JAMA Ophthalmology Clinical Challenge

An Unusual Upper Eyelid Mass

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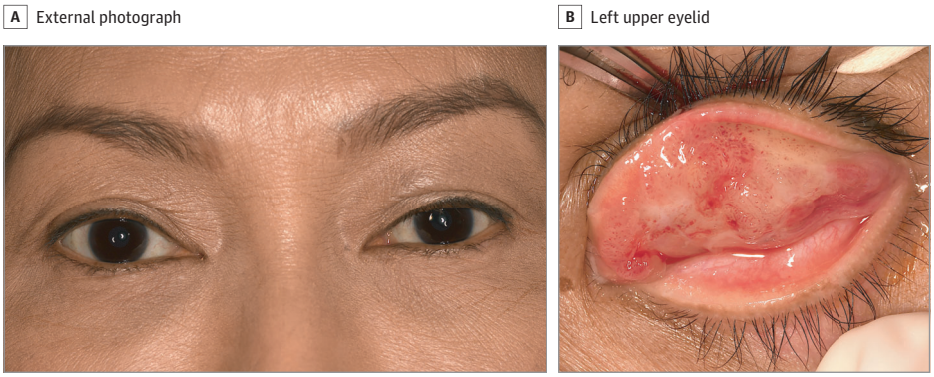

Figure 1. A, External photograph demonstrating left upper eyelid fullness. B, Everted left upper eyelid showing a diffuse, multinodular tarsal mass with hairpin-shaped vessels.

A 67-year-old Asian woman presented to her local ophthalmologist because of swelling of the left upper eyelid. Surgical drainage was performed, but the swelling persisted. Two months later, nontender preauricular fullness was noted, which prompted the patient to seek a second opinion. In the interim, magnetic resonance imaging of her orbit and parotid glands revealed focal enhancement in the left upper eyelid and superficial lobe of the left parotid gland. She was referred to us for diagnosis and treatment.

On examination, visual acuity was 20/20 in the right eye and 20/30 in the left eye. Ocular motility was full, and proptosis was absent. The left upper eyelid was thickened (**Figure 1**A). Eyelid eversion was not possible in the clinic because of mass effect, but a small portion of the tumor was visualized at the supranasal eyelid margin. All eyelashes were intact. A nontender left preauricular mass of 1 cm in diameter was palpable. The remainder of the findings on eye examination were unremarkable. After eyelid eversion in the operating room, the mass measured 35 mm in basal diameter and 5 mm in thickness and demonstrated an amelanotic appearance with a hairpin-shaped vascular pattern suggestive of an epithelial tumor (Figure 1B).

WHAT WOULD YOU DO NEXT?

- **A.** Administer topical interferon alfa-2b
- **B.** Start external beam radiotherapy
- **C.** Administer intralesional fluorouracil
- **D.** Perform a complete posterior lamellar resection

Diagnosis

Adenosquamous carcinoma of the conjunctiva

What to Do Next

D. Perform a complete posterior lamellar resection

Discussion

The differential diagnosis of a tarsal conjunctival mass includes various inflammatory or degenerative conditions, such as chalazion, trachoma, ligneous conjunctivitis, and foreign body inflammation as well as neoplastic conditions including conjunctival papilloma, squamous cell carcinoma, sebaceous carcinoma, and melanoma.[1] A persistent unilateral chalazion in an older woman raises concern for neoplasm. The nodular appearance, yellow color, and upper eyelid location should further narrow the differential to squamous cell carcinoma, sebaceous carcinoma, or amelanotic melanoma. The preservation of eyelashes somewhat argues against sebaceous carcinoma. A definitive diagnosis should be made by means of histopathologic examination. Therefore, complete posterior lamellar resection (choice D) would be both diagnostic and therapeutic in this situation, eradicating the mass and potentially providing diagnosis. A previous small incisional biopsy elsewhere suggested neoplasia, but complete excision provides greater tissue analysis, staining, and information for diagnosis. Other choices, such as topical interferon alfa-2b (choice A) and fluorouracil injections (choice C), would be recommended for minor squamous neoplasia, but not for disease as advanced as this case. External beam radiotherapy (choice B) would be appropriate in a case of potential metastatic disease.

