Concurrent Leiomyosarcoma and Basal cell carcinoma of the Conjunctiva: A Case report

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Citation

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Abstract

The most common conjunctival mass causing corneal blindness in countries with an arid dry hot climate is a pterygium. Lesions, which may clinically resemble a pterygium include benign and malignant tumors like choristomas, squamous cell and basal cell carcinomas.1,2,3,4 Primary basal cell carcinomas of mucosal surfaces, particularly the conjunctiva, are very rare with only 4 reported cases in the last 20 years.1,2,3,4

Four out of every million people develop leiomyosarcoma, which can affect the lungs, liver, blood vessels, or other soft tissues in the body. A benign leiomyoma of the conjunctiva has been identified once, however, occurrence of leiomyosarcoma of the conjunctiva in conjunction with a basal cell carcinoma has to our knowledge not been observed or reported so far. 4, 5, 6

CASE REPORT

A 74 year old man presented with a history of a slow-growing painless mass in his left eye of 4 months duration. His right eye had been amblyopic. He gave no history of trauma or systemic illness.

On examination, the best corrected visual acuity in his right eye was OD -2.50/-1.25/170° = 0.1, and hand movements in his left eye. External ocular examination revealed a fleshy mass about 7.0x5.0x2.5mm attached to the nasal conjunctiva by a broad-based peduncle. The mass extended across the limbus, overhanging on the adjacent cornea and covering the pupillary area. The surface of the mass was nodular and showed dilated blood vessels. Appositional closure of the lids in the left eye was defective due to the mass. The temporal conjunctiva and adjacent cornea appeared desiccated. Plica semilunaris and caruncle were normal. There was a nuclear cataract in his right eye and trachomatous scarring of the conjunctiva and cornea in both the eyes, worse in the left eye than in the right eye. Intraocular pressure in the right eye was 12 mmHg and could not be assessed in the left eye due to the mass. Extraocular muscle movements were normal. Fundus examination in the right eye was normal. The fundus of the left eye could not be evaluated due to the corneal scarring and the overhanging mass. A B scan ultrasound examination of both eyes revealed normal posterior segments. General examination and systemic examination was unremarkable. There was no localized or generalized lymphadenopathy.

Laboratory investigations revealed a normal complete blood count. Serology was negative for retrovirus, cytomegalovirus, and herpes simplex virus. CD4 cell count was 0.9×10^{9} /l (normal =$0.5\text{-}1.3\times10^{9}$ /l)and CD 8 cell counts (0.6×10^{9} /l) (normal=$0.3\text{-}1.0\times10^{9}$ /l) Liver function and renal function tests were normal. Stool tested negative for occult blood. Ultrasound abdomen and X-ray chest were normal. Mantoux test was reported negative. Computed cranial tomography showed no intraocular or orbital or intracranial extension of the growth.

The tumor was excised with a lamellar keratectomy on the corneal and a lamellar sclerectomy on the scleral side with a margin of 1 mm on the corneal and 2 mm on the scleral sides, and application of cryotherapy (- 80? C for 60 sec) to the tumor bed.

Histopathological evaluation revealed epithelial parakeratosis, basal cell proliferation and nuclear atypia, loss of polarity and occasional mitoses involving the basal layers of the conjunctiva indicating solar elastosis. There was a basophilic degeneration of the subepithelial collagen. The epithelium was thinned with downward extensions of basaloid cells in strands with peripheral palisading of nuclei, interspersed with horn cysts. The stroma showed a mucoid degeneration suggesting a basal cell carcinoma. Trichrome stain revealed an unencapsulated nodular lesion with

indistinct margins composed of spindle cells with oval nuclei, and typical cytoplasmic fibrils without necroses. Nuclear atypias, giant cells and 4-6/10 mitoses/hpf were noted. (Fig 1,2&3).These cells showed a strong expression of smooth muscle actin and were negative for melanocytic and epithelial markers (S-100, HMB 45, cytokeratins). Immunostaining was negative for human papillomavirus or Epstein-Barr virus. The tumor cells tested negative for human papillomavirus DNA on in situ hybridization. Alterations of bcl -2, c-erb -b2 and Rb oncoproteins were not found immunohistochemically. Overexpression of p53 was detected by immunohistochemistry in both tumors, but p53 gene mutations were not found by polymerase chain reaction. Local recurrence was noted after 3 years.

Figure 1

Figure 1: HE stain (magnification ×100) shows epithelial proliferations composed of basaloid cells with keratin cysts. Small clusters of epithelium are seen to invade underlying tissue with evidence of connective tissue tumor inferiorly

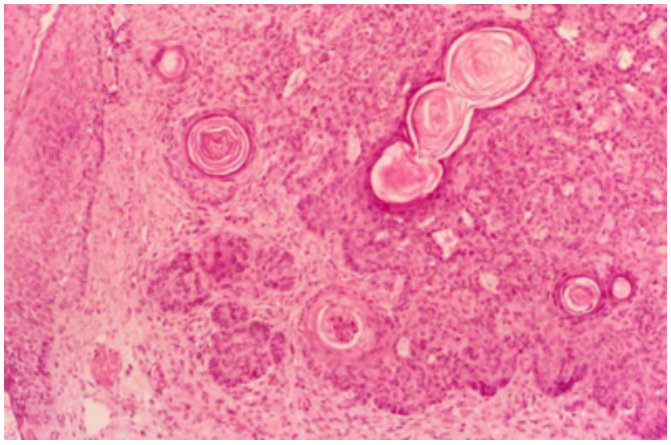


Figure 2

Figure 2: HE stain (magnification:×200): Details of the connective tissue tumor showing pleomorphism and normal as well as abnormal mitoses.

