

Case 9

History of Present Illness

A 22-year-old woman developed pressure pain in the right eye \times 3 months. It was tender to palpation and she felt like there was lump in the right upper eyelid. She notes that she had mild double vision when looking to the right and that her vision seems mildly blurred in that right eye. She tried hot compresses and a steroid-antibiotic ointment on the right upper lid, but the pain and the lump did not improve. Overall the pain has not changed substantially. She has a history of migraines, but these have not changed recently. She denies any migraine accompaniments (light or sound sensitivity or nausea) with this eye pain. She has a history of a blocked tear duct in the right eye for which she underwent a probing and irrigation at 8 months of age.

<i>Past medical and ocular history</i> Per HPI	<i>Past surgical history</i> Per HPI
<i>Medications</i> Sumatriptan prn headache Ondansetron prn headache Birth control	<i>Family history</i> Grandfather—Macular degeneration
<i>Social history</i> Manages meetings and events Does not smoke or drink	<i>Review of systems</i> Completely negative

Examination

<i>Acuity with correction</i> Right eye: 20/150 improving with pinhole to 20/20 Left eye: 20/20
<i>Pupils</i> Normal
<i>Intraocular pressure</i> Right eye: 17 mmHg Left eye: 20 mmHg

External exam

3 mm proptosis, right eye (Fig. 9.1a)

Firm lump in the superotemporal area right eye, inferior to the eyebrow and superior to the tarsus

Eye alignment and motility

25% deficit of abduction and elevation, right eye

Right esotropia and hypotropia in right and upgaze

Orthophoric in primary gaze

Slit lamp examination

Normal

Visual field

Normal

Fundus examination

Choroidal folds in the macula, right eye

Neurologic examination

Normal facial sensation

Otherwise unremarkable

Discussion***Ophthalmic Perspective: Dr. Lee***

She has developed blurred vision likely from something pushing on the back of the eye causing hyperopia. She also has double vision and is unable to move her eye up and to the right, suggesting that she has a mass there. The photo shows mild edema of the fat above the eyelid and below the orbital rim. There is mild ptosis of the right upper eyelid. The eyes appear straight as she looks straight ahead (Fig. 9.1a). It seems likely that she has a lacrimal gland mass.

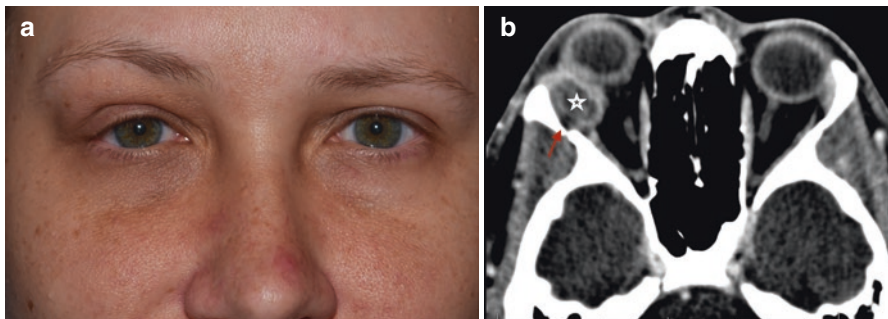


Fig. 9.1 (a) External images of the face show subtle enlargement of the brow fat above the eyelid. The right upper eyelid shows mild ptosis. (b) Axial CT of the orbit shows a cystic mass in the area of the lacrimal gland, lateral and posterior to the right globe (*star*). There is erosion of the lateral orbital wall (*red arrow*)

The most common would be a benign mixed tumor given the firmness of the tumor. Adenoid cystic carcinoma is in the differential, but these are usually much more painful because of neural invasion. The average age is 40 years, so our patient is young to have that. The differential could also include lymphoid hyperplasia, lymphoma (not this firm), sarcoidosis (not this firm), infection (usually more obvious), and other epithelial malignancies or metastases. See Fig. 8.1 for relationships of the lacrimal gland and adjacent anatomy. We should really start with CT of the orbit to look better at the bony orbital involvement and consider an excisional biopsy. MRI does not give us a good view of the bone, but would be acceptable to show lacrimal gland involvement. If there were more pain, redness, or a hot looking abnormality then one might consider antibiotics for an orbital cellulitis or corticosteroids for lacrimal gland inflammation. This would not be consistent with a preseptal lesion, because it is causing double vision. The double vision pushes us that there is something in the orbit.

Neurologic Perspective: Dr. Digre

First, her decreased vision is related to her choroidal folds in her macula; her vision was correctable to 20/20 with pinhole and she has no RAPD. This is one more reason why anyone with changes in the vision and eye pain needs a full, dilated eye examination. Then she has diplopia—just looking at her, you can see something is wrong because there is fullness over the lateral aspect of her orbit and that this diplopia is not from a cranial nerve abnormality. Imaging is the first step in determining the cause and CT of the orbit with bone windows would be what I would order. Fortunately for her, the slight discomfort she is experiencing is not making her migraines worse. Many of my patients with any kind of secondary pain will have worsening of migraines. If the discomfort got worse, I would suggest an anti-inflammatory medication (e.g., ibuprofen or naproxen).

Non-ophthalmic/Non-neurologic Perspective

This patient started with mild “eye” ache with some tenderness to touch of the right upper eyelid area. Just hearing the story, the most common thing to consider would be a stye. She tried hot compresses and eye ointment without benefit and now we are 3 months later. Early on styes can present with just some mild eyelid edema, but these are usually very close to the eyelid margin. In this case, she has some mild edema of the area inferior to the eyebrow, which would not be a stye since it does not involve the tarsus. This is the area of the right lacrimal gland. If you pull up on the eyelid and have the patient look down, you can get a better idea of whether the lacrimal gland is enlarged. In many cases MRI is better than CT, but with lacrimal gland disorders, it is better to get a CT orbit.

Follow Up

The patient underwent an orbital CT (Fig. 9.1b). This showed a $2.5 \times 2.0 \times 2.6$ cm enhancing mass in the area of the lacrimal gland with adjacent orbital roof erosion. She underwent an excisional biopsy of the lacrimal gland, which showed adenoid cystic carcinoma, basaloid type.

Historically, the 5-year survival was 17–20% with chemotherapy, radiation, and exenteration. With neoadjuvant intra-arterial doxorubicin and cisplatin, the survival rate has improved to 57–100%. The patient underwent intra-arterial therapy followed by orbital exenteration (removal of the eye and all of the orbital contents) and radiation therapy. Two years later, she experienced a recurrence in her right mandible. This was resected and she remained in remission at 4 years follow up. *Final diagnosis: adenoid cystic carcinoma of the lacrimal gland.*

Digre comment: Adenocystic in a 22-year-old is very rare. This case reminds us that we must be ever vigilant and that all tumors of the orbit and lacrimal gland need work up.

For Further Study

1. Tse DT, Kossler AL, Feuer WJ, Benedetto PW. Long-term outcomes of neoadjuvant intra-arterial cytoreductive chemotherapy for lacrimal gland adenoid cystic carcinoma. *Ophthalmology*. 2013;120:1313–23.
2. von Holstein SL, Rasmussen PK, Heegaard S. Tumors of the lacrimal gland. *Semin Diagn Pathol*. 2016;33:156–63.