Case 14

A 17-year-old-Thai woman from Nakhonsawan.

Chief complaint: Asymptomatic tiny papules on face, chest, and back for 8 years.

Present illness: The patient developed multiple brownish hyperkeratotic papules on face, neck, upper chest, and back since she was 9 years old. The lesions were not pruritic. She noticed that the rashes worsened by sweating, heat, and sunlight. She was treated with topical steroid without improvement.

Past history: She was previously healthy.

Family history: None of her family member has similar skin lesions.

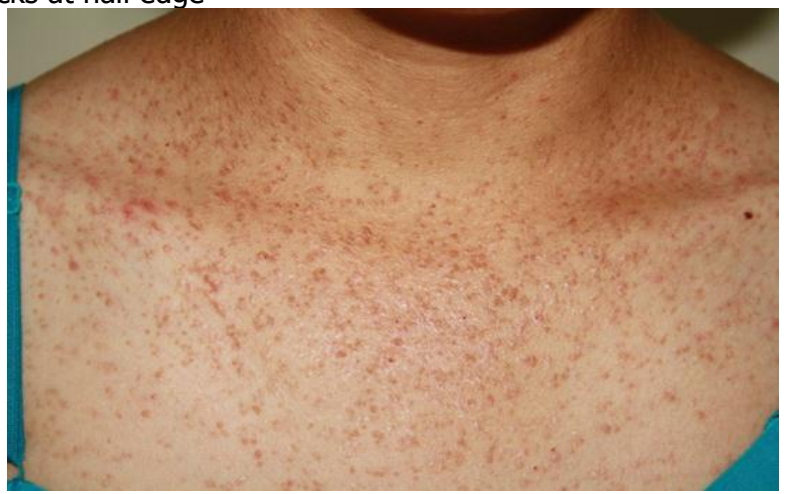
Skin examination

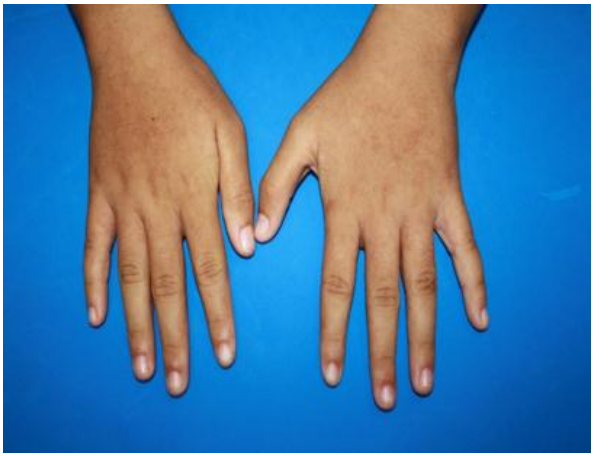
Skin: -Multiple discrete brownish hyperkeratotic papules on face,neck, upper chest, and back

-Multiple skin-colored flat-topped papules on both dorsum of hands and both palms

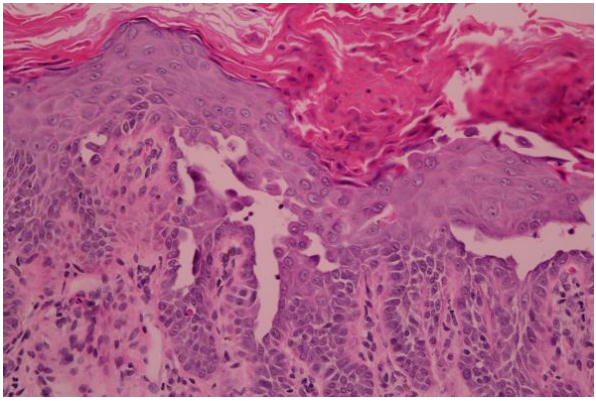
Oral mucosa: Multiple whitish papules on hard palate

Nails: Multiple red and white longitudinal bands, V-shaped nicks at nail edge





Other physical examination: unremarkable
Histopathology: (S13-015880, lateral neck)



• Scale-crust, hyperkeratosis, hypergranulosis and papillate epidermal hyperplasia with focal acantholytic dyskeratosis (FAD)
• Dense inflammatory-cell infiltrate of lymphocytes in the superficial dermis
Diagnosis: Darier disease
Treatment: Acitretin 25 mg/day, 10%urea cream bid
Presenter: Kulsupa Nimmannitya, M.D.
Consultant: Vasanop Vachiramon, M.D.

Discussion

Darier disease is a genetic disorder of keratinization with autosomal dominant inheritance, caused by mutation in the ATP2A2 gene resulting in sarcoplasmic/endoplasmic reticulum calcium ATPase 2 (SERCA2) defect that exhibits impaired epidermal cell-to-cell adhesion and altered differentiation.[1, 2] Clinical features are usually in the form of greasy, crusted, keratotic yellow-brown papules found particularly on seborrheic areas of the body, dystrophic nails, palmo-plantar pits, and papules on the dorsum of the hands and feet. Oral and esophageal involvement have been described.[3, 4] A case of basal cell carcinoma developing in a patient with type 2 segmental Darier disease has been reported, however, malignant transformation is rare.[5] A number of clinical studies have also described the co-occurrence of various neurological and psychiatric symptoms with Darier disease, including mood disorders, epilepsy, mental retardation and a slowly progressive encephalopathy.[6]

Darier disease is a chronic and unremitting disease for most patients. It is commonly misdiagnosed as seborrheic dermatitis or eczema.[7] Histologically, the lesions present suprabasal clefts in the epithelium with acantholytic and dyskeratotic cells.[3]

The disease is frequently aggravated by sun exposure therefore sunscreen and sun avoidance are essential. One report suggests that sunscreen and topical ascorbic acid would be very helpful in preventing the aggravation of Darier disease caused by sun exposure.[8] Patients with mild Darier disease may find topical isotretinoin helpful, but it is likely that most patients with widespread disease will require treatment with systemic retinoids.[9] Oral retinoids are the most effective treatment but their adverse effects are troublesome. Topical retinoids, topical corticosteroids, surgery, and laser surgery have their advocates but evidence for efficacy is sparse.[10] Other topical agents such as adapalene, 5-fluorouracil, or

tacrolimus have been reported to be effective in small numbers of cases.[11-13]

References

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