Intraocular Neoplastic Cyst From Mucoepidermoid Carcinoma of the Conjunctiva

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An 89-year-old woman with recurrent conjunctival mucoepidermoid carcinoma developed intraocular inflammation and an elevated fundus lesion that simulated choroidal detachment in her affected right eye. Intraocular invasion of squamous cell carcinoma was suspected and the eye was enucleated. Pathologic examination of the enucleated eye showed intraocular invasion by conjunctival mucoepidermoid carcinoma that formed a supraretinal cyst lined with malignant epithelial cells. The patient developed an orbital recurrence 1 year later and underwent orbital exenteration. She died 2 years later from an unrelated cause. Conjunctival mucoepidermoid carcinoma can exhibit intraocular invasion and produce an intraocular neoplastic cyst.

Mucoepidermoid carcinoma arises mainly in the salivary glands, but has also been noted in the upper respiratory tract and rarely in other sites. In the ocular region, it can affect the eyelid, conjunctiva, caruncle, lacrimal gland, lacrimal sac, and the paranasal sinuses. Conjunctival mucoepidermoid carcinoma has a tendency to recur after excision and invade the eye and the orbit. We describe a patient with recurrent conjunctival mucoepidermoid carcinoma that demonstrated intraocular invasion and formed a large intraocular cyst lined by neoplastic cells.

REPORT OF A CASE

An 89-year-old woman developed an exophytic papillary conjunctival lesion in her pseudophakic right eye, affecting the nasal bulbar and limbal conjunctiva between the 2-o’clock and 4-o’clock positions. The lesion was treated with wide excision and cryotherapy. Histopathologic examination revealed conjunctival squamous cell carcinoma with focal mucin production, consistent with mucoepidermoid carcinoma (Figure 1). She had a recurrence 3 months later (Figure 2), and was again treated with wide excision and cryotherapy. A wound leak through scleral dehiscence was discovered during the second surgery but was successfully treated with a patch graft of Tenon fascia and conjunctiva.

The patient had ocular pain 2 months later. Ophthalmic examination disclosed a red epibulbar mass at the prior site. There was no wound leak or hypotony, but marked intraocular inflammation was found. An elevated fundus lesion resembling choroidal detachment was observed ophthalmoscopically through hazy, inflamed media. The mass was located inferiorly between the 5-o’clock and 7-o’clock positions and measured approximately 10×10 mm in basal diameter. B-scan ultrasonography showed a 9.5-mm-thick, acoustically hollow cystic lesion in the supraretinal space (Figure 3). A neoplastic cyst caused by intraocular invasion of the conjunctival malignant neoplasm was suspected based on these findings. The patient underwent modified enucleation with wide tenonectomy and conjunctivectomy to include the recurrent bulbar tumor.

Results of a histopathologic examination showed that the conjunctival carcinoma had invaded the interior of the eye through a scleral wound. The tumor had grown posteriorly through a cyclodialysis...
cleft into the supraciliary and suprachoroidal space, forming a neoplastic cyst (Figure 4). The cyst was lined by tumor cells growing on the inner surface of the sclera and the outer surface of the detached uvea. No mucin production was identified intraocularly or in the tumor on the outer surface of the globe.

The patient developed massive orbital recurrence 1 year after enucleation. An eyelid-splitting orbital exenteration was performed. Histopathologically, the tumor exhibited squamous cell carcinoma but no mucus-producing cells were evident. The patient had no further recurrence and died 2 years later of cardiac disease.

**COMMENT**

Squamous cell carcinoma of the conjunctiva generally occurs in older individuals with a history of excessive sun exposure. The tumor typically is located in the interpalpebral region near the corneoscleral limbus. Squamous cell carcinoma of the conjunctiva tends to be superficially invasive. Deep intraocular invasion is uncommon.6-8 Mucoepidermoid carcinoma, a variant of squamous cell carcinoma, is more locally aggressive. In a review of 12 cases of conjunctival mucoepidermoid carcinoma from the English-language literature, 6 cases were found to have intraocular invasion and 4 cases had orbital involvement. No intraocular or orbital involvement was found in the remaining 2 patients.5 Intraocular invasion in conjunctival squamous cell carcinoma may simulate intraocular inflammation.8 Similarly, the intraocular invasion was heralded by marked intraocular inflammation in our patient.

The recommended treatment of conjunctival squamous cell carcinoma and mucoepidermoid carcinoma includes alcohol corneal epitheliectomy, wide surgical excision with tumor-free margins, and cryotherapy of the remaining uninvolved margins.9 Radiation treatment, either with B-irradiation or plaque radiotherapy, has been advocated to reduce the risk of recurrence in conjunctival mucoepidermoid carcinoma.5

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*Figure 1. Conjunctival biopsy specimen shows conjunctival squamous cell carcinoma with focal mucin production (arrow) (periodic acid–Schiff, original magnification ×50).*

*Figure 2. Anterior segment view shows the recurrent mucoepidermoid carcinoma at the nasal conjunctiva and limbus (arrows).*

*Figure 3. B-scan ultrasonogram shows a 9.5-mm-thick, acoustically hollow intraocular cyst (arrow).*
Our patient had 2 unusual features. In the reported cases of intraocular invasion by conjunctival mucoepidermoid carcinoma, the tumor generally involved the anterior uvea as a solid mass and there was no evidence of a fundus lesion. The occurrence of a neoplastic cyst in the supravuveal space that simulated a solid mass and there was no evidence of a fundus lesion. The occurrence of a neoplastic cyst in the supravuveal space that simulated a solid mass. Double arrows indicate retinal pigment epithelium; S, sclera.

Recurrent conjunctival mucoepidermoid carcinoma generally exhibits fewer mucus-secreting cells than the original tumor. The recurrent conjunctival and orbital tumors were composed entirely of squamous carcinoma cells in our patient. A prior report noted that the scleral defect from prior surgery predisposed the eye to the development of intraocular invasion through recurrent conjunctival squamous cell carcinoma. The scleral defect observed during our patient’s second conjunctival excision probably also played a role in the development of intraocular invasion. We believe that the defect was related to scleral necrosis from invasive tumor, as the margins were irregular and attenuated. Intraocular inflammatory signs in a patient with prior excision of conjunctival mucoepidermoid carcinoma should raise suspicion of intraocular invasion by the tumor.

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REFERENCES


A look at the past . . .

An hereditary cause for diseases or malformations of the eyes was found by Laqueur most frequently with retinitis pigmentosa and congenital cataract, and less frequently with albinism, total and partial iridocoria, microphthalmos, juvenile cataract, coloboma of the macula lutea, ectopia lentis, extreme myopia, and buphthalmus. Retinitis pigmentsa can almost always be traced back to consanguinity of the parents (a grossly exaggerated statement.—Herman Knapp). Congenital cataract is mostly due to direct transmission, and in albinism and congenital buphthalmus consanguinity plays the chief role. Congenital malformations of the iris are transmitted from generation to generation.