Conjunctival Mucoepidermoid Carcinoma: Clear Cell Variant

Mucoepidermoid carcinoma is a rare conjunctival neoplasm\(^1\) that is a common malignancy in the salivary glands.\(^2,3\) In the conjunctiva it often mimics squamous cell carcinoma.\(^3\) Histologically the tumors are believed to arise from pluripotential basal cells and, possibly, mucus-secreting elements, and therefore are composed of mucus-secreting cells intermixed with epidermoid cells.\(^1\)

The clear cell variant of mucoepidermoid carcinoma has been described in the major and minor salivary glands, the oral cavity, the skin, and the upper respiratory tract.\(^4-6\) We report a case of a clear cell variant of mucoepidermoid carcinoma of the conjunctiva. To our knowledge, this variant has not been previously reported in the conjunctiva.

Report of a Case. A 65-year-old Hispanic man was first seen by us with a 2-month history of a large, multilobulated conjunctival tumor on the lateral epibulbar surface of the left eye. His ocular history was unremarkable. His medical history was pertinent for uncontrolled diabetes mellitus for 5 years and “fluid removal from the right lung” within the past year.

On examination, his best-corrected visual acuity was 20/50 OU and his intraocular pressure was 11 mm Hg OD and 18 mm Hg OS. External examination showed a large temporal mass without adenopathy. In the left eye, an elevated mass with a multilobular surface, measuring 15.0 × 15.0 × 8.0 mm, extended from the lateral canthus to the cornea (Figure 1). The remaining superior bulbar conjunctiva was unremarkable, and no masses were seen on the lid or in the fornix. No orbital masses were palpable. The remaining anterior segment examination was unremarkable except for mild nuclear sclerosis. Gonioscopy demonstrated an open anterior chamber angle.

An incisional biopsy was performed in an attempt to debulk the mass, and cryotherapy was applied to the margins. Extensive bleeding occurred during the procedure, and it was difficult to dissect the mass to the posterior extent of the lesion.

Histopathologic examination of the conjunctival tissue revealed a large, multilobular tumor in the substantia propria that focally infiltrated the superficial epithelium. The epithelium was acanthotic in the areas of tumor invasion and replaced by clear tumor cells (Figure 2). In
the substantia propria, the tumor lobules were composed of a mixture of clear cells and areas of squamous differentiation (Figure 2 and Figure 3). The clear cells exhibited large, hyperchromatic, highly pleomorphic nuclei. Frequent mitotic figures were seen (Figure 4). Squamous cells with hyperchromatic nuclei and eosinophilic cytoplasm were observed focally. The clear cells showed intracytoplasmic periodic acid–Schiff–positive granules that were sensitive to diastase (Figure 5). Rare clear cells deep within the substantia propria showed positive staining with mucicarmine (Figure 6) and Alcian blue, indicating the presence of mucus-secreting cells. Immunohistochemical studies showed no staining of the tumor cells for S100 protein. The tumor extended to the margin of the resection.

The patient subsequently underwent exenteration of the left orbit. Further investigation revealed emphysema, enlarged mediastinal lymph nodes, and an area of pleural thickening. Other clinical and radiologic details of his follow-up and the pathological diagnosis of the mediastinal lymph nodes were not available.

Comment. This case highlights the clinical and histologic findings of an unusual variant of mucoepidermoid carcinoma—the clear cell variant in the conjunctiva. The most common clinical presentation of mucoepidermoid carcinoma in the conjunctiva is a limbal/perilimbal mass that is usually interpalpebral, similar to squamous cell carcinoma of the conjunctiva.3

The grading of mucoepidermoid carcinoma is dependent on the number of mitotic figures, nuclear pleomorphism, perineural invasion, necrosis, and solid vs cystic morphologic characteristics and not on the proportion of cell types.2 Although the presence of clear cells does not affect the grading of mucoepidermoid carcinoma, it has been noted that clear cells usually predominate in high-grade tumors.7 The origin of these clear cells in mucoepidermoid carcinoma has been debated. They may be a result of hydropic degeneration of the epidermoid cells, but may also result from fixation artifacts, sparse organelles, or the presence of mucin, glycogen, or lipid.2,4

The other unusual feature of this tumor was the infiltration of the superficial epithelium by the neoplastic cells, akin to a “pagetoid growth pattern.” The apparent pagetoid change and the presence of clear cells made it difficult to differentiate this
tumor from sebaceous cell carcinoma. However, sebaceous carcinoma cells do not exhibit the abundant periodic acid–Schiff–positive granules in the cytoplasm and, instead, stain positive with lipid stains.8

Apart from sebaceous cell carcinoma, the differential diagnosis of clear cell carcinomas in the conjunctiva includes balloon cell melanoma. The lack of intracytoplasmic melanin pigment and S100 protein immunostaining made balloon cell melanoma unlikely. The differential diagnosis may also include clear cell oncocytoma, eccrine acrospiroma, acinic cell adenocarcinoma, and metastatic carcinomas from the thyroid, kidney, or lung.2,4,6 In our patient, metastatic clear cell carcinoma from the lung was considered, but histologic sections showing transitional areas that included squamous and mucus-secreting cells suggested that the tumor was conjunctival in origin.

