:



(a) (4x view)At places tumor show aggregates of inflammatory cells intermingling with meningothelial cells.



(b) (20x view)Showinglymphoplasmacytic infiltrate with intervening meningothelial cells.

After resection of the tumor all the haematological parameters were within normal limits within 1 month.

Discussion:

Although most meningiomas are benign and are graded into WHO grade I, the chordoid variant is associated with a less favourable clinical outcome and is graded into WHO grade II&has significant recurrence rate(10).

Further chordoid meningioma many times have significant mucoid back ground & may be confused with more commonly occurring chordoma,metastatic mucinous carcinoma and chordoidglioma. So close attention to tumor cell cytology help not to miss diagnosis of rare chordoid meningioma, whenever one comes across a tumor with significant mucoid or myxoid background.

As sometimes extensive lymphoplasmacytic infiltrate is associated with chordoidmeningioma,a close examination for foci of mucoid area containing meningothelial cells should be searched for,in tumors resembling inflammatory pseudotumor.

In conclusion, chordoid meningioma is a rare morphologic variant of meningioma and may be associated with systemic or hematologic abnormalities. Thepotential histopathologic misdiagnosis can be avoided by establishment of histologic features.

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