A 27-year-old man referred for progressive right eye ptosis since one year. Inferior globe displacement and proptosis of the right upper lid was apparent. His left eye had congenital constant esotropia and was amblyopic with only finger count vision. Limitations in adduction and elevation, in addition to poor levator function were noted upon examination (Figure 1). Macular wrinkling in the OD and a cup-to-disc ratio of 0.8 with intraocular pressure of 18 mmHg in both eyes were additional findings.

Orbital imaging was performed to evaluate his progressive proptosis. There was an isodense mass with bone erosion in the superotemporal area of the right orbit with intracranial extension that remained extradural despite its extension and size as noted on CT scan (Figure 2). Indentation and displacement of the globe was visible. For better evaluation, we performed an MRI scan, which revealed a non-enhancing hyperintense lesion seen in T1-weighted and T2-weighted MRI (Figure 3). The lesion had a sharp border with no cranial extension of the tumor but the duramater was obviously intact. Orbitotomy through the lateral upper lid crease was performed and revealed a large cyst that had a chocolate color mixed with yellow pointy material. Pulses conducted through the brain were visible due to bone erosion and the connection between the cranial fossa and the cyst. The pathology slide is presented in Figure 4.

What is your diagnosis?
See the next page for diagnosis.
Cholesterol granuloma is a granulomatous foreign body reaction around cholesterol crystals with or without a surrounding fibrous capsule. There is a positive history of previous trauma in some cases. Different organs, such as the breasts, lungs, kidneys, peritoneum, petrous apex and orbit are previously reported locations of presentation. An osteolytic lesion most often located in the superior temporal part of the orbital cavity with involvement of the frontal bone is a typical presentation for orbital cholesterol granuloma. Erosion of the orbital roof and extradural spread into the anterior cranial fossa is a possibility. Of the theories that attempt to explain its etiology, hematocele formation is the most popular. In this theory, when a hematoma is formed in the subperiosteal space of the frontal bone and is not absorbed in time, the blood becomes degraded and organized. Giant cells approach this area and a granulomatous reaction occurs. Cholesterol crystals and multinucleated giant cells can be found in the specimens of cholesterol granuloma.

Diplopia on upgaze, aching pain in the orbit, progressive proptosis, inferior displacement of the globe and limited upgaze are the prevalent reported manifestations. Intracranial extension can cause neurologic findings such as headaches, which were absent in the current case despite the large intracranial extension.

The typical finding on CT scan is a non-calcifying mass lesion isodense with brain. No sclerotic margins are apparent. On MRI, a hyperintense lesion on both T1-weighted and T2-weighted images is found, which does not enhanced with gadolinium.

Drainage and total removal of granulomatous tissue via an anterior or occasionally lateral orbitotomy is one management option. Curettage of the lesion from the bone and peristomeum is essential for the prevention of recurrence; however, some authors believe that complete curettage is not necessary.

References