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Case Report

# Conjunctival Mucoepidermoid Carcinoma: a Case Report and Review of the Literature

**Nogba Pawoo DO[1], Neena Mirani[1,2] and Paul D Langer FACS[2]***

[1]*Department of Pathology and Laboratory Medicine, Rutgers New Jersey Medical School, Newark NJ, New Jersey*

[2]*Institute of Ophthalmology and Visual Science, Rutgers New Jersey Medical School, Newark, NJ, New Jersey*

***Corresponding Author:** Paul D Langer FACS, Institute of Ophthalmology and Visual Science, Rutgers New Jersey Medical School, Newark, NJ, New Jersey.

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## Abstract

Mucoepidermoid carcinoma is a malignant tumor composed of an admixture of squamous and variable proportions of mucus-secreting cells. Conjunctival mucoepidermoid carcinoma (CMEC) is a rare neoplasm of which, to our knowledge, there has only been 43 previously reported cases. It is a highly aggressive tumor with the tendency to recur locally. This neoplasm is anatomically, clinically and histopathologic ally closely related to conjunctival squamous cell carcinoma (CSCC) and represents diagnostic and therapeutic challenges for the ophthalmologist and pathologist. Here we report an additional case of CMEC in an anterior exenteration specimen.

***Keywords:*** *Conjunctival Mucoepidermoid; Carcinoma*

## Case Report

A 58-year-old Haitian man had undergone two previous penetrating keratoplasties on the right side, the most recent one 15 years earlier, for complete corneal vascularization of unknown etiology. After both transplants, he had gradual recurrence of corneal vascularization. The patient was eventually referred to another corneal surgeon who performed a biopsy of the cornea and conjunctiva which was interpreted as squamous cell carcinoma. The patient was placed on topical 1% 5-fluorouracil drops 4 times daily, 5 days per month per cycle, for 6 months (6 cycles) total duration. 6 months after the completion of this treatment cycle, he had clinically persistent disease and was placed back on the topical 5-fluorouracil. He was then lost to follow up for nearly three years. He subsequently returned to the 2nd corneal surgeon with apparent massive recurrence and was referred for management.

On presentation, vision was light perception vision in the right eye and 20/25 in the left eye. The right upper eyelid was completely ptotic and grossly thickened. The right bulbar conjunctiva was densely thickened, markedly injected with papilliform growths laterally, and contained multiple excrescent nodules (Figure 1A). The cornea was covered with vascularized tissue precluding a view into the anterior chamber. The patient underwent an eyelid-sparing anterior exenteration without complication.

The right anterior exenteration specimen (Figures 1B, 1C, and 1D) revealed a 1 cm raised, grey-white, fleshy firm mass sitting on the medial bulbar conjunctiva and a portion of the cornea. In addition, there were nodular growths on the lateral bulbar and lower palpebral conjunctiva (Figures 1B, 1C). The lacrimal gland and intraocular structures were unremarkable, and a prosthetic intraocular lens was in place.

Microscopic examination of the nodular mass on the medial bulbar conjunctiva reveals predominantly moderately differentiated squamous cell carcinoma with keratinization and a few foci of intracytoplasmic vacuoles (Figure 2A), confirmed to be mucin by mucicarmine

stain, PAS and Alcian blue stains (Figures 2B, 2C). The squamous component of the tumor reveals immunoreactivity with P63 and CK 5/6. The mucinous component of the tumor is immunoreactive to CK7 (Figure 2D), CEA and EMA. The tumor is negative for CK20 and P16. These findings support the diagnosis of mucoepidermoid carcinoma. The lateral bulbar conjunctival growth is predominantly papillary with focal glandular and squamous differentiation. Similar morphology is evident in the previous incompletely excised conjunctival lesion in 2009.

