XANTHOGRANULOMA (XG) MASQUERADING AS TUMOUR OF SELLAR REGION- A DIAGNOSTIC CHALLENGE!!

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PRESENTATION OF CASE

Xanthogranuloma of sellar region is rare and poses a diagnostic challenge for neurosurgeons, radiologists and pathologists. We report a rare case of sellar xanthogranuloma in a 25-year-old man who presented with complaints of retro-orbital headache along with progressive diminishing vision. Clinical examination of the right eye showed atrophy of the right fundus. MRI showed a well-defined cystic mass in sella region with clinical impression of pituitary macroadenoma. Surgical excision was performed and diagnosis was confirmed by histopathology. The spectrum of cystic pathology occurring in the sellar region includes many entities including xanthogranulomas. They should be included in differential diagnosis of intrasellar tumours.

Xanthogranulomatous reaction or otherwise called as "cholesterol granuloma" is characterised by presence of cholesterol clefts, lymphoplasmacellular infiltrates, foreign body multinucleated giant cells around the cholesterol clefts foamy macrophages (xanthoma cells), haemosiderin deposits, and fibrous proliferation and small epithelial cell clusters.[1,2]

Xanthogranulomas (XGs) are observed at various sites like including middle ear, mastoid bone, paranasal sinus and intracranial region; most common site being choroid plexus located in trigone of lateral ventricle.[3,4] Its location in the sellar region is extremely rare.[1]

Majority of intracranial XGs are benign and asymptomatic lesions that are usually found in 1.6-7% of CNS autopsies.[5] They are seen usually in adolescents and adults.[6]

Since reports on XG are rare, further reports are required, so that clinicians can gain insight into the course, management and outcome of XG.

A 25-year-old male presented was admitted with complaints of severe headache that was localised to retro-orbital region along with progressive diminishing vision. Clinical examination of eye revealed bilateral optic atrophy of fundus (Figure 1). MRI showed well-defined cystic mass in sella (app. 24 x 25 x 23 mm) with suprasellar extension reaching upto floor of the third ventricle and stretching the optic chiasma with impression of pituitary macroadenoma. It was hyperintense on T1w and T2w images with fluid-filled level suggestive of internal haemorrhage (Figure 2). There was no hormonal pathology. The mass was homogenously enhancing and was soft and fragile. The lesion was removed by surgical exploration via transsphenoidal selective tumorectomy 5 months after clinical onset. Histopathological examination showed a circumscribed area consisting of foamy macrophages, hemosiderin-laden macrophages, cholesterol clefts, lymphoplasmacytic infiltrate with small epithelial cell clusters. Final histopathology diagnosis of xanthogranuloma of sellar region was given Figure (3-6).

CLINICAL DIAGNOSIS

XGs often present with hormonal deficit (hypopituitarism), headache, visual field deficits (bitemporal hemianopia), hydrocephalus (due to foramen of Monro obstruction), generalised fatigue, weight loss, polyuria, polydipsia and changes in consciousness.

There are no typical radiological features for XG of sellar region. As per the previous literatures, majority (approx. 70%) of them show marked hyperintensity of the lesion on T1 weighted images, while 65% reveals iso to hyperintensity of the lesion on T2 weighted images. However, it's difficult to differentiate XGs from other sellar mass lesions preoperatively. On CT scan, craniopharyngioma appears as a partially-calcified mass, whereas no calcification is seen in XG. Cholesterol clefts appear as high-intensity signal in T1 scans and low in T2 scans, hemosiderin-T1 isointense and T2 low signal fibrosis shows both T1 and T2 low signals.[3]

The spectrum of tumours in the sella region that can come as differential diagnosis includes pituitary adenomas, craniopharyngiomas, Rathke's cleft cyst, metastatic neoplasms and granulomatous inflammation.

Compared to classical craniopharyngiomas, xanthogranulomas characteristically occur in adolescents and young adults predominantly at intrasellar location. They are smaller in size with more severe endocrine deficiencies, but they are easily resected and have better outcomes. On the contrary, craniopharyngiomas arise in the pituitary stalk mainly in the suprasellar space.

