



Case Report

## Xanthomatous Meningioma - A Rare Case Report

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### ABSTRACT

Xanthomatous meningioma is an extremely rare variant of meningioma. The histopathological hallmark of Xanthomatous meningioma is the presence of Xanthomatous tumour cells and these are considered as neoplastic meningotheial cells and not macrophages. We describe sixth documented case of xanthomatous meningioma in the temporo-parieto-occipital region. A 24- years- old Indian female presented with diminished vision, multiple episodes of vomiting and occasional episodes of headache. Magnetic resonance Imaging revealed a large, ill-defined tumor arising from the dura in the temporo-parieto-occipital lobe measuring 6.2x7.26.7cm with supratentorial as well as infratentorial extension. The histopathological examination revealed Xanthomatous meningioma.

The special stains were done for PAS which was negative for foamy cells which ruled out macrophages. IHC results showed tumor cells expressing Epithelial Membrane Antigen (EMA) as well as the foamy cells were EMA positive. MIB-1 (KI67) proliferation index approximately 3.2% in highly cellular areas. The final diagnosis of Xanthomatous meningioma was made. We are presenting this case for its clinical, radiological, histopathological and IHC study as this is an extremely rare case.

**Key words-** xanthomatous meningioma, metaplasia, EMA, meningeal tumors.

### INTRODUCTION

Meningiomas are benign tumors of the central nervous system originating from meningotheial cells i.e. the arachnoid cap cells located in the cranium and spinal canal. Meningioma accounts for about 24 % of primary CNS tumors [1] The incidence of this tumor is higher in females and increases with age. [2] A recent World Health Organisation Classification of Tumors of Central Nervous System divides grade I meningiomas into 9 subtypes:

meningotheial, fibrous, transitional, psammomatous, angiomatous, microcystic, secretory, lymphocyte-rich and metaplastic. [3]

Metaplastic meningioma is one of the rarest subtypes, and is defined as a tumor containing striking focal or widespread mesenchymal components including osseous, cartilaginous, lipomatous, myxoid, xanthomatous singly or in combinations. [3] Metaplastic meningioma with xanthomatous change, namely

“xanthomatous” meningioma has been extremely rarely reported. [4-7]

Xanthomatous meningioma should be differentiated from two entities- clear cell meningiomas and macrophages because clear cell meningiomas are biology aggressive Grade II tumors and presence macrophages indicates an inflammatory condition.

## CASE REPORT

A 24 years old Indian female presented with diminished vision, multiple episodes of vomiting and occasional episodes of headache. Fundoscopy revealed

grade IV papilloedema. Magnetic Resonance Imaging revealed an ill defined heterogeneously enhancing solid space occupying lesion measuring about 6.2cm (craniocaudal) x 7.2(AP) x6.7cm (transverse) epicentered over right temporo-parieto-occipital region with supratentorial as well as infratentorial extension.

**Gross examination:** We received multiple, irregular, grey white to grey brown, soft tissue pieces aggregating to 4.5x3x0.9cm. Cut-section revealed soft, grey- white to grey- brown areas.

Table1: Clinicopathological features of previously reported cases of Xanthomatous Meningioma

Case No.	Age/Gender	Location	Transition between conventional meningioma and xanthomatous area	Immunohistochemistry/special stain.	Reference
1	24/Male	Right parietal	+	CD68(+)	(3)
2	2/Female	Posterior pyramid	Not available	EMA (+)	(4)
3	10/Male	Right frontal	+	CD68 (+)	(5)
4	61/Female	Right occipital to parietal	+	EMA (+),CD68 (+),Ki-67 LI 2.2%	(6)
5	76/Male	Left parasagittal to frontal	+	EMA (+), CD68 (+), adipophilin (+), Ki-67 LI 2.1%	(7)
6-present case	24/ Female	Temporal-parieto-occipital region.	+	EMA (+), Special stain – PAS – Negative, Ki-67-3.2%	

EMA epithelial membrane antigen; Ki-67 labelling index.

