Case 8

A 27-year-old Thai woman, from Surajthani

Chief complaint Generalized pruritic hyperkeratotic papules on face, neck, upper chest and back for 10 years

Present illness Ten years ago, she had pruritic hyperkeratotic papules on face,neck,upper chest and back. Her skin lesions were exacerbated by sunlight and heat. She also had nails abnormality.

Past History She is healthy She had no history of any disease.

Family History No one in her family has the same skin lesions.

Physical examination

Skin: Generalized hyperkeratotic erythematous to brownish papules on face, neck, upper chest and back

Oral mucosa: Multiple cobblestone-like papules on hard palate

Nails: Multiple longitudinal red and white bands ,V-shaped nicks on right index finger.

Otherwise are unremarkable

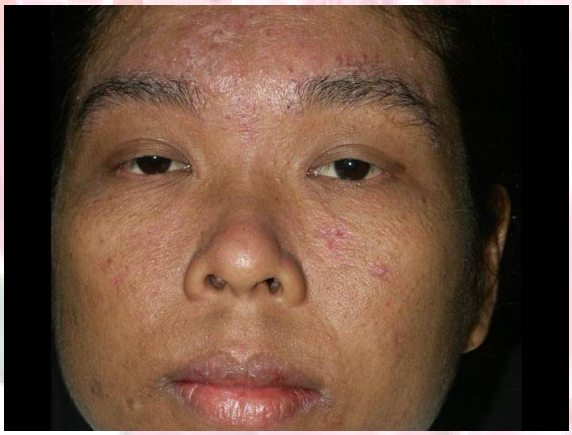

Fig. 1

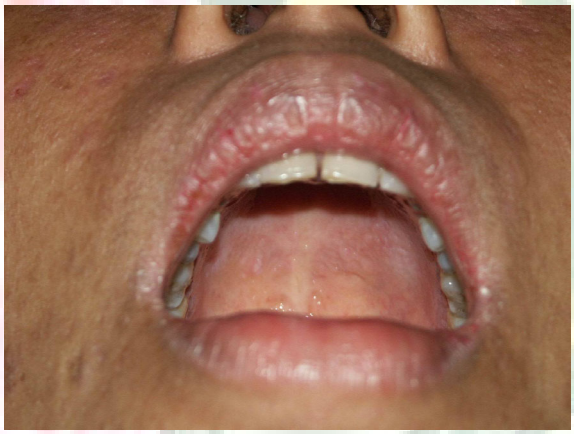

Fig. 2

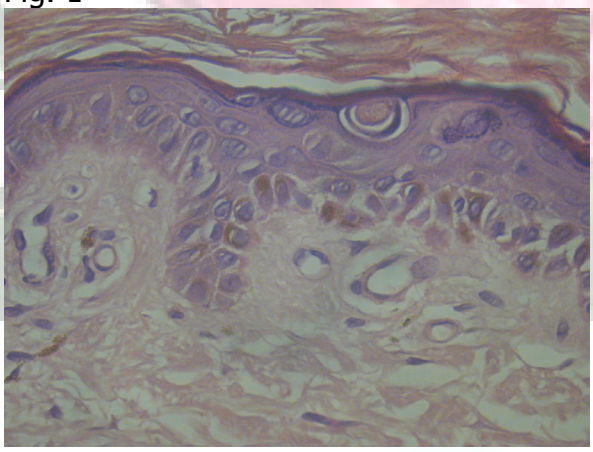

Fig. 3

Diagnosis Darrier's disease

Treatment Neotigason 25 mg/day
10% urea cream/ Triamcinolone cream

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Discussion

Darrier's disease is a dominantly inherited skin disease. The disease is characterized by hyperkeratotic papules,predominantly on the upper trunk and scalp.The molecular

defect is associated with 12q23-24.1 (ATPA2 gene,encoding the sarco/endoplasmic reticulum CA2+ATPase type 2 isoform,SCRCA2).

Histopathologic and ultrastructural revealed breakdown of the desmosome-keratin filament complex between keratinocytes,loss of cohesion between suprabasal epidermal cells.

Immunocytologic studies have demonstrated that the major desmosomal proteins and epidermal keratins are present within clinically uninvolved and involved epidermis.

The onset is usually at first and second decade. Darrier's disease is a chronic and unremitting for most patient.Topical therapy sometimes provided relief of symptoms but had no effect on the progress of the disease.Oral retinoids were effective ,but long-term therapy was tolarated poorly.

Reference

1. Burge SM ,Wilkinson JD,Darrier-White disease; review of clinical features in 163 patients.J Am Acad Dermatol1992;27:40-50.
2. Kastl J, Horacek J.Problem of concomitant occurence of Hopf's acrokeratosis verruciformis and Darrier's disease.Cesk Dermatol.1994;49:387-91.
3. Marron Gasca J,Darrier's disease with an extensive hypertrophic component. Actas dermofisiliogr1995;66:131-42.