GENETICS AND GENODERMATOSES

PROGRESSIVE SYMMETRIC ERYTHROKERATODERMIA: CASE REPORT

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Background: Progressive symmetric erythrokeratodermia (PSEK) includes both clinically and genetically heterogeneous group of disorder characterized by well-demarcated erythematous and hyperkeratotic plaques, symmetrically distributed over the extremities, buttocks, face and the trunk. The treatment of PSEK is generally unsatisfactory. The cases are preferably treated with topical retinoids, calcipotriol, emollients, keratolytics and topical corticosteroids with limited or no success. There has been reaported some cases in which oral retinoids were used with success in patients with PSEK. We report here a typical case of PSEK treated with Acitretin because of its rarity in dermatological literature and its satisfactory response to the Acitretin treatment.

Observation: A 20-year-old woman presented with dry skin and persistent erythematous, hyperkeratotic plaques. She had a history of dry skin since infancy. The patient developed persistent erythematous, hyperkeratotic plaques at the age of 17.Dermatologic examination revealed dry skin and symmetric, hyperkeratotic,erythematous plaques on the dorsal side of hands, forehands, elbow, neck, and trunk. She had also palmoplantar keratoderma.Skin biopsy revealed marked hyperkeratosis, irregular acanthosis, papillomatosis in some areas and perivascular lymphocyte infiltration. Topical treatment of emollients, keratolytics and topical corticosteroids were ineffective. The patient was given Acitretin at the dose of 10 mg/day (0.03 mg/kg) for 2 months and showed a significant improvement.

Key Message:We report here a rare case of PSEK who responded well to Acitretin treatment with no significant side-effect. Therefore,Acitretin can be evaluated as a potential therapeutic alternative for those PSEK patients who are unresponsive to the other topical treatment options.