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Case Report

Pathology Section

Orbital Cholesterol Granuloma: A Rare Case Report

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ABSTRACT

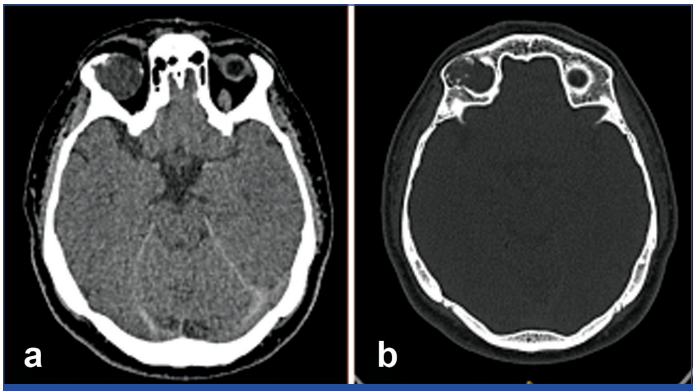
Orbital cholesterol granuloma is a rare entity which can cause proptosis, diplopia and extraocular movement restriction. It is often mistaken for other entities such as epidermoid, dermoid cysts and lacrimal gland tumours. Hereby, authors report a case of a 36-year-old male, who presented with proptosis of right eye and diplopia on upward gaze. Computed Tomography (CT) scan showed an extraconal lesion in superotemporal compartment of right orbit with erosion of bony wall. On imaging findings, epidermoid and dermoid cysts were considered as probable diagnoses. On histology, the lesion showed cholesterol clefts, foamy and haemosiderin laden macrophages and foreign body giant cells. Lesion was diagnosed as cholesterol granuloma. While being very rare at this site, this lesion carries a good prognosis with complete surgical excision. Patient showed marked postoperative improvement and was asymptomatic at discharge.

Keywords: Cholesterol clefts, Dermoid, Extraconal, Orbit, Proptosis

CASE REPORT

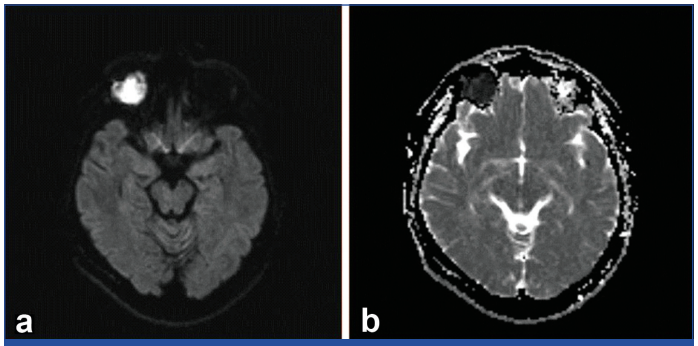
A 36-year-old male patient presented with gradually progressive proptosis involving right eye since three years. Patient also complained of occasional headache in right temporal region and double vision on upward gaze since two months. The thyroid function tests were within normal limit and he had no significant past history or co-morbidities. General and systemic examination conducted was within normal limits. Tests for visual acuity and colour vision were normal. Perimetry was normal for both eyes. He had upward gaze restriction on extraocular muscle function testing. Rest of the cranial nerves examination was within normal limits.

Computed tomography scan showed a well-circumscribed hypodense lesion with smooth margins in the extraconal superotemporal compartment of right orbit. Peripheral nodular calcifications were seen along its anterior and medial walls with erosion of the superolateral bony wall of the right orbit at the lacrimal fossa. No intracranial extension was seen [Table/Fig-1].

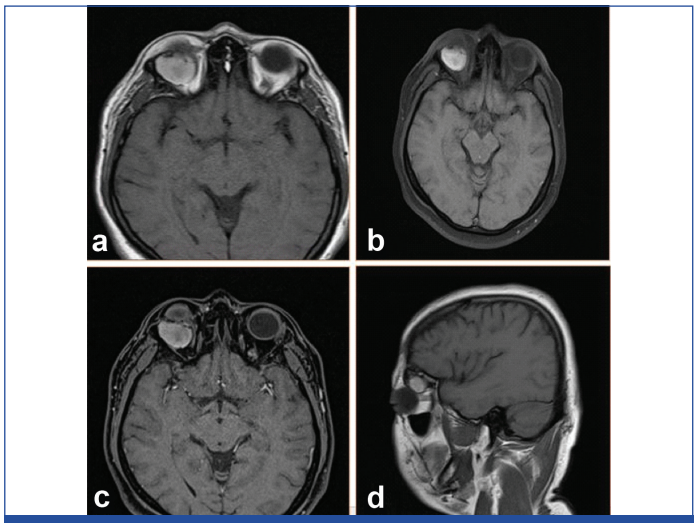


[Table/Fig-1]: Axial CT scan, (a) Brain window and (b) Bone window showed a well circumscribed hypodense lesion in the right orbit superotemporal extraconal compartment with peripheral nodular calcification causing erosion of the lacrimal fossa.

