

Mucoepidermoid carcinoma of the conjunctiva with lung metastasis

*Pukhraj Rishi, Rashi Sharma¹,
Krishnakumar Subramanian², Nirmala Subramaniam¹*

A 36-year-old lady presented with redness and decreased vision in right eye since 6 months. She was earlier diagnosed of cavitary lung lesion, presumed secondary to tuberculosis and treated with anti-tubercular treatment for 4 months. Examination of affected right eye revealed nil light perception, conjunctival congestion with an exuberant mass in the inferotemporal bulbar conjunctiva, proptosis, iris neovascularization, 360° closed angles, intraocular pressure of 48 mm Hg, exudative retinal detachment, uveal mass and orbital extension. A diagnostic needle biopsy of uveal mass revealed malignant cells. Computed tomography-guided lung biopsy revealed squamous cell carcinoma (SCC), indicating metastatic spread from the orbit. She underwent lid-sparing exenteration of the right eye. Histopathological examination of the orbital tissue revealed mucoepidermoid carcinoma arising from the conjunctiva with extensive invasion into the orbital tissue, muscle fibers, sclera, choroid and optic nerve. Multiple tumor emboli were seen in the lumen of orbital blood vessels. In conclusion, mucoepidermoid carcinoma of the conjunctiva is a rare, aggressive variant of SCC. Early intervention is essential to prevent intraocular invasion and systemic metastasis.

Key words: Biopsy, conjunctiva, lung, metastasis, mucoepidermoid carcinoma, tumor

Mucoepidermoid carcinoma predominantly originates from salivary glands. Ocular tissues affected with this neoplasm include conjunctiva, lacrimal sac, and lacrimal gland.^[1] The aggressive nature of the tumor and its tendency for intraocular

invasion are well-known.^[2] Though distant metastasis to preauricular/cervical lymph nodes and liver has been reported, the incidence is quite low.^[3] However, there have been no reports of mucoepidermoid carcinoma metastatic to the lung. Hereby, we report a case of a 36-year-old lady with mucoepidermoid carcinoma of the conjunctiva (MECC) presenting with intraocular invasion, orbital extension and metastasis to the lung.

Case Report

A 36-year-old lady presented with redness and decreased vision in right eye for 6 months. She was earlier diagnosed of a cavitary lung lesion, presumed secondary to tuberculosis and treated with anti-tubercular treatment for 4 months. At presentation, vision was nil light perception in the affected right eye. The left eye was essentially normal. Examination revealed conjunctival congestion with a diffuse vascularized conjunctival mass in the inferotemporal bulbar conjunctiva and proptosis [Fig. 1a]. Iris neovascularization, 360° closed angles with intraocular pressure of 48 mm Hg, and a choroidal mass with total retinal detachment were noted. Ultrasound of right eye revealed diffuse choroidal thickening with uveal mass, widening of the tenon's space and diffuse orbital tumor [Fig. 1b]. Magnetic resonance imaging scan revealed uveal mass and diffuse orbital tumor [Fig. 1c]. A diagnostic needle biopsy of the uveal mass revealed the presence of atypical cells in necrotic background suggestive of malignancy. The patient underwent lid-sparing exenteration of the right eye [Fig. 1d]. Histopathological examination of the exenterated specimen revealed the presence of mucoepidermoid carcinoma with multiple tumor emboli within the blood vessels of the orbital tissue [Fig. 1e and f]. Extensive invasion into the orbital tissue, muscle fibers, sclera, choroid and optic nerve was noted. Postoperative period was uneventful. Before orbital exenteration, she also underwent computed tomography-guided biopsy of the cavitary lesion in the left lung that revealed squamous cell carcinoma (SCC) [Fig. 2a and b]. In view of the systemic spread, radiotherapy was advised. At 3 months follow-up, two cycles of radiotherapy were completed, and the right socket was healthy.

