ABSTRACT

Objective
To report a patient with cholesterol granuloma and describe the distinct clinical, radiologic, and histopathologic findings.

Methods
This is a case report.

Results
A 33-year-old man presented with a 12-month history of diplopia on left upward gaze. He underwent repair of wound laceration at the right fronto-temporal area 25 years prior to consultation due to a head bump sustained in a vehicular accident. Examination revealed nonaxial proptosis, inferior globe displacement, and mild limitation on left upward gaze. Orbital imaging revealed an ovoid extraconal expansile soft-tissue mass in the left orbit, slightly compressing the globe inferiorly, and thinning and widening of the superior portion of the orbital wall. Excision biopsy of the orbital mass was done through a lateral orbitotomy with bone flap. Histopathology revealed characteristic features of cholesterol granuloma including abundant cholesterol clefts, foreign-body giant cells, lipid-laden histiocytes, and hemosiderin macrophages with absence of epithelial components.

Conclusion
Orbitofrontal cholesterol granulomas have typical clinical, radiologic, and histopathologic features. Surgical excision has a high success rate with a low incidence of recurrence.

Keywords: Cholesterol granuloma, Orbit, Orbital lesion, Diplopia, Proptosis
CHOLESTEROL granuloma is a rare expansile orbital lesion with an unclear pathogenesis. It presumably arises from organization of incompletely resorbed orbital hemorrhage caused by trauma, orbital surgery, foreign body, hemorrhagic diathesis, or vascular lesion, with subsequent granulomatous response to blood-breakdown products, fibrous encapsulation, and recurrent hemorrhage into the cyst.1, 2

We present a patient with cholesterol granuloma of the left orbit, resulting in proptosis, globe displacement, and limitation in ocular movement. This report describes the clinical, radiologic, and histopathologic features of a case of orbital cholesterol granuloma.

METHODOLOGY
A 33-year-old-man presented with a 12-month history of diplopia on left upward gaze. He had a history of gout with regular intake of allopurinol and colchicine. He underwent repair of wound laceration at the right fronto-temporal area 25 years prior to consult due to a head bump sustained from a fall. There was no history of diabetes, hypertension, tuberculosis, asthma, or allergies. On examination, best corrected visual acuity was 20/20 bilaterally; there was no periorbital edema, erythema, or tenderness. A 2.5-mm left nonaxial proptosis with inferior globe displacement was noted, as well as mild limitation on left upward gaze. There was also prominence of the left lateral brow area, noted by the patient for 20 years to have slowly progressed with time. The remainder of the ocular examination was unremarkable.

Computed tomography (Figure 1) showed an ovoid extraconal expansile soft-tissue mass with a slightly enhancing rim in the superolateral portion of the left orbit measuring approximately 2.0 x 2.5 x 2.9 cm and slightly compressing the globe inferiorly, with thinning and widening of the superior portion of the orbital wall. The bony walls of the right orbit were unremarkable. The globes, extraocular muscles, optic nerves, and visualized portions of the brain were within normal limits.

All blood-test results were normal. Radiography of the chest, electrocardiogram, and urinalysis were also normal.

The patient underwent excision biopsy of the orbital mass through a lateral orbitotomy with bone flap. After reflection of the periorbita from the orbital roof, a reddish brown mass adherent to the irregular bony defect was encountered. This was accompanied by the release of moderately thick reddish-brown fluid.

RESULTS
Histopathologic examination disclosed fragments of yellowish brown tissues measuring 2 x 2 x 1 cm in aggregates. Microscopic sections showed a nonencapsulated dense fibrous connective tissue with no evidence of epithelial elements or keratin. There were numerous cholesterol clefs scattered within a chronic inflammatory infiltrate. Foreign-body giant cells enveloped most cholesterol clefs. Aggregates of birefringent hematoidin crystals surrounded by giant cells were present. Large foamy histiocytes, hemosiderin-laden macrophages and occasional fragments of bone were scattered throughout the connective tissue (Figure 2).

