

■ Case Reports

Eyelid squamous cell carcinoma in xeroderma pigmentosa: A therapeutic challenge

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Abstract

A 10-year-old male, Muslim child diagnosed with xeroderma pigmentosa with a right upper lid malignancy presented as a therapeutic challenge because of the non-availability of normal facial skin for grafting and lid reconstruction. The lid lesion had been present for the last one month. The disease had been present for past five years with polymorphous, erythematous lesions all over the body. On examination, there was a hard reddish yellow mass in the lower lid with features of secondary infection along with corneal xerosis in both eyes. With a clinical diagnosis of malignancy, the lesion was subjected to a wide excision after the investigations. The resultant large tissue defect in the right lower lid region was managed by a full thickness trans-midline lateral forehead flap based on superficial temporal artery. Post operatively, there was a satisfactory graft uptake. The paucity of surgical options for the plastic reconstruction coupled with the large area to be reconstructed made our case worth reporting.

Key words: erythematous, malignancy, forehead flap, excision

Introduction

Xeroderma pigmentosa is a rare autosomal recessive disorder. This disease leads to a 1000 fold increase in the risk of cutaneous and ocular neoplasms. The prognosis for life is poor in young age due to early superimposed malignancies. After the excision of lid lesions, surgical options for the reconstruction of lid defects are limited. In this case report we share our experience in managing a lid lesion diagnosed in a known case of xeroderma pigmentosa and its therapeutic outcome.

Case history

JM, a muslim, male child presented with right lower lid mass at the age of 10, 5 years after the initial diagnosis of xeroderma pigmentosa. In these last 5 years, he had developed photophobia, skin lesions comprising of large red spots changing to dark lesions with blistering, freckles on limbs and face, hypopigmented and hyper-pigmented lesions covering both sunexposed and covered part of skin and was being treated with vitamin-A supplements and sunscreen lotions. He presented to our ophthalmology O.P.D. with a history of right lower lid mass for 1 month with relentless worsening of vision in his both eyes for the last 5 years.

At presentation, there was a large reddish yellow mass arising from the right lower lid, about 2×10 mm size with irregular surface, hard in consistency and with multiple points of hemorrhage with superadded infection. The conjunctiva was diffusely congested, cornea showed xerosis along with a macular-leucoma grade central corneal opacity. The posterior details were not visible.

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In the left eye there was conjunctival congestion with corneal xerosis. The posterior segment did not reveal any significant abnormality. The best corrected visual acuity in the RE was counting fingers close to face whereas in the LE , it was 20/40.FNAC of the lower lid mass revealed a differentiated squamous cell carcinoma .

After the investigations, patient was subjected to surgery under general anaesthesia, in which the tumour involving complete right lower lid was excised. The resultant surgical defect extended from inner canthus to the outer canthus. A full thickness lateral forehead flap beyond midline was taken based on the superficial temporal artery. The flap was mobilized through a skin tunnel and end to end closure was adapted for reconstruction [Figure1]. The inner lining of the flap was not lined with amniotic membrane or buccal mucosa. Post –operatively, patient had a satisfactory uptake of the graft [Figure 2].



Figure 1: Per-operative photograph showing the placement of the trans-midline lateral forehead flap over the lower lid defect created after the excision of the lid malignancy.



Figure 2: Post- operative follow-up photograph of the patient showing the satisfactory graft uptake.

Discussion

Numerous techniques for the reconstruction of isolated periocular defects have been described, including the ‘ rotation cheek flap (Hornblass, 1988), ‘Tenzels semicircular flap (Hornblass, 1988), ‘Hughes procedure’ (Hornblass, 1988), ‘bipedicle upperlip flap’ (Hornblass, 1988), ‘temporal flap’ (Hornblass, 1988), etc . Long standing xeroderma pigmentosa is associated with telangiectasias, shiny irregular areas of atrophy and warty areas may appear on the exposed skin of face and may rapidly change into malignancies. Therefore, most of the reported reconstruction techniques may not be used in these patients due to the lack of availability of normal skin tissue for grafting.

Therefore, the current case was a therapeutic challenge, as a normal site for skin muscle flap could not be identified. Choice of site of flap was limited to the forehead as this was the least involved site. Distance flaps could not be considered as patient’s father did not consent for the same.

To cover the entire lower lid the flap had to be extended beyond vertical midline on the forehead. As there was a good arterial base, no flap necrosis ensued. As the lower lid does not play a vital role in the resurfacing of the tear film the flap was not lined by mucosa. The flap healed well, without resultant ectropion or lower lid lag. Need for providing rigid support was not an issue as the lower lid tarsal plate is narrow. The pedicled flap provided a thick layer of muscle which remained in position.

References

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