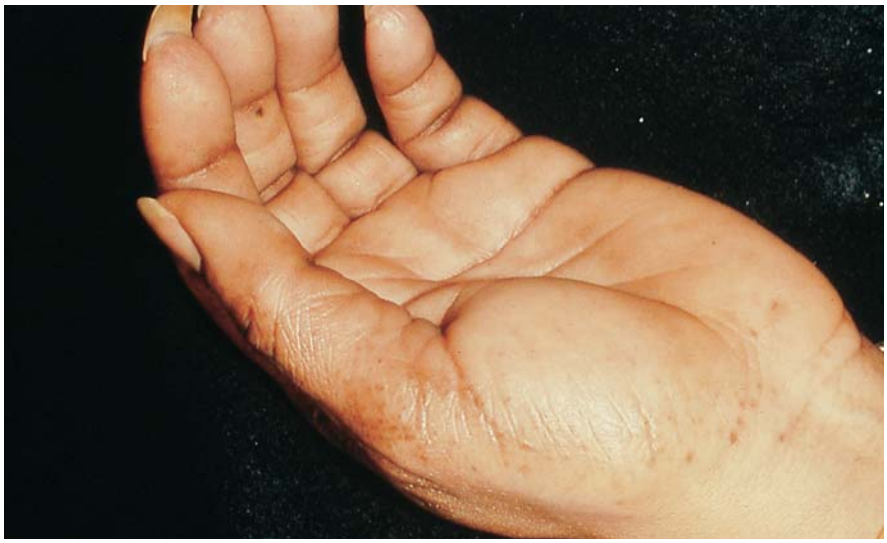
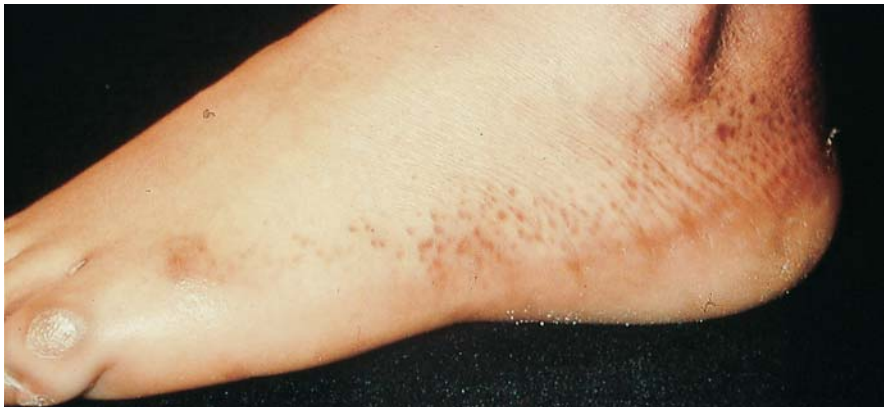
What Is Your Diagnosis?





An otherwise healthy 47-year-old black woman had a 3-year history of asymptomatic crateriform papules along the lines of transgredience on her hands and feet. Also, she had thickening and hyperpigmentation of the skin overlaying some of the metacarpophalangeal and proximal interphalangeal joints. She reported that her 22-year-old son developed similar papules on his hands and feet 2 years prior to this incident. Both the patient and her son denied hyperhidrosis of the palms and feet.

PLEASE TURN TO PAGE 29 FOR DISCUSSION

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The Diagnosis: Focal Acral Hyperkeratosis

In 1983, Dowd and colleagues[1] reported on 15 patients with focal acral hyperkeratosis (FAH), a condition characterized by oval or polygonal crateriform papules along the junction of the dorsal and palmar or plantar surfaces (lines of transgredience)(Figure 1). These papules gradually increase in number and may resemble acrokeratoelastoidosis.[1] Patients may develop thickening and hyperpigmentation of the skin associated with hyperkeratotic papules (calluslike lesions) over several of the interphalangeal joints. Discrete hyperkeratotic lesions over the heels also may be seen.[1-3]

FAH is more common in blacks and in women.[1] Most cases develop in the second and third decades of life, and patients may notice a gradual increase in lesion count over many years.[1] Although some cases of acrokeratoelastoidosis are associated with hyperhidrosis,[4] no association has been made between FAH and hyperhidrosis. Unlike acrokeratoelastoidosis, FAH involves histologic changes confined to the epidermis and exhibits focal hyperkeratosis overlaying a clavuslike depression and acanthosis[1] (Figure 2). Mild dilation of upper dermal vessels is evident; however, there is no dermal inflammation. Both collagen and elastin fibers are normal. There are no altered elastic fibers in the dermis as seen in cases of acrokeratoelastoidosis.[1]

