Cholesterol granuloma of the orbit: An atypical presentation

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Cholesterol granuloma is a rare, well-defined lesion of the orbit. In the orbit, diploe of the frontal bone is involved almost exclusively. We report an atypical case of cholesterol granuloma involving superomedial quadrant of orbit. A 42-year-old male presented with progressive, painless, proptosis with infero-temporal displacement of left eye. A large mass was felt beneath the bony orbital margin in the superomedial quadrant of the left orbit. Computerized tomography (CT) scan revealed an extraconal superomedial, heterogeneous enhancing mass which was isodense with brain and pushing the globe inferolaterally and anteriorly. Excision biopsy of the tumor revealed the typical features of a cholesterol granuloma without any epithelial elements. Cholesterol granuloma of the orbit is a rare entity, but it can be diagnosed and differentiated from other lesions of the superior orbit by its characteristic clinical, radiological and histopathological features. An appropriate intervention in time carries a good prognosis with almost no recurrence.

Key words: Atypical, cholesterol granuloma, orbit

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Cholesterol granuloma is a foreign body response to the presence of crystallized cholesterol.[1] It is a rare but well characterized orbital entity which occurs almost exclusively within the frontal bone overlying the lacrimal fossa.[2] Other regions of the head, namely; zygomatic bone, and petrous bone, have also been found to be involved.[1,3] The few cases that have been reported in the last decade have a superolateral presentation.[2-5] We report an atypical case of cholesterol granuloma in a 42-year-old male, involving superomedial quadrant of orbit. Detailed search on search engine MEDLINE could not find any such case in the literature.

Case Report

A 42-year-old male presented with progressive, painless, proptosis of left eye for the past 18 months. The onset was insidious and had been associated with progressive diminution of vision in the same eye. No history of trauma or previous surgery could be elicited.

Orbital examination revealed 4 mm of proptosis with inferolateral displacement of the globe [Fig. 1]. A large mass was felt beneath the bony orbital margin in the superomedial quadrant of the left orbit. The mass was firm in consistency. Ocular movements were restricted in the upgaze. On ocular examination, the best corrected visual acuity of right eye was 20/20, whereas in the affected eye it was 3/200. Near vision in the right eye was N6, while in the left eye it was N60. Also,

color vision was impaired and Amsler grid showed distortion of horizontal and vertical lines in the left eye. Choroidal folds were present on fundoscopy.

Computerized tomography (CT) scan of left orbit revealed an extraconal superomedial, heterogeneous enhancing mass measuring 3.7 × 3.2 × 2.6 cm which was isodense with brain and pushing the globe inferolaterally and anteriorly. There was thinning of cortices in medial wall, lateral wall, and roof of left orbit with bony deficit in the lateral aspect of roof of left orbit [Fig. 2]. No mass was felt or seen in the typical superolateral area of the orbit either on radiological or on clinical examination. Routine investigations were within normal limits.

The mass with golden brown content was completely excised by anterior orbitotomy approach through an upper eyelid crease incision. Curettage of the frontal bone was also done to remove the residual granulomatous material. Histopathological examination shows cholesterol crystals surrounded by foreign body giant cells, packed macrophages, lymphocytes and extravasated RBCs [Fig. 3a and b]

Discussion

Orbitofrontal cholesterol granuloma is a rare entity; mostly occurring in middle-aged men and generally presents with a gradual mass effect in the superolateral orbit.[2] The first orbital case was reported by Denig in 1902.[6]

The common sites of occurrence are the middle ear, mastoid antrum and petrous apex.[7] Less commonly, it has also been reported in frontal bone, zygoma, paranasal sinuses, breast, peritoneum, testes, intima of atheromatous arteries, lung and within tumors such as thyroid adenoma and odontoma.[2]

Though the exact event responsible for such granuloma has not yet been elucidated, different mechanisms have been suggested for its formation.

Trauma with hemorrhage has been put forward as the initiating event in the cholesterol granuloma of the orbit. The predilection of frontal bone and the male preponderance further support the trauma theory.[3] However, there was no history of trauma in our case. There are also some reports where no history of trauma could be elicited.[3]

A ventillatory obstruction in the petrous apex has also been proposed as a cause of cholesterol deposition.[8] However, this mechanism cannot be applied to orbital cholesterol granuloma as they are quiet separate from frontal sinus.[4]

The typical CT findings are of a non-calcifying extraconal mass in the supero-temporal quadrant of the orbit which is isodense with brain and is associated with thinning of the inner and outer tables and ragged areas of bone erosion with destruction.[4] On rare occasions, the mass can be hypodense in nature.[9] In our case, the CT findings are typical of cholesterol granuloma except the atypical site of presentation which was superonasal quadrant of orbit.

