Case Letter

Plaques: A Rare Presentation of Acrokeratoelastoidosis

To the Editor:

Acrokeratoelastoidosis (AKE) is a rare disease first described by Costa[1] in 1953. Typically it is only a cosmetic nuisance in the majority of patients and presents as asymptomatic, small, firm, flesh-colored to yellowish, round to polygonal papules with occasional keratosis or umbilication on the radial and ulnar margins of the hands and/or feet.[1-3] In some cases, the lesions occur on the anterior aspects of the wrists, fingers, or lower legs.[1] The lesions are always bilaterally distributed. Acrokeratoelastoidosis is a chronic skin disorder that commonly presents during childhood or adolescence, but presentation in adulthood also has been described.[3] Histologically, AKE always shows hyperkeratosis, acanthosis, decrease of elastic tissue, and elastorrhexis of remaining elastic fibers. Plaque-type lesions are rare. We describe a patient who presented with plaques on the radial and ulnar margins of the hands.

A 36-year-old Chinese woman presented with asymptomatic, small, firm papules of 6 months' duration that initially developed on the hands and gradually increased in number, coalescing into plaques. The feet were spared. She had no medical history of hyperhidrosis, chronic trauma, friction, or excessive sun exposure, and no family history of similar symptoms. No prior therapy had been attempted.

Physical examination showed nonconfluent, firm, flesh-colored to yellowish, translucent, smooth papules with wavy edges that were symmetrically distributed on the radial and ulnar margins of the hands; some papules had coalesced into plaques (Figure 1). A biopsy specimen taken from a plaque on the hypothenar eminence of the right hand revealed focal hyperkeratosis, hypergranulosis, acanthosis, and mild chronic inflammation with hematoxylin and eosin stain (Figure 2A). Aldehyde fuchsin staining showed fragmented and rarefied elastic fibers in the reticular dermis (Figure 2B). The patient was diagnosed with AKE. Oral tretinoin 10 mg twice daily was initiated and resulted in an evident response after 2 weeks of treatment. However, the patient stopped taking the medication because of pruritus and dry skin and the lesions then reappeared.

Acrokeratoelastoidosis is a rare keratotic disorder. It seems to have no racial or ethnic predilection and occurs more frequently in women.[4,5] It also is rare in China, with few cases reported, all women.[5] The reason for the gender predilection in China remains unknown. The course is chronic, but it may rapidly progress during pregnancy.[6]

The pathogenesis of AKE is still unresolved.[2,3] Although many cases are sporadic,[5] it appears to be inherited in an autosomal-dominant fashion, most likely related to chromosome 2.[7] Typically, AKE presents as papules that are discrete and bilaterally distributed in the palmoplantar margins,[2,3] but some of the papules in our patient coalesced into plaques, which is unique. The histologic hallmarks indicated that the lesions were AKE.

The differential diagnosis of AKE includes hereditary papulotranslucent acrokeratoderma, focal acral hyperkeratosis, and keratoelastoidosis marginalis.[8] Hereditary papulotranslucent acrokeratoderma also is inherited in an autosomal-dominant fashion and shares similar acral, translucent, keratotic papules with AKE, but there is no chronic inflammatory cell infiltrate, degeneration of collagenous

Figure 1. Nonconfluent, firm, flesh-colored to yellowish, translucent, smooth papules distributed on the radial margin of the hand; some of papules coalesced into plaques.

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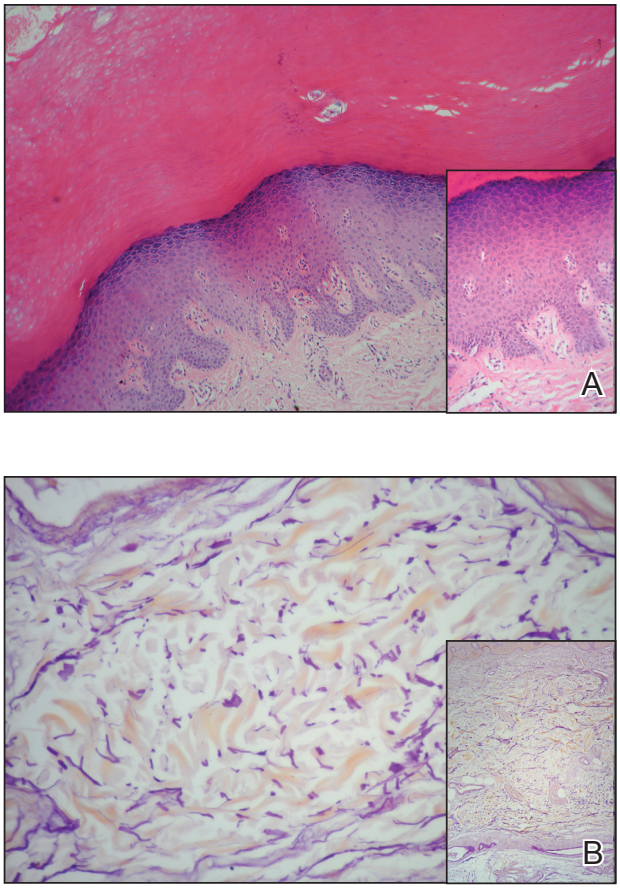

Figure 2. Histopathology revealed hyperkeratosis, hypergranulosis, acanthosis, and mild chronic inflammation (A) (H&E, original magnification ×250 [inset, original magnification ×400]), as well as fragmented elastic fibers in the reticular dermis (B) (Aldehyde fuchsin, original magnification ×400 [inset, original magnification ×100]).

fibers, or fragmentation of elastic fibers. The clinical appearance of focal acral hyperkeratosis is similar to AKE, but no changes are revealed in the elastic tissue.[9] Because AKE, focal acral hyperkeratosis, and hereditary papulotranslucent acrokeratoderma have similar lesions and overlapping histologic changes, they may be considered variants of the same entity.[4] Keratoelastoidosis marginalis, also called degenerative collagenous plaques of the hand, mainly affects white individuals aged 40 to 60 years with a history of prolonged sun exposure. Papules often are distributed over the junction of the dorsal and palmar skin and less often on the ulnar sides of the hands. The clinical lesions are similar to those in our patient, but histopathology of keratoelastoidosis marginalis shows amorphous, basophilic, elastotic masses and thickened, fragmented, calcified elastic fibers in the upper and mid dermis.

Therapies including liquid nitrogen, topical salicylic acid, methotrexate, dapsone, tar, cryotherapy, systemic prednisone, retinoic acid, clobetasone cream,[5] and erbium:YAG laser[10] have been applied. Thus far, no optimal treatment has been recommended and no tendency of spontaneous resolution has been previously reported in the literature. Our patient responded to tretinoin, but the lesions recurred after withdrawal of the medication; therefore, tretinoin may not be an optimal treatment option. Because the lesions are limited to the skin and AKE is only considered a cosmetic problem with a good prognosis, we recommend a wait-and-watch approach.

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