Erosion of the Orbital Plate (Frontal Bone) by a Benign Tumor of the Lacrimal Gland

Albert Hornblass, M.D.
Alan H. Friedman, M.D.
Arnold Yagoda, M.D.

SUMMARY
A benign mixed tumor of the lacrimal gland of long standing duration caused complete erosion of the orbital roof and dura. At the time of surgery, a large cerebrospinal fluid leak communicated into the orbit after removal of the tumor.
This was treated with gelfoam, bed rest and antibiotics. The patient had a complete recovery.

INTRODUCTION
Lacrimal gland tumors as a group are a relatively uncommon cause of proptosis. Benign mixed tumors of the lacrimal gland are usually slow growing and symptomless. Invasion of adjacent bone and erosion of the dura mater of the brain, while occurring with malignant lacrimal gland tumors, have not been noted to occur with the benign mixed tumors.

We present a case of a benign mixed tumor of the lacrimal gland. This resulted in dissolution of the orbital plate of the frontal bone, producing a direct communication of the frontal lobe of the brain with the orbit. Following surgical excision of the tumor, the patient developed a transitory cerebrospinal leak into the orbit and face. To our knowledge, these complications of benign mixed tumors of the lacrimal gland have not been previously reported.

CASE REPORT
J.M., a 69-year-old white female, presented with a history of chemosis in the left eye for approximately ten years, and with actual proptosis for six years (Figure 1). She was remarkably free of complaints despite her gross deformity and consented to seek medical attention only because of the admonishment of friends and relatives. A thyroid work-up had been negative on several occasions in the past. Her general medical condition was good except for a well-compensated hypertension. A general medical examination and routine lab tests were within normal limits.

Ocular examination revealed a visual acuity of 20/30+ in the right eye and 20/100 in the left eye with correction. There was marked proptosis with downward and inward displacement of the left globe (Figure 2). A bony, hard, nontender, smooth, fixed mass was palpable beneath the superior orbital rim laterally. (Figures 3 & 3A). The extraocular movements were full without diplopia. There was chemosis of the conjunctiva in the left eye with mild corneal staining. The anterior chamber was deep and clear.

From the Division of Ophthalmology, Department of Surgery, State University of New York, Downstate Medical Center, and the Department of Oculoplastic Surgery, Manhattan Eye, Ear and Throat Hospital (Dr. Hornblass); the Departments of Ophthalmology, Lenox Hill Hospital (Drs. Hornblass, Friedman and Yagoda); and the Mount Sinai School of Medicine (Dr. Friedman), New York, New York.

Requests for reprints should be addressed to Albert Hornblass, M.D., 903 Lexington Avenue, New York, New York 10021.
and the lens was normal. Applanation tonometry was 13 mm of mercury in the right eye and 19 mm in the left eye. Fundus examination was normal. The right eye was within normal limits. Preoperative x-rays (Figure 4) and CT scan revealed bony disruption of the lateral and posterior-medial walls of the orbit and the orbital roof (Figures 5 & 6).

The patient was brought to the operating room for exploration and excision of the lesion. At surgery, a lateral and superior orbitotomy was performed, revealing an encapsulated tumor (Figure 7). A bony defect in the roof of the orbit, approximately 25 mm in diameter, was noted (Figures 8 & 8A). The frontal lobe of the brain covered by its dura communicated with the orbit through this defect. In addition, the tumor was adherent to the orbital bone laterally. The entire tumor was removed within its capsule, including fragments of the adherent orbital bone (Figure 9). The bony defect in the orbital roof was packed with gel foam only. No prolapse of the neural tissue into the orbit or invasion of the neural tissue by the tumor was noted at the time of surgery.

Postoperatively, the patient was placed on Keflin intravenously, and complete bed rest. There was cerebrospinal fluid leakage, resulting in edema of the face ipsilaterally (Figure 10). This subsided spontaneously within a few days with complete resolution of the facial swelling (Figure 11). The ptosis resolved and the vision in the left eye returned to 20/30. (Figures 12 & 13).

Microscopic examination of the specimen showed tubular structures arranged in an irregular pattern within a myxoid stroma. The tubular structures were lined by a double layer of epithelium. Several areas of the tumor displayed squamous metaplasia (Figure 14). The tubes or ducts contained PAS-positive material while the myxoid stroma contained acid mucopolysaccharide (Figure 15). Fragments of bone showed true invasion by tumor.

DISCUSSION

Lacrimal gland tumors usually present as a slowly developing fullness of the upper outer lid with eventual ptosis. With time, a downward and inward displacement of the globe occurs, accompanied by proptosis. A hard mass is often palpable at the superolateral orbital rim. Although

FIGURE 2 (Hornblass et al.): Note the downward displacement of the left globe. There is a significant fullness of the left lacrimal area.

FIGURE 3 (Hornblass et al.): Lateral view shows the large tumor projecting anteriorly and laterally.

FIGURE 4 (Hornblass et al.): X-ray shows deterioration of the roof of the orbit and lateral orbital roof.

FIGURE 3A (Hornblass et al.): Artist's depiction of position of tumor.
FIGURE 5 (Hornblass et al.): CT scan in P-A view shows the defect in the roof of the orbit.

