

Acrokeratoelastoidosis and Knuckle Pads Coexisting in a Child

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PRACTICE POINTS

- Acrokeratoelastoidosis presents as small, firm, translucent, linear papules on the ventral-dorsal palmoplantar junction.
- Acrokeratoelastoidosis does not require treatment but can be treated topically with keratolytics such as tretinoin and salicylic acid.
- Knuckle pads may respond to urea cream, salicylic acid cream, or intralesional corticosteroids.

Acrokeratoelastoidosis (AKE) is a marginal papular keratoderma that typically presents in childhood and young adulthood. Childhood cases have exhibited autosomal-dominant inheritance. Acrokeratoelastoidosis is distinct from other palmoplantar marginal papular keratodermas because of its characteristic dermal elastorrhexis with an overlying epithelial dell. We report an 11-year-old boy with multiple translucent hyperkeratotic papules in a linear fashion on the bilateral palmoplantar surfaces characteristic of AKE. He also presented with knuckle pads on the proximal and distal interphalangeal joints that rarely have been reported with AKE, suggesting that AKE and atraumatic knuckle pads may coexist.

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Case Report

An 11-year-old boy presented with atraumatic thickening of the skin on the bilateral distal and proximal interphalangeal joints of 1 year's duration. The patient also noted small bumps of unknown duration across the bilateral palms and soles with prominence on the lateral aspects. The patient previously used over-the-counter topical wart removal treatment and topical salicylic acid with minimal improvement. The patient reported no pertinent medical or surgical history, although there was a family history of Alport syndrome, predominantly in male relatives. The patient's father and paternal grandfather were noted to have similar lesions on the palms.

On physical examination, multiple pink to flesh-colored hyperkeratotic plaques were noted over the proximal and distal interphalangeal joints of the bilateral hands (Figure 1A). Upon close inspection, there were small flesh-colored and slightly translucent papules in a linear distribution on the palmar surfaces of the hands (Figure 2A) with predominance on the thenar and hypothenar eminences. The flexural creases of the bilateral wrists also revealed linear flesh-colored papules. The same small flesh-colored and translucent papules also were noted on the plantar surfaces of the bilateral feet (Figure 2B).

A biopsy was obtained from one of the small translucent papules on the left palm. Hematoxylin and eosin-stained sections revealed elevated compact orthokeratosis with an underlying central epidermal dell (Figure 3). A diagnosis of marginal papular keratoderma was made and further elastin staining was completed. Elastin stains showed marked thinning of the elastin fibers throughout the reticular dermis. Many elastin fibers in the reticular dermis demonstrated a fine arborizing pattern that normally is only evident in the papillary dermis (Figure 4). Acrokeratoelastoidosis (AKE) was diagnosed histopathologically, and knuckle pads were diagnosed clinically.

Because the patient was asymptomatic, he did not want treatment of AKE. He had marked improvement of the knuckle pads after 1 month with daily application of urea cream 10% (Figure 1B), and intermittent use was required for maintenance.

Comment

Etiology—Acrokeratoelastoidosis was first described in 1953 and is considered a type of palmoplantar marginal papular keratoderma.¹ There is overlap within the marginal papular keratodermas that makes precise diagnosis difficult within this group. The marginal papular keratodermas on the palms and soles are a group of disorders that include AKE, focal acral hyperkeratosis (FAH),

