MELANOMA AND MELANOCYTIC NAEVI

SCHIMMELPENNING SYNDROME WITH RARE UNUSUAL ASSOCIATIONS

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Background : Schimmelpenning syndrome is a rare multi-system disorder characterised by congenital sebaceous nevus associated with other neurological, ocular and skeletal abnormalities. Choristomas are congenital lesions representing normal tissues in an abnormal location. We report a case of an 18 yr old male patient with a large Nevus Sebaceous measuring 18.5×5.5 cm on the left cheek extending till the angle of mouth, involving the vermillion & upper lip extending to the neck and the suprastrenal notch. Ocular involvement in the form of nevus mass completely occupying the left eye with an upper lid coloboma & a schwanomma at the lateral canthus. The patient also right sided hemiparesis(CNS) & lower limb shortening on the left side(Skeletal) which lead to a limping gait.

Observation : We present the case for the multidisciplinary management which was done in the form of complete mid-dermal excision of the nevus with primary closure of the midline areas & full thickness grafting on the cheek & temporal area. A left eyelid & ocular surface resection & reconstruction using amniotic membrane graft was done. Prosthetic correction for the gait was planned. At 1 month follow up, the midline lesions had healed well with primary intention & the graft had taken up well at the cheek & temporal area. The lid position was also near normal. It was a very favourable outcome with a significant improvement in the patient's QLI.

Key message : We present this case to bring to light the possible management of ENS even with extensive cutaneous & extracutaneous involvement thereby giving a highly improved QLI to the patient.

KEY WORDS Epidermal nevus syndrome, schimmelpenning syndrome, coloboma, choirstoma, skingrafting