

Hyperkeratotic Papules on the Medial Aspects of the Feet

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PRACTICE POINTS

- Acrokeratoelastoidosis is a rare autosomal-dominant genodermatosis characterized by firm yellow papules along the margins of the hands and feet.
- Most therapies generally are ineffective but have included urea, salicylic acid, and tretinoin.

To the Editor:

A 43-year-old woman with recently diagnosed diabetes mellitus and a history of thrombotic thrombocytopenic purpura on chronic oral steroids presented with a several-year history of small bumps and bilateral hyperpigmentation on the feet. On physical examination 2- to 3-mm dark brown, hyperkeratotic, firm papules were present on the medial aspects of the feet as well as the dorsal and medial aspects of the thumbs (Figure 1). There also were brown thickened firm plaques on the heels and soles of the feet.

A punch biopsy of the medial aspect of the right foot was performed (Figure 2). Microscopic examination revealed acral skin with hyperkeratosis, parakeratosis, mild hypergranulosis, mild basilar pigmentation, and mild dermal fibrosis (Figure 2A). A periodic acid–Schiff stain for fungus was negative. An elastic van Gieson stain showed fragmentation of the dermal elastic fibers (Figure 2B). The patient was diagnosed with acrokeratoelastoidosis (AKE).

Acrokeratoelastoidosis is a rare autosomal-dominant genodermatosis characterized by firm yellow papules and plaques that appear along the



Figure 1. Dark brown firm plaques and 2- to 3-mm hyperkeratotic firm papules on the lateral aspects of the feet (A) and hyperkeratotic firm papules (2–3 mm) on the dorsal and medial aspects of the thumbs (B).

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The authors report no conflict of interest.

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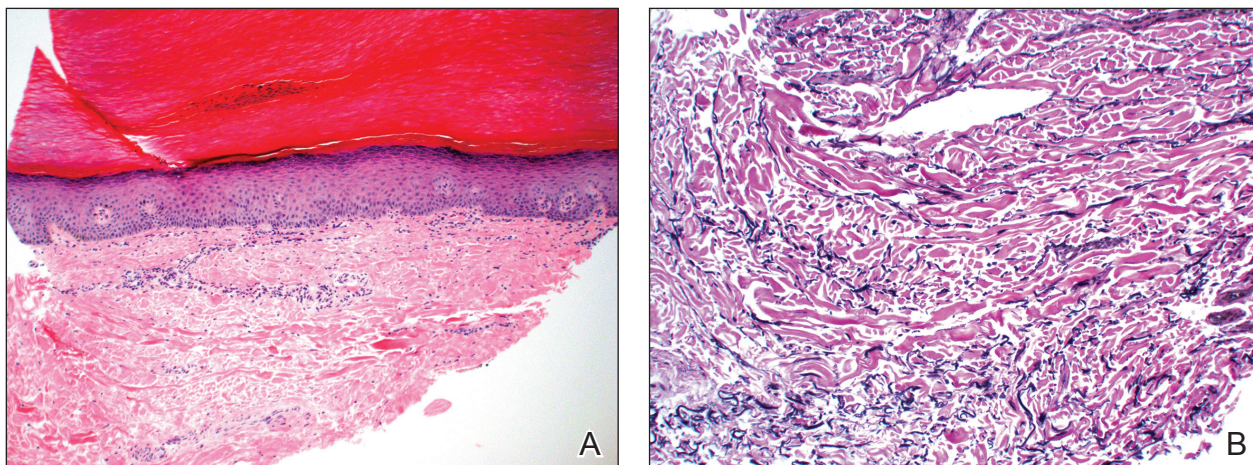


Figure 2. Histopathology showed hyperkeratosis, parakeratosis, and mild hypergranulosis (A)(H&E, original magnification $\times 4$). Mildly decreased elastic fibers with fragmentation were seen with elastic van Gieson stain (original magnification $\times 20$).

margins of the hands and feet and increase in number over time.¹ Histopathologically, hyperkeratosis with hypergranulosis and acanthosis can be seen. Elastorrhexis, resulting in fragmentation of elastic fibers within the dermis, typically is present, a feature that distinguishes AKE from focal acral hyperkeratosis.² Also, the dermis may be normal with hematoxylin and eosin stain or slightly thickened with mild depression and thin elastic fibers. There is no reported racial or sex predilection, but rapid progression of the disease during pregnancy has been observed.³

The pathogenesis of AKE is not completely understood. However, it has been implicated that abnormalities in the secretion of elastic fibers from fibroblasts may be involved in disease pathogenesis.^{4,5} Electron microscopy has demonstrated fibroblasts with dense granules at the periphery of their cytoplasm and an absence of surrounding elastic fibers. Genetic studies have linked underlying mutations in chromosome 2 to the disease.⁶ Defects in keratinization and overproduction of filaggrin also may be involved in the disease process.⁷

Most therapies generally are ineffective but have included urea, salicylic acid, prednisone, and tretinoin.⁸ Six-month treatment with etretinate 25 to 50 mg has shown promising results, though recurrences occurred with dosage reduction or

discontinuation.⁹ Our patient demonstrated mild improvement with urea cream 30%.

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