

Case report

Orbital cholesterol granuloma: A report and discussion of orbital findings

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ABSTRACT

Purpose: To report a case of orbital cholesterol granuloma and discuss the orbital findings seen in this entity.

Observation: A 38-year-old male presented with an 8-month history of progressive left upper lid ptosis and hypoglobus. Clinical examination was significant for 3 mm of hypoglobus and restricted supraduction in the left eye. Contrast-enhanced computed tomography imaging revealed a well-circumscribed lesion in the superotemporal orbit causing extensive bone erosion that appeared to arise from the lacrimal gland. An incisional biopsy was performed, and histopathological evaluation demonstrated fibrovascular tissue surrounding a mixture of histiocytes and cholesterol clefts, consistent with a cholesterol granuloma.

Conclusions and importance: Orbital cholesterol granulomas are rare lesions that are predominantly found in the superotemporal orbit. These lesions can be associated with marked bony changes in the superotemporal fossa that can be mistaken for a lacrimal gland neoplasm; however, bony erosion is a hallmark of this lesion and should be considered on the differential diagnosis of any lacrimal gland mass with extensive bony erosion.

1. Introduction

Cholesterol granuloma describes an accumulation of organized blood byproducts, including cholesterol clefts, hemosiderin, and hematoxin, that lacks both an epithelial and endothelial lining.¹ These lesions arise in a broad range of anatomical locations, including the petrous apex, lung, and breast.^{2–4} In the orbit, the inciting event for cholesterol granuloma formation is an orbital hemorrhage from any cause, including minor trauma or spontaneous hemorrhage in a patient on anticoagulation.⁵ Although histopathologically synonymous, “cholesterol granuloma” has traditionally been applied to lesions throughout the body, whereas “hematic cyst” has only been applied to orbital lesions. Classically, hematic cysts were thought to be secondary to remote trauma, whereas cholesterol granulomas were ascribed an etiology based on anatomical location.⁶ Over time, reports of hematic cysts not associated with antecedent trauma became more common, and the term no longer necessarily implies a traumatic etiology.

Despite this, the terms “hematic cyst” and “cholesterol granuloma” both continue to pervade the literature, leading to confusion, fragmented descriptions, and incomplete characterization of this synonymous lesion.^{1,7–9} We report a case of an orbital cholesterol granuloma

and discuss characteristic findings and presentations for this entity after reviewing all reported cases in the literature to date under its various nomenclatures.

2. Case report

A 38-year old male presented with an 8-month history of progressive left upper lid ptosis and hypoglobus. He denied pain, paresthesias, diplopia, or decreased vision. On clinical exam, visual acuity and intraocular pressures were normal and there was no axial proptosis. However, there was 3 mm of left hypoglobus associated with upper lid ptosis (Fig. 1) and limited supraduction.

Computed tomography with contrast revealed a non-enhancing, well-circumscribed lesion in the superotemporal orbit with extensive erosion of the adjacent frontal bone and lacrimal gland fossa (Fig. 2). Magnetic resonance imaging with gadolinium showed a non-enhancing mass with intermediate to high signal intensity on T1- and T2- weighted imaging that appeared to arise from the lacrimal gland (Fig. 2).

Given the aggressive bone erosion, an orbitotomy was performed. Intraoperatively, the lateral orbital wall was found to be discolored with diffuse bone pitting. A sizeable lytic bone lesion measuring 12 × 6

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Fig. 1. Above: A 37-year-old male presents with progressive left hypoglobus and ptosis. Below: Three months status post orbitotomy and incisional biopsy of the orbital lesion, there is complete resolution of hypoglobus.

mm was found in the superotemporal orbit (Fig. 3). The periosteum was adherent to the pitted orbital bone and the adjacent orbital fat appeared infiltrated. Histopathologic evaluation disclosed fibrovascular tissue surrounding an admixture of histiocytes, cholesterol clefts, and hemosiderin. No malignancy was identified, and the findings were consistent with a cholesterol granuloma (Fig. 4). Postoperatively, the patient had complete resolution of hypoglobus and lid ptosis and regained to full extraocular motility (Fig. 1).

3. Discussion

For the purposes of this discussion, we will use the term “orbital cholesterol granuloma” as a unifying term to discuss the histopathologically identical terms *blood cyst*, *hematocele*, *xanthomatosis of the orbit*, *cholesteatoma*, *hematic cyst*, and *orbitofrontal cholesterol granuloma*. In a systemic review of orbitofrontal cholesterol granulomas, Hughes et al. focused on lesions found only in the superotemporal orbit.⁷ However, further review of the literature shows that these lesions can also be found in the intraconal space and orbital floor. A systematic PubMed search was performed using the above synonymous nomenclature and these additional cases are compiled in Table 1 (any case not involving the orbit or not meeting the core histological definition was excluded).

