Images in Clinical Practice

Xeroderma Pigmentosum

A 10-year-old male child, born of consanguineous parents presented with an 8-year history of freckle-like pigmented macular lesions interspersed with atrophic, hypopigmented macules over the face *{Fig. 1)* and sun exposed areas of neck, forearms, hands, legs and feet. Ocular mucosa and mucocutaneous junctions of the lips were also involved. In addition he had a generalized, dry (xerotic) skin. The child had severe photophobia and there was a history of recurrent, superficial ulcerations over the bridge of nose which healed slowly leaving behind atrophic scars (*Fig. 2).* His elder brother had similar disorder and died at the age of 18-years after developing multiple skin malignancies including squamous cell and pigmented basal cell carcinomas (*Fig.* 3). Survival beyond the second decade is unusual in this rare autosomal recessive disorder where there is a defect of DNA excision repair.

In our patient, we have advised strict protection from sunlight including wearing of two layers of full sleeved clothing, sunglasses and application of a thick layer of physical sunblock cream like lactocalamine over all uncovered areas of the skin. The child has been forbidden to play out in the sun and the parents have been asked to look for and report immediately any tumors that may develop so that early surgical excision may be undertaken. In addition the child has been given topical retinoic acid (0.5%) over the sun exposed parts.

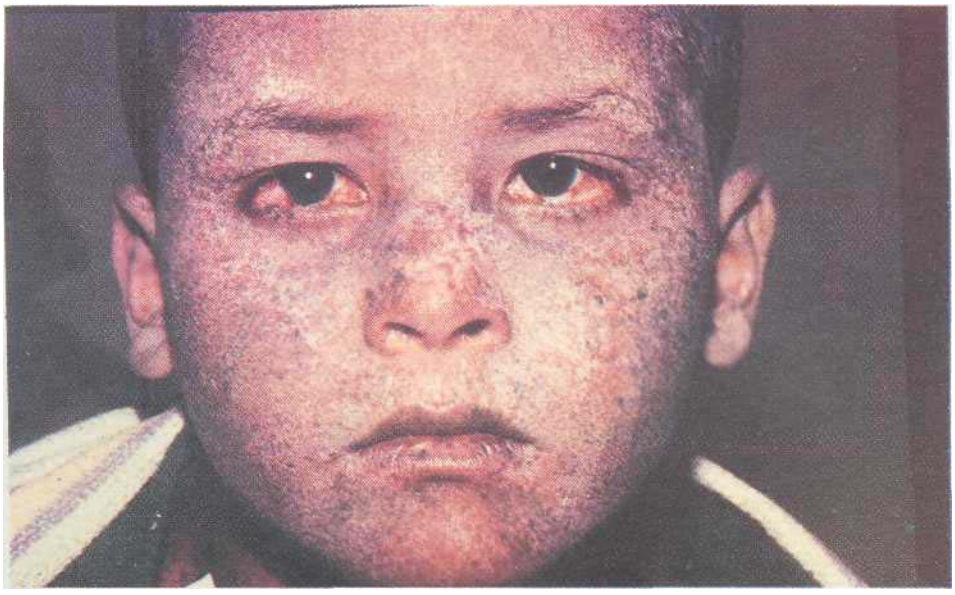

Fig. 1. Freckle-like pigmented macular lesions interspersed with atrophic hypopigmented macules.

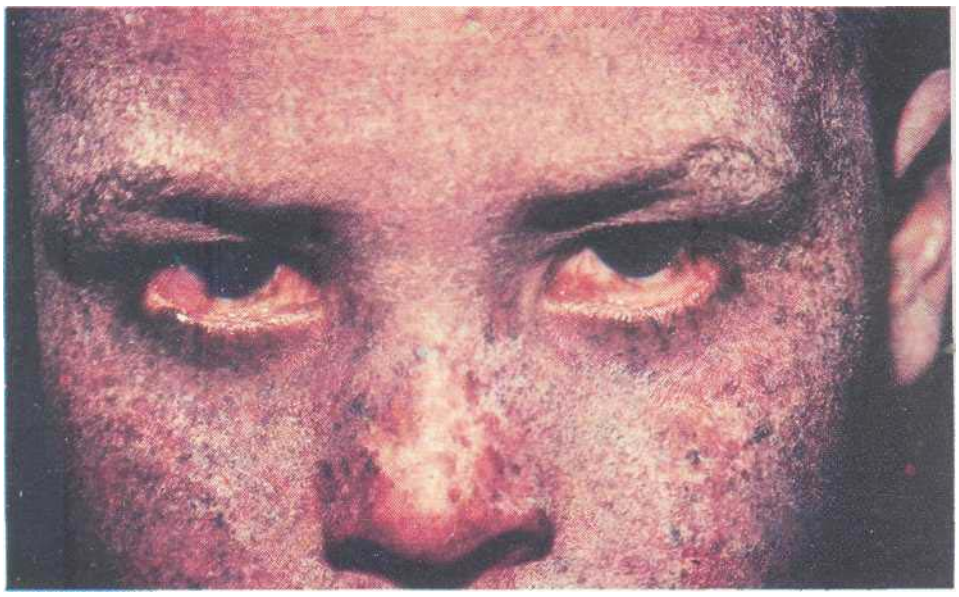

Fig. 2. Recurrent, superficial ulcers which heal with atropliic scars.

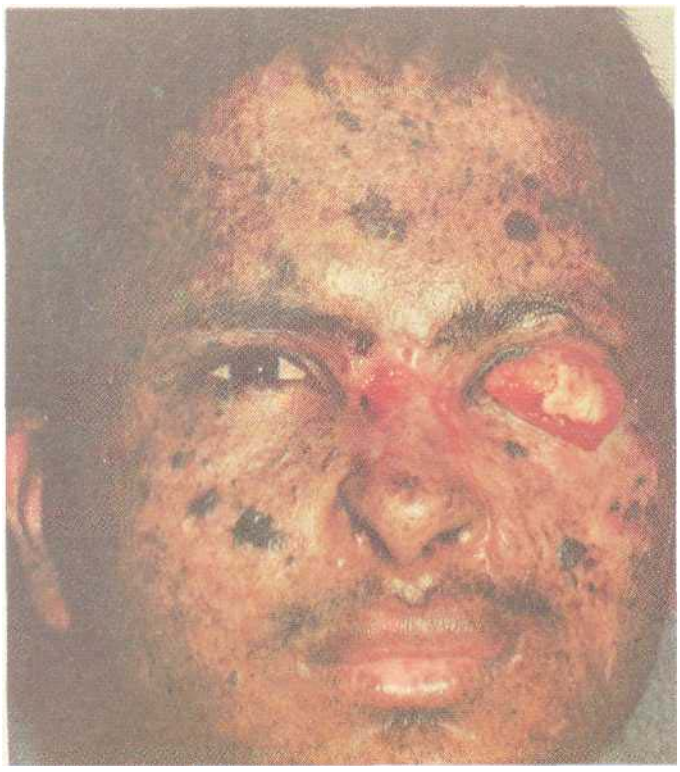

Fig. 3. *Elder sibling who developed multiple cutaneous malignancies.*

Oral retinoids have been tried elsewhere and there is evidence to suggest that they reduce the incidence of skin cancers in this condition.

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