

An Uncommon Case of Noninvasive Ocular Surface Squamous Neoplasia Involving the Entire Cornea

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ABSTRACT

Ocular surface squamous neoplasia (OSSN) is relatively rare with an incidence of 0.13–1.9/100,000 population. OSSN includes dysplastic lesions involving the squamous epithelium of the conjunctiva or cornea. Epibulbar squamous cell carcinoma and epithelioma have been noted commonly, but cases in which the tumor is primary on the cornea are sufficiently rare to warrant reporting in each instance. We describe a rare case of noninvasive OSSN involving the entire cornea in a human immunodeficiency virus-negative patient. The patient was successfully treated with no recurrence, after intact surgical removal, mitomycin C treatment, and cryotherapy.

Keywords: Noninvasive ocular surface squamous neoplasia, ocular surface squamous neoplasia involving entire cornea, squamous cell carcinoma cornea

INTRODUCTION

Ocular surface squamous neoplasia (OSSN) is uncommon with an incidence of 0.13–1.9/100,000 population, and it primarily occurs in older males.^[1,2] OSSN includes dysplastic lesions involving the squamous epithelium of the conjunctiva or cornea, which includes squamous papilloma, conjunctival-cornea intraepithelial neoplasia, carcinoma *in situ* (CIS), and invasive squamous cell carcinoma (SCC). Epibulbar SCC and epithelioma have been noted commonly, but cases in which the tumor is primary on the cornea are sufficiently rare to warrant reporting in each instance. We describe a rare case of noninvasive OSSN involving the entire cornea

in a human immunodeficiency virus (HIV)-negative patient. The patient was successfully treated with no recurrence, after intact surgical removal, mitomycin C (MMC) treatment, and cryotherapy.

CASE REPORT

A 38-year-old female patient, a farmer by occupation, presented to the OPD with complaints of foreign body sensation, redness, and diminution of vision in the right eye since 1 year. She also noticed a mass in the right eye, which gradually increased in size [Figure 1]. There was no history of trauma or foreign body in the right eye and no history of any ocular surgery in the past.

There was no history of weight loss. Systemic examination revealed no abnormality. Ocular examination revealed a 12 mm × 11 mm spherical mass all over the cornea. The mass was grayish, white, and

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granular. It appeared to be adherent to the underlying cornea. Almost entire cornea was covered, only a rim of the cornea was visible nasally. The mass was limited only to the cornea. The details of the anterior chamber, iris, pupil, and lens could not be visualized. The visual acuity of the right eye was the only perception of light and projection of rays in all the quadrants. Intraocular pressure was digitally normal. Ocular movements were full and free in all directions of gaze. The left eye was normal in all aspects. Preauricular and submandibular lymph nodes were not palpable. The patient was not positive for HIV. Clinical diagnosis of OSSN of the cornea was made. The patient was posted for a surgical excision of the mass.

A disposable plastic drape was sutured around the limbus over the surrounding conjunctiva so that only the cornea was exposed. This was done to prevent seedling. Whole corneal mass was dissected out with lamellar dissection of superficial cornea using a crescent blade. The mass was excised in toto, along the surgical plane, so the extent was well demarcated. The excised mass was sent for histopathological examination. The histopathological report confirmed the existence of SCC of the cornea [Figure 2]. Double freeze-thaw cryotherapy was applied with conical probe of 1 mm, to cover the entire peripheral cornea. Postoperative treatment included topical application of steroid-antibiotic combination and atropine 1% eye drop 3 times a day. The eye was patched for a week till complete epithelialization of the cornea took place. Punctal occlusion was achieved with punctal plugs before starting mitomycin therapy. MMC eye drop (0.02%) was advised 4 times a day after the completion of epithelialization, with 1 week on and 1 week off, in alternating cycles for 8 weeks. The best corrected visual acuity recovered to 6/18. The patient did not show any signs of recurrence during the follow-up period of 1 year [Figure 3].