Mucoepidermoid carcinoma of the conjunctiva often presents as a limbal or bulbar conjunctival mass with symptoms of chronic irritation. It was reported for the first time in conjunctiva by Rao and Font in a series of five cases in 1976, all the five cases had recurrence within 6 months.^[4] Clinical morphology can range from simple conjunctival nodule to leukoplakic, infiltrative, ulcerative, and papillomatous lesions. Histopathology of MECC is variable. Our case revealed tumor cells arranged in lobules with fine septa separating them. The cells had squamoid morphology with duct-like structures and were seen infiltrating sclera and orbital tissue. Tumor emboli were seen in blood vessels. The tissue was positive for alcian blue staining [Fig. 3].

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Ocular Oncology Service, Departments of ¹Oculoplasty and ²Larsen and Toubro Ocular Pathology, Sankara Nethralya, Chennai, Tamil Nadu, India

Correspondence to: Dr. Pukhraj Rishi, Ocular Oncology Service, Sankara Nethralya, 18 College Road, Chennai - 600 006, Tamil Nadu, India. E-mail: docrishi@yahoo.co.in

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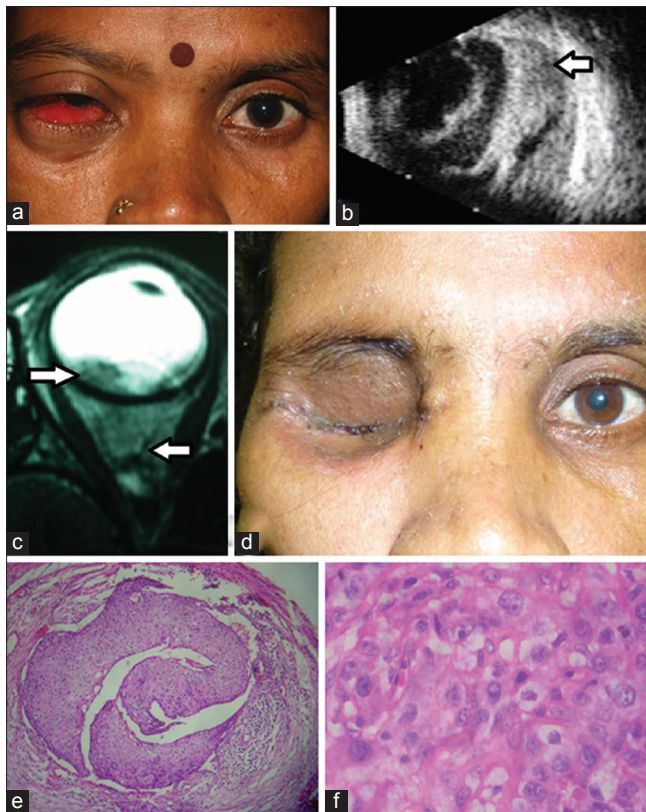


Figure 1: (a) At presentation, external photo reveals conjunctival congestion, fullness of right lower lid and nonaxial proptosis. (b) Ultrasound B-scan shows diffuse choroidal thickening, widening of tenon's space and diffuse orbital mass (left arrow). (c) Magnetic resonance imaging of right orbit shows intraocular mass lesion (right arrow), thickened posterior ocular coats, irregular and enhancing soft tissue around and temporal to optic nerve head (left arrow). (d) External photo 1-week after lid-sparing exenteration of right eye. (e) Photomicrograph (H and E, $\times 40$) shows tumor cells with squamoid features with multiple duct-like structures containing mucin. Tumor emboli are seen within blood vessel. (f) Photomicrograph (H and E, $\times 100$) shows tumor cells with clear spaces that contain mucin

