Case report

Ocular surface squamous neoplasia in a young adult – its nature and unusual course

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Abstract

Background: We describe a case of a recurrent ocular surface squamous neoplasia (OSSN) in a 37-year-old male with a mass at the infero-temporal quadrant along with an isolated primary lesion at the upper nasal quadrant for the last five months with a past history of surgical excision 7 years ago for a nodular mass in the same eye. The mass showed delayed response to Mitomycin C (MMC) therapy and finally developed limbal stem cell deficiency.

Case report: A 37-year-old male presented with a five-month history of foreign body sensation and localized conjunctival hyperemia and two progressively enlarging bumps over the limbus in the left eye. The past history stated a surgical excision, done 7 years ago, for a nodular mass in the same eye. A clinical diagnosis of recurrent OSSN was made, and confirmed by impression cytology. The patient received initial treatment with 0.02 % MMC, but did not show any improvement even after 3 cycles, but later showed marked chemoreduction with 0.04 % MMC on two cycles and a complete resolution of the neoplasia after two more cycles. Impression cytology at six months revealed no abnormality. But at the 10-month follow-up, limbal stem cell deficiency was observed.

Conclusion: While examining, managing and follow-up of a case of OSSN, one needs to know the atypical nature and response of the tumour. Long-term follow-up in these cases is mandatory.

Introduction

Ocular surface squamous neoplasia (OSSN) is a disease of the elderly[1], having predilection for the interpalpebral area, mostly the corneoscleral limbus[1] in 87.8 % of cases. Management can be either surgical or by topical mitomycin C (MMC) drops. We report a case of OSSN in which the inferior and superior limbus were involved in a young adult, showed delayed recurrence after surgery, took an unusually prolonged time for complete resolution with topical MMC and finally resulted in limbal stem cell deficiency (LSCD).

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Case report

A 37-year old male shopkeeper presented with two gradually increasing, masses in the left eye over five months. The past history revealed temporal limbal mass excision in the same eye seven years ago for a nodule at the temporal limbus. On examination, best corrected visual acuity (BCVA) was 20/30 left eye (LE) and 20/20 right eye (RE). Slit-lamp (S/L)

examination of the LE revealed two separate, raised masses, pale pink in colour with a scaly surface at the inferior (5.5mm x 4mm) and superior (5mm x 3.5mm) limbus, invading 2mm into the cornea (Fig.1a). The anterior chamber (AC) was clear. Intraocular pressure was 16 mm in both eyes. There was no lymphadenopathy or any systemic abnormalities.

A provisional diagnosis of recurrent OSSN was made, and confirmed by impression cytology (IC). Intraocular invasion was ruled out by ultrasound biomicroscopy (UBM) (Fig.1b). The ELISA test for Human Immunodeficiency Virus (HIV), done after taking patient's informed consent, was negative.

Topical MMC (0.02 %), four times a day, in a cyclic manner, one week on the drug and one week off it was started. As there was no improvement after three cycles, Interferon alpha-2 b (INF á-2b) drops (1 million IU/ml) was suggested but refused by the patient due to its cost. Topical MMC concentration was increased to 0.04 %. After two subsequent cycles, a remarkable chemoreduction was noticed and complete resolution occurred after two more cycles, following which the drug was discontinued. The patient did not show any signs or symptoms of drug intolerance during the therapy except for a mild irritation off and on which, however, subsided with topical non-steroidal anti-inflammatory drops twice daily and a tear substitute four times a day. Pre- and post-treatment specular microscopic findings were within normal limits. The BCVA was stable. The IC at six months revealed no abnormality.

At the ten-month follow-up, the cornea was lustureless with multiple superficial vascularisation indicating limbal stem cell deficiency (Fig. 2a) and with a BCVA of 20/40. IC revealed the presence of a few conjunctival goblet cells (Fig 2b). Instillation of frequent topical lubricant drops, two hourly, was advised. There was no recurrence after two years of follow-up.

Discussion

In the management of OSSN, excision biopsy, though considered the gold standard, may not eradicate the subclinical lesion if no frozen section has been adopted. As per the patient's old records, there had been an excision of the mass but without frozen section. This might have had an implication of incomplete eradication, and consequently recurrence. More recently, topical medical therapy (Fucht et al 1997; Chen et al 2004; Holocombe et al 2006) is preferred over surgical, as it treats potentially abnormal cells on the entire ocular surface while conserving the limbal stem cell population.

The present case is interesting in many ways. (i) It occurred in a 30 year-young individual with no exposure to ultraviolet rays, unlike a field worker. It was not associated with HIV. (ii) The primary lesion had occurred seven years ago, at the inter-palpebral area, but the classical area was not involved in the recurrent lesions. (iii) The previous records document the initial lesion as a nodular form and the present lesions do not fit into either of the two morphological profiles (nodular or flat), precluding classification on this basis. (iv) Recurrence usually occurs within the first two years of resolution but in the present case, it was delayed for seven years, which could be due to the presence of subclinical tumor cells. (v) The unusual but significant finding was the development of LSCD. LSCD following MMC is less expected than after wide surgical excision where there is the possibility of direct damage of the limbal stem cells (Khong et al 2006). If it does occur, it may be attributed to the diffuse tumor location rather than to the MMC per se (Khong et al 2006). The delayed response in the case under report could be multifactorial: (i) the use of less concentrated drops, or (ii) less potent drops and (iii) a poor compliance of the patient. The first factor was ruled out as the use of less concentrated drops 0.02 % is universally accepted for OSSN (Panda et al 2008; Sepulveda et al 2010). The second may not be a possibility as the drug is routinely prepared in our own pharmacy adopting

the same formulation. Regarding the third, though a firm opinion cannot be given, the patient appears to be compliant.

In summary, the concept that OSSN is a disease of the elderly and in immunocompromised persons does not hold good for the present case. Young and healthy individuals may be affected by OSSN even without being associated with HIV. Recurrence following incomplete removal of the neoplasia does occur, but recurrence after seven years is apparently rare for which constant follow-up for these patients is a must. Complete resolution after topical MMC, which usually takes four cycles, may be delayed in some situations (Lee et al 1995; Fucht et al 1997; Chen et al 2004). Whether this delay in the present case is as a result of its atypical and recurrent nature or due to an inadequate drug concentration cannot be commented on the basis of a single case report. Long-term use of the prescribed drug, more than four to five cycles, is further a matter of concern as far as complications are concerned.

Thus, we would like to reiterate that if there is a delayed response to MMC, efforts should be made to find out the exact cause and the patient should be monitored constantly both during therapy and following complete resolution (Lee et al 1995). Though more aggressive surveillance is recommended in the first two years of follow-up, long-term follow-up is essential as recurrences can occur even many years later, as evident in the present case.

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