

OCULAR SURFACE SQUAMOUS NEOPLASIA (OSSN) IN A 17-YEAR-OLD – A CASE REPORT

Case Report

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A young female of 17 years presented with progressive growth in her left eye with loss of vision. She had previously received topical medication with no effect. She was on anti-retroviral therapy for seven years. She had a fungating squamous cell carcinoma mass covering the entire ocular surface for which total orbital exenteration was done.

Case Scenario

A 17-year-old female from rural district in Eastern province of Zambia presented to the University Teaching hospitals Eye Hospital complaining of a progressive growth on her left eye for six months. It was associated with white eye discharge. The growth was of spontaneous onset, covering whole eye such that she was unable to see with her left eye. She had received treatment with topical medication at local clinic with no improvement. She had no history of ocular surgery or any other growths on the eye or the body. Significant medical history was that she had been on Anti-retroviral therapy for the past nine years for vertically transmitted Human Immunodeficiency Virus (HIV). There was a positive family history of non-healing leg ulcer in her mother who had passed on some years back. She was an orphan living with her aunt.

On examination, she appeared appropriate for age and of fair nutritional status. She had no pallor, no jaundice but had a small 1x1 cm mobile, non-tender left submandibular

lymphadenopathy. General systemic examinations were otherwise normal. Visual acuity was 6/6 for the right eye and no light perception for the left eye. Her right eye had normal anterior and posterior segment findings. Left eye findings included restriction of extra ocular movements in all planes of gaze. She had a huge fungating mass filling entire palpebral fissure and covering bulbar to forniceal conjunctiva. Ocular structures were not discernible.

A clinical diagnosis of left eye squamous

cell carcinoma was made. The patient was admitted and counselled on the diagnosis and prognosis as well as need for orbital exenteration. Full blood count, renal function and liver function test parameters were all within normal range.

She underwent left eye total orbital exenteration and tissue was sent for histopathology confirmation which confirmed the diagnosis of squamous cell carcinoma (SCC) of the left eye.



Figure 1: Clinical appearance of the patient's left eye with tumour covering entire ocular surface.

Discussion

Ocular surface squamous neoplasia (OSSN) is a term that encompasses a spectrum of precancerous and cancerous lesions of the conjunctiva ranging from conjunctival intraepithelial neoplasia (CIN) to frankly invasive squamous cell carcinoma (SCC) with destruction of the orbit and intracranial invasion (1). Classically, OSSN has been associated with light skinned elderly males living in regions with excessive sunlight (ultraviolet) exposure. Other identified risk factors include immunosuppression, xeroderma pigmentosa, and infection with Human Papilloma Virus (HPV) types 16 and 18 and with Human Immunodeficiency Virus (HIV) (2).

Current recommendation is to test for HIV in all patients presenting with OSSN especially those younger than fifty years. A recent review of studies showed high incidence of HIV in patients presenting with OSSN and also a high relative risk of OSSN in patients with HIV (1). HIV has also been associated with more aggressive OSSN lesions. Mean age of patients with both OSSN and HIV has been found to be lower than with OSSN alone. However, still the mean age ranges in various studies for occurrence of OSSN in HIV patients have been in the 30's to 50's (3), (4). There is no documented evidence of occurrence of OSSN in patients younger than 20 years

of age. At the UTH Eye a situation was encountered where a patient aged 17 years was diagnosed with SCC and this was confirmed histologically.

Patients with OSSN commonly present with ocular redness and irritation, in addition to the lesion on the ocular surface which may grow within weeks to years; commonly of several months (5). Visual acuity is only reduced if lesion grows over the cornea. Clinically, CIN, carcinoma in situ and invasive SCC may have similar appearance. Lesions commonly arise within the interpalpebral fissure, mostly at the limbus. OSSN may appear gelatinous, papilliform, leukoplakic, or nodular lesion (5). The lesion may or may not be pigmented. If the condition is neglected, the lesion may invade the intraocular structures and spread to systemic circulation resulting in metastasis (2). It may also extend to involve the orbital structures. This was the case with the patient under discussion, hence exenteration was done. The patient was young, sexually inactive and HIV positive which she got vertically. There was no history of trauma or eye infection prior to the onset of the eye growth.

Fluorescein, Rose Bengal or Lissamine green staining have been used to help clearly delineate the borders of the lesion. Exfoliative cytology may be used to

identify neoplastic cells, and ultrasound biomicroscopy to determine depth of invasion; though definitive diagnosis still remains histology (5).

Treatment of OSSN lesions includes surgical and medical options. Surgery includes excision biopsy using the no-touch technique with cryotherapy. Recurrence rates range from 33% to 50% (2). Medical options include topical chemotherapy with 0.02%- 0.04% mitomycin, or 1% 5-fluorouracil, or interferon-alpha2b. In cases of orbital extension of invasive SCC orbital exenteration is done with or without adjuvant radiotherapy (2). Topical antibiotics must never be used as the treatment option for conjunctival growths. This patient was initially given this treatment option which was very wrong. This calls for sensitisation of health workers and the community so that such lesions are not taken for granted at all.

This case highlights a very young age at occurrence of OSSN in a patient with HIV. She presented with an advanced OSSN lesion at 17 years old. She had a six months history of a growing lesion in her left eye which was of an aggressive nature and also a seeming delay in identifying the possibility of OSSN led to a loss of the eye in order to preserve life.

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