Lacrimal Sac and Nasolacrimal Duct Non-Keratinizing Squamous Cell Carcinoma: Diagnostic Challenge

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Abstract

Educational Objective: At the conclusion of this presentation the participant should be able to recognize the rarity in primary nasolacrimal duct and sac tumors as well as the diagnostic challenges associated in obtaining a preoperative tissue diagnosis.

Study Design: Case report.

Methods: Review of clinical case records.

Results: We report a 30 year old male who presented with one year history of left epiphora and an expanding subcutaneous medial canthal mass. A CT scan was obtained which demonstrated a left fungating lacrimal sac mass lesion with extension into nasolacrimal duct resulting in dilation of nasolacrimal and evidence of osseous remodeling with erosive changes. Several biopsies were obtained over the course of three visits given the difficulty in establishing a diagnosis. On the final biopsy, the pathology team was unable to make a final diagnosis stating that the tissue was consistent with most with atypical epithelial proliferation favoring squamous papilloma malignant epithelial transformation given the presence of atypia within the sample however direct invasion was observed. Given MRI finding demonstrating invasion of the orbital fat and possible impingement of the inferior orbital muscle with pathology favoring malignancy, a multidisciplinary tumor board recommended surgical excision. The patient underwent extended medial maxillectomy, lateral rhinotomy with orbital exenteration and reconstruction using split calvarial bone graft, superficial temporal-parieto-parietal flap with split thickness skin graft. The final pathology was consistent with primary lacrimal sac p16 negative and EBV negative non-keratinizing squamous cell carcinoma involving the orbital fat.

Conclusions: Our case demonstrates the importance of a multidisciplinary approach in managing these rare tumors. Furthermore, it highlights the difficulty in establishing a preoperative diagnosis and imaging finding characteristic of lacrimal system tumors.

Introduction

Lacrimal sac and nasolacrimal duct malignancies are rare and require a high index of suspicion. The incidence is reported between 1.3% of patients evaluated for acquired obstruction of the lacrimal drainage system.1 The majority of lacrimal sac neoplasms are epithelial in origin, and the majority of epithelial tumors are malignant.2

Most common presenting symptoms:
- Recurrent epiphora
- Palpable mass superior to medial canthal ligament
- Bloody reflux with atrophic iritis

Treatments:
- DCR with tumor excision or complete dacryocystectomy
- Perioperative radiotherapy
- Wide surgical excision of tumor and the entire lacrimal drainage system (canaliculus, sac, and nasolacrimal duct), orbital exenteration, lymph node dissection, and reanastomosis of the nasolacrimal system is indicated in larger, widespread malignant lesions.3,4

No current evidence-based clinical guidelines exist.

Discussion

Diagnostic Dilemma:
- Rare nature and insidious presentation lead to misdiagnosis/treatment delay
- No definitive invasion on pathology: biopsy exhibited epithelial proliferation with foci of pleomorphic cells and numerous mitosis suggestive of non-keratinizing SCC. Interval non-inflammatory increase in size of mass after biopsy.
- Imaging reviewed by multidisciplinary team: tumor involvement of the anterior orbital fat and inability to clear the tumor from the orbit without compromising globe function and recurrence.
- Little clinical data comparing orbital exenteration survival, recurrence rates or visual function after radiation therapy.
- Malignant tumors with wide excision and lateral rhinotomy have a recurrence rate of 12.5% contrasted to 43.7% in patients with localized lacrimal sac excision. The gain provided by orbital exenteration in terms of local control and longterm survival is difficult to assess when orbital invasion is limited because of the low number of cases reported in the literature.1,5

Epithelial malignancies of the lacrimal sac are quoted to have a recurrence rate of 50%, 50% of recurrences these are fatal.6

Conclusion

Our case demonstrates the importance of a multidisciplinary approach in managing these rare tumors. Furthermore, it highlights the difficulty in establishing a preoperative diagnosis, and imaging finding characteristic of lacrimal system tumors.

Case Description

A 30-year-old male without significant PMH presented to Otolaryngology clinic with a left orbital mass near the medial canthus which had grown rapidly over the previous month.
- He presented with a firm, painless, 1cm lesion over the medial canthus with overlying erythema (figure 1); no visual changes or ulceration was noted.

Figure 1. Patient at presentation with initial nasolacrimal duct biopsy specimen. Pathology reported bland cytology with no discrete atypical mitoses. Slight nuclear hyperchromatism and hyperplasia suggestive of atypia.

Figure 1a. Patient 1 month after presentation. 2b Coronal CT orbits shows fungating mass within left lacrimal sac which projected into the nasolacrimal duct & anteriorly from the orbit. Dilation of osseous remodeling of the left nasolacrimal duct was noted. There was no evidence of orbital disease (figure 2b)
- MRI demonstrated a soft tissue mass centered on the left lacrimal sac and nasolacrimal duct with extension into the medial aspect of the extraocular fat and potential involvement of inferior oblique muscle (figure 2c, 2d)
- Incisional biopsy and repeat nasolacrimal duct biopsies revealed atypical epithelial proliferation favoring non-keratinizing SCC (figures 2e, 2f).

The lesion began to rapidly expand in the interim.

Figure 2. 2a Patient 1 month after presentation. 2b Coronal CT orbits shows fungating mass within lacrimal sac which projects inferorly from the NLD and anteriorly from the orbit. 2a Coronal and 2d Axial T1 MRI show a soft tissue mass in left lacrimal sac extending to the orbit clearly violating the extraocular fat. There potential violation of the infraorbital margin.

Figure 2a.
- Incisional & nasolacrimal duct biopsy shown nuclear hyperchromatisit, cellular pleomorphism with cellular atypia which imparts the impression of malignant change within a squamous papilloma. Due to the fragmented nature, invasion cannot be identified, but also cannot be ruled out.
- After multidisciplinary tumor board discussion, he underwent an extended medial maxillectomy, lateral rhinotomy, orbital exenteration, left anterior ethmoidectomy and left frontal sinusotomy. He was reconstructed using split calvarial bone graft, a superficial temporoparietal flap and a split thickness skin graft.
- Final pathology was consistent with primary lacrimal sac p16 negative and EBV negative non-keratinizing squamous cell carcinoma involving the orbital fat.

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References

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