After surgical resection, histopathologic examination revealed squamous neoplasia of the tarsal conjunctiva with invasion of the underlying tarsus, adjacent orbicularis, and superior orbit. The neo-

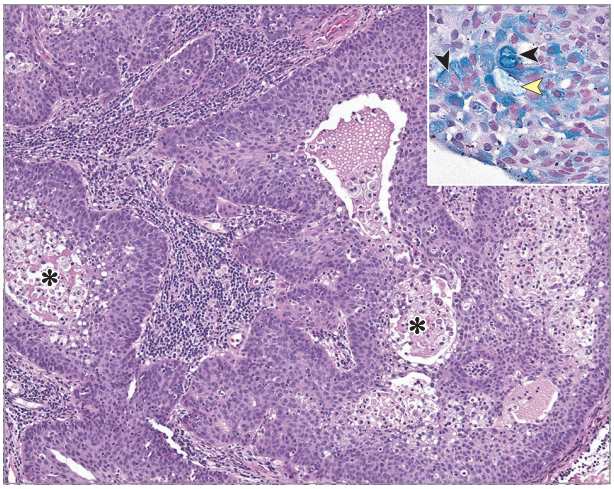

Figure 2. Adenosquamous carcinoma (squamous cell carcinoma with mucinous differentiation). Invasive neoplasm, composed of irregular nests of pleomorphic, mitotically active squamous cells with central acantholysis (asterisks) and comedonecrosis (hematoxylin-eosin, original magnification ×50). Inset, Focal intracytoplasmic (black arrowheads) and extracellular (yellow arrowhead) mucin (Alcian blue, original magnification ×100).

plastic cells featured pleomorphic nuclei with central nucleoli and abundant cytoplasm with intercellular bridges, compatible with squamous differentiation, but lacked keratinization. Alcian blue stain highlighted rare goblet cells with intracytoplasmic mucin in the invasive component of the tumor. No nuclear or cytoplasmic features of sebaceous carcinoma were seen. Focal lymphovascular invasion was observed (**Figure 2**). These features were compatible with adenosquamous carcinoma (formerly termed *mucoepidermoid squamous cell carcinoma*).

Adenosquamous carcinoma is a rare, highly aggressive malignant neoplasm.[2] When adenosquamous carcinoma was first identified in the conjunctiva, it was termed *mucoepidermoid carcinoma*, because of its morphologic similarity to mucoepidermoid carcinoma of major salivary glands.[3] Mucoepidermoid carcinoma is a neoplasm of glandular derivation featuring 3 cell types: epidermoid, intermediate, and mucinous.[4] In contrast, conjunctival adenosquamous carcinoma, a neoplasm of surface epithelial derivation, lacks intermediate cells and frequently displays prominent squamous differentiation with keratinization. Thus, the terminology has been redefined to adenosquamous carcinoma, a tumor arising from surface epithelium demonstrating both squamous and glandular differentiation.[5,6]

Regardless of the nomenclature, these lesions are aggressive with a potential for metastasis, as observed in this case. Thus, careful evaluation by pathologists for mucinous cells in conjunctival carcinoma is essential for appropriate diagnosis and timely management.

Patient Outcome

After surgical excision of the posterior lamella of the upper eyelid with anterior orbit, negative surgical margins were found and the patient subsequently underwent sentinel lymph node biopsy with head and neck dissection, confirming suspected lymph node metastasis. Because of the presence of intraparotid lymph node metastasis, the patient received adjuvant proton beam therapy to the parotid gland, targeting any residual tumor. Concurrent cisplatin chemotherapy was also administered to prevent systemic recurrence. She remained disease free at the 1-year follow-up.

ARTICLE INFORMATION

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Published Online: October 22, 2020. doi:10.1001/jamaophthalmol.2020.2859

Conflict of Interest Disclosures: None reported.

Funding/Support: This study was supported by the Eye Tumor Research Foundation (Dr Shields).

Role of the Funder/Sponsor: The Eye Tumor Research Foundation had no role in the design and conduct of the study; collection, management, analysis, and interpretation of the data; preparation, review, or approval of the manuscript; and decision to submit the manuscript for publication.

Additional Contributions: We thank the patient for granting permission to publish this information. The authors acknowledge Antonio Yaghy, MD, Ocular Oncology Service, Wills Eye Hospital, Thomas Jefferson University, for his contribution to the article's written composition. Dr Yaghy was not compensated for his contribution.

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