Ind J Dermatol Venereol Leprol 1995; 61 : 116-7

PROGRESSIVE SYMMETRIC ERYTHROKERATODERMA

A Ghorpade, C Ramanan

Progressive symmetric erythrokeratoderma (PSEK, Gottron's Syndrome) in a 10 year-old-boy is reported. The diagnosis was confirmed by histopathology.

Key Words: Erythrokeratoderma, Symmetric progressive erythrokeratoderma, Gottron's syndrome

Introduction

Gottron's syndrome is a rare cornification disorder inherited as an autosomal dominant trait with variable penetrance and characterized by rapid epidermal cell proliferation.[1] There are scanty reports of this condition in Indian literature.

Case Report

A 10 year-old-boy presented to us with history of skin lesions on both the knees, elbows, hands and feet. The asymptomatic lesions had started about 5 year ago and had gradually increased in size initially for one year, after which they became stable. There was no family history of similar lesions and no seasonal variation. Cutaneous examination revealed symmetrical, well defined, erythematous, raised plaques with fine scales on both the knees and dorsa of hands (Fig. 1). There were similar lesions on both the upper arms, extensor aspect of elbows, and lateral malleoli. There was no palmoplantar keratoderma, and no lesions on trunk. The scalp, nails and mucous membranes were free. The systemic examination was normal. Histopathology from the plaque over knee revealed hyperkeratosis, focal parakeratosis, an intact granular layer, acanthosis (Fig. 2) and mild chronic inflammatory infiltate in upper dermis around blood vessels. Some of the prickle cells had perinuclear vacuolation. The patient improved with topical keratolytic ointment but the lesions recurred after

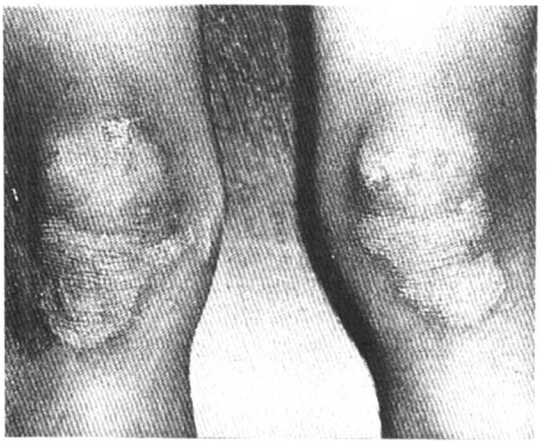

Fig. 1. Erythematous, hyperkeratotic, sharply defined plaques distributed symmetrically on the knees.

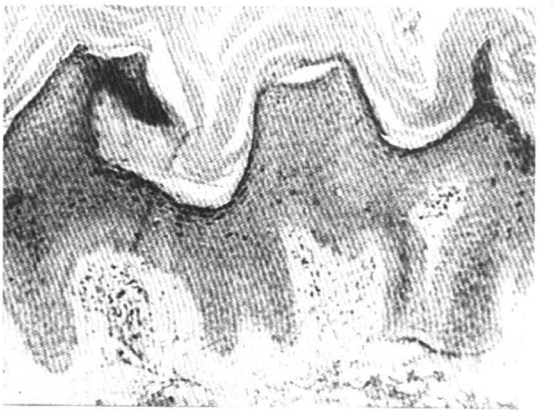

Fig. 2. Histopathlology showing hyperkeratosis patchy parakeratosis, intact granular layer, acanthosis and superficial dermal inflammatory infiltrate (H&E x100).

From the Department of Dermatology & Venereology JLN Hospital & Research Centre, Bhilai Steel Plant, Bhilai - 490006, India.

Address correspondence to : Dr Ashok Ghorpade 1B/Street 13/Hospital Sector, Bhiliai-490006.

discontinuing treatment.

Discussion

In PSEK, the symmetric, finely scaly, erythematous plaques start in early childhood and are stable after initial progression. The lesions commonly involve shoulder girdle, buttocks and face, ankles and wrists. Palmoplantar keratoderma may be seen in half the cases and in some, the lesions regress after puberty. Both the sexes are affected equally and the general health is good.[2] Our clinical impression was confirmed by the classical histopathological features in our patient.

Erythrokeratoderma variabilis (EKV) which may resemble PSEK differs from the latter, as the lesions of EKV continuously changes and may be induced by external mechanical pressure and temperature changes. The lesions may improve in summer and aggravate during pregnancy.[4] In addition, the lesions in EKV tend to involve abdomen and thorax also, unlike PSEK, EKV may be associated with deafness, physical retardation and peripheral neuropathy.[5] Localised pityriasis rubra pilaris differs from PSEK, in that the former shows follicular change as follicular erythema and keratosis.[2] The treatment is mainly symptomatic. However, oral retinoids have also been found effective.[2,5]

References

1. Hopsu-Havu VK, Tuohimata P. Erythrokeratodermia congenitalis progressive symmetrica (Gottron): An analysis of kinetics of epidermal cell proliferation. Dermatologica 1971; 142: 137-44.
2. Nazzaro V, Bardon CB. Progressive symmetric erythrokeratodermia: Histological and ultrastructural study of patients before and after treatment with etretinate. Arch Dermatol 1986; 122:434-40.
3. Gewritzman GB, Winkler NW, Dobson RL. Erythrokeratodermia variabilis. A family study. Arch Dermatol 1978;14: 259-61.
4. Beare JM, Nevin NC, Frogatt P, et al. Atypical erythrokeratodermia with deafness, physical retardation and peripheral neuropathy. Br J Dermatol 1972; 87: 308-14.
5. Tamoyo L, Ruiz-Maldonado R. Oral retinoid (RO 10-9359) in children with lamellar ichthyosis, epidermal hyperkeratosis and symmetrical progressive erythrokeratodermia. Dermatologica 1980; 161 : 305-14.