

Syringoid eccrine carcinoma of the eyelid presenting as cicatricial entropion

Manjool M. Shah ¹, Eric A. Steele ^{1,2}, Kevin P. White ³, David J. Wilson ¹

¹Department of Ophthalmology, Casey Eye Institute, Oregon Health & Science University, Portland, Oregon 97239, USA

²Portland Veteran's Affairs Medical Center, Portland, Oregon 97201, USA

³Department of Dermatology, Oregon Health & Science University, Portland, Oregon 97203, USA

Correspondence to: Eric A. Steele. Department of Ophthalmology, Casey Eye Institute, 3375 SW Terwilliger Blvd., Portland, Oregon 97239, USA.

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Dear Sir,

My name is Manjool M. Shah, from the Casey Eye Institute at Oregon Health & Science University in Portland, Oregon, USA. I wish to write to you with regards to an interesting presentation of a rare eyelid malignancy.

A 41-year old Caucasian female was referred to the oculoplastic service by her comprehensive ophthalmologist for evaluation of symptomatic left lower lid entropion. The patient had been experiencing approximately six months of foreign body sensation and ocular surface irritation.

At her initial visit, the patient denied previous history of trauma or surgery involving the lid. Her examination was notable for a cicatricial left lower lid entropion, with distortion of her left lower lid margin architecture and nearly complete madarosis of the nasal half of the lid (Figure 1). The entire margin in this area was rotated in towards the eye, with keratinized tissue rubbing on the cornea. There was a slightly pearly character with fine telangiectasias. Slit lamp examination of the anterior segment showed epithelial erosions on the inferior aspect of the left cornea, and was otherwise unremarkable. The vision was intact, and ocular motility was full. There was no lymphadenopathy. Carcinoma was suspected, and an incisional biopsy was performed at this time.

The changes in the initial biopsy were subtle, and diagnosed descriptively as epithelial hyperplasia and inflammation, without evidence of carcinoma. The clinical suspicion for carcinoma remained high, and a subsequent full thickness pentagonal wedge biopsy measuring 6 mm in width was undertaken and the lid was reconstructed primarily. The histopathological findings in this second biopsy were



Figure 1 Clinical photograph demonstrating the patient's left lower lid entropion.

consistent with scar.

With two biopsies interpreted by the dermatopathology service as negative for malignancy, the decision was made to proceed with more definitive scar excision and reconstruction of the lower lid, as the patient continued to be symptomatic from the lower lid entropion. A full thickness excision of approximately half of the lower lid was performed, and a Hughes tarsoconjunctival pedicle flap was used to reconstruct the posterior lamella. The undermined anterior lamella was draped over the defect. The excised tissue was sent to pathology for permanent sections.

Surprisingly, this excisional specimen showed findings not apparent in the initial two biopsies, with changes suggesting syringoid eccrine carcinoma (Figure 2). Because this excision and reconstruction had not been performed with attention to margin control, the patient agreed to proceed with additional resection and reconstruction soon thereafter. The newly-constructed Hughes flap was excised, and additional lid tissue was removed with frozen section margin control. The resultant defect comprised nearly the entire left lower lid. A hard palate mucosal graft was placed to reconstruct the posterior lamella of the lid, and a Mustarde rotational facial flap was utilized to reconstruct the anterior lamella of the lid. Nine months later, the patient underwent a revision of the lid, and a representative biopsy at that time was negative for recurrent malignancy. With 12mo of follow up since complete resection of the carcinoma, the patient has done well, although she continues to have intermittent symptomatic eye irritation that is controlled with lubrication. Syringoid eccrine carcinoma (SC) is an extremely rare malignant neoplasm of eccrine sweat glands, with relatively few reports in the literature describing involvement in the periocular structures [1-5]. Historically, SC has suffered from inconsistency in nomenclature, often being described as syringomatous carcinoma, microcystic adnexal carcinoma, malignant syringoma, sclerosing sweat duct carcinoma, clear cell syringoid carcinoma, and eccrine carcinoma [1]. In general, the well differentiated subtype is the most commonly

