

The Case of the Ex-Marine With Exophthalmos

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After serving in the Marines as a mechanic, Luis Cantalejo* held a series of jobs, most recently managing an auto body repair shop. The 36-year-old Hispanic man had been in this position for little more than a year, and for much of that time, his workmates, friends and family had been telling him that his right eye “did not look normal.” They had noticed that it was bulging forward and downward. Eventually, after much urging from his wife, he saw a physician and was then referred to us.

We Get a Look

When we spoke to Mr. Cantalejo, he denied trauma, double vision, blurry vision or change in vision. He had no associated eye pain, redness, irritation, tearing or photophobia. Old photos from four years prior showed no proptosis or displacement.

On review of systems, he told us

that he hadn’t experienced any fever, weight loss, nausea or vomiting, headache, or nasal or sinus problems. He also hadn’t noticed any dizziness, tingling, tremors, numbness or weakness. His medical history included seasonal allergies and he had been taking over-the-counter Claritin (loratadine) as needed. He had no known drug allergies and denied tobacco use. His

ocular history included soft contact lens wear, and he had not undergone any ocular surgery.

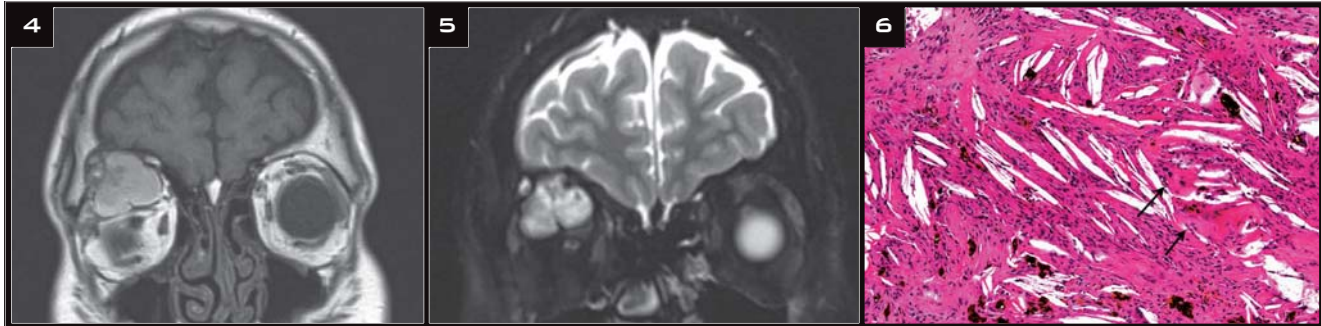
On exam, his BCVA was 20/25 in the right eye and 20/20 in the left. The cycloplegic refraction was $-1.25 +1.50 \times 085$ in the right eye and -3.75 sphere in the left. Goldmann tonometry applanation readings were 20 mmHg in both eyes. On upgaze, his applanation reading was 27 mmHg in the right eye and 20 mmHg in the left. Confrontation visual fields and extraocular motility were full in both eyes. His pupils were equally round and reactive, and there was no afferent pupillary defect. Ishihara color vision was normal in both eyes.

On slit-lamp exam, the lids and lashes were normal. In each eye, the cornea, lens and vitreous were clear,

What’s Your Diagnosis?



HIS RIGHT EYE. (1) Fundus photography reveals choroidal folds extending from the optic disc to the macula. (2) The CT scan shows a nonenhancing, low density mass in the superotemporal orbit. Note the bone spicule within the mass. (3) There also is a well-defined remodeling of the right orbital roof, with 2.5 cm of bone loss at the caudal margin of the frontal bone.



MORE CLUES. (4) On T1, the lesion is mostly hyperintense with some areas of decreased signal superolaterally. Intraorbital extension shows inferior displacement and mild compression of the globe. (5) On T2, we note a well-defined, heterogenous soft tissue mass with a hypointense peripheral rim. (6) The H&E stain reveals cholesterol clefts, hemosiderin deposits and granulomatous inflammation with giant cells.

the anterior chamber was deep and quiet, and the conjunctiva and sclera were white and quiet.

The external exam showed gross right hypoglobus and exophthalmos. Compared with the left eye, the right eye showed 5 mm of proptosis and was displaced 5 mm inferiorly. We also noted the following measurements for his right eye and left eye, respectively: inferior sclera show of 1 mm and 0.5 mm; vertical interpalpebral distance of 9 mm and 8 mm; levator excursion of 15 mm and 15 mm; and marginal reflex distance, which measures the distance between the central corneal light reflex and upper lid, of 3 mm and 2 mm. There was 2 to 3+ fullness on retropulsion of the right eye. We saw that he had a 5-mm linear scar above the right brow. Corneal and periorbital sensation was within normal limits. There was no lagophthalmos and no local lymphadenopathy.

