

## Case Report

### Apocrine Adenocarcinoma of Lacrimal Sac Presenting as Chronic Dacryocystitis

Amber Hoang<sup>1\*</sup>, Kay T Khine<sup>2</sup> and Renzo Zaldivar<sup>2</sup>

<sup>1</sup>Department of Internal Medicine, New Hanover Regional Medical Center, Wilmington, North Carolina, USA

<sup>2</sup>Department of Ophthalmology, University of North Carolina School of Medicine, Kittner Eye Center, Chapel Hill, North Carolina, USA

#### Abstract

The objective of this paper is to report a case of lacrimal apocrine adenocarcinoma which presented as chronic dacryocystitis. A 62-year-old male was referred by his primary ophthalmologist for evaluation of chronic dacryocystitis of the right eye for 3 months. Biopsy showed a rare and aggressive type of cancer: apocrine adenocarcinoma of the nasolacrimal sac and paranasal sinuses. The patient was treated with orbital exenteration followed by chemoradiation therapy. Apocrine adenocarcinoma, a rare tumor of the eyelid glands, can masquerade as chronic dacryocystitis.

**Keywords:** Adenocarcinoma; Apocrine; Dacryocystitis; Lacrimal

#### Introduction

Apocrine adenocarcinomas are tumors originating from apocrine glands that line the follicles of the eyelashes called the glands of moll [1]. The primary site for apocrine adenocarcinoma is the axilla with about 17% of cases in the head and neck [2]. There are only 21 cases of ocular adnexal apocrine adenocarcinoma reported to date [3].

The mean age of patients with primary cutaneous apocrine adenocarcinoma is 67 years [2]. The most common surgical approach was local excision (50%), followed by wide excision (30%), mastectomy (4%), and amputation (0.6%) [2]. The prognosis of cutaneous apocrine adenocarcinoma is intermediate with a median survival of 51.5 months [2]. The presence of lymph node metastases significantly decreased survival to 33 months [2].

#### Case

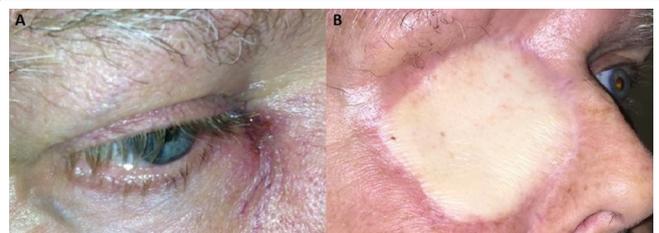
A 62-year-old Caucasian male was referred by his primary ophthalmologist for chronic dacryocystitis of the right eye (OD) for the

**\*Corresponding author:** Amber Hoang, New Hanover Regional Medical Center, 2131 South 17<sup>th</sup> Street, Wilmington, North Carolina, USA, Tel: +1 9106677000; Email: a.hoang@knights.ucf.edu

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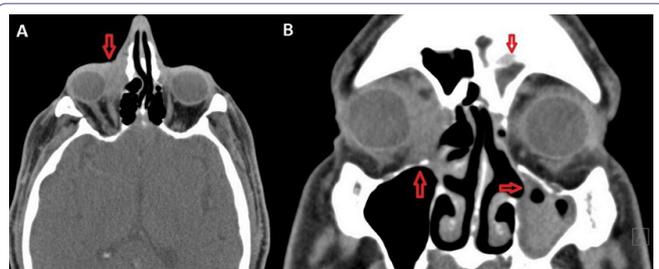
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past three months. In the summer of 2014, the patient first noticed a cystic lesion in the right medial canthus which he attributed to an eye infection after swimming in a hot tub. Around December 2014, he developed conjunctival injection and discharge and was prescribed Cephalexin by his primary ophthalmologist. His symptoms improved with antibiotics but he continued to have redness and discharge. He denied loss of vision or ocular pain. Past medical history was unremarkable. Ophthalmic examination at our institution revealed a firm ulcerative nodule at the right medial canthus above the medial canthus tendon with erythema and spontaneous pus drainage (Figure 1A). Visual acuity was normal and intraocular pressure was 21 mmHg OD. The patient was placed on Sulfamethoxazole and Trimethoprim. Culture of the pus drainage was positive for coagulase-negative *Staphylococcus aureus* species.



**Figure 1:** A) Ophthalmic examination at our institution revealed a firm mild nodule at the right lacrimal sac above the medial tendon with erythema and pus drainage. B) The patient was treated with a right orbital exenteration and neck dissection followed by right latissimus dorsi free flap reconstruction.