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PATHOLOGICAL DISCUSSION

XG of sellar region was first reported in 1988 with only 4 of 211 sellar and juxtasellar tumours (1.9%) showing typical features of cholesterol granulomas.[7] First case series of 37 cases were described by Paulus et al in 1999, which classified xanthogranuloma of sellar region as a distinct entity following, which WHO (2000) accepted that XG of sellar region as a specific brain tumour separating it from the classical adamantinomatous craniopharyngioma, which differs in its histological features, age of onset, symptoms and prognosis.[8]

The exact histogenesis of XG in sellar region is still on debate, but several aetiologies have been postulated. It is generally considered to be a secondary tissue reaction to degeneration, haemorrhage or inflammation.[3,9] Certain degenerative changes in pituitary adenomas and extreme adeno or infundibuloneurohypophysitis also play triggering factors in formation of giant cell granuloma.

Most xanthogranulomatous reactions at the sellar region are associated with epithelial lesions that bear some degree of overlap with adamantinomatous craniopharyngioma and Rathke's cleft cyst.[3] In adamantinomatous craniopharyngioma, the epithelial cells form stellate reticulum along with an intervening layer of polygonal cells, which are squamous in nature and the presence of wet keratin clearly differentiates craniopharyngioma from a XG lesion.

Role of IHC is minimal and maybe used to differentiate the lesion from pituitary adenoma. Surgery using transsphenoidal or transcranial approach is the best treatment option.[4] Surgical removal of the mass is necessary to achieve correct diagnosis and reduce the mass effect.[10] Xanthogranuloma of sella has a favourable outcome without any relapse after complete resection. In our case, the patient was relieved from his symptoms of headache and visual disturbance at the time of discharge.

FINAL DIAGNOSIS

So, a final diagnosis of xanthogranuloma of sellar region was opined in this case based on distinct histomorphological findings. The clinicopathological aspects of sellar xanthogranuloma and its differential diagnosis are dealt in this case report in a hope to understand the true nature and cause of this rare entity. It is difficult to diagnose XG preoperatively as there is no specific clinical and radiological feature and diagnosis mostly depends on histopathological examination after surgical intervention. It should be included in the preoperative DD of intrasellar tumours and should be operated at the earliest to improve the visual and endocrinological outcomes.

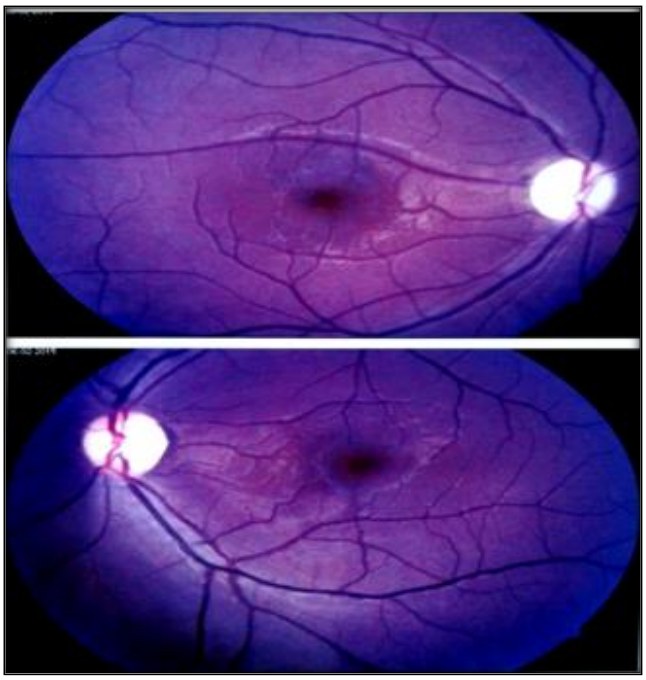

Figure 1. Fundoscopy Showing Optic Atrophy of Right Eye (above) and Left Eye (Below)

