Management of Transitional Cell Carcinoma of the Lacrimal Sac: A Multidisciplinary Approach to Orbit Sparing Treatment

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OBJECTIVES
- To review the presentation of lacrimal sac transitional cell carcinoma.
- To advise otolaryngologists of warning signs concerning for lacrimal sac malignancy.
- To discuss new multidisciplinary treatment advances for this exceedingly rare tumor.

INTRODUCTION
Transitional cell carcinoma (TCC) of the lacrimal sac is an extremely rare entity; however, it is critical to consider because a delay in diagnosis can be fatal. If discovered early, advances in proton beam therapy have made orbit sparing surgery a more feasible option for treatment. Preoperative involvement of radiation oncology and ophthalmology helps determine the extent of surgery needed. Postoperatively, chemoradiation is important for maximizing locoregional control.

CASE REPORT
A 56-year-old man was referred with a six-month history of a painless medial canthal mass and epiphora. He denied any purulent or bloody discharge, and he had no changes in visual acuity. There was no antecedent trauma or illness. He had no recent fevers, chills, weight loss, or night sweats.

On exam, the mass was firm and not reducible. Prior to presentation, a CT had been obtained which demonstrated a mass based in the region of the lacrimal sac, extending down the nasolacrimal duct and possibly into the anterior medial portion of the orbit. A biopsy was performed which was initially interpreted as inverted papilloma and subsequently re-interpreted as transitional cell carcinoma. He was thus referred for further treatment.

The patient underwent magnetic resonance imaging (MRI) for further characterization of the mass. This demonstrated a hypointense soft tissue mass, expanding the right nasolacrimal duct and extending to the lamina papyracea and through the lamina cribrosa, ~3cm in craniocaudal dimension. There was no intracranial extension. (See Figures 1A, B, C).

Otolaryngology and ophthalmology performed orbit sparing en-bloc excision of the mass and lacrimal system with a medial maxillectomy, total ethmoidectomy and resection of peribulbar contents. The infraorbital nerve was identified and preserved. The medial cut ends of the upper and lower lids were sutured to reform the medial canthus, and this was anchored to the periosteum of the medial orbital wall to provide support to the orbit.

Postoperatively, he received proton beam radiation (70Gy) and concurrent weekly carboplatin/Taxol chemotherapy. He is currently being followed with serial exams and imaging, and at nineteen months after treatment, he has no evidence of recurrence. His vision remains excellent, with the exception of mild lateral gaze diplopia.

Histologic evaluation revealed transitional cell carcinoma, consistent with lacrimal gland origin, with a high mitotic rate (>30/10 HPF) and squamous differentiation focally. (See Figures 2A-B).

HISTOLOGIC FINDINGS

REFERENCES & ACKNOWLEDGMENTS
8. Thanks to Dr. Jonathan Lukens, BIDMC Pathology, for assurance with the pathology findings in the case.

FIGURES

PREOPERATIVE IMAGING

Figure 1A. T1-weighted axial MRI demonstrating a mass expanding the lacrimal sac, eroding through lamina and compressing orbital fat.

Figure 1B. T1-weighted axial post-gadolinium MRI demonstrating heterogenous enhancement.

Figure 1C. T2-weighted coronal MRI demonstrating 3cm craniocaudal dimension.

HISTOLOGIC FINDINGS

Figure 2A. H&E stain (10x) demonstrating undulating fronds of TCC consistent with lacrimal gland origin, with infiltration into bone.

Figure 2B. H&E stain (40x) demonstrating high mitotic rate (>30/10).

CONCLUSIONS
- Transitional cell tumors of the lacrimal sac are very rare but potentially fatal.
- Clinicians should have high index of suspicion for neoplasm if symptoms are persistent, as a delay in diagnosis affects prognosis and chance for cure.
- Treatment should include a multidisciplinary team of otolaryngologists, ophthalmologists, oncologists and radiation oncologists.
- Multimodality treatment advances now enable more orbit sparing resections and greater survival chances.

DISCUSSION
Tumors of the lacrimal sac are very rare. To date, approximately 100 tumors have been reported, and of these, 75% are malignant. Overall, lacrimal sac tumors most commonly present in the fifth decade of life.

Lacrimal sac tumors can be divided into:
- Epithelial (73%)
  - Benign: squamous and transitional cell papillomas most common
  - Malignant: SCC and TCC most common, adenocarcinoma, mucouseropithelial, adenoid cystic, poorly differentiated carcinoma
- Non-epithelial (27%)
  - Fibrous histiocytoma, lymphoid lesions, malignant melanoma most common

The most common presenting signs and symptoms are:
- Epiphora (53%)
- Recurrent dacrocystitis (38%) and/or Lacrimal sac mass (36%)
- Proptosis
- Firm, incompressible masses
- A mass located above the medial canthal tendon
- Lacrimal hemorrhage, epistaxis, pigmented ocular discharge

Diagnostic tests should start with dacrocystogram to look for a filling defect or delayed drainage of contrast. The triad of an irreducible mass, epiphora, and positive dacrocystogram is very concerning for malignancy. Computed tomography (CT) can demonstrate expansion or erosion into the lacrimal sac fossa, and any invasion into neighboring structures. Ultimately, the definitive diagnosis of TCC requires tissue, and excisional biopsy is ideal. If complete excisional biopsy is not easily performed, incisional biopsy should be made into deep tissue tumor; a superficial biopsy carries risk of misdiagnosis of chronic inflammation or pseudotumor.

Pathology of TCC shows spindly, elongated cells, with nuclear polymorphism, increased cellularity, and mitotic figures. Histopathologically, TCC can be confused with inverted papilloma, thus it is critical that the histopathology be interpreted by an experienced head & neck pathologist. The management of the two entities which was initially interpreted as inverted papilloma and subsequently re-interpreted as transitional cell carcinoma. He was thus referred for further treatment.

The patient underwent magnetic resonance imaging (MRI) for further characterization of the mass. This demonstrated a hypointense soft tissue mass, expanding the right nasolacrimal duct, and extending to the lamina papyracea and through the lamina cribrosa, ~3cm in craniocaudal dimension. There was no intracranial extension. (see Figures 1A, B, C).

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