Histologically, it is characterized by cholesterol clefts surrounded by granulomatous inflammation with predominance of foreign body giant cells and blood-derived debris.[1] Epithelial elements are characteristically absent.[2] This absence of epithelial elements differentiates cholesterol granuloma from one of its important differential diagnosis, the dermoid cyst.[4] Other important conditions which have to be differentiated from orbital cholesterol granuloma include

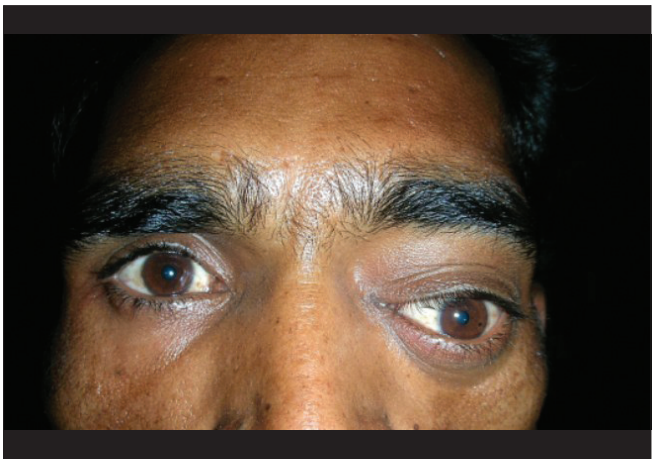

Figure 1: Superomedial mass with inferolateral displacement of globe

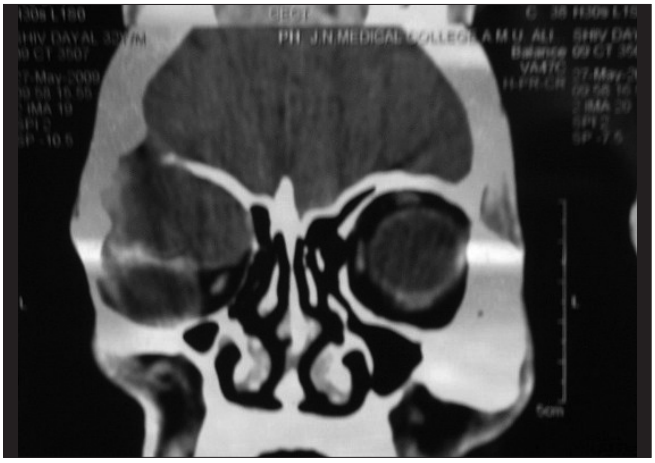

Figure 2: CT scan with an extraconal superomedial heterogeneous enhancing mass which is isodense with brain

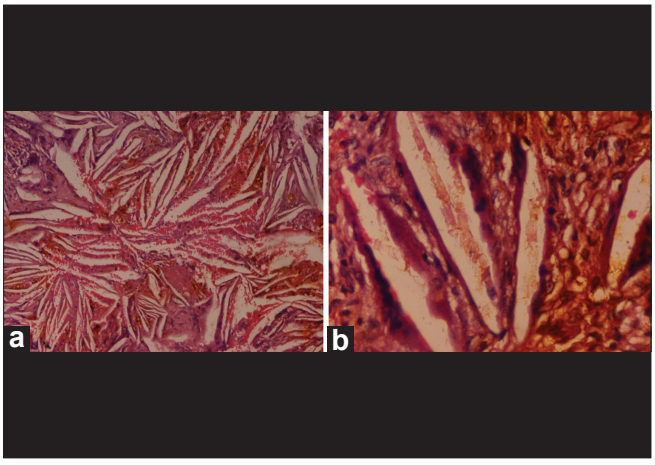

Figure 3: Photomicrograph showing (a) cholesterol crystals surrounded by foreign body giant cells, packed macrophages, lymphocytes and extravasated RBCs (H and E, ×100). (b) Magnified view of cholesterol crystals with cholesterol clefts (H and E, ×400)

lacrimal gland tumor and aneurysmal bone cyst. However, in our case, a differential diagnosis of lacrimal gland tumor was not considered due to the superonasal presentation of the mass.

The majority of aneurysmal bone cysts present in the second decade with a female preponderance of 5:3.[1] The characteristic histological feature of aneurysmal bone cyst includes cavernous blood-filled spaces that lack endothelial lining, pericytes, or smooth muscle, and also cholesterol crystals are absent.[1]

Definitive management of cholesterol granuloma includes removal of the granulomatous mass via anterior or antero lateral orbitotomy, with curettage of the residual material adherent to bone.[4] Although recurrence is quiet rare, the curettage helps in preventing the recurrence.[10]

To conclude, cholesterol granuloma of the orbit is a rare entity and should be differentiated from other lesions of the superior orbit, i.e. lesions of both superolateral and superomedial quadrant. An appropriate intervention in time carries a good prognosis with almost no recurrence.

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