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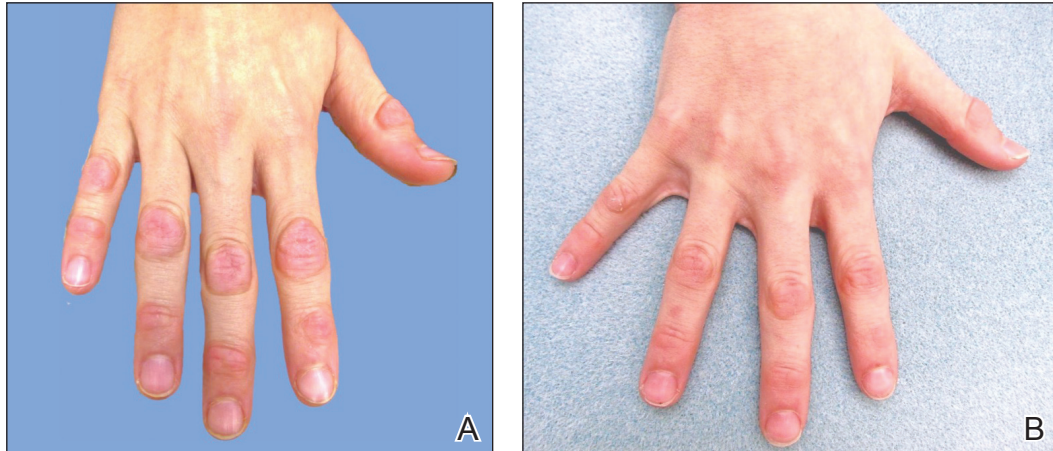


FIGURE 1. Hypertrophic knuckle pads over the proximal and distal interphalangeal joints on the right hand before (A) and after daily treatment with urea cream 10% for 1 month (B).

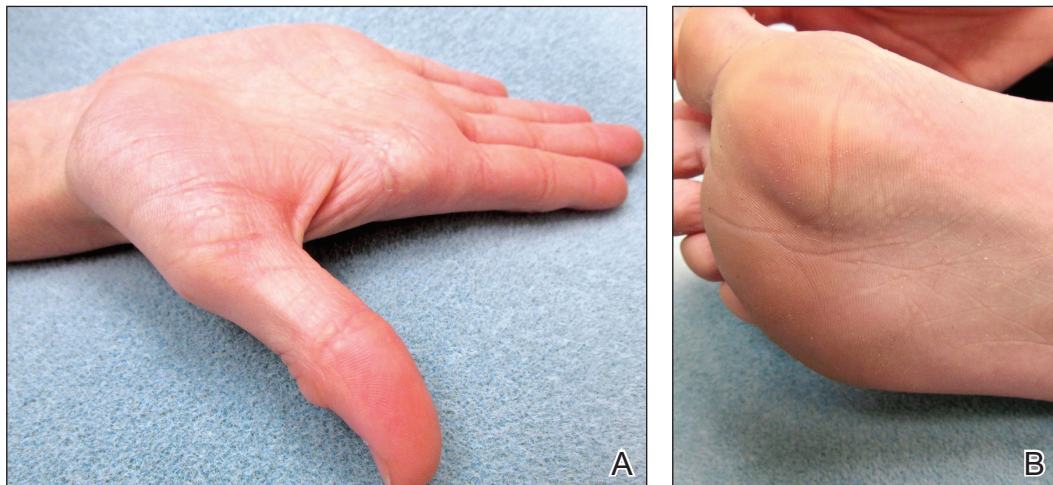


FIGURE 2. Small flesh-colored, slightly translucent papules were linearly distributed on the palmar surface of the right hand (A) and the plantar surface of the right foot (B).

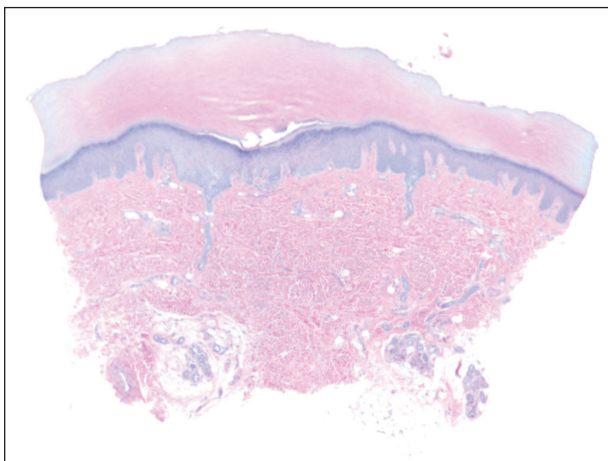


FIGURE 3. Histopathology revealed elevated compact orthokeratosis with an underlying central epidermal dell (H&E, original magnification $\times 4$).

