

Ocular Surface Neoplasia in a 35yr Old Female HIV Patient - A Case Report and Study of Literature

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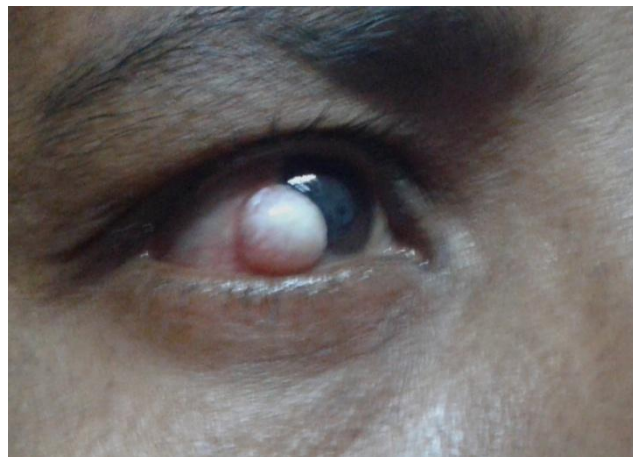
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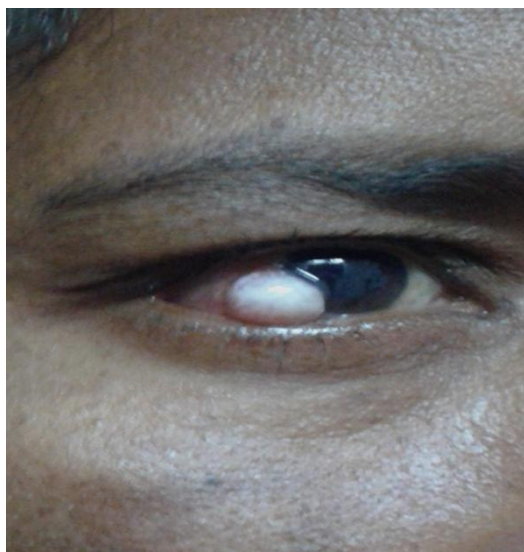
Abstract: A 35yr old female presenting with swelling over lateral limbal area 5x5x4mm of right eye since 2 months, initially small in size, gradual, progressively increasing in size.

Keywords: Ocular surface neoplasia



1. Introduction

Here we are presenting a case of Ocular surface neoplasia of right eye.



2. Case Report

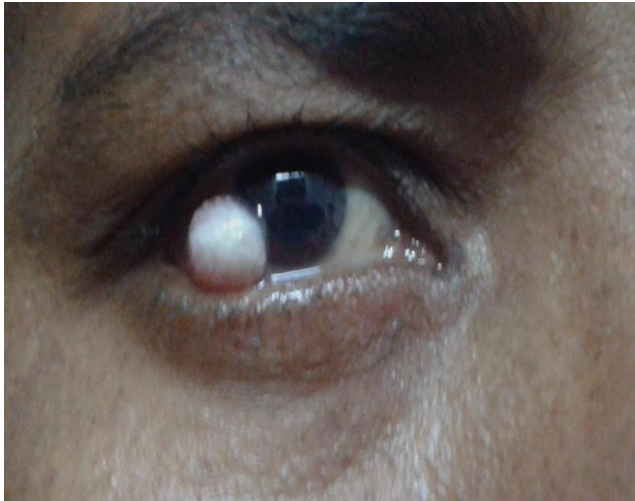
A 35 yr old female, farmer by occupation, presented with swelling right lateral limbal area 5x5x4mm of 2 months

duration. Swelling was initially small in size, gradual progressive increase in size, and attained the present size. No history of trauma. No H/O of pain, redness, photophobia. History of watering, on exposure to air, present. She is non-hypertensive & non diabetic. Family history: Her husband died 10yrs back due to Ca throat & HIV. Personal history: Pt take mixed diet, Bowels and micturition: normal, appetite and sleep: normal, not a smoker, nor alcoholic. General examination: Pt moderately built & nourished, BP: 120/80mmhg, PR:82/min, CVS: S1S2 present, RS: Clear. Ocular examination: Head Posture: midline, Facial symmetry: Bilaterally symmetrical, Extra Ocular Movements: Mechanical restriction of extreme lateral gaze present in right eye. Right eye: eye brows, eye lids, eye lashes normal, swelling of 5x5x4mm present. 2.5mm (half) of the lesion is over the conjunctiva and the rest is overhanging on to the cornea, cornea is clear, Anterior chamber contents & depth: normal, Iris colour & pattern: normal, Pupil: NSRL, Lens: Clear. Left eye findings are normal. Vision both eyes: 6/6, Fundus: Both eyes normal. B scan: normal study of posterior segment, Complete Haemogram done. Hb: 9 gm%, RBC: 3.10 millions / cumm, TC:8200 cell/cumm, DC p:79%, L: 17%, E: 4%, M:0%, ESR: 65MM after 1st hr, platelets: 1,56,000 cell/cumm, PCV 27ml%, BT 1 mt 50 secs, CT 3mt 40 secs, Perypheral smear: microcytic hypochromic anaemia with mild neutrophilia, RBS:140mg%, blood urea: 27mg%, HIV positive, CD4 count 238.

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3. Discussion

Ocular surface squamous neoplasia includes a spectrum of benign, pre-malignant, and malignant unilateral slowly progressive epithelial lesions of conjunctiva & cornea. Risk factors include UV light exposure, Human papilloma virus (type 16) infection, AIDS, xeroderma pigmentosum and stem cell therapy. Common cysts found in conjunctiva are due to dilatation of lymph spaces, forming rows of little cysts when small (lymphangiectasis), occasionally small but multilocular cysts occur (lymphangiomas). Tumorous conditions in conjunctiva have a tendency to be polypoid owing to perpetual movements of the globe and lids. Congenital tumours: these include dermoids & dermolipomas. They are actually not true neoplasms but are in fact *choristomas* or a collection of heterotopic tissue (normal tissue in abnormal position), which grow as tumours or tumour like conditions.

