

## Pigmented Xerodermoid

A 5-year-old boy, born out of a consanguineous marriage, presented with pigmented skin lesions involving sun-exposed areas for 2 years (*Fig. 1* and *Fig.2*). The child was born full term at home, with an uneventful postnatal period; though the milestones of the child were delayed. The skin lesions had appeared on face two years ago and gradually involved upper trunk, and exposed parts of extremities. His younger sister too had similar, but milder skin lesions. His height for age, weight for age and weight for height were less than 3 standard deviation. Skin was dry. Multiple hyperpigmented as well as hypopigmented macules were noted on sun-exposed areas. There was decreased lacrimation while crying. Ophthalmological examination showed corneal xerosis. Neurological examination did not reveal any abnormality. On the basis of clinical findings, diagnosis of pigmented xerodermoid was made. Strict sun-protection and liberal use of sunscreen were advised.

Xeroderma pigmentosum (XP), is a rare autosomal recessive disease characterized by sun sensitivity, photophobia, early onset of freckling, and subsequent neoplastic changes on sun-exposed skin (median age of onset of non-melanoma skin cancer is 8 years). The median age of onset of the cutaneous symptoms is 1-2 years and the skin abnormalities result from genetically acquired inability to repair UV induced DNA damage. Pigmented xerodermoid, also known as XP variant (XPV), shows a typical XP phenotype, but is characterized by a significantly later onset (after 30 years of age), generally milder symptoms, and a protracted course of the disease. Notably, XPV may appear earlier in the tropics. Treatment includes strict photo-protection of the skin and eye, and use of lubricating eye drops. Regular whole body examination by a dermatologist is needed to identify and treat premalignant and malignant lesions. Regular ophthalmologic examination too is desirable. Since strict photo-protection is advocated, Vitamin D should be given prophylactically.

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**FIG. 1** Multiple hyperpigmented macules on face



**FIG. 2** Multiple hyperpigmented macules with hypopigmented macules. Skin appears atrophic.