On Magnetic Resonance Imaging (MRI), the lesion was well-defined, round to oval, smoothly marginated with altered signal intensity. It measured 2.4×2.8×2.2 cm. It appeared predominantly hyperintense on T2 Weighted (T2W) and Fluid Attenuated Inversion Recovery (FLAIR) images with few discrete hypointensities at the periphery. It appeared hyperintense on T1W images and T1W fat saturated images. Nodular areas of blooming were seen at the periphery of the lesion representing calcifications. The lesion showed strong diffusion restriction on Diffusion-Weighted Imaging (DWI) with low Apparent Diffusion Coefficient (ADC) values. No significant post contrast enhancement was seen. No intracranial or intraconal extension was seen [Table/Fig-2,3].



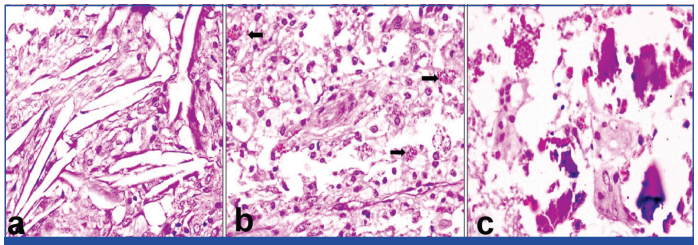
[Table/Fig-2]: (a) Axial diffusion weighted image: (b) Showed strong diffusion restriction within the lesion with reduced ADC values.



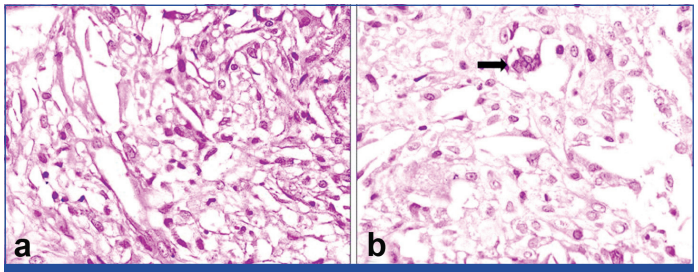
[Table/Fig-3]: (a) Axial T1W image showed T1W hyperintense contents of the lesion; (b) Axial T1W fat saturated images showed persistent high T1 signal without suppression; (c) Axial T1W postcontrast images showed no obvious enhancement (just intrinsic high T1 signal). (d) Sagittal T1W image showed extraconal location of the lesion in the superior compartment.

On imaging findings, epidermoid and dermoid cysts were considered as probable diagnoses. Patient underwent right sided frontotemporal incision and excision of the extraconal lesion. Intraoperatively, cheesy, chocolate coloured thick fluid with solid flakes came out under pressure from the junction of orbital capsule and the supraorbital ridge. The periorbital space was widened and more cheesy material was evacuated under pressure. The orbital capsule became lax. Fine dissectors were used to scrape the cheesy material off the bony wall of the orbit which was ragged due to the underlying pathology. Sample was received for histopathological examination in the form of multiple cheesy yellowish tissue bits. No cyst wall was identified grossly.

Microscopically, fibrous tissue was seen with dense aggregates of foamy macrophages along with prominent cholesterol clefts. Foreign body giant cells and haemosiderin laden macrophages were also seen along with areas of calcification [Table/Fig-4,5]. There was no evidence of cyst lining, epithelial elements or any neoplastic lesion. Considering the histology and clinico-radiological features, the lesion was diagnosed as cholesterol granuloma of the orbit.



[Table/Fig-4]: a) On histology, the lesion showed numerous cholesterol clefts (H&E, 40X). b) Sheets of foamy macrophages were seen along with haemosiderin laden macrophages (black arrows) (H&E, 40X). c) Foci of calcification within the lesion (H&E, 40X).