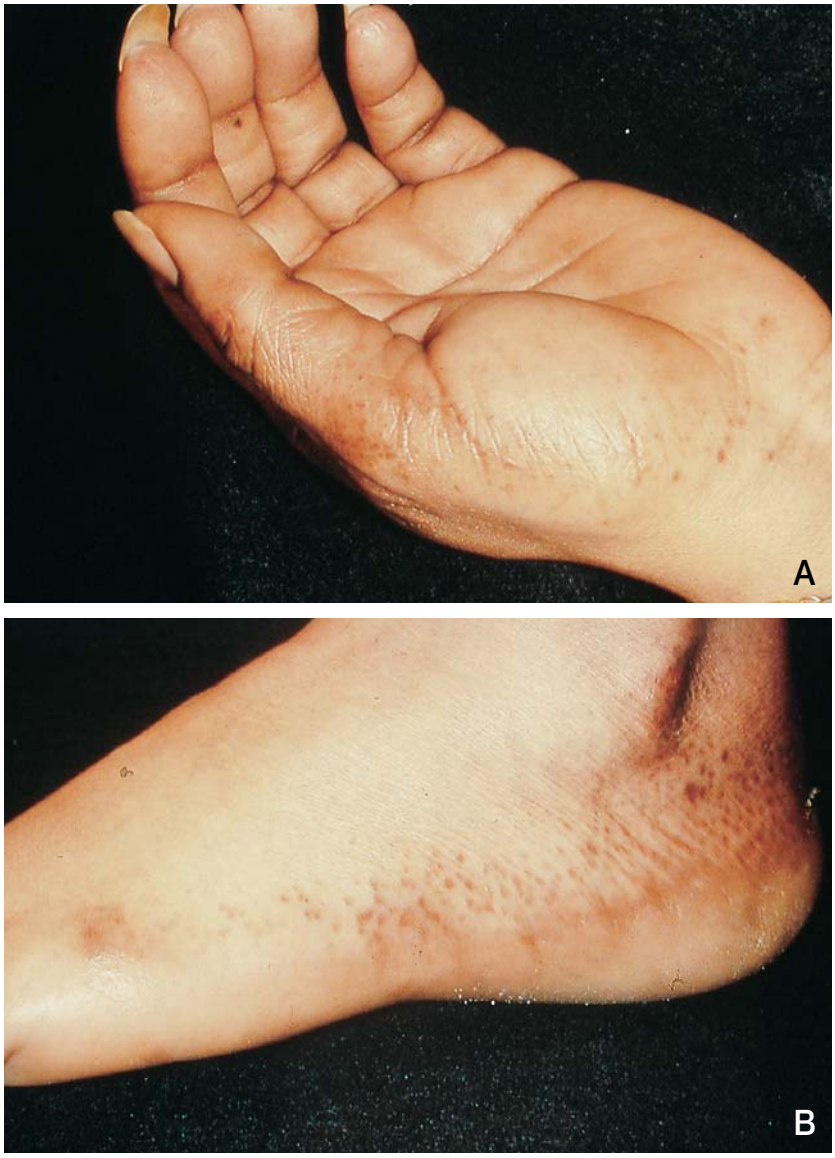

Figure 1. Oval and polygonal crateriform papules along the side of the left hand (A) and left foot (B). The left foot also has a clavuslike lesion over the fifth interphalangeal joint.

Acrokeratoelastoidosis should be included in the differential diagnosis of FAH. Acrokeratoelastoidosis has been associated with an autosomal-dominant chromosomal aberration of chromosome 2p in familial cases.[5] This condition presents as small pearly papules with a smooth surface, but occasionally these papules may be hyperkeratotic. Typically, there is involvement on the sides of the hands and feet, thenar and hypothenar eminences, and malleoli. Some patients complain of palmar or plantar hyperhidrosis.[4] The original case report on acrokeratoelastoidosis described involvement of the lower two thirds of the anterior legs or lateral margin of the wrists.[4] In contrast to FAH,

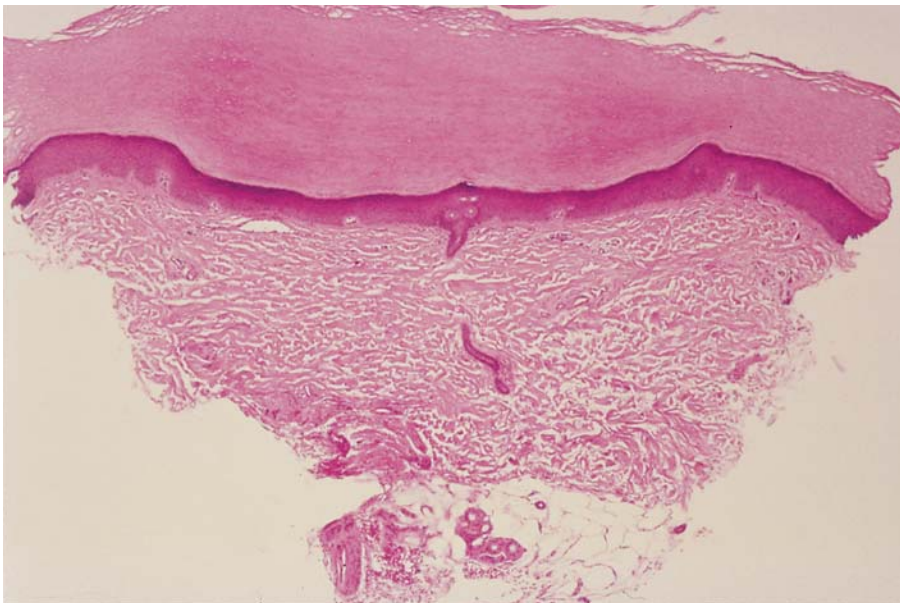

Figure 2. The clavuslike depression in an acanthotic epidermis, mild dilation of upper dermal vessels, absence of dermal inflammation, and normal collagen fibers (H&E, original magnification ×20).

histopathology of acrokeratoelastoidosis reveals marked orthokeratotic hyperkeratosis, fragmented and decreased numbers of dermal elastic fibers, and homogenized dermal collagen.[2,6]

Other diseases that may be included in the differential diagnosis of FAH are verruca plana, acrokeratosis verruciformis of Hopf, epidermodysplasia verruciformis, punctate porokeratosis, palmoplantar lichen planus,[1] punctate palmoplantar keratoderma, dyshidrotic eczema, and xanthoma.[2]

Treatment of FAH often is disappointing. Multiple treatment modalities have been unsuccessful, including the use of liquid nitrogen, salicylic acid, ammonium lactate, tretinoin, oral prednisone, methotrexate, dapsone, antibiotics, tar, silver nitrate, and grenz ray, and only partial response was noted with etretinate.[1,7] Our patient did not respond to ammonium lactate or urea.

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