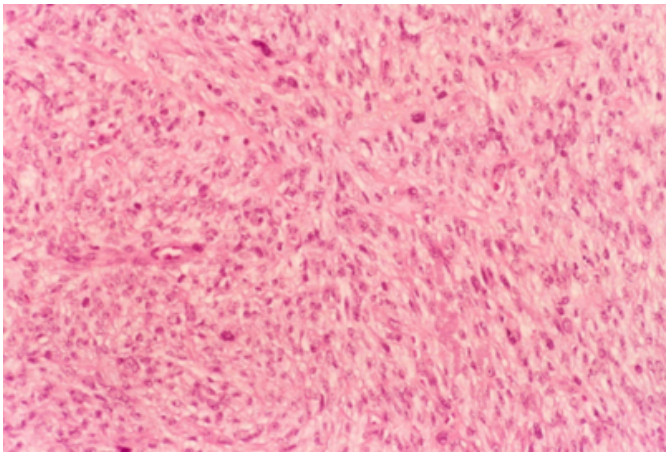
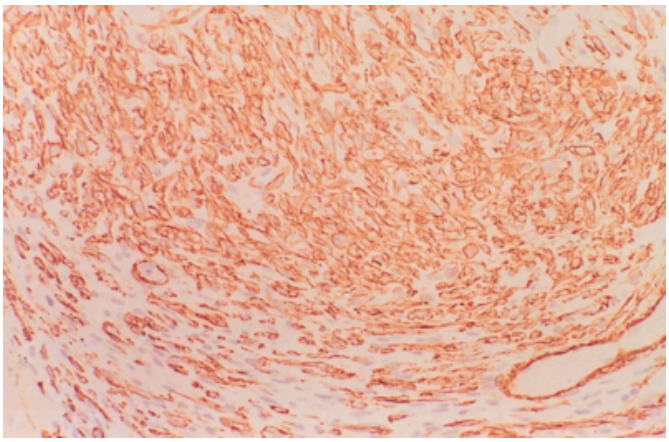


Figure 3

Figure 3: (magnification:×200) Trichrome stain of the connective tissue tumour showing spindle shaped cells with oval nuclei, and typical cytoplasmic fibrils without necroses, nuclear atypias, and giant cells .



DISCUSSION

In this report, we describe an elderly patient suffering from a slow-growing conjunctival mass composed of a low-grade leiomyosarcoma and a basal cell carcinoma, which extended on the cornea.

Basal cell carcinomas originating from the basal layer of the nasal limbal conjunctival epithelium have been reported earlier [1,2,3,4,5] .

Ash and Wilder observed one case of basal cell carcinoma initially in a series of 93 corneoscleral epithelial tumors. Later Ash reported 53 cases of basal cell carcinoma in a series of 1120 patients with epibulbar tumors. [4,5,6] However,

it is not clear if these lesions were extensions from adjacent adnexal structures or developed primarily from the conjunctiva. Aftab and Percival, Apte et al and later Hussein et al reported the occurrence of a primary basal cell carcinoma as fleshy growth on the nasal interpalpebral conjunctiva. None of the lesions were invasive. Complete excision of the lesions was performed. None recurred. [4,5,6]

Cable et al reported on a morpheaform type of basal cell carcinoma originating from the basal layer of the nasal limbal conjunctival epithelium with intraocular extension and secondary glaucoma. [7] Our patient had no evidence of intraocular, intraorbital or intracranial extension.

The most important risk factor for basal cell carcinomas is UV-B irradiation being abundant in the desert country of Oman and may have been involved in our patient, since histopathology features of solar elastosis are evident. 30-50% of basal cell carcinomas have been found to express UV specific mutations in p53 tumor suppressor genes. [6,7,8,9] In our patient the tumor tested negative for p53 mutations, but p53 appeared overexpressed. A possible facilitation of tumor development has been postulated involving an infection with human papillomavirus 16 and 18 and immunosuppression. [8] However, no human papillomavirus infection was detected in our patient, who was HIV-seronegative as well.

In summary primary basal cell carcinomas of the conjunctiva are rare. The lesions tend to occur in older individuals, in the actinically exposed areas of the interpalpebral conjunctiva. The lesion can be locally invasive and destructive but usually does not metastasize. Local recurrence may occur as in our patient . [4,5,6,7]

Leiomyomas are mesenchymal tumors of smooth muscle origin. A conjunctival leiomyoma, but not a leiomyosarcoma, has been reported in a 35 year old man presenting with a slow-growing episcleral mass similar to our patient, however, without extending on the cornea. [10] Simultaneous occurrence of a squamous cell carcinoma and a leiomyosarcoma of the oral cavity has also been reported before. [11] The lack of smooth muscles in the conjunctiva raises the possibility of its origin from either the pluripotent germ cells of the epithelium or the smooth muscle layer of the conjunctival blood vessels. [10,11] . No distant metastasis was noted in our patient.

Three rare features are presented in this report – a primary basal cell carcinoma of the conjunctiva (4 cases reported in the last 20 years), a leiomyosarcoma of the conjunctiva (no case reported so far to our knowledge) and the simultaneous occurrence of both.(one case reported so far to our knowledge). The need for curative removal of epithelial tumors and an exact histopathological work-up of conjunctival masses is highlighted in order to identify a malignant potential and to counsel the patient accordingly. [1,11]

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