

Darier Disease

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A 60-year-old woman presented with a generalized pruritic rash since adolescence. She reported her son had a similar rash. Physical examination revealed greasy brown and keratotic papules and plaques in a seborrheic distribution, which included her chest, back, forehead, scalp margins, and ears (Figures 1 and 2). Histopathology revealed acantholytic dyskeratosis. The clinicopathological features along with the patient's family history were consistent with Darier disease. The patient's skin disease markedly improved with acitretin.

Darier disease, also known as keratosis follicularis, is an autosomal dominant disorder affecting the *ATP2A2* (ATPase sarcoplasmic/endoplasmic reticulum Ca^{2+} transporting 2) gene and typically manifests during adolescence. Clinically, the disease presents with greasy hyperkeratotic papules and plaques in



FIGURE 2. Keratotic papules and plaques affecting the back.



FIGURE 1. Greasy papules and plaques affecting the scalp margin, neck, and ear.

a seborrheic distribution, along with palmo-plantar pits and acral keratosis.^{1,2} Skin lesions may be pruritic, painful, and/or malodorous. Additionally, characteristic nail findings include alternating white and red longitudinal streaks (candy-cane nails) and V-shaped nicking of the free nail edge. Oral involvement can also be seen with cobblestoning of the oral mucosa, gingival hypertrophy, and obstructive sialadenitis.^{3,4} The diagnosis can be confirmed through a skin biopsy revealing acantholytic dyskeratosis in the appropriate clinical setting, although in some instances, genetic testing may be required.

Treatment goals include patient education on exacerbating factors such as ultraviolet light, heat, and friction. Additionally, patients are susceptible to superimposed viral and bacterial skin infections; thus, surveillance,

prevention, and treatment of infection is paramount. Although there are no curative treatments of Darier disease, topical corticosteroids and systemic retinoids may be used to control inflammation and hyperkeratosis, respectively.²⁻⁴ Benefits from surgical interventions have also been reported.^{1,4}

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