Progressive and symmetric erythrokeratoderma of adult onset: A rare case

Sir,

A 28-year-old farmer presented to us with dry, scaly plaques over dorsa of hands, knees, sides of trunk and anterolateral thighs, of 6 years duration, with a progressive increase in the past 1 month. The condition initially started as dryness and fissuring of dorsa of hands, followed by scaling. The lesions used to heal with hypopigmentation. He also developed diffuse scalp scaling and itching over the lesions in the past 1 month. He did not give any history of oozing, vesiculation, pustulation, photoaggravation, allergy to fertilizers or plants, or any systemic complaint. He used to consume alcohol occasionally. There was no similarly affected member in the family. He did not take any long-term oral medication prior to the onset of the lesions. He was a product of non-consanguineous marriage, with normal birth history and milestones.

Systemic examination was normal. Skin examination revealed dry, scaly, well-demarcated plaques with fissures

over dorsa of both the hands, elbows, knees, ankle, and shin [Figures 1 and 2], associated with palmoplantar keratoderma. There were less demarcated and less scaly plaques over both axillae and scapular regionswith diffuse scaling over scalp, waist, and buttocks. We came to a differential diagnosis of pityriasis rubra pilaris, psoriasis and progressive symmetric erythrokeratoderma; an initial diagnosis of pellagra was later abandoned in view of his detailed history. All routine blood investigations were within normal limits, and a 4 mm skin punch biopsy specimen helped us to rule out psoriasis and pityriasis rubra pilaris and clinch the diagnosis to symmetric progressive erythrokeratoderma. The H&E stained biopsy section showed hyperkeratosis, parakeratosis and acanthosis with mild spongiosis and a patchy lymphohistiocytic infiltrate in the upper dermis, which was consistent with progressive symmetric erythrokeratoderma (PSEK) [Figure 3]. He was started on Acitretin 25 mg/day perorally, and he improved considerably on follow-up at 1 month.

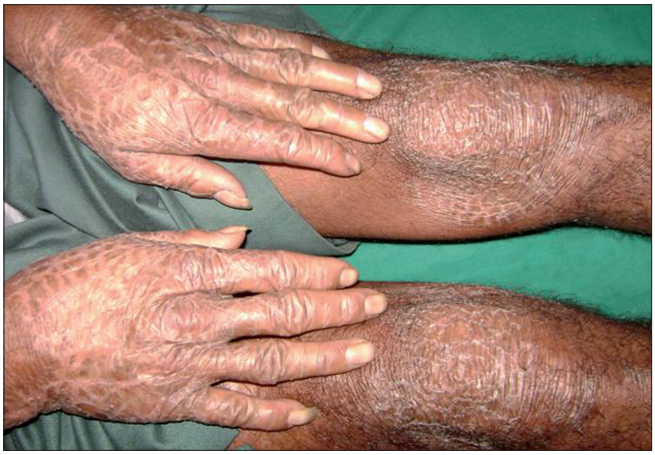

Figure 1: Erythematous hyperkeratotic scaly plaques over knees and dorsae of hands

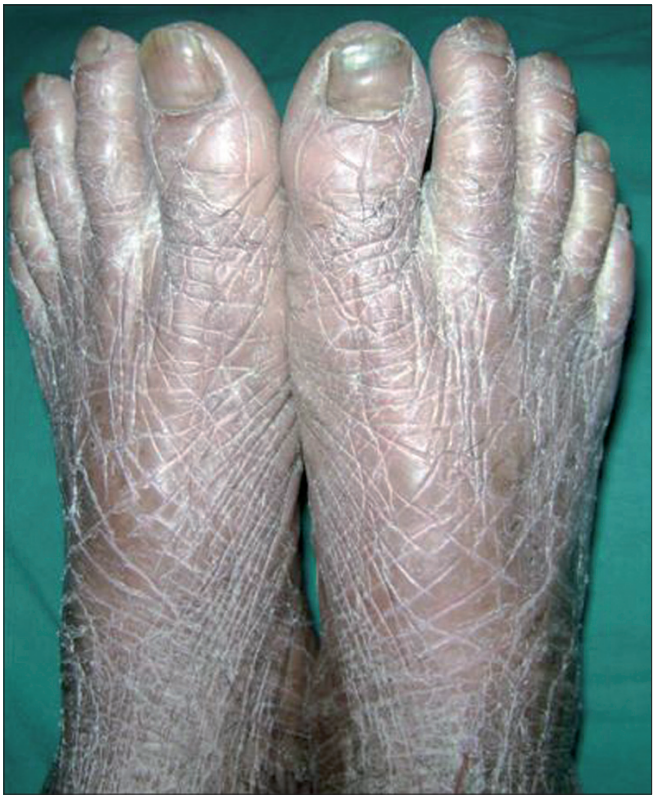

Figure 2: Dorsae of feet showing erythematous hyperkeratotic plaques with dystrophic toe nails

The erythrokeratodermas are a rare group of genodermatoses, characterized by well-defined hyperkeratotic erythematous plaques, either constant or migratory. There are three well-recognized clinical types, all occurring in childhood with varying genetic mutation and clinical presentations: erythrokeratoderma variabilis, erythrokeratoderma en cocardes, and PSEK. All have similar, nonspecific histopathology showing hyperkeratosis, parakeratosis, acanthosis, and papillomatosis.[1] Progressive symmetric erythrokeratoderma or Gottron's disease is a rare autosomal disorder of cornification with variable penetrance associated with frame shift mutations in the loricrin gene, characterized by epidermal hyperproliferation, which was first reported by Darrier in 1886 and further elucidated by Gottron in 1922. Since then, less than 50 cases have been reported in the literature.[2,3] It is usually seen in first decade as well-demarcated erythematosquamous plaques symmetrically distributed over head, cheeks, shoulder girdle, and buttocks, along with limited plaques over ankles and wrists.[1] The disease progresses over the next few years and then remains stable over time, with morphology, color, and site remaining constant.[4] Positive family history can be obtained in only about 50% patients and rest of the cases are due to spontaneous insertional mutation in Loricrin gene. The diagnosis is invariably clinical, as loricrin gene analysis is difficult in hospital settings and microscopy is nonspecific at best. The treatment includes systemic and topical retinoids, keratolytics and glucocorticoids, which have variable efficacy.

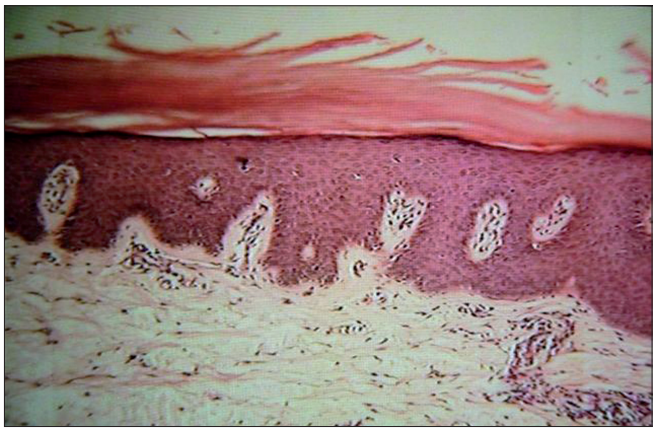

Figure 3: Photomicrograph showing hyperkeratosis, parakeratosis, and acanthosis with mild spongiosis of epidermis, and a patchy lymphohistiocytic infiltrate in the upper dermis (H&E, ×10)

The peculiarity of this case is its relatively late onset, which has not been mentioned in literature, though an adult onset case of erythrokeratoderma variabilis resistant to oral retinoid therapy has been reported.[5]

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