

## PhotoDermDiagnosis

# **Skin-colored, waxy, hyperpigmented, crusted, keratotic papules in seborrheic areas of an adult male**

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A 35-year-old, otherwise healthy man, presented with multiple asymptomatic waxy, crusted greyish brown, hyperpigmented papules on the face, neck, chest, and upper back since adolescence. The eruption was associated with occasional itching and mild stinging. There was history of recurrent secondary staphylococcal infection treated with antibiotics. There was no family history of similar dermatosis.

On physical examination, there were multiple skin-colored, waxy, hyperpigmented, crusted, keratotic papules admixed with hypomelanotic macules predominantly affecting the seborrheic areas including the face, neck, chest, and the upper back. (**Figure 1**) The eruption was malodorous. The fingernails showed longitudinal white and red streaks, subungual hyperkeratosis, and marginal V-shaped nicking. On oral examination, multiple whitish papules were found on the palate and gums.

His routine blood and urinalysis were unremarkable.

What is your diagnosis?

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**Figure 1** Multiple greyish brown keratotic papules on neck and chest area.

## **Diagnosis**

Darier's disease

## **Discussion**

Darier's disease was first described by a French dermatologist, Jean Darier, and an American dermatologist, James C. White in 1889.<sup>1</sup> It is an autosomal dominant genodermatosis typified by the development of a peculiar mucocutaneous papular eruption of keratotic papules and onychodystrophy.<sup>1,2</sup> The cutaneous eruption predominantly affecting the seborrheic areas including the scalp, face, neck, chest, and the back. With no sex predilection, it begins to appear with the onset of puberty as small firm skin colored waxy papules which subsequently evolve into greyish brown keratotic papules and confluent plaques.<sup>1-5</sup> Palmoplantar areas may be involved in 50% of the cases as multiple focal hyperkeratotic papules and pits filled with keratin.<sup>1</sup> Multiple verrucous flat papules may appear on the dorsa of the hands and shins. In a small percentage of cases, intertriginous areas including the axillae, inframammary areas, groins, and post auricular areas may be involved where the lesions tend to become papillomatous and vegetative.<sup>6</sup> The eruption is malodorous. Multiple whitish papules may appear on the palate, tongue, and the gums.<sup>7</sup> The associated nail dystrophy consists of alternating longitudinal red and white streaks, subungual hyperkeratosis, splintering, fragility, and marginal V-shaped nicking.

The eruption is bilateral and symmetrical, but rarely acral hemorrhagic and segmental cases have been reported.<sup>1</sup> The eruption is usually worse in the summer and exacerbated with heat, sweating, and occlusion. It may also be triggered by intense UV exposure. Lithium carbonate has also been reported to exacerbate the eruption in a few cases.<sup>1,2</sup>

Histopathology shows suprabasal clefting, acantholysis, and dyskeratosis with appearance of corps ronds and grains. There is superficial hyperkeratosis and papillomatosis. A perivascular predominantly lymphocytic dermal infiltrate is present.<sup>1,2,8</sup>

The disease is inherited in autosomal dominant pattern. A mutation in the gene ATP2A2 which encodes for sarco-endoplasmic reticulum Ca<sup>2+</sup> ATPase, ATP2A2 (SERCA2). The mutation results in deficiency of the endoplasmic calcium stores which may impair normal processing of the proteins required for the cell-to-cell adhesion (desmoplakin I and II, plakoglobin, and desmoglein) ultimately resulting in acantholysis.<sup>9,10</sup>

The diagnosis is based on the clinical and histopathologic features. The differential diagnosis includes acrokeratosis verruciformis of Hopf, Grover's disease, Hailey-Hailey disease, and pemphigus vegetans.<sup>1</sup>

Darier disease is a lifelong disease with remissions and relapses. Treatment consists of daily skin care with antiseptic cleansers, light weight clothes, avoidance of sun and heat exposure, topical corticosteroids, and retinoids.<sup>1-5,11,12,13</sup> Oral retinoids are very effective but their use is limited by their adverse effects.<sup>14</sup> Intermittent therapy during the summer is more advisable than continued therapy. Cyclosporin may be effective in resistant cases.<sup>1,2</sup> Treatment with laser ablation has been found successful in segmental Darier's disease.<sup>17,18,19</sup>

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