With the addition of cases within the intraconal space and orbital floor, we still see that 92% (172 cases) of cholesterol granulomas localize to the superior or superotemporal orbit.⁷ This location is not only a common site of impact in blunt trauma but also has the weakest attachment of the periorbita to the bone and constitutes the largest continuous concave surface within the orbit.^{10,11} Common presenting signs and symptoms included proptosis, globe displacement, limitation of extraocular movement, diplopia, pain, and decreased visual acuity.⁷ Our patient developed painless, progressive hypoglobus over the course of several months. Interestingly, while many patients have choroidal folds on examination, this finding was not always associated with a decreased visual acuity worse than 20/20, highlighting the importance of a dilated fundus examination.

Lesions found along the orbital floor represent 6% (11 patients) of all cases, and all were associated with prior orbital fracture repair with an implant. Most implants were smooth alloplastic materials, such as silicone, nylon foil, and polytetrafluoroethylene.^{8,12–14} Unlike lesions in the superior and superotemporal orbit, lesions along the orbital floor were not characterized by bony erosion or expansion. The lack of bony involvement may be related either to the initial loss of bone in the

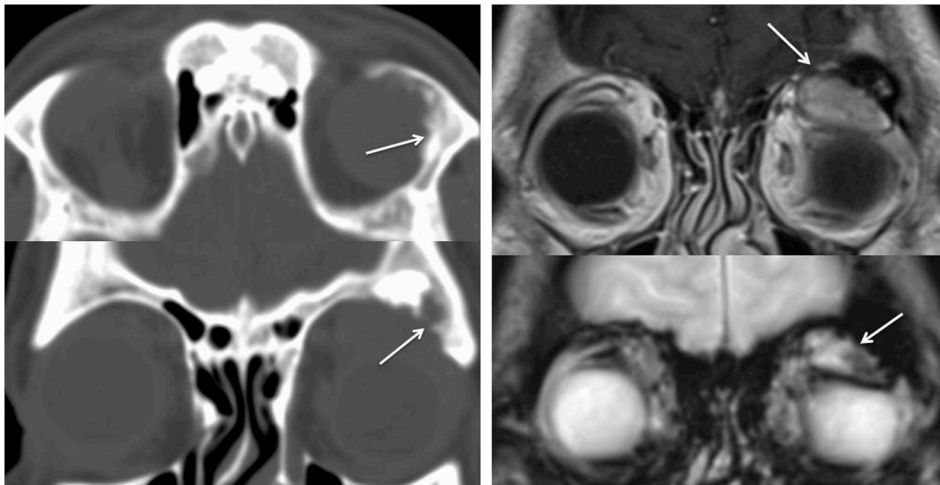


Fig. 2. Upper left: Axial CT image showing a cystic lesion in the superotemporal orbit with associated erosive bony expansion of the superotemporal orbital wall. Lower left: Coronal CT image showing a 12 mm × 6 mm lesion with bony expansion and adjacent sclerosis. Upper right: T1-weighted MRI with gadolinium shows a non-enhancing mass with intermediate to high signal intensity that appears to arise from the lacrimal gland. Lower right: T2-weighted MRI with gadolinium shows a lesion with high signal intensity in the superotemporal orbit.

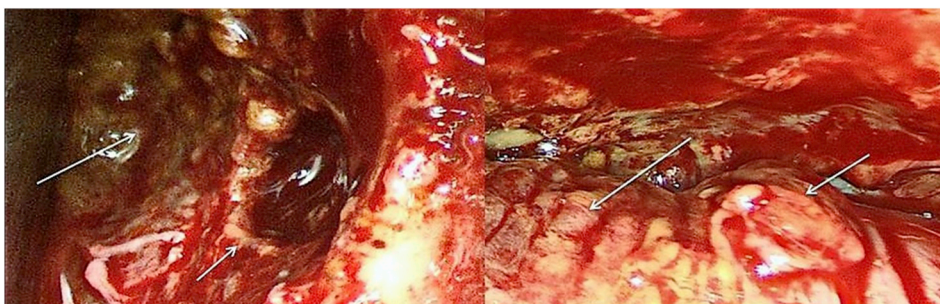


Fig. 3. Left: Intraoperatively, a large 12 mm × 6 mm lytic bone lesion (short arrow) was visualized with diffuse pitting of the superotemporal orbital bone (long arrow). Right: The orbital cholesterol granuloma (short arrow) lacked a clearly defined pseudocapsule, and the adjacent orbital fat (long arrow) appeared abnormal and potentially infiltrated by a malignancy.

