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Quick Response Code:

Childhood porphyria causing lifelong multilating photosensitivity

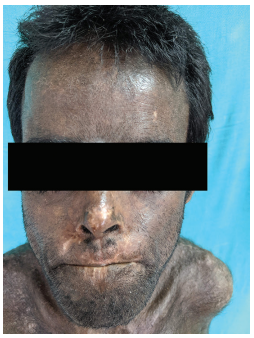

Fig. 1. Clinical photograph demonstrating mutilating scars over the face.

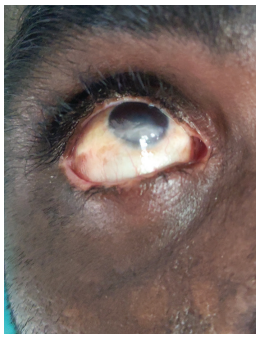

Fig. 2. Clinical photograph demonstrating corneal opacity.

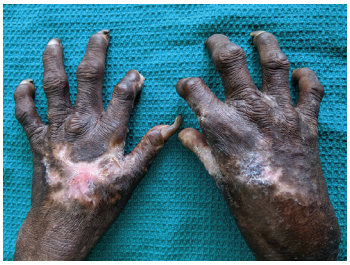

Fig. 3. Clinical photograph demonstrating shortening of fingers and mutilating scars over the hand.

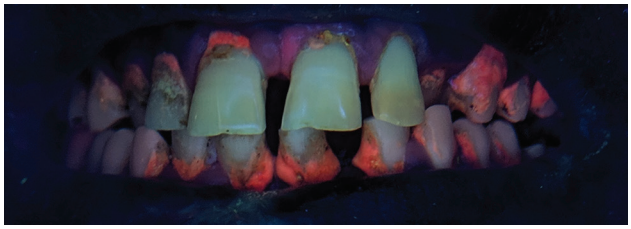

Fig. 4. Wood's lamp photograph demonstrating erythrodontia.

Fig. 5. Wood's lamp photograph of the patient's urine demonstrating red-orange fluorescence.

A 31 yr old male[†] was admitted for otherwise benign, itching and burning sensation over the sun-exposed sites for two weeks in the department of Dermatology, Madras Medical College and Rajiv

[†]Patient's consent obtained to publish clinical information and images.

Gandhi Government General Hospital, Chennai, India. The patient was first presented in October 2004; since then, he was under follow up. He was diagnosed with congenital erythropoietic porphyria at the age of eight months, a severe and rare childhood porphyria causing lifelong mutilating photosensitivity and haematological disease, an autosomal recessive inherited deficiency of uroporphyrinogen III cosynthase enzyme. There were mutilating scars over the face (Fig. 1), corneal opacity (Fig. 2), hypertrichosis of the chest, resorption of the terminal phalanx of the extremities (Fig. 3) and brown-coloured teeth and teeth, urine and skin lesions showed reddish-orange fluorescence under Wood's lamp (Figs 4 and 5). The patient remained on regular photoprotection follow up for ulceration, photodamage, scar tissue and haemolytic anaemia. Bone marrow transplant is the only cure.

***Conflicts of Interest*:** None

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