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Nasolacrimal Duct Obstruction Caused by Primary Lacrimal Sac Adenocarcinoma

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Lacrimal Sac Adenocarcinoma Excision and Reconstruction

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Abstract

Purpose: To report surgical management of a rare case of lacrimal sac adenocarcinoma.

Methods: Interventional case report.

Results: A 55-year-old woman presented to the oculoplastics service with 6 months of epiphora. Lacrimal irrigation confirmed right nasolacrimal duct obstruction. Symptoms resolved after a dacryocystorhinostomy (DCR). Histopathology of the lacrimal sac was negative for malignancy. One year later she had recurrent epiphora and a painful medial canthal swelling. CT orbit showed a right nasolacrimal duct mass. Biopsy by nasal endoscopy revealed poorly-differentiated adenocarcinoma. A dacryocystectomy with medial maxillectomy including orbital periosteum provided clear margins. A radial forearm osteocutaneous free flap was used to reconstruct the vertical nasomaxillary buttress with the skin lining the nasal passages. The orbit was reconstructed with polyethylene coated titanium mesh and miniplates. Adjunctive radiation was given and she has been disease free with good eye function for 4 months post resection with satisfactory aesthetic result.

Conclusion: Malignancy of the nasolacrimal drainage system is rare and early diagnosis is key to maximizing patient outcomes.

Background

Nasolacrimal drainage system tumors are rare and diagnosis is difficult as symptoms often mimic those of the more common primary acquired nasolacrimal duct obstruction (PANDO). Common to both are symptoms of epiphora and stasis of tears in the lacrimal sac. Acute dacryocystitis can result- a painful abscess in the lacrimal sac just inferior and posterior to the medial canthal tendon. Examination findings that are more suspicious for malignancy include chronic painless dacryocystitis, bloody tears or bloody reflux on irrigation, epistaxis, numbness in the infraorbital nerve region or a firm, immobile mass extending superior to the medial canthal tendon. Tumors are typically diagnosed when suspicious findings lead to CT imaging or when dacryocystorhinostomy (DCR) is performed and the lacrimal sac is biopsied. Some surgeons only biopsy during DCR when suspicious operative findings are present while others biopsy the lacrimal sac routinely.

Most malignant lacrimal sac tumors are epithelial in origin (75%), most commonly squamous cell and transitional cell carcinomas (1). Adenocarcinoma is exceedingly rare, as there are only 16 previously published cases at the time of this writing (2-8).

These tumors typically require wide resection with en bloc excision of the mass, canaliculi, nasolacrimal duct, the lacrimal fossa, adjacent lacrimal ethmoid cells, and the periosteum of the lacrimal sac and nasolacrimal duct. Despite aggressive treatment, lacrimal sac tumors have a high rate of recurrence (1-7).

Case Report

- A 55-year-old woman was initially seen by oculoplastics for a 6 month history of right sided epiphora and episodic swelling adjacent to the medial canthus. The patient had no history of trauma, sinus surgery, bloody tears, epistaxis, difficulty with nasal breathing, nor cheek numbness.
- Lacrimal irrigation confirmed nasolacrimal duct obstruction (NLDO).
- Two weeks later, she underwent an external DCR and a routine biopsy of the lacrimal sac. Histopathology was consistent with chronic inflammation and negative for malignancy supporting a diagnosis of PANDO.
- She reported 90% resolution of tearing after her 2 month follow up.
- One year later, she presented with recurrent constant epiphora and painful swelling at the medial canthus. Lacrimal irrigation suggested recurrent NLDO.
- Oral antibiotics reduced swelling and pain of the apparent acute dacryocystitis, however epiphora and medial canthal fullness inferior to the canthal tendon remained. Her vision, extraocular movements, eye and orbit examination were normal.
- A CT scan of the orbits to evaluate for a reason for failed DCR was ordered.

Case Report

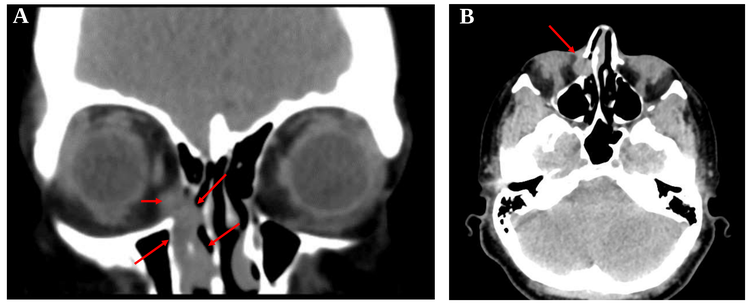

Figure 1. CT orbit coronal (A) and axial (B) views showing ill defined soft tissue density extending from the right lacrimal sac fossa and rhinostomy site through an enlarged, remodeled right nasolacrimal duct with extension to the inferior meatus.

