43

A 35-Year-Old Malawian Woman With a Painful Ocular Tumour

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Clinical Presentation

History

A 35-year-old woman from Malawi presents to the outpatient department of a local tertiary hospital. She was referred by an ophthalmic clinical officer from a district hospital for exenteration of the left eye because of an ocular tumour.

The first symptoms started 8 months prior when she noticed a whitish lesion growing on the conjunctiva of her left eye. She presented at a health centre and was prescribed non- specified eye drops. Yet, over the following months the lesion grew bigger and turned reddish. She went to a traditional healer who prescribed herbal eye drops, which did not help either. The lesion grew constantly bigger and she finally lost her eyesight in the affected eye. Pain also increased which made her present at her local district hospital.

The patient is known to be HIV-positive. She has been on antiretroviral treatment for the past 3 years. The CD4 count is unknown.

Clinical Findings

Localized swelling of the left eyeball and orbit, lid closure incomplete (Fig. 43.1). The visual acuity on the right side is 6/6, whereas the left eye has no light perception. Her left preauricular lymph nodes are swollen. She is afebrile and the rest of her physical examination is unremarkable.

Questions

1. What is the suspected diagnosis?
2. How would you manage the patient?

Discussion

An HIV-positive Malawian woman presents with a painful tumour of her left eye. It started as a whitish lesion on her conjunctiva several months ago. The lesion continued to grow and the affected eye eventually turned blind.

Answer to Question 1

What is the Suspected Diagnosis?

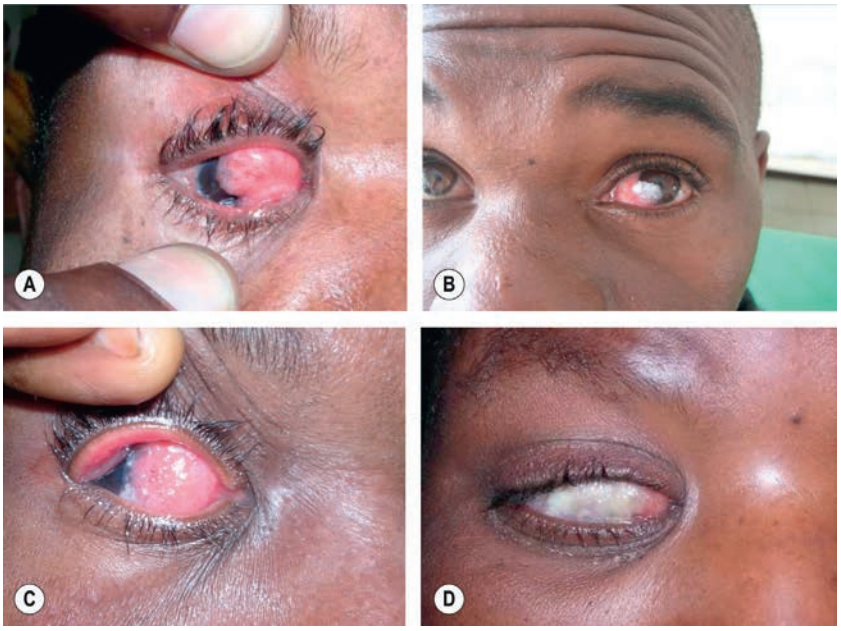
The lesion most likely is an advanced ocular surface squamous neoplasia (OSSN). OSSN are commonly seen in HIV-positive individuals in sub-Saharan Africa. They start as discrete whitish conjunctival lesions (Fig. 43.2) and may develop into large tumours if left untreated. Early stages may be confused with pterygium, with an amelanotic naevus, or a lipoma. Malignant lesions such as amelanotic melanomas, lymphomas or adenocarcinomas may look similar as well.

Answer to Question 2

How Would You Manage this Patient?

The patient should be started on analgesic treatment with nonsteroidal antiinflammatory drugs. She should be counselled and booked for surgery. An extended exenteration of

• **Fig. 43.1** Left eye with marked axial proptosis, nasal upper lid covered with tetracycline eye ointment.



