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SYMMETRICAL PROGRESSIVE ERYTHROKERATODERMA

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A 6-year-old boy had gradually progressive, asymptomatic, well-defined, bilaterally symmetrical, erythemato-hyperkeratotic plaques over dorsa of hands, feet, shins, popliteal fossae, natal cleft, both axillae and anterior aspect of abdomen, suggestive of symmetrical progressive erythrokeratoderma. Palms and soles were spared. Involvement of multiple flexural areas in addition to the classical sites was unusual.

Key words : Erythrokeratoderma, Progressive, Symmetrical.

Symmetrical progressive erythrokeratoderma (SPE) or Gottron's syndrome is a rare autosomal dominantly inherited genodermatosis.[1] The disease usually begins in infancy but onset may be delayed till adult life. It manifests as bilaterally symmetrical sharply demarcated, asymptomatic plaques of erythema with hyperkeratosis and scaling, situated over the feet, front of shins, back of hands and fingers. At times thighs, upper arms, shoulders, neck and face may also be involved.[2] In a few cases, Koebner phenomenon is also observed.[3] We are reporting a case of symmetrical progressive erythrokeratoderma who had unusual distribution of lesions.

Case Report

A 6-year-old male child had gradually progressive, well-defined erythematous hyperkeratotic plaques for the past four years. The plaques first appeared on the dorsum of hands, feet and shins, and subsequently involved popliteal fossae, natal cleft, both axillae, neck and anterior aspect of abdomen (Figs. 1 and 2). Palms and soles were free. Hair, teeth and nails were normal. Auspitz's sign and Koebner phenomenon were negative. Repeated scrapings and culture for fungus were negative.

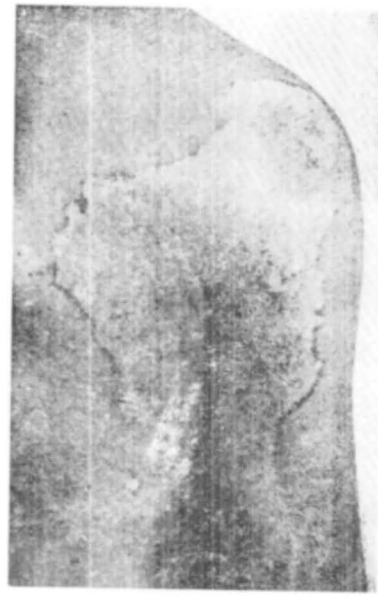

Fig. 1. Hyperkeratotic plaques involving axilla.

Out of three brothers, one had similar illness at the age of 4 years which disappeared spontaneously within 2-3 years.

Histopathological examination revealed marked hyperkeratosis, acanthosis and patchy parakeratosis. Granular cell layer, basal cell layer and dermis were essentially normal.

Comments

Early onset, positive family history, bilateral, symmetrically distributed, asymptomatic erythemato-hyperkeratotic plaques suggest the diagnosis of symmetrical progressive erythrokeratoderma.

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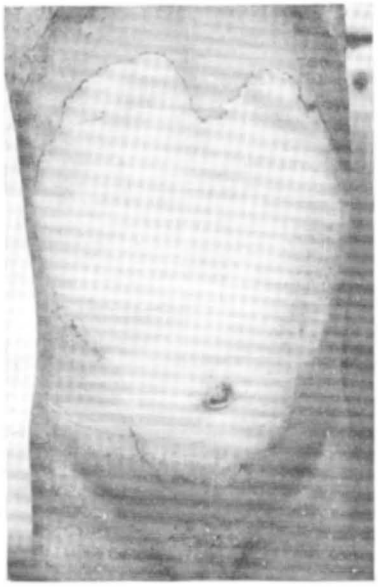

Fig. 2. Hyperkeratotic plaques involving anterior aspect of abdomen.

Relatively widespread distribution and involvement of the multiple flexural areas in addition to the classical sites[4-6] was unusual in this case.

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