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 suprauveal 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.

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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 suprauveal 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 tenectomy 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 su-
prachoroidal space, forming a neo-
plastic 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 identi-
fied intraocularly or in the tumor on
the outer surface of the globe.

The patient developed mas-
vie orbital recurrence 1 year after
enucleation. An eyelid-splitting or-
bital exenteration was performed.
Histopathologically, the tumor ex-
hibited squamous cell carcinoma but
no mucus-producing cells were evi-
dent. The patient had no further re-
currence and died 2 years later of
cardiac disease.

COMMENT

Squamous cell carcinoma of the con-
junctiva generally occurs in older
individuals with a history of ex-
cessive sun exposure. The tumor
typically is located in the interpal-
pebral region near the corneo-
scleral limbus. Squamous cell car-
cinoma of the conjunctiva tends to
be superficially invasive. Deep in-
traocular invasion is uncom-
mon.6-8 Mucoepidermoid carcinoma, a vari-
ant of squamous cell carcinoma, is
more locally aggressive. In a review
of 12 cases of conjunctival muco-
epidermoid carcinoma from the En-
glish-language literature, 6 cases
were found to have intraocular in-
vansion and 4 cases had orbital in-
volve. No intraocular or or-
bital involvement was found in the
remaining 2 patients.5 Intraocular in-
vansion in conjunctival squamous cell
carcinoma may simulate intraocu-
lar inflammation.8 Similarly, the in-
traocular invasion was heralded by
marked intraocular inflammation in
our patient.

The recommended treatment of
conjunctival squamous cell carci-
noma and mucoepidermoid carci-
noma includes alcohol corneal epi-
thelektomy, wide surgical excision
with tumor-free margins, and cryo-
therapy of the remaining unin-
volved margins.9 Radiation treat-
ment, either with β-irradiation or
plaque radiotherapy, has been ad-
vocated to reduce the risk of recur-
rence in conjunctival mucoepider-
moid carcinoma,5

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 suprachoroidal space that simulated a solid mass and there was no evidence of a fundus lesion. 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... 

A 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 irideremia, microphthalmus, juvenile cataract, coloboma of the macula lutea, ectopia lentis, extreme myopia, and buphthalmus. Retinitis pigmentosa 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.