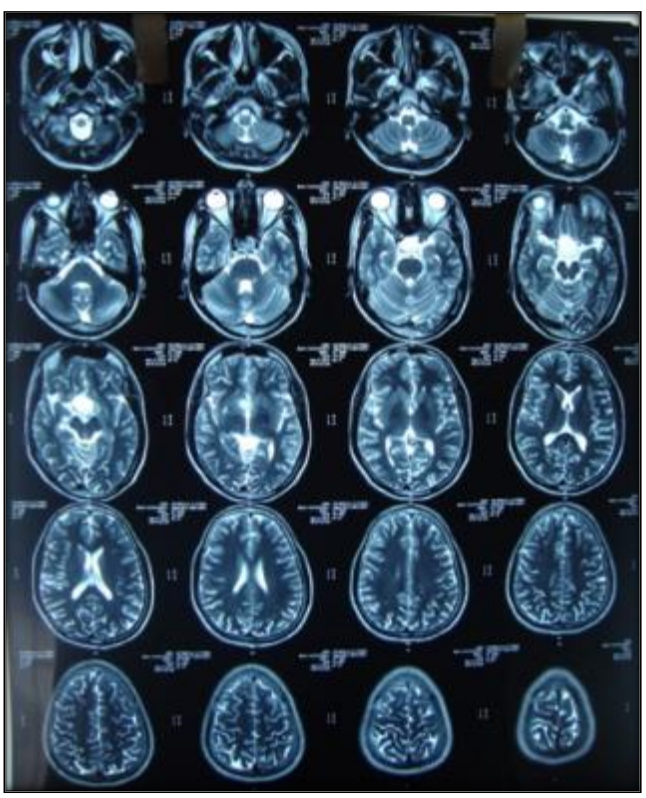

Figure 2. MRI Scan- Well-Defined Cystic Mass in Sella with Suprasellar Extension, Stretching the Optic Chiasma

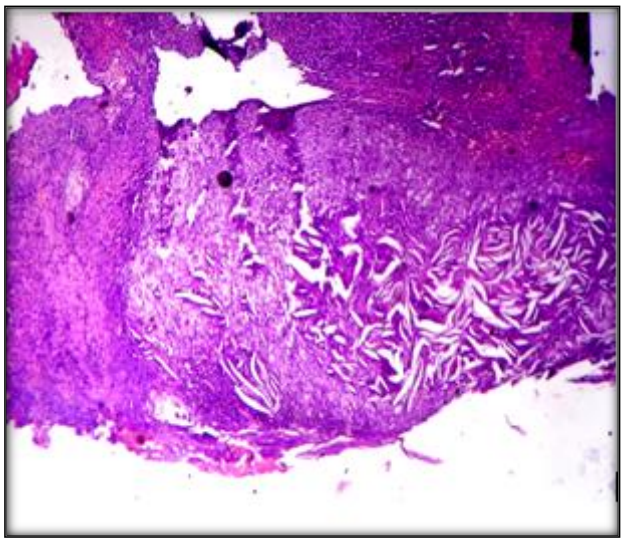

Figure 3(a). Photomicrograph (Scanner View 40X) Showing Areas of Granulation Tissue with Cholesterol Clefts

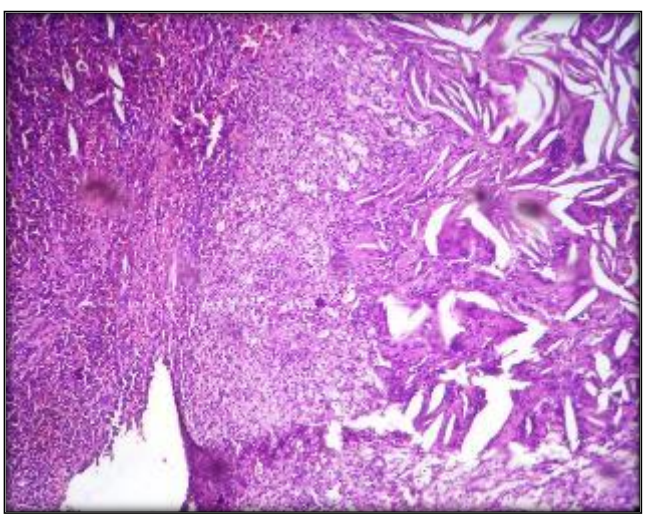

Figure 3(b). Photomicrograph (Scanner 40X)- Showing Band of Foamy Macrophages, Cholesterol Clefts and Lymphocytes

