Reconstructive surgery in a case of advanced sebaceous cell carcinoma of the eye Lid: a case report

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Case Report

A fifty year old female patient presented in the ophthalmology OPD with history of swelling over the left upper lid of one year duration. Initially the swelling was very small and painless, but gradually over a period of six months, it increased to the size of a peanut and was associated with mild pain. She consulted a local practitioner for the same and received treatment in the form of excision of the swelling, after which the swelling rapidly increased in size and was accompanied by severe pain. The swelling ruptured and was associated with purulent discharge. On general examination, she was averagely built and nourished, afebrile. Blood Pressure was 120/70 mm of Hg, Pulse was 78/minute. Preauricular lymph node was palpable, the size of which was about 3cm x 2cm x 2cm.It was hard in consistency, nontender, with irregular surface and was adherent to the underlying structures. Systemic Examination of the patient, including the cardiovascular system, respiratory system, central nervous system and per abdominal examination were within normal limits.

Ocular Examination

Right Eye was within normal limits.

Left Eye : No perception of light was present. There was a huge mass covering the left eyeball. The size of the swelling was 6cm x 5cm x4cm.

The swelling had two parts:

Upper part was reddish, hard and tender with irregular margins. Lower part was ulcerated. The size of the ulcer was 4cm x 3cm x 0.5cm with indurated edges, irregular margins. The floor of the ulcer was necrosed and was associated with a foul smelling purulent discharge. The ulcer bled on touch. The eye ball could not be examined as the upper lid swelling could not be retracted.

Fig 1. Patient at the time of presentation

Laboratory Investigations :

Hb: 8.9gm/dl. TLC: 13,700/cu.mm. Renal Function tests, Liver Function tests, Blood sugar fasting were within normal limits. She tested negative for and HIV antibodies and HBsAg. The biopsy from the upper lid swelling was suggestive of Sebaceous gland carcinoma. CT scan of the orbit or the head could not be carried out due to financial constraints of the patient.

Management

After the biopsy report, surgery was planned. Patient underwent left eye Exenteration surgery which included resection of entire contents of the orbit including the eyelids, the globe, extra ocular muscles, optic nerve, orbital fat and periosteum along with the tumour mass. The exposed bony orbit was covered with full thickness skin graft taken from the left thigh of the patient. The skin graft was placed with the raw surface towards the bony orbit. The graft was kept in place by the orbital pack made from dental mould followed by end to end tight suturing of the orbital margin skin with underlying muscles. The first skin graft became infected and the procedure had to be repeated after a period of two weeks. Antiseptic dressing was continued for two months. With a well taken skin graft, an orbital prosthesis was designed for the patient with the help of the Prosthodontics

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department for aesthetic reconstruction. Subsequently three cycles of adjunctive chemotherapy with injection Cisplatin 100 mg IV and injection Doxorubicin 60mg IV at three weekly intervals was given to the patient.

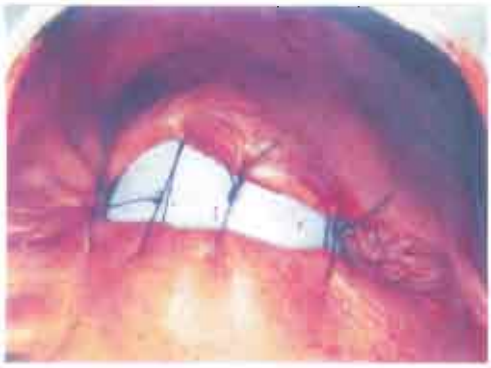

Fig. 2. Skin graft with mould

Fig. 3. Empty socket with dry skin graft

Discussion:

Sebaceous gland cell tumour of the eye lid is are rare tumours, making up to 0.2 - 0.7% of all eye lid tumours and 1-5.5% of all eye lid malignancies[1]. Incidence also shows great geographic variation, with 0.2-4.7% of all malignant eye lid tumours in the USA to about 28% in the Chinese population[2]. Male to female ratio is 1:1.5[3].

Clinical presentation depends on the gland of ori-gin (glands of Zies, Meibomian or the sebaceous glands of the lid skin), stage of the disease and its manner of spread. The clinical diagnosis of sebaceous carcinoma may be difficult, partly because it is rarely encountered and partly because of its propensity of the lid skin), stage of the disease and its manner of spread. The clinical diagnosis of sebaceous carcinoma may be difficult, partly because of it is to simulate other eyelid lesions.

Fig 4. After cosmetic reconstruction

The common clinical diagnosis often made are chalazion, unilateral blepharoconjunctivitis (masquerade syndrome), nodule at the lid margin, anterior orbital or lacrimal gland tumour, basal cell carcinoma, sebaceous cyst and keratosis[4]. In exceptional patients the presenting sign may be an enlarged cervical or preauricular lymph node, the usual primary lid lesion being inconspicuous and overlooked[5]. These diagnostic difficulties warrant a biopsy in all suspicious inflammatory conditions which do not respond to treatment[6]. The ability of sebaceous carcinoma cells to spread within the conjunctival epithelium has been appreciated from the earliest clinicopathological series. It is believed that the spread occurs via the ducts of the primarily involved glands of Zeis or meibomian glands and thereafter within the epidermis or toward the tarsal conjunctival epithelium, with progressive intraepithelial extension farther afield.

On histopathology the distinctive cytological features to be looked for and which form the basis of diagnosis are individual cells or cluster of cells with a finely vacuolated frothy cytoplasm surrounded by interstitial xanthomatous or granulomatous reaction (due to the large amount of lipids synthesized by the tumour cells and released in to the extra cellular space). Numerous features are associated with poor prognosis. Statistically, a delay in diagnosis (i.e., duration of symptoms) and a tumour size greater than 1 cm appear to be ominous signs. Involvement of both upper and lower lids result in 83% fatality rate[7]. The various pathologic features that indicate a poor prognosis are vascular or lymphatic invasion, orbital involvement, poor differentiation, highly infiltrative pattern and multicentric origin. The decrease in tumour related deaths and fatality rate due to sebaceous gland cell carcinoma in the last three decades can be attributed to the increased awareness of the clinician of the need to perform wide local excision of the tumour

with frozen section control of the margins[8]. Conjunctival map biopsies are also performed at many centres. Both methods, however can lead to errors. Microscopic evaluation of the permanent sections of the margins provide the most reliable confirmation of the adequacy of excision[9].

Wide local excision of the tumour with a 4 mm clear margin is the most widely accepted form of treatment for small tumours. However, patients with a large and disfiguring growth, tumours in-volving the orbit, multifocal tumours and cases with extensive pagetoid spread through the conjunctiva, are treated by exenteration, a form of radical local surgery[10]. The extent of orbital exenteration is gov-erned by the depth of invasion of the orbital com-partment. Total exenteration involves the removal of all the contents of the orbit including the periosteum, both eye lids and surrounding tissue so as to achieve a tumour free margin as mentioned before. Cryotherapy has been advocated by some authors as a means of treating diffuse conjunctival involvement, but can lead to troublesome dry eye syndrome post operatively. Radiotherapy is used only when patient is too ill to undergo surgery, or if it is unwilling for adequate radical surgery. Patients must be followed up at short intervals post-operatively as the tu-mour has a fast growth potential and are known to recur in 6 to 29% of cases[7], and majority of all recurrences appear within the first 4 years of treatment." Most patients after radical surgery are left with only the skin graft covering the orbital cavity, without any cosmetic reconstruction of the missing orbital structures. This puts the patient in deep mental and social agony. Our patient was unique in a way that we could achieve complete cosmetic reconstruction.

Acknowledgement

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