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Sebaceous Cell Carcinoma of the Eyelid: A Rapidly Enlarging Lesion with Massive Xanthogranulomatous Inflammation

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Abstract

A 76-year-old man presented atypically with 4 week history of a rapidly enlarging ulcerated nodular lesion of the left upper eyelid that was found to be sebaceous cell carcinoma. Further investigation showed no metastatic disease, and Mohs surgery was performed to resect the tumor. Histopathologic analysis showed features diagnostic of sebaceous cell carcinoma. However, most of the mass consisted of xanthomatous granulomatous inflammatory reaction vastly out of proportion with the tumor burden. The patient was spared from orbital exenteration, and no evidence of recurrence was present 6 months after resection.

Sebaceous cell carcinoma (SebCCa) of the eyelid is an uncommon and challenging tumor to recognize and treat successfully. The initial likelihood of recognition is only 18.6% among physicians in general and 50% among ophthalmologists.¹ The classic clinical picture is a painless and yellowish nodule of the upper eyelid with associated destruction of microarchitecture and disproportionate eyelash loss. SebCCa, however, is notorious for masquerading as other entities, resulting in delayed diagnosis of 6 months to 1 year or longer contributing to increased morbidity and mortality. A case of rapidly enlarging SebCCa of the eyelid with unique histopathologic characteristics is reported.

Case Report

A 76 year-old man presented to his ophthalmologist with a 4 week history of mild erythema and edema of the left upper eyelid. The patient was initially treated conservatively for a chalazion with topical antibiotic/steroid ointment and asked to return in 3 weeks. On his follow up visit, a marked increase in the size of the lesion and features suspicious for a malignancy were noted. An incisional biopsy at the time yielded a preliminary diagnosis of SebCCa, prompting a referral for further management. A week later at oculoplastics consultation, the patient reported a 4 week history of a rapidly enlarging lesion with yellowish discharge, episodes of hemorrhage, and decreased vision of the left eye due to mechanical obstruction (Figure 1A).

Examination showed visual acuity of 20/40 OD and 20/60 OS. A very aggressive appearing 20mm × 14mm × 6mm pedunculated, multilobulated, yellowish, and ulcerated mass with

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areas of necrosis and telangiectasia protruded from the posterior lamella of the left upper eyelid. The rest of the ophthalmic exam was normal except for moderate bilateral nuclear sclerosis, and no palpable lymph nodes were present in the head and neck. Subsequent PET/CT showed the hypermetabolic left upper eyelid mass (Figure 1B). No regional or distant metastatic disease was identified. Conjunctival map biopsies were deferred owing to the normal clinical appearance of the bulbar conjunctiva, the inferior fornix, and the inferior palpebral conjunctiva.

The patient was promptly referred for resection of the tumor with a modified Mohs technique with additional margins using frozen and paraffin embedded sections. The histopathology showed a tumor with papillomatous epithelium that was infiltrated by malignant cells with abundant vacuolated cytoplasm and small nuclei, thereby confirming SebCCa with pagetoid spread (Figure 2A) and mitotic figures (Figure 2B). However, the peripheral margins of the tumor showed widespread xanthogranulomatous reaction without any evidence of SebCCa (Figure 2C). Further evaluation of the center of the tumor by immunocytochemistry showed cytokeratin positive SebCCa infiltrating the mass and small foci in the subepithelial stroma (Figure 2 D & E). These tumor cells were surrounded by a much larger dense collection of CD 68 positive foamy histiocytes (Figures 2F & 2G) that composed the bulk of the tumor.

Discussion

This patient's clinical presentation of SebCCa was unusual for the extremely rapid progression. Additionally, the histopathologic findings were unique in these authors' experience. There have been reports of lipid from tumor inciting a foreign body giant cell reaction that may superficially resemble a chalazion, but, in general, SebCCa appears to generate a less intense inflammatory response with T helper cells than basal cell carcinoma.² A literature review performed on PubMed with keyword "sebaceous cell carcinoma" and a review of several atlases of eyelid tumors,^{3,4,5} revealed no cases with histopathology similar to findings seen in this report. One recent report showed macrophage infiltration localized to tumor cell nests in 57% their cases,⁶ but none seemed to have presented with massive reaction as noticed in our case. In one author's experience (DMA), with over 200 cases of SebCCa, this was the only case with massive xanthomatous granulomatous reaction to the lipid laden tumor cells.⁷ These tumor cells were stained positive on cytokeratin and appear distinct from CD68 stain positive foamy macrophages. What was initially believed to be intraorbital extension of the tumor near the insertion of the left medial rectus muscle, was found to be xanthomatous granulomatous inflammatory reaction to the tumor with clear margins. Careful long-term follow-up will be necessary to observe for local recurrence or metastatic disease. In this very unusual case, a reasonable, but incorrect, assumption that the orbital findings observed on imaging were tumor invasion could have led to unnecessary exenteration of the orbit with resultant disfigurement and loss of sight.

Précis

A case of sebaceous cell carcinoma of the eyelid presenting with rapidly enlarging ulcerated nodular mass and found to have widespread xanthomatous granulomatous inflammatory reaction surrounding the tumor cells.

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Figure 1.

A. Clinical photograph of patient showing yellowish ulcerated mass of the posterior lamella of the left upper eyelid. **B.** PET/CT of the region showing hypermetabolic left upper eyelid mass that had a maximum standard uptake value (SUV) of 10.6 abutting the nasolacrimal apparatus with thickening of the skin extending onto the nasal sidewall and postseptal extension involving the attachment of the medial rectus muscle, and the Tenon's capsule thought to be orbital extension of the tumor.

Figure 2.

A. H & E stained section of the lesion showing papillomatous conjunctival epithelium infiltrated by SebCCa cells with cytoplasmic vacuolations. **B.** Enlarged view of malignant SebCCa cells with mitotic figures (arrows)(400X) **C.** Enlarged view of foamy histiocytes present in the deep orbital portion of the tumor (400X) Inset photograph showing a multinucleated giant cell (arrow) **D.** Cytokeratin (high molecular weight) immunostaining of the lesion showing tumor cells in the center (arrows) and within the epithelium (arrowheads) (2X) **E.** Enlarged view of cytokeratin stained positive SebCCa cells within the epithelium (200X) **F.** CD68 immunostaining of the lesion showing a dense mass of CD 68 positive histiocytes in the subepithelial stroma (2X) **G.** Enlarged view of CD 68 positive macrophages (400X)