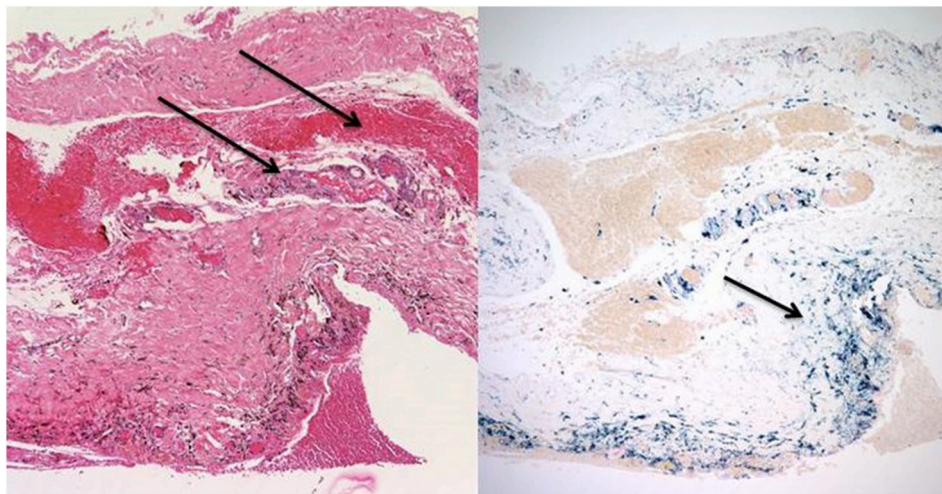


Fig. 4. Left: On Hematoxylin and eosin staining (100X), the lesion shows a fibrovascular wall surrounding inflammation and hemorrhage. No endothelial or epithelial structures are noted. Right: Prussian blue staining (100X) shows hemosiderin and hematoid deposition within the fibrovascular tissue.

Table 1

Additional 15 cases of orbital cholesterol granulomas found in the orbital floor or intraconal space included in the “orbitofrontal cholesterol granuloma” review of 172 cases by Hughes et al.⁷

Author (Year)	Age	Location	Bony Erosion	Notes
Wolter et al. (1966) ²¹	54M	Floor ^a	N	No Trauma
Mauriello et al. (1984) ¹⁵	33M	Floor	N	Trauma s/p repair
	33F	Floor	N	Trauma s/p repair
Milne (1987) ¹²	17M	Floor	N	Trauma s/p repair
Cameron (1988) ¹⁹	24F	Intraconal	N	No Trauma
Amrith et al. (1990) ¹⁸	3F	Intraconal	N	N/A
Goldberg et al. (1992) ²⁰	35M	Intraconal	N	Trauma
McCannel et al. (1996) ¹³	49M	Floor	N	Trauma s/p repair
Kang et al. (1996) ¹⁴	27M	Floor	Y	Trauma s/p repair
Iwata et al. (2000) ¹⁶	22M	Intraconal	N	Trauma
Glavas et al. (2004) ⁸	89F	Floor	N	Trauma s/p repair
Lee et al. (2011) ²²	34M	Floor	Y	s/p repair
Chao et al. (2015) ²³	N/A	Floor ^a	N	Trauma s/p repair
Shrirao et al. (2016) ²⁴	48M	Floor	Y	No Trauma
Ochoa-Escudero et al. (2018) ²⁵	28M	Floor/Maxilla ^a	N	Trauma s/p repair

^a Histology unclear.

trauma or the implant itself acting as a barrier to bone changes from an expansile hemorrhage. Although some reports describe an interval as long as 20 years from the time of fracture repair to presentation of a cholesterol granuloma, an acute hemorrhagic episode may occur with rapid onset of symptoms.^{14,15} This contrasts with the insidious and gradual development of lesions in the superior, superotemporal, and intraconal locations.^{10,16–18}

Lesions found within the intraconal space are extremely rare with only four reported cases representing about 2% of all documented cases.^{16,18–20} All patients presented with decreased visual acuity and choroidal folds. Consistent with their location, these lesions were not associated with bone erosion.

Interestingly, the superonasal and medial orbit appear to be spared by this entity. Although previous authors have described reports of a cyst occurring in the superonasal orbit, further review of the associated histology demonstrated an endothelial lining to the lesion, thus eliminating it from the definition of a cholesterol granuloma.²¹

Most patients do well following surgical resection or debulking of cholesterol granulomas, with improvement in globe position, proptosis, diplopia and restriction of extra-ocular movements. In our case, an incisional biopsy was sufficient to precipitate complete resolution of the patient's signs and symptoms. Amrith et al. found that even in cases of severe visual impairment, vision can improve from hand motion to 20/20 post-operatively; only one patient in their cohort had a poor visual

outcome and this patient presented with no light perception vision.¹⁸

Ultimately, these lesions are rare if found intraconally or at the orbital floor. When found in the superotemporal orbit, these lesions may commonly be mistaken for a malignant neoplasm. However, superotemporal orbital cholesterol granulomas are almost always found with prominent bony changes and should be considered in the differential diagnosis for any mass presenting in the lacrimal gland fossa.

Patient consent

Patient consent was obtained to publish these images.

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Conflicts of interest

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Authorship

All authors attest that they meet the current ICMJE criteria for

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.ajoc.2019.100468>.

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