FIGURE 6 (Hornblass et al.): CT scan shows mass in the lacrimal gland area causing severe proptosis.

FIGURE 7 (Hornblass et al.): Lacrimal gland tumor being excised completely encapsulated from supero-lateral aspect of the orbit.

FIGURE 8 (Hornblass et al.): Frontal lobe is seen through defect of the roof of the orbit.
FIGURE 8A (Hornbliss et al.): Artist's depiction of orbital roof defect.

FIGURE 9 (Hornbliss et al.): The specimen consisted of several firm white fragments of tissue with pieces of bone.

FIGURE 10 (Hornbliss et al.): Postoperatively there was periorbital emphysema and ecchymosis. Note the mottled appearance of the skin.

FIGURE 11 (Hornbliss et al.): Postoperatively the CSF is completely absorbed. The eye is no longer proptotic.

FIGURE 12 (Hornbliss et al.): A close-up postoperative view reveals a superior palpebral furrow scar.
retro-orbital pressure can be exerted on the optic nerve and the central retinal vessels coursing through it, this is the exception rather than the rule. In fact, visual symptoms, except for possible diplopia, are usually strikingly absent. Pain is also uncommon and if present, usually indicates malignancy. It is of interest that the patient often finds his way to the physician’s office only after many admonitions from friends and relatives.

Lacrimal gland tumors as a whole account for only a small percentage of patients presenting with proptosis. In one study by Lloyd, only 8 out of 212 cases of proptosis were caused by lacrimal tumors. The relative paucity of these lesions contributed to the lack of understanding of their true nature until Forrest classified them in 1954.

A variety of tumors may be encountered in the lacrimal gland. These include primary tumors, secondary tumors (lymphoma and metastatic carcinoma and inflammatory pseudotumors, e.g., reactive lymphoid hyperplasia, sarcoidosis, etc.). Primary lacrimal gland tumors are of epithelial origin and are on a statistical basis, evenly divided into

**FIGURE 13** (Hornblass et al.): Laterally, there is no proptosis and the fullness in the lacrimal area is resolved.

**FIGURE 14** (Hornblass et al.): High power view. Several areas of the tumor show squamous metaplasia. (Hematoxylin-Phloxine-Safranin x 250)
benign and malignant neoplasms. The "benign mixed tumor" is the only histological type of non-malignant primary lacrimal gland tumor and accounts for approximately 50% of all lacrimal neoplasms. The malignant lacrimal epithelial neoplasms, however, comprise a group of several histologically distinct types: adenoid cystic carcinoma, the most common type; malignant mixed tumors; pure adenocarcinomas; mucoepidermoid carcinoma; squamous cell carcinomas and oxyphil adenomas (oncocytomas), are all common types of lacrimal gland tumor, except for the benign mixed tumor.

Definitive diagnosis and treatment of these tumors rests in the hands of the surgeon. A thorough physical and laboratory examination, including x-rays, ultrasound and CT scan of the orbit, is undertaken preoperatively to delineate as best as possible the nature and extent of the lesion. Thus prepared, the surgeon must then explore the lesion and decide at the time of surgery what course of action to pursue. If an encapsulated, noninvasive lesion is discovered and the preoperative evaluation is not suggestive of a malignant lesion or a pseudotumor, the diagnosis is almost certainly to be a benign mixed tumor, although a frozen section would help to confirm the clinical impression.

In this case, an attempt at complete excision of the lesion is indicated. Failure to remove all of the lesion is responsible for subsequent local recurrences.

If an invasive lesion is present, however, a frozen section must be taken and if the presence of carcinoma is confirmed, complete exenteration has usually been advised. Failure to do so almost inevitably leads to the subsequent demise of the patient due to recurrent tumor. Recently, en block excision of tumor and bone has been advocated by Henderson instead of exenteration.

The excellent prognosis for the benign mixed tumor of the lacrimal gland has been amply demonstrated by Zimmerman and Forrest. Of 54 patients with benign mixed tumors reported by Zimmerman and followed for at least five years, only one tumor-related death occurred. In the study by Forrest, there was only one tumor-related death among 22 patients studied. Unless the tumor is completely excised, however, local recurrences requiring reoperation can be expected.

The case presented here has an unusual feature not previously reported. Although it fit most of the above criteria for a benign mixed tumor both pre-operatively and from its appearance at surgery, it caused loss of bone in the
adjacent orbit. Malignant lacrimal tumors can certainly invade bone, causing both reactive hyperostosis and destructive bony loss; in fact, this is one of their hallmarks. Benign tumors, however, do not usually have this feature. In our case, dissolution of the orbital bone by this large benign tumor did occur over a period of many years, and was noted on preoperative CT scan. Histopathological study revealed invasion of contiguous bone by tumor. More important, the dura had been eroded and the brain exposed. It is pointed out, therefore, that a bony defect of the orbit noted preoperatively does not necessarily imply malignancy. Furthermore, postoperatively, the problem of cerebrospinal fluid leak was encountered because of the communications between the intracranial cavity and orbit created by the tumor and its removal. This was treated conservatively with bed rest and antibiotics and spontaneously sealed off after four days.

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REFERENCES