In the conjunctiva, mucoepidermoid carcinomas are aggressive tumors that are locally and regionally invasive.3,9 Recurrence rates after excision are almost 100%.3 Therefore, early diagnosis is essential. Lymph node involvement has been described, but distant metastasis is unusual if the tumor is treated aggressively.9 The clear cell variant in the conjunctiva may prove to be more aggressive. The treatment includes wide excision, adjuvant cryotherapy, and enucleation/exenteration in addition to radiotherapy for recurrent lesions.9,10

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2. Ellis GL. Clear cell neoplasms in salivary glands.
Epithelial Downgrowth Complicating Evisceration With Orbital Implant Exposure

We describe an unusual case of implant exposure with epithelial downgrowth into the scleral pouch following evisceration. The patient’s clinical course and treatment as well as a histologic analysis of the excised scleral pouch are detailed. Excision of the implant and scleral pouch followed by diamond-shaped dermis-fat graft replacement was eventually required. The internal aspect of the sclera was completely covered with nonkeratinized stratified squamous epithelium. Epithelial downgrowth following evisceration is a rare complication that should be considered when orbital implant exposure occurs or when cysts overlying the sclera are discovered. Epithelial downgrowth needs to be adequately treated to allow for successful socket reconstruction and ocular prosthesis retention.

Evisceration is a common opthalmic procedure that has historically been used for cases of severe ocular trauma, cosmetic improvement of a disfigured eye, endophthalmitis, and blind, painful eye.1 Evisceration is considered a safe and effective procedure with few postoperative complications. Reported complications include prolonged postoperative pain and swelling, hemorrhage, suppurative necrosis and extrusion of sclera, orbital cellulitis, meningitis, extrusion of implants, and sympathetic ophthalmia.2,3 We present a case of epithelial downgrowth into the scleral pouch following evisceration. To our knowledge, it is only the third case described in the peer-reviewed literature and the first case described since 1965.2,3

Figure 1. Large anterior orbital conjunctival cyst with underlying area of orbital implant exposure in October 2002, occurring 18 months following scleral patch graft and approximately 2 years following the original evisceration.

Report of a Case. In October 2000, a 28-year-old white man had a blind, painful right eye. The patient had sustained a penetrating injury by scissors to the right eye at 2 years of age. The patient did not have details of the original injury or treatment, and the previous records were not available. Ophthalmic examination in October 2000 showed significant retinal scarring with stromal haze of the cornea in the right eye and an unremarkable left eye. Uncomplicated evisceration of the right eye with removal of the cornea and intraocular contents was performed in standard fashion 1 month later. The internal aspect of the sclera was swabbed with alcohol, and a 20-mm silicone implant was inserted. Histopathologic analysis of the excised cornea showed band keratopathy and endothelial decompensation with evidence of a previous, healed, full-thickness corneal wound. The intraocular contents showed uveal tissue, atrophic retina, and mature bone consistent with osseous metaplasia of the retinal pigment epithelium.

Postoperative follow-up at 1 week and 1 month showed healthy conjunctiva and socket, and the patient was fitted for an ocular prosthesis 2 months later. Examination 1 month after this showed a 5- to 6-mm area of centrally located orbital implant exposure. One week later, in March 2001, the patient underwent right orbital implant reinsertion with scleral patch graft reinforcement and conjunctivoplasty. Histologically, the excised conjunctival tissue revealed epithelial thickening and scarring with chronic inflammation. Examination 1 week and 1 month postoperatively revealed no implant exposure and well-healed conjunctiva. Vaulting of the ocular prosthesis was advised to provide a better fit.

Eighteen months following the scleral patch graft (2 years following the original evisceration), the patient reported displacement of the prosthesis. On examination, a fairly large anterior orbital cyst was noted (Figure 1). Excision of the cyst was performed in October 2002. The cyst was noted to be thin walled, containing a predominantly clear, slightly mucinous fluid. A 3- to