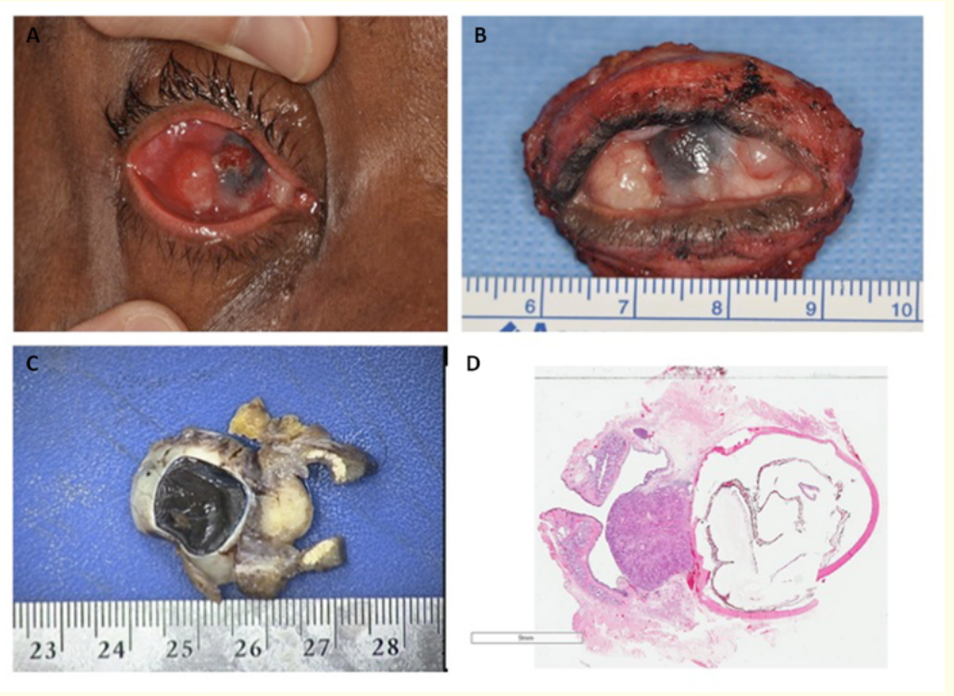


***Figure 1:*** *(A) Right eye with thick nodular growths on the lateral and medial bulbar conjunctiva; (B) Right anterior exenteration specimen (hematoxylin and eosin 100X); C) Firm tumor involving the cornea and conjunctiva with sparing of the intraocular structures; D) Whole mount of tumor.*

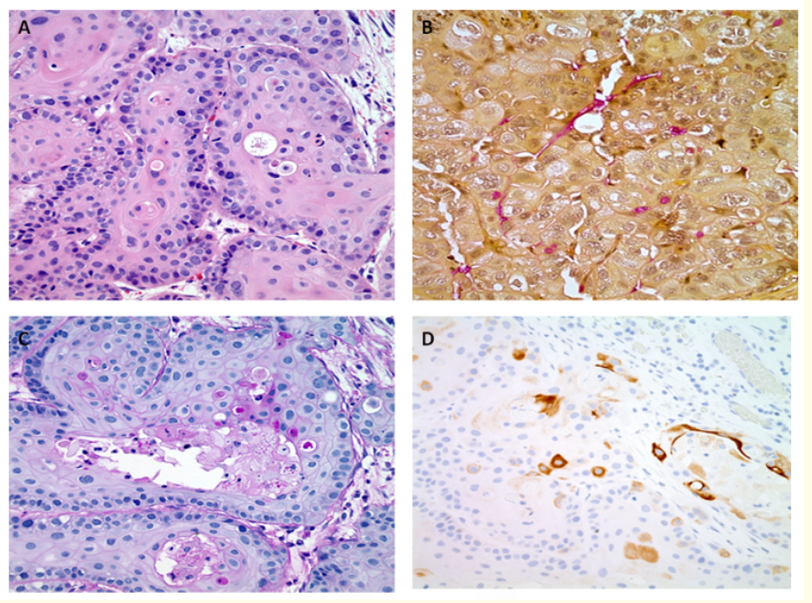


***Figure 2:*** *(A) Squamous cell carcinoma with intracytoplasmic vacuoles (hematoxylin and eosin 200X); B) Mucicarmine stain (200X); C) PAS stain (200X); D) Cytokeratin 7 staining glandular epithelial cells (200X).*