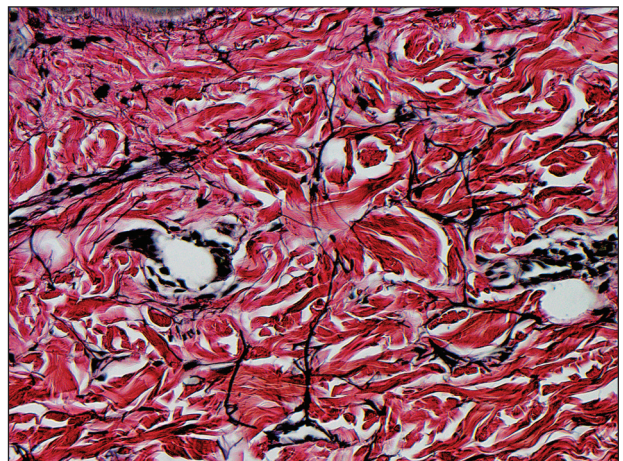


FIGURE 4. Elastin stain showed arborizing thin elastin fibers throughout the reticular dermis (original magnification $\times 40$).

mosaic acral keratosis, degenerative collagenous plaques on the hands, and digital papular calcific elastosis. These diseases are similar in clinical and histopathological features; some argue these diseases are the same entity.²

Acrokeratoelastoidosis has been hypothesized to originate from altered elastic fiber synthesis from fibroblasts.³ Because AKE is rare, most cases of common knuckle pads do not coexist with AKE; therefore, it is unknown if the underlying etiology remains the same for both entities. Unlike AKE, knuckle pads are often associated with Dupuytren contractures, repetitive trauma, or friction to the area.^{1,2}

Presentation—Acrokeratoelastoidosis is a rare disease with onset in childhood or young adulthood. Childhood cases are inherited in an autosomal-dominant fashion.¹ Adulthood onset suggests a sporadic form of inheritance. Acrokeratoelastoidosis has no gender or racial predilection.⁴ It presents over the thenar and hypothenar eminences, as well as the lateral digits, calcaneal tendon, and dorsal digits.¹ Most often, AKE occurs symmetrically along the border separating the ventral and dorsal aspects on the palms and soles. These lesions present as small, firm, translucent papules that align linearly on the ventral-dorsal palmoplantar junction in a pattern resembling paving stones.¹ Coalescence of papules into plaques has been reported. Extension of lesions to the dorsal and palmar surfaces can occur. Small circumscribed callosities may develop over the metacarpophalangeal and interphalangeal joints resembling knuckle pads.²

Histopathology—Histopathologically, AKE is distinguished by elastorrhexis—thinning, fragmenting, and rarefaction of elastin fibers—in the epidermis and reticular dermis layers.³ Acrokeratoelastoidosis also presents with orthokeratosis overlying a cuplike epithelial depression and possible epithelial acanthosis.^{2,5} Many cases exhibit hypergranulosis at the base of the epidermal dell. Dense

basophilic granules may be seen in the peripheral cytoplasm of fibroblast cells coming from the hypothesized defect in elastin secretion.^{1,3,4}

Differential Diagnosis—The main differential diagnosis of AKE is FAH. Clinically and histopathologically they appear identical; both diseases have cuplike epidermal depressions with overlying orthohyperkeratosis and prominent hypergranulosis.⁵ The elastin stains, Verhoeff-van Gieson or acid orcein stain, are imperative for distinguishing these two diseases. Although AKE demonstrates elastorrhexis and reduced elastic fibers, FAH reveals no alteration of elastic fibers. It has been suggested that FAH is a clinical variant of AKE and should be titled AKE without elastorrhexis.¹

Treatment—Acrokeratoelastoidosis is asymptomatic except for mild palmoplantar hyperhidrosis and typically does not require treatment⁴; however, the condition can be of cosmetic concern for patients. Lesions can be treated topically with keratolytics such as tretinoin and salicylic acid. A wide variety of systemic treatments including methotrexate, prednisolone, dapsone, and acitretin have been reported with variable clinical response.²⁻⁴ Copresenting knuckle pads can be treated with urea cream, salicylic acid cream, or intral-lesional corticosteroids.¹

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