CASE REPORTS

ERYTHROKERATODERMIA PROGRESSIVUM SYMMETRICUM

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A case of progressive symmetric erythrokeratodermia (PSEK) in an Indian male is being reported. He presented with slowly progressive bilateral symmetrical plaques with peripheral rim of erythematous halo, which were limited only to the limbs. Koebner's phenomenon was positive. Family history was negative. Histopathology ruled out psoriasis and confirmed the diagnosis of PSEK.

Key words: Erythrokeratodermia, Keratinization

Erythrokeratodermias (EK) are a heterogenous group of inherited cornification disorders.[1] EK were classified as (i) confluent and reticular EK of Gougerot and Carteaud, (ii) variable EK of Mendes Da Costa, and (iii) symmetric progressive EK.[2] Progressive symmetric erythrokeratodermia (PSEK) was first reported by Darier in 1911.[3] PSEK is characterised by bilateral symmetrical hyperkeratotic plaques with sharp borders, surrounded by narrow (5-8 mm) erythematous halo on knees, elbows, palms, soles, back of hands and feet.[4] Familial cases have not been reported. Onset is between early childhood and adolescence.[4] Koebner's phenomenon was elicited in PSEK.[5] Histopathology reveals hyperkeratosis with scattered mounts of parakeratosis, preserved granular layer, absence of suprapapillary thinning and proliferation of papillary vessels accompanied by scant diffuse/perivascular infiltrate.[4]

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Case Report

A 45-year-old male had moderately itchy, slowly progressive bilateral symmetrical, large, well-defined, purplish-brown plaques covered with fine scales over dorsa of hands, wrists, feet and lower legs since 20 years. Plaques had peripheral rim of erythema. Koebner's phenomenon was positive as plaques extended proximally in the form of multiple broad bands seen on medial side of forearms and on anterior, lateral and back of lower legs (Fig.1). Plaques around ankles showed accentuation of skin creases and a few scattered flat/verrucous papulas. Patient had atopic constitution as he had nasobronchial allergy, photosensitivity, recurrent urticaria and early bilateral cataract which was operated 3 years back. Past history of seborrhoea capitis was present. Family history of atopy and ichthyosis

was negative.

Hair, nails, teeth, palms and soles were normal. Routine investigations were within normal limits. Biopsy showed massive hyperkeratosis with a few mounts of parakeratosis, delling filled with keratin and marked acanthosis with upward extension of epidermis which at places enclosed keratin. Stratum granulosum was normal. Epidermal cells showed vacuolisation. Papillomatosis without suprapapillary thinning was observed. Upper dermis showed mild to moderate perivascular mononuclear infiltrate.

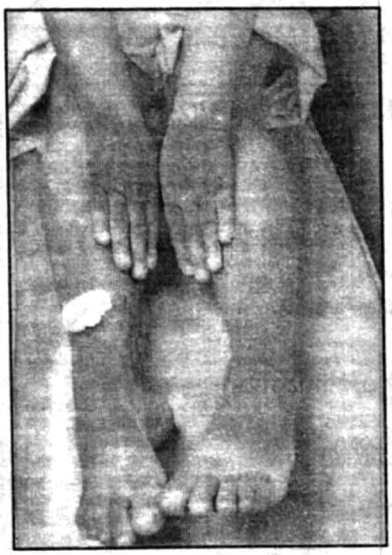

Fig.1. Typical large plaques with peripheral erythematous halo and multiple linear bands representing positive Koebner's.

Discussion

PSEK is a very rare type of EK characterised by plaques limited to limbs, late onset as compared to other 2 types of EK and negative family history. In our patient onset was at the age of 25, family history was negative, koebner's phenomenon was positive, typical plaques progressed very slowly and were limited only to dorsa of hands, feet and adjoining areas of limbs. Histopathology helps in diagnosis of PSEK and to exclude psoriasis.

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