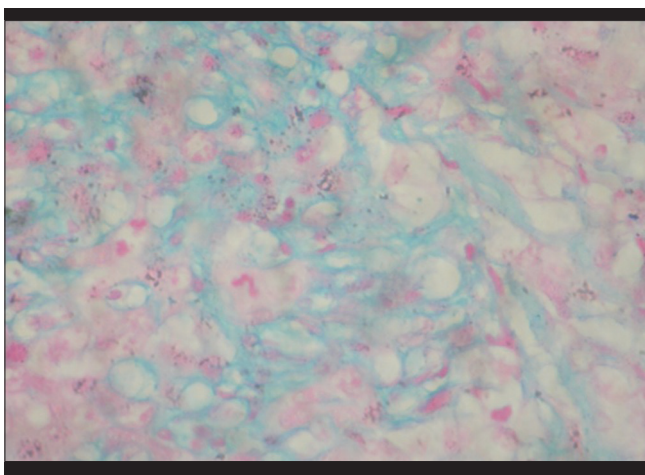


Figure 3: Photomicrograph of orbital biopsy tissue (alcian blue, $40\times$) with positive staining for acid mucopolysaccharide

Review of literature suggests the role of intraocular invasion by tumor cells to be implicated in the mucinous

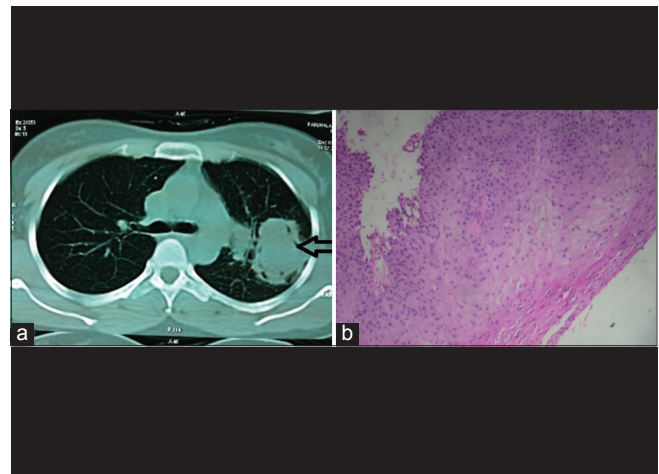


Figure 2: (a) Magnetic resonance imaging lung shows cavitary lesion in left upper lobe (arrow) with a large necrotic center and nodular enhancing soft tissue toward the hilum with thin, uniform wall. (b) Photomicrograph of lung biopsy tissue (H and E, $40\times$) shows squamous metaplasia with nonspecific chronic inflammation

differentiation in the tumor, and perhaps mucinous metaplasia is due to intraocular factors.^[2,5,6] Lymph node involvement has been described, but distant metastasis is unusual if the tumor is treated aggressively.^[7] Seitz and Henke, suggested that early enucleation including normal appearing tissue next to the globe should be considered for treatment of primary mucoepidermoid carcinoma to avoid later exenteration or metastasis.^[8] Johnson *et al.* and associates reported two cases of MECC presenting with cervical lymph node metastases, in a series of 30 invasive secondary orbital SCC that were treated with exenteration and radical neck dissection.^[9] Recurrence is also a common feature of MCC occurring to the extent of 79%, most commonly in the first 6 months of follow-up.^[10]

In our patient, extensive orbital disease at the time of diagnosis necessitated exenteration and vascular invasion was seen histopathologically. There was a large cavitary lesion in the left upper lobe of lung with central necrotic area and nodular enhancing soft tissue toward the hilum with a fairly thin, uniform wall. Systemic metastasis was treated with radiation therapy. The patient was recovering well. Given the high frequency of misdiagnosis and incorrect management, it is essential that proper histopathological examination be carried out.^[10] The treatment strategy is decided once the status of intraocular spread as well as distant metastasis is known.

Conclusion

Mucoepidermoid carcinoma of the conjunctiva is rare, aggressive variant of SCC, characterized histopathologically by both mucoid and epidermoid cells, which requires urgent intervention to prevent metastasis. Increased awareness of subtle clinical and pathologic findings and rigorous sequential treatment strategies should help clinicians and surgeons in the treatment of MECC for improved outcomes and better prognosis.

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