Pleomorphic Adenoma of the Lacrimal Gland With Extensive Calcification

Pleomorphic adenoma is the most common epithelial tumor of the lacrimal gland, and when this tumor is suspected, removal of the mass in its entirety without biopsy should be performed. Clinical, radiological, and ultrasonographic characteristics, such as extended duration of symptoms, absence of pain, a smooth and well-defined border, absence of bony erosion, and lack of calcification, suggest the diagnosis of pleomorphic adenoma. We present an unusual case of pleomorphic adenoma, initially appearing as a lacrimal fossa mass with dense calcification on ultrasonographic and computed tomographic evaluations.

Report of a Case. A 35-year-old white man was seen with a drooping left upper eyelid that he had had for 1 to 2 years (Figure 1). He denied pain or diplopia. His medical history was significant for trisomy 21 and hypothyroidism for 18 months for which he took levothyroxine sodium. His surgical history included a laceration repair of his right brow at age 8 years.

Ocular examination results disclosed corrected visual acuity of 20/30 OU, normally reactive pupils without an afferent pupillary defect, and full ductions. He was orthophoric at distance. Mild left upper eyelid ptosis accompanied left globe displacement, including 0.5 mm of axial proptosis and 5 mm of inferior displacement (Figure 1). Anterior segment and dilated ophthalmoscopic examination findings were unremarkable.

Computed tomographic imaging studies revealed a well-circumscribed soft tissue mass in the superotemporal left orbit, with internal calcification and without bone destruction (Figure 2). Ultrasonographic studies of the orbit showed a round, well-circumscribed, high-reflective and calcified lesion (Figure 3).

The patient underwent surgical excision of the mass through a lateral orbitotomy with replacement of the lateral orbital wall. At the time of surgery, the tumor appeared smooth, gray, and oval. Normal-appearing lacrimal gland tissue was observed anterior to the mass. The tumor was removed without violation of the tumor capsule. The patient’s recovery was uneventful.

Histopathologic examination results disclosed an encapsulated tumor composed of numerous lobules and cords of squamoid cells separated by spindle cells in a mucoid matrix. Areas of squamous dif-
differentiation with keratin production were present as well as numerous areas of calcification measuring up to 250 µm in diameter (Figure 4). Areas of chondroid differentiation were also present. The tumor capsule was intact.

Comment. Pleomorphic adenomas of the lacrimal gland are typically associated with an excellent prognosis, provided they are initially removed in their entirety.1,2,6 Dispersion of tumor cells by biopsy or incomplete excision is associated with late recurrence and occasional malignant transformation.7 Preoperative identification of pleomorphic adenoma by clinical and radiological evidence allows planning of an en bloc excision via a lateral orbitotomy.

Various hallmarks of pleomorphic adenoma have been described to aid in their identification. Symptoms present for more than 1 year and an absence of pain are suggestive of this tumor.8 An oval, encapsulated appearance without bony destruction or irregular edges on computed tomographic studies is characteristic.9 On ultrasonographic studies, pleomorphic ad-
enomas are medium to highly reflective, with a regular structure and moderate sound attenuation.  

This tumor was characteristic of pleomorphic adenoma in several ways. The patient had a painless change in appearance for at least 1 year. The lesion was oval, well circumscribed, and appeared encapsulated on both computed tomographic and ultrasonographic studies. However, it showed significant, diffuse calcification on both imaging studies.

In general, the presence of calcification is a radiological sign of malignancy, and calcification of masses of the lacrimal gland fossa usually suggests malignant disease. Only 3 histologically proven cases of pleomorphic adenoma with calcification can be found in the literature.  

Even in pleomorphic adenomas of the parotid gland, calcification is only rarely found and suggests an older tumor age.  

In summary, although calcification is more common in malignant lacrimal gland fossa tumors, the presence of calcification should not exclude a diagnosis of pleomorphic adenoma. Other clinical and radiological characteristics should be considered when planning surgical management.

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Bony Hamartoma of the Inferior Orbital Rim in a Patient With Tuberous Sclerosis

Named by Bourneville, one of its earliest discoverers, tuberous sclerosis is a heritable neurocutaneous syndrome that is classically manifested by the Vogt triad of mental deficiency, epilepsy, and adenoma sebaceum.  

Despite the classic triad of findings, tuberous sclerosis is a protean disorder characterized by the presence of hamartomas (benign neoplasms composed of cellular elements normally present in tissue) in multiple organ systems, including the brain, kidneys, heart, spleen, lungs, and eye.  

Osseous involvement is known to occur in the skull, long bones, pelvis, and metacarpal and metatarsal bones. We report a case of tuberous sclerosis that manifested as a bony mass arising from the inferior (maxillary bone) orbital rim. Histopathologic examination of the excised lesion revealed a hamartoma composed of mature bone. Based on our MEDLINE review of the English-language literature, this is the first reported case of a bony hamartoma arising from this location of the facial skeleton in a patient with tuberous sclerosis.

Report of Case. A 6-year-old girl with known tuberous sclerosis had a mass involving the right inferior orbital rim. The lesion was first noticed by her parents at 1 year of age, and it slowly enlarged during the next 5 years. The patient was diagnosed as having tuberous sclerosis at the age of 6 months based on the characteristic symptoms and signs of a seizure disorder, developmental delay, and classic skin lesions, including adenoma sebaceum along the bridge of the nose and an ash leaf spot on the back of the right calf.

There was no other significant medical or family history. The patient’s fraternal twin was unaffected. Although the mass did not cause any functional symptoms, the patient’s parents were concerned about the progressive increase in size and the obvious facial deformity.

On initial examination, a 1.5 × 1.0-cm firm, nontender, immobile mass was identified along the right inferior orbital rim and cheek area (Figure 1). In addition, a 1.0 × 0.5-cm hyperpigmented, slightly elevated skin lesion was noted overlying the inferior orbital rim mass. There were also several small skin lesions involving the midface and nose, which appeared to be adenoma sebaceum, and an ash leaf

Figure 1. Patient with a large elevated mass along the right inferior orbital rim and cheek area measuring 1.5 × 1.0 cm.