Conjunctival Sebaceous Carcinoma Masquerading As Chronic Blepharoconjunctivitis

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INTRODUCTION

The masquerade syndrome was first described by Theodore (1), and Irvine (2) independently in 1967, as chronic blepharoconjunctivitis due to an underlying conjunctival carcinoma. Conjunctival intraepithelial neoplasia and sebaceous cell carcinoma have been reported to show intraepithelial (pagetoid) spread to the conjunctiva mimicking chronic blepharoconjunctivitis, or cicatrizing conjunctivitis (3,4). These lesions have no distinct borders and therefore, may be clinically indistinguishable from the uninvolved tissue. Patients with this form of the disease are more prone to having multiple tumor recurrences and requiring multiple surgeries with poorer outcomes, largely because of the difficulty and delay in diagnosis and treatment.

We describe herein a patient who was initially diagnosed with chronic conjunctivitis/blepharoconjunctivitis. Biopsy that was performed due to the unresponsiveness to medical treatment and persistent inflammation uncovered underlying conjunctival sebaceous carcinoma.

REPORT OF CASE

Figure 1: Left eye of the patient. Slit lamp photograph shows severe blepharitis with thickening of eyelid margins, subepithelial fibrosis with fornix foreshortening, and a fibrovascular pannus extending over the peripheral cornea.

In February of 1994 a 74 year-old woman with a 5 year history of blepharitis and dry eyes was referred for evaluation of her cicatrizing conjunctivitis. The patient's past medical history revealed eczema dating from childhood, radiation therapy for that with subsequent development of multiple skin cancers throughout most of her adult life. She also had had bilateral cataract surgery followed by bilateral retinal detachment and repair. Her visual acuities were 20/25 and 20/200 right and left eyes, respectively. She was noted to have mild lower lid laxity and moderate mucoid discharge in both eyes. Slit lamp biomicroscopy of the right eye revealed severe blepharitis, lid margin telangiectases and madarosis. Mild conjunctival hyperemia and superficial punctate keratopathy with mild anterior stromal scarring were also noted. The patient's left eye had more severe involvement with blepharitis and thickened eyelid margins, marked subconjunctival fibrosis with fornix foreshortening and symblepharon formation (**Figure 1**). The entire lower forniceal conjunctiva had an unusual appearance, with "frosting" of opalescent epithelium. The epithelium of the left cornea, was drawn in a vascularized pannus which decreased the patient's visual acuity. The patient was diagnosed with atopic blepharokeratoconjunctivitis and associated sicca syndrome and was treated with topical cromolyn sodium eye drops, artificial tears, and dexamethasone ointment as well as systemic antihistamine per os. A conjunctival biopsy performed for definitive diagnosis disclosed intraepithelial atypical cells with vacuolated cytoplasm (**Figure 2**). Lipid stains were positive establishing the diagnosis of sebaceous carcinoma. Several conjunctival map biopsies revealed extensive, diffuse involvement of the palpebral as well as bulbar conjunctiva. A left orbital exenteration was recommended. The patient decided against any surgery and was lost to follow-up. In July 1995, we received a report stating that she had bilateral conjunctival sebaceous carcinoma with systemic metastasis. In April 1997, she died due to metastatic disease.

Figure 2: Histopathology shows pagetoid spread of malignant sebaceous cells in the conjunctival epithelium (Hematoxylin-Eosin stain; original magnification, x 100).

DISCUSSION:

Sebaceous carcinoma most often originates from the meibomian glands, glands of Zeis, and caruncle (5). It predominantly occurs in elderly people in the sixth decade of life and is more common in women (6). Bilateral sebaceous cell carcinoma has been described in patients who had previous radiation treatment after a latent period of 8 to 56 years (7,8). Our patient had a past medical history of radiation therapy in childhood for eczema and allergies with subsequent development of bilateral sebaceous carcinoma at the age of 74. Sebaceous carcinoma of the eyelid is a relatively rare condition comprising about 1 % of all malignant tumors of the lid (5). It is an important lesion however, since it is one of the most malignant primary tumors of the eye, with 5-year and 10-year actuarial tumor death rates of 15% and 28% respectively (6). A delay in establishing the correct diagnosis is the key factor for this high mortality. It has been shown that the 4-year mortality rate increased from 13 % to 43% when the duration of symptoms before the excision of the tumor was greater than 6 months (6). Unfortunately, the average time between the presentation and diagnosis is 1 to 3 years (9). The lesion usually presents as a slowly enlarging, firm, painless mass mimicking a chalazion. But the firmness of the mass, loss of eyelashes, nodularity and thickening of the eyelid margins are clues indicative of probable sebaceous carcinoma rather than chalazion. The diagnosis is based on the presence of sebaceous elements within the tumor. The demonstration of these elements by lipid stains on frozen sections is helpful in differentiating this tumor from squamous cell carcinoma. Particularly in cases with diffuse epithelial involvement, the intraepithelial tumor cells in paraffin-embedded conjunctival specimens may be easily overlooked or misinterpreted, since the lipid inside the sebaceous cells is removed during specimen processing. In suspected cases, a full thickness lid biopsy and alerting the pathologist to the possibility of malignancy are essential for maximizing the likelihood of correct diagnosis. Exenteration is indicated in cases with documented orbital invasion.

In conclusion, all cases of persistent external ocular inflammation should be viewed with a high index of suspicion for a possible underlying malignancy. Early histopathologic examinations should be performed on all cases of chronic conjunctivitis that are unresponsive to the usual medications. The specimens should be handled and processed appropriately, and examined by an experienced pathologist who has been alerted to the possibility of malignancy. Close follow-up of patients to detect local recurrences and systemic involvement is mandatory in the care of patients with ocular malignancy.

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