

Title : Cutaneous Malignancies in Xeroderma Pigmentosum: Earlier Management Improves Survival.

Author Name : Sudhir M. Naik

CO Author Name : Ashok M. Shenoy

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Department : Department of Head and Neck Oncology

Asbtract :

Xeroderma pigmentosum (XP) is a rare autosomal recessive disease, characterized by hypersensitivity of the skin to ultraviolet (UV) radiation leading to high incidence of skin cancer and progressive neurological complications. It results in premature development of neoplasias due to an exacerbated hypersensitivity to UV radiation causing premalignant and malignant lesions leading to death in early adulthood. Two cases of clinical features of xeroderma pigmentosa with skin lesions were managed in our department. One had multiple clusters of basal and squamous cell carcinomas and the other had malignant melanomas and right neck nodes. Both were treated with multiple wide excisions and the neck node were surgically managed with radical neck dissections. Skin malignancies were common in the sun exposed areas and patients were advised regular 3 months follow up. The disease is ultimately fatal, life can be prolonged by simple preventive measures to minimize sun exposure. Comprehensive multimodality management includes patient education and counselling for the psychosomatic disorder and genetic counseling remains the most important preventive measure.