Dermoids are lenticular yellow tumours, usually astride the corneal margin, most commonly on the outer side. They consist of sebaceous glands and epithelium with sebaceous glands and hairs, which may cause irritation. They tend to grow at puberty & should be dissected off the globe if they cause excessive astigmatism, encroach on visual axis or cause an unacceptable cosmetic blemish. After removal the site of attachment to the cornea remains opaque. The area is early disguised by tattooing but presently replacement by a lamellar graft is preferred.

Dermolipoma or fibrofatty tumours are congenital tumours found at the outer canthus sometimes associated with the accessory auricles and other congenital defects in babies. They consist of fibrous tissue and fat, sometimes with dermoid tissue on the surface, and are not encapsulated. The main mass may be removed with care if cosmetically unacceptable, but it will be found that the fat is continuous with that of the orbit. Both limbal dermoids and dermolipomas are more common in children with congenital development anomaly known as Goldenhar syndrome (oculoauriculovertebral dysplasia). This syndrome affects structures derived from first branchial arch leading to pre auricular tags, deformities of external ear and vertebral anomalies. Colobomas of lids and iris and Duane retraction syndrome are other associated ocular abnormalities.

Papillomata: These occur at inner canthus, in the fornices or at the limbus. They may become malignant and should be removed.

Simple granulomata: Consists of exuberant granulation tissue, generally polypoid in form, often grow from tenotomy wounds or the sites of foreign bodies. They are common in empty sockets after excision, and at the site of chalazia which have been insufficiently scraped. They should be removed with a pair of scissors and send for histopathological examination.

Squamous cell carcinoma (epithelioma): It is a fleshy pink papillomatous mass with feeder vessels or occasionally may exhibit diffuse growth & masquerade as chronic conjunctivitis. Corneal involvement may occur. This occurs where one kind of epithelium passes into another; therefore in the conjunctiva it occurs chiefly at the limbus. Papillomata in the old people often take malignant proliferation. Bowen's intraepithelial epithelioma or carcinoma in situ is also seen. Squamous cell carcinoma spreads over the surface and into the fornices, rarely penetrating the globe. Metastatic disease is extremely rare. They must be removed as freely as possible, the base been cauterised by diathermy or treated with cryotherapy; & the diagnosis should be microscopically confirmed. On the slightest sign of recurrence with invasive squamous cell carcinoma, the eye must be excised & if recurrences take place the orbit must be exenterated & radiation therapy given.

Basal cell carcinoma (Rodent ulcer): may invade the conjunctiva from the lids.

Lymphomas: Conjunctival lymphomas occur on the bulbar conjunctiva or in the fornix. They are typically described as painless slow growing salmon coloured i.e. light pinkish mildly elevated homogenous lesions which can sometimes have small round aggregations that look like fish eggs. An excision biopsy must be done. Systemic lymphoma may be associated hence a thorough systemic evaluation is mandatory. Radiotherapy is effective.

Kaposi sarcoma: Kaposi first described this special type of sarcoma in 1872. It is a tumour of vasculoformative mesenchymal tissue with pericytes and endothelial cell components. This tumour is seen in HIV pts. It affects the skin or any organ & is common in southern Mediterranean regions, eastern Europe & Africa. The lesion may also affect eye lids and face. The tumour is highly vascular, with bluish red colour & presents as an elevated nodule resembling subconjunctival haemorrhage. The lesion is malignant & the treatment is palliative with radiotherapy. The treatment is based on subtypes. Localized lesions can be treated with cryotherapy, intralesional injection of Vinblastine, Alitretinoin gel, radiotherapy, topical immunotherapy or surgical excision. Extreme cutaneous disease may require IV chemotherapy or immunotherapy. With the AIDS related Kaposi sarcoma highly active antiviral therapy has been shown to prevent or reduce regression. Some AIDS patients have complete resolution of the lesion and prolonged remission while continuing the therapy.

Pigmented tumours: These constitute important type of neoplasia which introduces difficult clinical decisions: some are malignant. They are grey gelatinous or pigmented nodules situated at limbus or near the plica semilunaris. They have the same structure as in the skin-groups, often alveolar, or naevus cells in close connection with the epithelium. They are congenital and tend to grow at puberty, rarely becoming malignant. In view of this they should be excised completely before puberty, lest malignant changes follow the operative disturbance. It should be noted that pigmentation at limbus occurs normally in people with dark complexion. Precancerous melanosis is diffusely spreading pigmentation of the conjunctiva, rare in elderly people, may also involve skin of lids & cheek. It is liable to spread slowly & may eventually assume malignant characteristics, giving rise to metastasis. The condition should be viewed precancerous & though radiosensitive at this stage, but in malignant phase becomes radioresistant in which case the only effective treatment is wide excision with exenteration of the orbit & extensive reconstitution by skin grafting. Beta irradiation, either primary or after tumour excision, is the treatment of choice for conjunctival melanomas.

Malignant melanoma is rare. It typically occurs at limbus, is usually pigmented, & occurs mostly in elderly. It spreads over the surface of the globe but rarely penetrates it; recurrences & metastasis occur as elsewhere in the body. The neoplasm may be alveolar (derived from naevi) or round or spindle-celled. The treatment is by excision of the globe or exenteration of the orbit.

4. Summary

In view of the above clinical features age of the patient and HIV status, this is a case of Kaposi Sarcoma arising from conjunctiva in the right eye. Patient has been advised instillation of mitotycin eye drops 6th hrly.

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