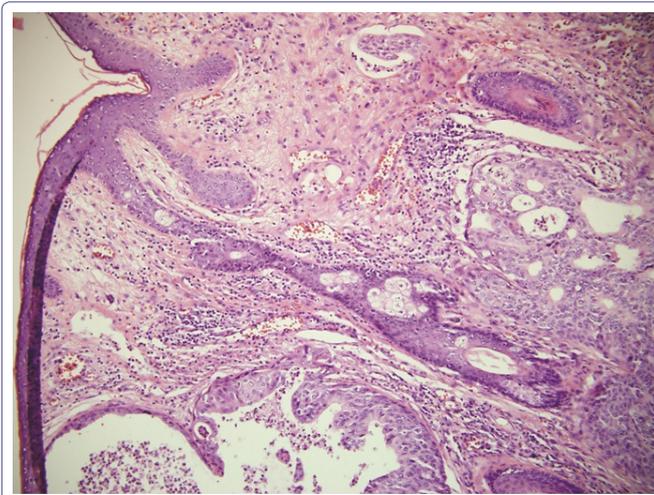
Given the severe skin erosion and concern for malignancy, radiologic imaging was obtained to assess the orbit and nasolacrimal system. Computed Tomography (CT) of the orbit revealed 2.8 × 1.5 × 1.6cm soft tissue mass seen in the right nasolacrimal duct region abutting the right globe with mild adjacent bone erosion (Figure 2).



**Figure 2:** Axial View. A) Non-contrasted CT of orbit reveals 2.8 × 1.5 × 1.6cm soft tissue mass seen in the right nasolacrimal duct region abutting the right globe with mild adjacent bone erosion concerning for malignancy (left). (coronal) B) Polypoid mucosal thickening is seen in the right maxillary sinus. There is mucosal sinus disease with near complete opacification of the left maxillary sinus and complete opacification of the left frontal sinus (right).

Biopsy of the lesion was immediately performed following CT imaging and pathological examination revealed apocrine adenocarcinoma of the right lacrimal sac and sinus mucosa (Figure 3).

Positron Emission Tomography (PET) scan revealed moderately avid bilateral level Ib/submandibular lymph nodes consistent with TNM stage of T3N2bM0. The patient underwent radical surgery including open craniofacial resection with frontal craniectomy, subtotal



**Figure 3:** Histopathological examination revealed poorly differentiated apocrine glandular cells consistent with apocrine adenocarcinoma of lacrimal sac extending to the peripheral edge and base.

maxillectomy, partial rhinectomy, right orbital exenteration and neck dissection followed by right latissimus dorsi free flap reconstruction. Irrigation was not performed since there was an active infection. He also completed 6 cycles of cisplatin (30mg/m<sup>2</sup>) and paclitaxel (60mg/m<sup>2</sup>) with concurrent 200cGy in 30 fractions of radiation therapy. In December 2015, the patient underwent a CT scan of the sinuses, Magnetic Resonance Imaging (MRI) cranial nerve V protocol, and PET scan which all demonstrated no recurrence of disease (Figure 1B).

## Discussion

According to a review of 19 cases of eyelid apocrine adenocarcinoma by Figueira et al., the mean age of patients affected was 61.4 years (range 36-85 years); 26% of cases were women [3]. The most common presentation was a painless, blue-brown eyelid lump [3]. About 47% of cases reported local orbital and periorbital invasion. Another 35% of patients had local lymph nodes involvement [3]. Distant metastases to the pelvic bone, lung, and brain were also reported [4,5].

The histopathological diagnostic criteria of apocrine adenocarcinoma described by Kipkie and Haust requires: 1) Pathognomonic "decapitation secretion," 2) Periodic Acid-Schiff (PAS) positivity, 3) Iron-positive intracellular pigment, 4) Eosinophilic cytoplasm, 5) Tissue from an area normally containing apocrine glands [6]. Immunohistochemistry is a useful diagnostic tool for differentiating apocrine adenocarcinoma from other subtypes such as apocrine hidrocystomas and apocrine cystadenoma. While benign tumors can also possess PAS-positivity and decapitation secretion, they distinctly stain positive for cytokeratins and human milk fat globulin 1 [7]. Furthermore, GCDFP-15 is helpful in cases where the characteristic "decapitation secretion" of apocrine epithelium is not easily observed, and it has been shown to stain positive in 84% of apocrine-related neoplasms [8]. Although GCDFP-15 was negative in our case, histopathological examination revealed poorly differentiated apocrine glandular cells consistent with apocrine adenocarcinoma.

The recommended management is combined exenteration surgery and radiotherapy in eyelid apocrine carcinomas with advanced metastases [3,5]. Nonmetastatic cases can be successfully treated with

wide local resections and radiotherapy as an adjuvant treatment for local orbital or nodal disease [5]. Local lymph node dissections have been reported to have variable success [4,9,10]. Chemotherapy was described in one of the cases with no follow-up report on survival [5]. This case report incorporated TMN staging, which is the current recommendation of the American Joint Committee's 7th Edition Eyelid Carcinoma Classification System on Cancer [11]. The most important survival determinants are the presence of lymph node involvement, local invasion, and systemic metastasis [3]. TMN staging can help physicians define prognosis and, subsequently, develop an appropriate treatment plan based on the extent of disease.

## Conclusion

Primary lacrimal sac tumor is extremely rare. Furthermore, the presentation of apocrine adenocarcinoma masquerading as chronic dacryocystitis makes this case unique. Given the aggressive nature of this tumor, it is critical that clinicians heighten their clinical suspicion for malignancy when assessing a case of chronic dacryocystitis with skin erosion and a palpable lacrimal mass.

## Declaration of Interests

The authors declare that they have no conflicts of interest.

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