[Table/Fig-5]: a) Sheets of foamy histiocytes with interspersed cholesterol clefts (H&E, 40X). b) Foamy macrophages along with foreign body type of giant cell (black arrow) (H&E, 40X).

Postoperatively, proptosis had completely disappeared. There were no immediate postoperative complications and the patient was asymptomatic at discharge and also during six months follow-up period.

DISCUSSION

Cholesterol granuloma is a foreign body granulomatous reaction to cholesterol crystals which are derived from breakdown of the lipid layer of the cell membrane of ruptured red blood cells [1]. Middle ear, mastoid, petrous apex are some of the common sites of occurrence [2]. In the orbit, it's a rare entity with poorly understood pathogenesis [3]. On imaging it mimics dermoid or epidermoid cysts and lacrimal gland neoplasms [4]. Lung, breast, peritoneum and testes are some of the other uncommon sites for this lesion [2,5,6]. In the orbit, it is predominantly found in superotemporal extraconal compartment and very rarely in superomedial quadrant [2], intraconal location [7] and orbital floor [8]. Middle aged men are most commonly affected [2,6].

Several different mechanisms of pathogenesis of orbitofrontal cholesterol granulomas have been proposed. Trauma with bleeding is one of the proposed mechanisms, however, in quite a few reports including the present one, no history of trauma could be elicited [2,7,8].

Proptosis, decreased visual acuity, diplopia, extraocular movement restriction and pain are some of the reported symptoms [5]. In present case, the patient presented with progressive proptosis of right eye, right temporal headache and diplopia on upward gaze. His visual acuity and perimetry by confrontation method were normal for both eyes.

The mass appears on CT as an extraconal lesion in superotemporal orbit often associated with marked bony changes in the form of bone erosion, destruction and thinning of inner and outer tables [2]. On MRI, the lesion shows high signal on T1W whereas, T2W images show central high signal intensity with or without low peripheral signal due to haemosiderin rim. The high intrinsic T1 signal, without suppression on fat saturated images is due to the paramagnetic effect of heme iron in methaemoglobin [9,10]. On imaging, the lesion mimics lacrimal gland tumours, dermoid and epidermoid cysts, aneurysmal bone cysts, eosinophilic granulomas, mucoceles and metastasis. Many of these can be differentiated based on their typical clinical and radiological features [11]. CT and MRI findings in present case were suggestive of epidermoid or dermoid cyst.

Intraoperatively, various reports have described a cystic lesion with yellow-grey wall containing green, mucinous fluid or curry sauce like fluid or golden-brown material [2,11]. In the present case cheesy, brownish material was aspirated. Bony changes were identified intraoperatively.

Histological features in present case were very typical of cholesterol granuloma. Cholesterol clefts, foamy macrophages, haemosiderin laden macrophages, foreign body giant cells and calcification have all been described in various case reports [2,5,11]. Since, the lesion did not have any epithelial elements or cyst lining authors did not consider any other differentials such as a dermoid cyst or a neoplastic lesion. Findings of the current case are compared with the other published case reports [Table/Fig-6] [2,4,5].

Author's name	Age/ Sex	Clinical presentation	Location in orbit	Histological findings
Rizvi SA et al., [2]	42/Male	Progressive proptosis	Extraconal superomedial	Cholesterol crystals, macrophages, foreign body giant cells.
Rong AJ et al., [5]	38/Male	Progressive ptosis, hypoglobbus	Superotemporal	Histiocytes, cholesterol clefts, haemosiderin.
Ugga L et al., [4]	73/Male	Proptosis, decreased visual acuity	Superior aspect	Chronic granulomatous inflammation, haemosiderin, calcification, red cell extravasation.
Vaidya MM et al., (Present case)	36/Male	Proptosis, diplopia on upward gaze	Extraconal, superotemporal	Cholesterol clefts, haemosiderin laden marcophages, foamy macrophages, foreign body giant cells.

[Table/Fig-6]: Comparison of findings of the present case with other case reports [2,4,5].

Complete removal of the lesion with drainage and curettage is curative with a low incidence of recurrence [11]. Surgical resection is often followed by improvement in proptosis, diplopia and EOM restriction [5].

CONCLUSION(S)

Orbital cholesterol granulomas are rare entities that are often mistaken for other benign or malignant lesions. It should always be considered in the differential diagnosis of a superotemporal orbital lesion with prominent bony changes. Complete surgical excision carries a good prognosis with a low chance of recurrence. Careful evaluation of clinico-radiological and histopathological features is required for accurate diagnosis and optimum management of the patient.

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