The dilated fundus exam showed normal discs in both eyes. There was no disc edema. In the right eye, there were choroidal folds (Fig. 1). There was no retinal detachment or mass.

We performed computed tomography of the orbits with and without contrast. This revealed a nonenhancing, expansile low-density mass arising from the right anterior frontal bone, lateral to the frontal sinuses (Fig. 2). The lesion measured 2.2 x 2.7 x 2.3 cm. There was well-defined remodeling of the right orbital roof without evidence of aggressive lytic bone destruction. A thin linear bone spicule was identified around a portion of the

lesion. There was associated dehiscence of the frontal bone with approximately 2.5 cm of bone loss (Fig. 3). There was no evidence of communication with the right frontal sinus or right ethmoid air cells.

MRI with and without contrast, revealed a well-defined soft tissue mass extending from the right orbital roof into the extraconal space. The mass was hyperintense on T1 images and did not enhance centrally (Fig. 4). On T2 images, the lesion was heterogeneous with a very hypointense peripheral rim (Fig. 5). There was no evidence of intracranial extension or adjacent dural enhancement.

The differential diagnosis was: cholesterol granuloma; chronic hemorrhage into a vascular abnormality, such as hemangioma or lymphangioma; orbital dermoid; epidermoid cyst or mucocele.

The patient underwent an anterior orbitotomy and excision of the right orbital mass. This was found to be cystlike and contained a yellow-brown viscous material with small bony fragments. The contents were sent for frozen section, cytology and permanent studies. Final pathology showed fibrin, cholesterol clefts, and hemosiderin deposits with foreign-body giant-cell reaction (Fig. 6). Cytology revealed no malignant cells. We made a diagnosis of orbital cholesterol granuloma.

Discussion

Orbital cholesterol granuloma is a rare osteolytic lesion that was first described by Denig in 1902. It localizes

mostly to the superotemporal orbit and predominantly affects males in the third to sixth decade of life.¹ It has also been referred to as lipid granuloma, chronic hematic cyst, hematocele, xanthomatosis and histiocytic granuloma, but, unlike a true cyst, cholesterol granuloma does not have epithelial elements.²⁻⁴

Cholesterol granuloma occurs most commonly in the middle ear, mastoid antrum and petrous apex.² When in the orbit, the most common presenting sign is progressive proptosis. Other signs include ptosis, periorbital pain and swelling, and globe displacement. Visual acuity may be reduced from induced astigmatism or the mass effect of choroidal folds in the macula.¹

Differential diagnosis of orbital cholesterol granuloma includes epidermoid or dermoid cysts, lacrimal gland tumor, frontal sinus mucocele, teratoma, metastasis, aneurysmal bone cyst, Langerhans' cell histiocytosis, lytic Paget's disease and cystic ossifying fibroma.²⁻³

Characteristic findings on CT are an osteolytic isodense or hypodense lesion that is nonenhancing, located extraconally and extending to the periorbitum. Proptosis and displacement of the globe inferiorly secondary to mass effect are also typical. Bony spiculation or small bone fragments within the lesion may also be seen.^{1,2} MRI shows a high signal intensity lesion on both T1- and T2-weighted images characteristic of chronic hemorrhage.^{1,2,5}

Histopathology shows cholesterol clefts with granulomatous inflamma-

tion and giant cells, surrounded by a fibrous capsule. Hemosiderin may be found extracellularly and intracellularly within histiocytes.^{2,3}

The pathogenesis of cholesterol granulomas is unclear. Trauma resulting in hemorrhage has been proposed as a possible mechanism. Cholesterol is produced as the result of the breakdown of blood components, and a foreign body reaction then forms around the cholesterol crystals. Bone resorption is believed to be initiated by the prostaglandins produced by the platelets within the hematoma.^{2,3,6} In our case, although the patient did not recall prior trauma, he did have a scar on the skin anterior to the lesion.

Recently, Yoshikawa et al. identified high tissue plasminogen activator (tPA) immunoreactivity in the endothelial cells of the vessels lining the wall. It is theorized that blood coagulation and hemostasis precede fibrinolysis in a mechanism similar to chronic subdural hematoma.⁷

A generally accepted treatment is complete excision via an anterior or lateral orbitotomy, followed by aspiration and curettage of the contents. Thorough drainage of the contents is necessary to prevent recurrence.^{1,2}

* Patient name is fictitious.

Dr. Gan is a resident and Dr. Lissner is an assistant professor in oculoplastic surgery; both are at Northwestern University, Chicago.

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