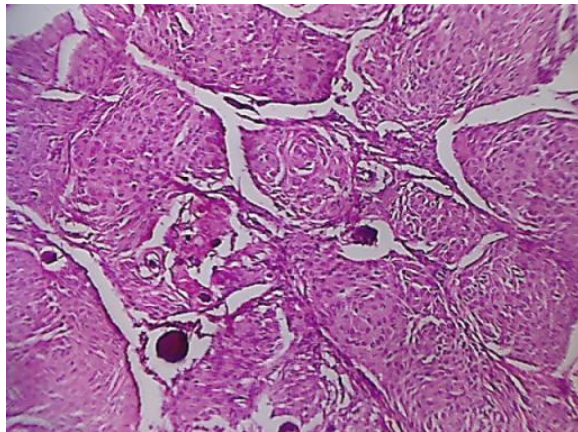


Figure 1: Microphotograph showing neoplastic meningotheial cells arranged in whorls and bundles & showing presence of psammomatous cells.

**Microscopic examination** revealed a benign meningeal tumor arranged in whorls and bundles. The cells were spindloid or round to oval having round to oval delicate

vesicular nuclei with inconspicuous nucleoli and moderate amount of pale to eosinophilic cytoplasm.(Figure1) The intervening stroma showed presence of round to oval cells containing small vesicular nucleus of moderate to ample amount of pale to granular cytoplasm arranged in diffuse sheets. Some of the cells exhibited clear cytoplasm. (Figure 2)Foci of psammoma bodies are also seen. The mitotic activity was not increased. The histopathological diagnosis of Xanthomatous meningioma was made.

Special stains were done, PAS was negative for foamy cells. IHC results showed tumor cells expressing EMA as well as the foamy cells were EMA positive. MIB1 (Ki-67) proliferation index approximately 3.2% in highly cellular areas.

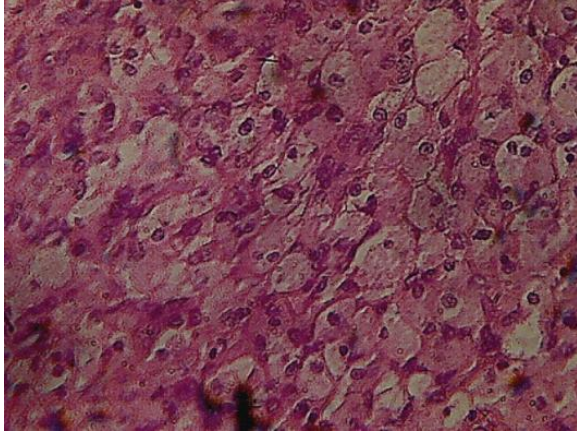


Figure2: Microphotograph showing neoplastic xanthomatous tumor cells having granular to foamy cytoplasm.

## DISCUSSION

The meningiomas most commonly occur at sites are the front half of the head that includes lateral cerebral convexities, midline along falx cerebri adjacent to the major venous sinuses parasagittally and olfactory groove. The term “Xanthomatous “ meningioma was first coined by Kepes in 1934.<sup>[3]</sup> Ours is the sixth documented case since then. Table 1 summarises the clinic-pathologic features of five previously reported cases of Xanthomatous meningioma. This tumor affects mainly children and young adults or elderly. (Table 1). The histopathologic hall mark was the presence of xanthomatous tumor cells<sup>[4-7]</sup> These xanthomatous cells were positive for Epithelial membrane antigen and negative for PAS. So they were not macrophages. Although extremely rare, the xanthomatous change has also been observed in atypical and anaplastic meningiomas as well as grade 1 meningiomas.<sup>[8,9]</sup>

Meningiomas are generally firmly attached to the dura and indent the surface of the brain but rarely invade it. Most meningiomas are benign and can be removed surgically.

## CONCLUSION

We have an extremely rare case of xanthomatous meningioma for its clinical,

histopathological and IHC findings. It is important to recognize the xanthomatous change occurring in a meningioma from the macrophages indicating inflammatory reaction. Secondly, some of these neoplastic cells show clear cell change in the cytoplasm which resembles clear cell meningioma grade II. It is important to recognize that xanthomatous change can occur in meningiomas and to avoid misidentifying these cells as macrophages.

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