DISCUSSION

OSSN is uncommon and it primarily occurs in older males (78.5%). The average age of occurrence has been noted to be 60 years, ranging from 20 to 88 years.^[3] A number of risk factors, including genetic predisposition, fair skin, pale irises, high propensity to sunburn, a past history of skin cancer, immunosuppression, human papillomavirus (HPV) infection, chronic infection by HPV, HIV, or trachoma, vitamin A deficiency, xeroderma pigmentosum, and smoking, have been associated with the appearance of OSSN. It is estimated that the risk of SCC increases 13-fold in patients with HIV.^[4] The disease behaves in a much more aggressive manner in patients with AIDS. Epibulbar SCC and epithelioma have been noted commonly, but cases in which the tumor is primary on

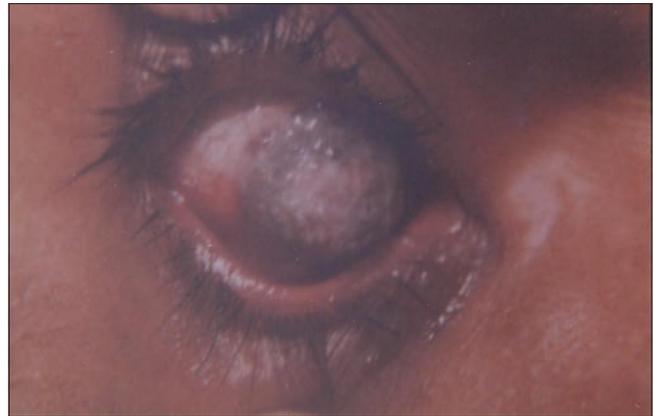


Figure 1: Preoperative clinical photograph of the entire corneal involvement of squamous cell carcinoma

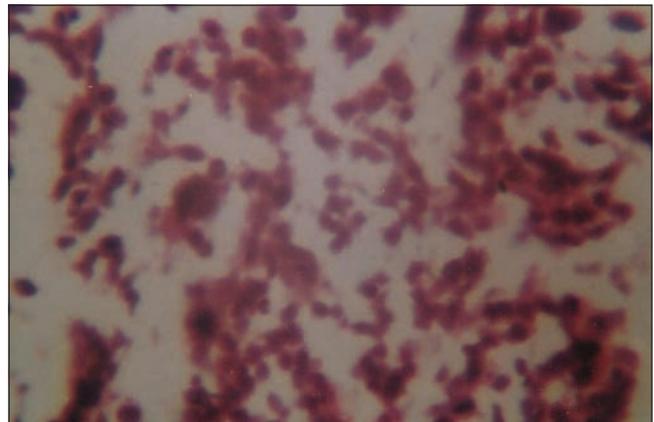


Figure 2: Histopathological slide confirming the diagnosis of squamous cell carcinoma

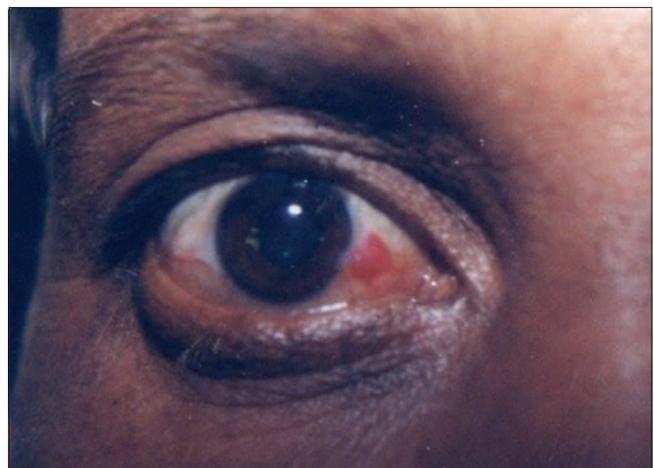


Figure 3: Postoperative clinical photograph of the eye free from squamous cell carcinoma

the cornea are sufficiently rare to warrant reporting in each instance.