The patient tolerated the procedure well and there were no complications. He was given oral antibiotics and analgesics postoperatively. There was resolution of the diplopia but persistence of the prominence of the left lateral brow area. At his last clinic visit 16 months after the surgery, there was no recurrence of the mass.

DISCUSSION
Cholesterol granuloma is an uncommon lesion that usually affects middle-aged men.3 The largest series of cases

Figure 1. Coronal (A) and axial (B) sections showing a left orbital mass with thinning and widening of the superior portion of the orbital wall (C).
Figure 2. Cholesterol granuloma. A: Cholesterol clefts (φ) (H & E x 40). B: Foreign body giant cells (φ) surrounding cholesterol cleft (H & E x 180). C: Hematoidin crystals (φ) with foreign-body-giant cell (↑) (H & E x 100). D: Lipid laden macrophages (φ) (H & E x 180). E: Bone fragment (φ) (H & E x 40). F: Hemosiderin-laden macrophages staining blue with Prussian blue stain x 80.
was reported by McNab et al., who described the clinical features and management of 27 patients with orbitofrontal cholesterol granuloma. The term “cholesteatoma” was introduced in 1838 by Muller for any lesion containing cholesterol crystals. Both epidermoid cholesteatoma and cholesterol granuloma consist of cholesterol clefts and foreign-body giant cells with blood-degradation products such as hemosiderin and hematoidin. Cholesterol granuloma is distinguished from an epidermoid cholesteatoma by the lack of epithelial elements.5

Several reports were published using different names to describe these lesions including chronic hematic cyst,6, 7, 8, 9 xanthomatosis of the orbit,10 and lipid granuloma of the frontal bone.11, 12 However, there should be a uniformity with regard to the nomenclature. Cholesterol granuloma is a better term as suggested by McNab et al. since the osteolytic changes is a result of a granulomatous reaction to cholesterol crystals.1 Hematic cyst should be reserved to describe intraorbital accumulations of blood. Cholesterol granuloma should also be distinguished from acute subperiosteal hematomas which often result from trauma in children13, 14 or in adults.15

The cholesterol in the lesion results from the breakdown of cell membranes of erythrocytes. The crystals stimulate granulomatous inflammation that sets in capillary growth and further accumulation of red blood cells and other breakdown products producing a mass effect. Suggested possible etiologies for the hemorrhage include facial or head trauma16 and predisposing intradiploic anomaly.3

The frontal bone is the largest concave bony surface in the orbit. Granulomas tend to arise in the diploe of the frontal bone, causing expansion and eventual erosion of the inner and outer tables. The underlying peristeam in this area is not firmly attached as compared to other areas of the orbit. This could explain the common subperiosteal location of these granulomas.14

Bone resorption present in computed tomography could be attributed to prostaglandins, specifically PGE2, as seen in malignant lesions as well as in mucoceles of the paranasal sinuses and benign dental cysts.17, 18 The transudation of red blood cells and other blood products specifically platelets provides a good source of these prostaglandins.

Osteolytic lesion with a density equivalent to the density of the brain and occasional intralesional bone fragments are characteristic features seen in computed tomography. Lesions present as high T1 and T2 signal intensities on MRI that could provide added information in difficult cases. Dermoid cysts and lacrimal-gland tumors are common lesions that should be differentiated from this entity.

Patients usually present with a superotemporal mass ranging from weeks to years as seen in our patient. This would progress and lead to displacement of the globe inferiorly and proptosis. Ocular motility restriction with associated diplopia is present in majority of patients with cholesterol granuloma.6, 8 There may be accompanying orbital pain, which should be differentiated from a malignant process such as adenocystic carcinoma of the lacrimal gland that invades the nerve early in the disease process.

In conclusion, orbitofrontal cholesterol granulomas have typical clinical, radiologic, and histopathologic features. Surgical excision has a high success rate and leads to a low incidence of recurrence of the condition.

References