- CT Orbits showed a soft tissue density mass extending from the right lacrimal sac fossa and DCR site through an enlarged and remodeled right nasolacrimal duct with extension to the inferior meatus of the right nasal passage.
- Nasal endoscopy by otolaryngology revealed a right friable mass along the anterior inferior meatus. Biopsy showed poorly differentiated glandular structures that were AE1/AE3 and CK7 positive, indicative of lacrimal sac adenocarcinoma.
- PET CT showed hypermetabolic activity confined to the right lacrimal fossa.
- Given her imaging findings, she was determined to have Stage III (T3N0M0) primary lacrimal sac adenocarcinoma. Tumor board recommended surgical resection.
- Surgical management to preserve the right eye was elected. A right lateral rhinotomy incision was modified to include an oval resection of the medial canthus encircling the medial upper and lower eyelid and their lacrimal canaliculi, the caruncle, the medial canthal skin and anterior medial canthal tendons. Dissection was carried into the orbit in the extraconal space, sharply dissecting the inferior oblique from its origin, and drawing the eye and orbital contents laterally from the lacrimal sac and the periosteum of the medial orbital wall and orbital floor. An *en bloc* dacryocystectomy with medial maxillectomy including the orbital periosteal barrier, the entire nasolacrimal duct, and inferior turbinate was performed to provide clear surgical margins. A radial forearm osteocutaneous free flap was used to reconstruct the vertical nasomaxillary buttress with the skin lining the nasal passages and helping to recover the medial canthal skin defect. The inferior orbital rim was reconstructed with a curved miniplate and polyethylene coated titanium mesh was molded to restore the medial orbital wall and medial orbital floor partitions. The eyelids were fixated to titanium mesh to recreated the medial canthal angle.
- Closest surgical margin was 1mm at the lacrimal sac periosteal lining adjacent to the orbit.
- The patient received 60 Gy of adjuvant radiation with fields designed to spare the globe
- She is currently disease free 4 months after resection.

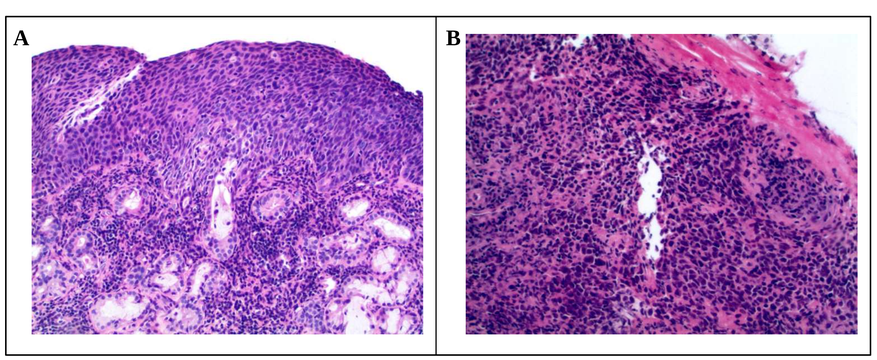

Figure 2. Hematoxylin and eosin staining of the resected lacrimal sac tumor. A. Poorly differentiated glandular structures. B. Magnified view showing a dense monoclonal infiltrate with high mitotic figures.

Discussion

- Our patient is one of the few reported cases of biopsy confirmed lacrimal sac adenocarcinoma, presenting unique management considerations.
- Epiphora and nasolacrimal duct obstruction due to PANDO is common and performance of DCR is a highly effective procedure to bypass obstruction and relieve epiphora. The standard of care is to suspect malignancy when symptoms, examination findings or surgical findings are out of the ordinary. There were no features to cause suspicion at initial presentation and this surgeon routinely biopsied the lacrimal sac as in all cases.
- Lacrimal sac malignancies are thought to arise de novo, and malignancy may not have been present when the initial biopsy was done. Chronic inflammation may have served as a substrate for malignant transformation.
- Alternatively, the malignancy could have been at an early stage initially but not detected on the biopsy due to sampling error (7, 9). Or, the malignancy could have arisen in the nasolacrimal duct, distal to the bypass site of the dacryocystorhinostomy.
- High index of suspicion for malignancy is essential in recurrent nasolacrimal duct obstruction, necessitating imaging or thorough examination during repeat DCR for suspicious lesions for biopsy (1,7,9).
- The use of radial osseous free flaps has been widely demonstrated in head and neck surgery. Radial free flaps are favored due to thin, pliable, soft tissue component, multiple skin islands, and the possibility of incorporating vascularized tissue (14). The successful use of radial osseous free flap in head and neck reconstructions inspired its use in reconstruction of the vertical nasomaxillary buttress in the present case of lacrimal sac tumor resection.
- This is the first documented case of orbital rim reconstruction with a radial osseous free flap. Resections reported in the literature for lacrimal sac tumor excisions typically use local flaps for reconstruction. Hardware has also been used with tailored contoured titanium in orbital reconstruction (10-12). Musculocutaneous free flaps have also been used such as in the case of a left temporalis muscle flap being used to reconstruct an orbital defect and the use of a radial forearm cutaneous free flap. To our knowledge, the flaps did not include osseous tissue (13, 14).

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