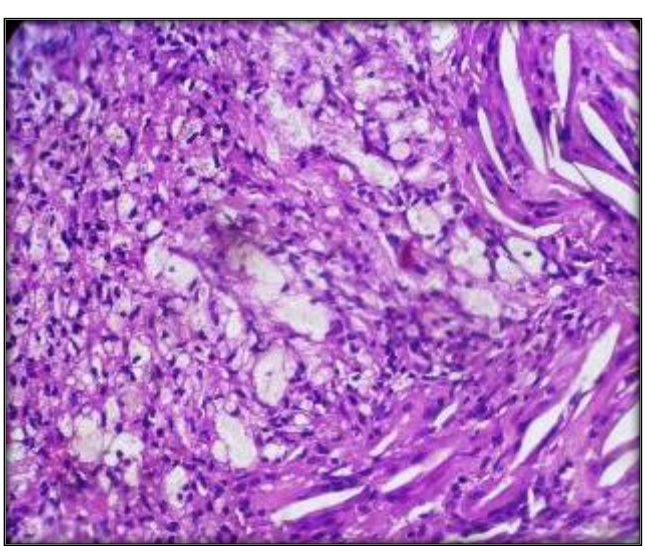

Figure 4. Photomicrograph (HP 100X)- Showing Foamy Macrophages Cholesterol Clefts and Lymphocytes

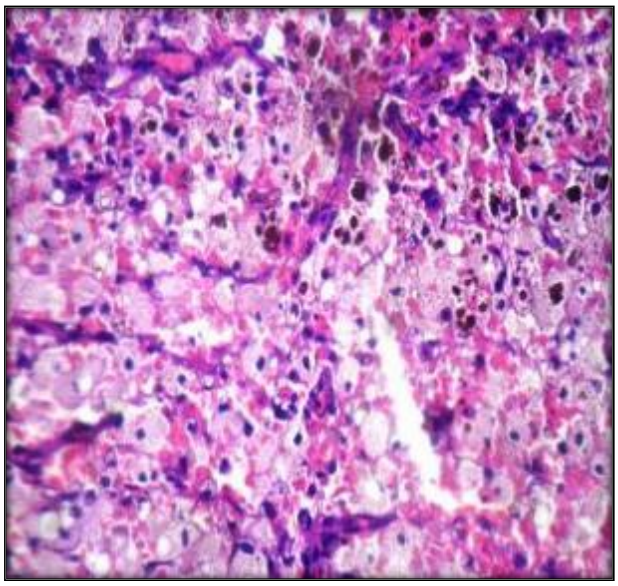

Figure 5. Photomicrograph (HP400x)- Showing Band of Foamy Histiocytes and Hemosiderin-Laden Macrophages

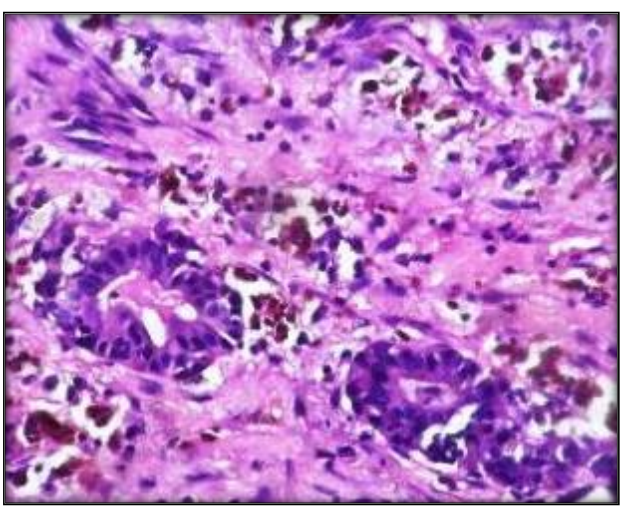

Figure 6. Photomicrograph (HP 400X)- Showing Epithelial Cell Cluster Lined by Low Columnar Epithelium and Hemosiderin-Laden Macrophages

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