Diffuse Non-epidermolytic Palmoplantar Keratoderma

A 12-year-old female presented to the Dermatology Outpatient Department with thickening of palms and soles. To start with, the palmoplantar skin was red at the age of 4 months. Subsequently, a thick yellowish hyperkeratosis appeared and expanded to attain the present status. On examination, yellowish hyperkeratotic plaques were found on the palms and soles, with sparing of the arches of feet and sharp cut-off at the wrist. (***Fig.* 1** and **2**) The dorsa of the fingers showed mild involvement, with a scleroderma-like thickening of the skin distal to the proximal interphalangeal joints. The oral mucosa, teeth and nails were normal. Systemic examination was non-contributory. Patient's father had similar lesions on palms and soles. A provisional diagnosis of Diffuse Palmoplantar Keratoderma (PPK) was made. Histopathological examination of the skin biopsy taken from the patient's sole revealed orthokeratotic hyperkeratosis, hypergranulosis and acanthosis without epidermolysis. Hyphae were not detected on examination of the squames in the scraping; with 10% potassium hydroxide. Based on clinical and histopathological findings, diagnosis of "Unna-Thost variety of Diffuse Non-epidermolytic Palmoplantar Keratoderma" (NEPPK) was done. Both, the patient and her father have been prescribed topical tretinoin and emollients, and at present, they are under follow-up.

Unna-Thost disease (Diffuse NEPPK) is inherited in an autosomal dominant manner, underlying gene defect being in 12q11-q13 or Desmoglein 1. The disease develops in early childhood and persists throughout life. Clinically, there is hyperkeratosis on the palms and soles. Clinical differential diagnoses include Howel–Evans syndrome (focal keratoderma on pressure sites, leukokeratosis of the oral mucosa and esophageal cancer); Clouston syndrome (hidrotic ectodermal dysplasia, hypotrichosis and nail dystrophy), Huriez syndrome (scleroatrophic changes on the dorsal aspect of the hands and hypoplastic nails in addition to the PPK). Secondary dermatophyte infection, leading to maceration and peeling of the palms and soles, is a common complication. These conditions persist for life and may be passed on to the next generation. Treatment options include salicylic acid 4-6% in petrolatum, 50% propylene glycol in water under plastic occlusion, lactic acid- and urea-containing creams and lotions.

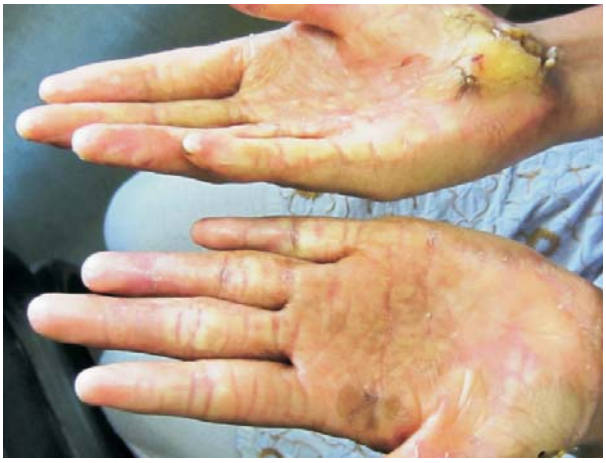

FIG. 1 *Yellowish hyperkeratotic plaques on palms.*

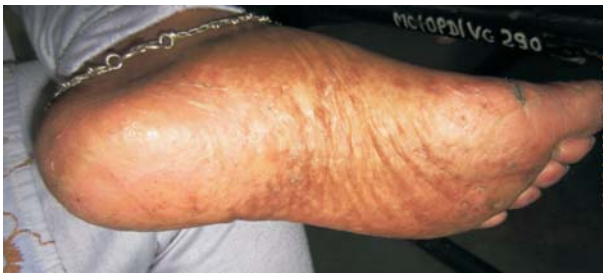

FIG. 2 *Hyperkeratosis sparing the arches of feet.*

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