## Discussion

Mucoepidermoid carcinoma is a malignant tumor composed of an admixture of squamous cells and a varying amount of mucous secreting cells. It is a rare conjunctival malignancy and accounts for 0.3% of premalignant and malignant squamous lesions in the conjunctiva [1]. Most of the reported cases are single case reports with a few small series [2-21]. The first five cases of CMEC were reported in 1976 by Rao and Font; they were noted clinically to be more aggressive than conventional SCC with the propensity for recurrence and intraocular invasion [8]. Recently Moloney et. al. reported a case of CMEC and found 42 previously reported cases in the English literature, however, they postulate that this number is likely an underestimate due to both clinical and histopathological misdiagnosis [17]. CMEC has a male predominance and is typically diagnosed between the third and eighth decade of life. CMEC often presents as a pearly white, nodular, ulcerated lesion or as a presumably inflamed pterygium. Clinically, CMEC may be indistinguishable from squamous cell carcinoma. In fact the most common presentation is a conjunctival mass with redness and irritation, usually first involving the limbal conjunctiva, then subsequently involving the bulbar and palpebral conjunctiva [8]. CMEC has been well documented to arise in patients with systemic immune and autoimmune diseases such as rheumatoid arthritis, HIV, ocular cicatricial pemphigoid and multiple sclerosis; in addition to persons with prior malignancy and surgery [2,6,9,10,18,19]. Moloney., *et al.* reported a case of CMEC in a patient with a pterygium for 10 years that increased in size, and Soong., *et al.* reported a case in an HIV-positive patient with a pterygium that became inflamed [10,17]. Early tumor recurrence, ulceration, scleritis, fixation of the lesion to the sclera, and corneoscleral thinning or perforation are all common features of CMEC [20].

CMEC is often a challenge to diagnose in a biopsy specimen and can be misdiagnosed as squamous cell carcinoma; several cases have been reported in which this misdiagnosis occurred. This common misinterpretation is due to the predominance of the epidermoid component, as the mucin-producing elements may not be expressed until there is recurrence or intraocular invasion [2,9,12,18-20]. Mucin stains may be useful when the histogenesis of a conjunctival epithelial tumor is in doubt. Other stains for confirmation of CMEC are Alcian blue, colloidal iron and PAS. The glandular component of the tumor is often immunoreactive to EMA, CEA, MUC19 and CK 7 [18,22,23].

CMEC demonstrates aggressive behavior with an extraordinary capacity for recurrence and invasion and demands more aggressive surgical management than the more prevalent SCC. The documented recurrence rate is 85% compared to 5% for CSCC [1,9]. Intraocular and/or orbital invasion are the norm, and cases of lymph node and liver metastasis have been reported [18]. Risk factors for local invasion include previous surgery, previous patch graft, and delay in diagnosis [20]. Conventional therapy used to treat SCC has been reported to be ineffective [2,5,8,9,19]. Due to the risk of recurrence and orbital invasion, at least wide local excision with frequent follow-up to detect early recurrence is recommended, however, disease control has been documented with cryotherapy and or keratoplasty. For tumor invading the globe or orbit, enucleation or exenteration is recommended [17,21].

Of note, in the salivary gland, mucoepidermoid carcinoma is graded as low, intermediate, or high grade based on a numerical scale that accounts for the presence of neural invasion, necrosis, mitosis, and anaplasia [22,23]. Whether this grading system is relevant in the conjunctiva remains elusive. MEC of the salivary gland also can possess at (11;19) (q21, p13) translocation causing fusion of CRTC-1 – MAML2 or CRTC-3, which has been reported to indicate a better prognosis [22,23]. To our knowledge this genetic anomaly has yet to be documented in CMEC.

In summary, CMEC is a rare neoplasm with poor prognosis compared to conjunctival squamous cell carcinoma and should be treated aggressively. It represents diagnostic and therapeutic challenges for the ophthalmologist and pathologist. Delay in treatment can be avoided by early clinical diagnosis with symptoms such as rapid growth with inflammatory signs. For pathologists, any rapidly growing conjunctival mass with squamous differentiation, or a recurrent conjunctival tumor with mucin production, should yield a high index of suspicion for CMEC; extensive sampling of the tumor and the use of specific glandular epithelial immunohistochemical stains are often vital in reaching the correct diagnosis.

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