OSSN is often diagnosed in the early stages due to the easily visible location. OSSN involves the conjunctiva

or the cornea individually but more commonly involves both. The clinical presentation of these lesions varies across a wide spectrum and can range from mild to severe dysplasia to full-thickness epithelial dysplasia (CIS) and invasive SCC. OSSN typically presents as a growth on the ocular surface and gives rise to symptoms such as foreign body sensation, redness or irritation and rarely, diminution of vision due to high astigmatism or involvement of visual axis. Over 95% of OSSN cases originate in the limbus, often in the interpalpebral region.^[5,6] The reason for the propensity of the lesions to this region is thought to be due to a combination of factors, including the presence of transitional type epithelium, high ultraviolet exposure, and a high mitotic rate.^[5]

Masses are initially mobile; the conjunctiva in later stages becoming fixed to the globe with deeper scleral infiltration. Through close, careful examination with slit lamp biomicroscopy, OSSN lesions can frequently be distinguished from other conjunctival lesions, such as pterygia and conjunctival lymphoma. The gold standard for the diagnosis of OSSN is the histopathological evaluation of the lesion after an incisional or excisional biopsy.

It is not unusual for invasive SCC to invade locally into the sclera, intraocularly, or into the orbit, with one study, estimating incidence rates to be 37%, 13%, and 11%, respectively.^[7]

The line of management mainly depends on the type of lesion and histopathologic findings, and can range from topical chemotherapy or excision alone for smaller lesions to a combination of surgical excision, cryotherapy, and chemotherapy for larger or invasive lesions. Rarely, radiotherapy, and in extreme cases, enucleation and even exenteration, may be required.^[5,7,8]

Dissection of all abnormal tissue with a wide surgical conjunctival margin of 3 mm is usually sufficient. Intraoperative cryotherapy is commonly used as adjunctive therapy as it is known to decrease the recurrence rate by the destruction of any residual tumor tissue beyond the horizontal or deep surgical margin of the wound. If the lesion extends onto the cornea, absolute alcohol can be used to remove the involved corneal epithelium. Copious irrigation needs to be performed following the application of absolute alcohol. After removal of larger tumors, conjunctival autografts or amniotic membrane grafts can be used to help close the conjunctival defect.^[5,8]

Chemotherapy has become the mainstay of adjunctive treatment with surgical excision and has shown to be

very effective. This is typically accomplished through topical eye drops of MMC, 5-fluorouracil, or interferon alpha 2B. MMC appears to produce cell death in OSSN by apoptosis and necrosis.^[9] MMC is used in the concentration of 0.02–0.04% 4 times a day, with 1-week on and 1-week off, in alternating cycles for a maximum of 8 weeks. The 1-week on, 1-week off regimen prevents damage to more slowly dividing epithelial cells and limbal stem cells, allowing them to repair their DNA. Allowing time for complete epithelial healing before application of MMC is important in avoiding the most serious complications such as corneal epitheliopathy, scleral ulceration, uveitis, cataract, and glaucoma. Side effects from topical chemotherapy include dry eye, superficial punctate epitheliopathy, punctal stenosis, and rarely stem cell deficiency. Punctal occlusion with punctal plugs is recommended before starting these regimens to prevent punctal stenosis.^[5,7,8]

In this case, whole corneal mass was dissected out with lamellar dissection of superficial cornea using a crescent blade. Double freeze-thaw cryotherapy was applied with a conical probe of 1 mm, to cover the entire peripheral cornea. MMC eye drop (0.02%) was advised 4 times a day after the completion of epithelialization, with 1 week on and 1 week off, in alternating cycles for 8 weeks. The peculiar points of this case are, the patient was HIV negative; the whole cornea was involved which could be removed intact successfully.

Squamous lesions of the cornea and conjunctiva are uncommon, but early diagnosis and initiation of treatment are key in prevention of visual loss and systemic morbidity and mortality. Our patient recovered well with no recurrence because of prompt treatment with complete excision followed by chemotherapy.

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Conflicts of interest

There are no conflicts of interest.

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