H.K. Dermatol. Venereol. Bull. (2004) **12**, 170-171

Dermato-venereological Quiz

LY Chan 陳來源

A 16-year-old Chinese girl presented with mildly itchy persistent rash over dorsa of her feet for four years. The rash began from her right big toe and increased in size progressively, causing difficulty in moving her ankle joints. She received various topical steroids and systemic antifungals from doctors but the response was poor. Her past health was good with normal developmental milestones. Her mother had similar rash over both legs since childhood. Physical examination revealed well-demarcated scaly erythematous plaques over her ankles, dorsal aspects of both feet, heels and soles (Figures 1 & 2).

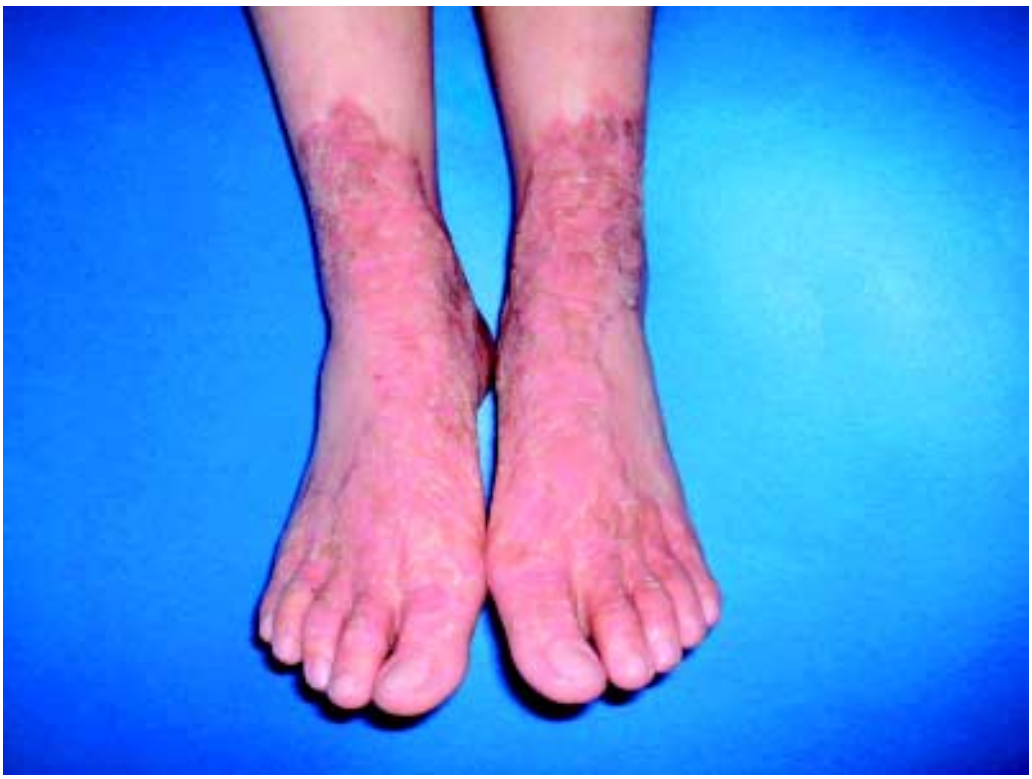

Figure 1.

Private Practice

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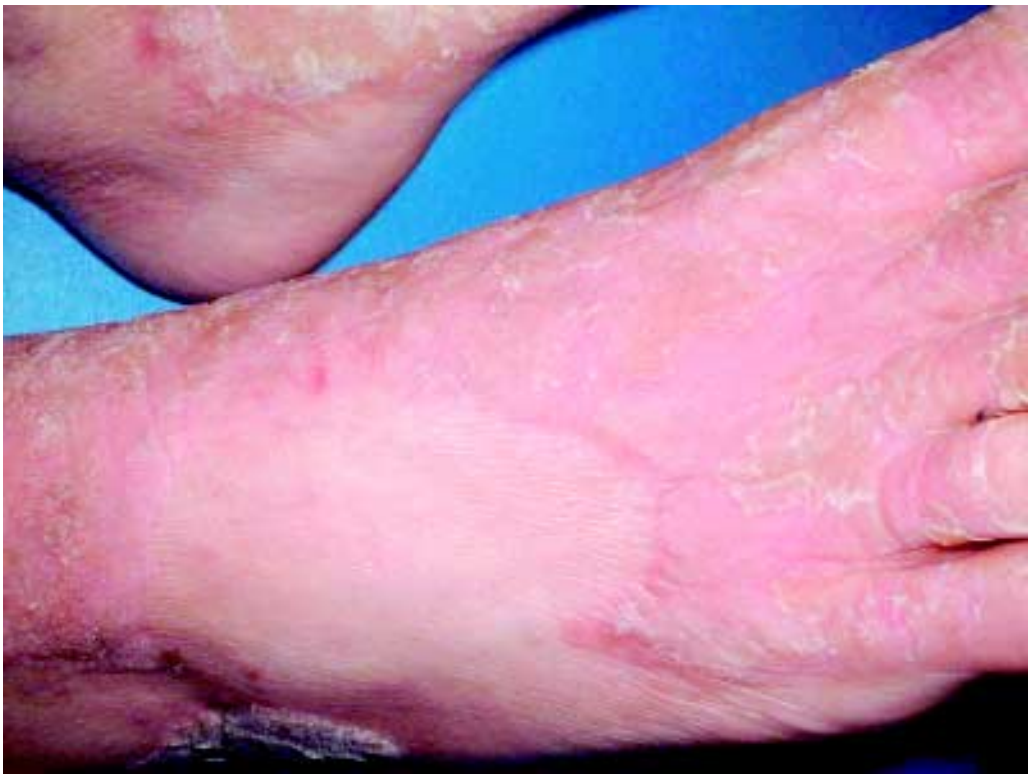

Figure 2. Close-up view.

Questions

1. What are the clinical differential diagnoses?
2. In our patient, the skin scraping for fungal microscopy and culture was negative. Skin biopsy showed features compatible with chronic eczema. What is the diagnosis?
3. What are the major clinical types of this condition?
4. What is the underlying genetic defect for this condition?
5. What is the management?

(Answers on page 180)

Answers to Dermato-venereological Quiz on pages 170-171

1. The clinical differential diagnoses include psoriasis, chronic eczema, fungal infection, and erythrokeratoderma.

2. With a positive family history, poor response to topical steroids, negative fungal culture and non-specific histologic features, the most likely diagnosis is erythrokeratoderma; progressive symmetric erythrokeratoderma (PSEK) type.

3. Erythrokeratodermas are a group of rare genodermatoses with well-demarcated hyperkeratotic and erythematous plaques. There are two major clinical types: erythrokeratoderma variabilis and PSEK. The histopathologic features of erythrokeratoderma are non-specific and included acanthosis and papillomatosis with compact hyperkeratosis, basketweave hyperkeratosis, and parakeratosis. Palmoplantar keratoderma and neurologic defects may be present in erythrokeratoderma.

4. PSEK is an autosomal dominant condition with incomplete penetrance and is characterised by non-migratory, hyperpigmented, symmetric, hyperkeratotic plaques on the extremities, buttocks, and face. Studies of large affected family have found mutation in loricrin gene in PSEK. Loricrin is the major constituent of the epidermal cornified cell envelope.

5. Topical keratolytics, retinoids, emollients and steroids produce variable response. Systemic retinoid has been reported to be useful. There was marked improvement in our patient after taking acitretin.