• **Fig. 43.2** Squamous cell carcinoma of the conjunctiva (SCCC). The lesion is commonly seen in HIV patients in the tropics and should not be missed during routine clinical examination. (Courtesy Nicholas A.V. Beare).

the left eyeball and orbit should be done. Control of the patient's HIV infection is crucial, and her HIV viral load should be checked.

Because the patient has lost her eyesight on the left side, the right eye should be carefully examined for possible growth of another OSSN that could be removed at an early stage. A fundoscopy should be performed in order to detect any abnormalities, especially an upcoming cytomegalovirus infection.

The Case Continued…

The patient was admitted to the hospital and counselled several times about the need for surgery. Yet she refused surgical intervention and was only willing to accept conservative treatment.

The patient's left orbit was covered with antibiotics and bandaged. When pain was sufficiently controlled, the patient was discharged with the offer to come back at any time.

She was asked to report to her antiretroviral therapy (ART) clinic for control of the viral load and possible switch of her antiretroviral therapy.

SUMMARY BOX

Ocular Surface Squamous Neoplasia

OSSN are commonly seen in HIV-positive individuals in tropical countries. Early diagnosis is crucial for successful treatment, and any clinician working in a tropical region with high HIV prevalence should be able to recognize an OSSN.

The term OSSN is used to describe dysplastic lesions of conjunctiva and cornea ranging from conjunctival intraepithelial neoplasia (CIN) to invasive squamous cell carcinoma of the conjunctiva (SCCC). The use of ultrasound biomicroscopy (UBM) in OSSN may help detect intraocular invasion. Prominent nodular tumours >5 mm thick can also be taken as risk factors for intraocular involvement. HIV infection, ultraviolet radiation (UV) and human papilloma virus (HPV) are strongly associated with OSSN. These factors, together with vitamin A deficiency, weaken the tumour surveillance system and allow DNA-damaged cells to proliferate into tumours. A five- to tenfold increase in incidence has been observed in parallel with the HIV epidemic. The current incidence of OSSN in sub-Saharan Africa is estimated to be 2.2 per 100 000, and it continues to rise (USA: 0.3 per 100 000). By which mechanism HIV infection favours development of OSSN is as yet unknown. Co-infection with human papilloma virus (HPV) has been implicated in the aetiology of OSSN, but the virus could only be detected in less than half of cases.

OSSN typically presents as a greyish, elevated, gelatinous mass surrounded by engorged conjunctival vessels. There is no explanation why the disease is mostly unilateral. It often starts to develop at the nasal side of the eye and spreads to involve the whole conjunctiva, lids, local tissue and lymph nodes. In the developing world, patients often present late with sometimes disfiguring lesions. There is a trend towards treating conjunctival lesions suspected to be OSSN based on clinical impression. However, clinical diagnosis by slit lamp – with or without gonioscopy – is difficult because of the overlap in clinical features of OSSN and non–OSSN lesions. Toluidine blue 0.05% vital staining is a good screening tool. Negative staining results indicate that OSSN is relatively unlikely. It does, however, not replace surgical biopsy with histopathological examination.

Early, non-invasive stages of OSSN can be treated with topical chemotherapeutic agents such as topical 5-flurouracil and mitomycin or subconjunctival interferon-α2b. Primary treatment is surgical excision. Inexpensive use of fluorouracil 1% eyedrops for 4 weeks substantially reduces the risk of recurrence. It is a low-cost option listed on the WHO's Essential Drug List. In more advanced disease, surgery may have to involve enucleation of the eye or exenteration of the orbit. Adjuvant treatment can decrease recurrence rate, which after simple excision is high (30%–40%). Possibilities for adjunctive treatment include topical chemotherapy, cryotherapy and intraoperative β-irradiation. Diagnosis of OSSN in an HIV-unknown individual should prompt the clinician to perform an HIV test. Because HIV seems to play a role in tumour development, HIV-reactive patients with OSSN should receive effective ART to achieve virological control.

Further Reading

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