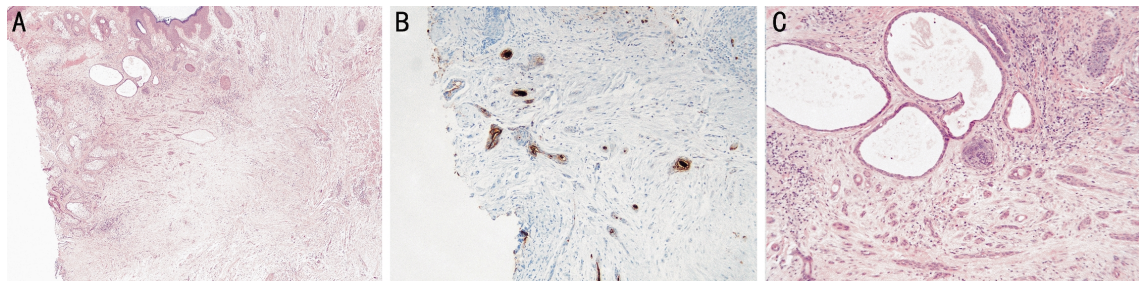


Figure 2 Photomicrographs demonstrating pertinent histopathological findings A: Hematoxylin and eosin (H&E) stained images (2.5×) revealing characteristic ductal structures with surrounding basaloid epithelial cells; B: Magnified image demonstrating positive carcinoembryonic antigen immunohistochemical staining of ductal structures; C: 10× magnification of H & E stained section.

encountered form of this entity.

SC typically presents in the fourth to fifth decade of life, predominantly in the head and neck region. Lesions often present similarly to morpheiform basal cell carcinoma, with a slow growing fleshy nodule or plaque with poorly-defined margins and fine telangiectasias. These tumors are highly invasive and locally destructive, with a predilection for perineural invasion and spread^[4]. Lymph node involvement has only been described in one case, but this was thought to have been secondary to direct perineural extension rather than true lymphatic spread.

Microscopically, there are usually small collections and cords of basaloid epithelial cells in the dermis, often surrounding small ductal structures reminiscent of those seen in syringoma. The neoplasm frequently extends throughout the dermis and into the underlying subcutis, and perineural involvement is common. In approximately 85% of cases, SC is encountered in the centrofacial region, but other facial tissues can be involved^[6].

The literature describes numerous case reports in which SC was misclassified on initial histopathological examination. As with our patient, these cases describe the utilization of numerous incisional and excisional biopsies before a definitive diagnosis was reached. This suggests the difficulty with which this entity is diagnosed, owing not only to its rarity but also to the fact that appropriate diagnosis requires assessment of architectural patterns, which may either be unavailable or unapparent in the initial tissue specimens.

Additional immunohistochemical stains were utilized to confirm the diagnosis of SC in this patient. Pancytokeratin highlighted the neoplastic collections, confirming epithelial origin, while carcinoembryonic antigen (CEA) highlighted small ductal structures within the aggregates (Figure 2). Of note, this latter finding suggesting ductal differentiation supports a diagnosis of syringoid eccrine carcinoma, and is not typical of basal cell carcinoma. Immunohistochemical staining has been described as quite variable, although cytokeratins and CEA are routinely noted to be positive^[7].

Treatment consists primarily of complete local excision, ensuring for tumor-free margins. Mohs micrographic surgery has been described for this entity as well^[8]. The literature suggests that given the predilection for perineural invasion, local recurrence rates approach 40% -60%^[1]. Radiotherapy

has been suggested as a potential adjunctive treatment modality, but local recurrence may still occur. Orbital extension has been described in cases of SC initially presenting as an eyelid finding, and orbital exenteration has been suggested given the aggressive nature of the malignancy^[5].

This case represents a particularly novel presentation of this malignancy resulting in cicatricial entropion. The differential diagnosis for cicatricial entropion is broad, and typically includes autoinflammatory, infectious, surgical, and traumatic etiologies. Malignant neoplasia are rarely included. Despite this, SC should be considered in cases of atypical cicatricial entropion given its aggressive behavior and high rate of recurrence. The clinical suspicion should be conveyed to the dermatopathologist to ensure the specimen is evaluated with this rare entity in mind.

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