Acrokeratoelastoidosis

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A 45-year-old woman presented with asymptomatic roughness of the palms since seven years. Her medical history was noncontributory and she had no other symptoms. There was no history of trauma, excessive sun-exposure, or hyperhidrosis. There was no history of similar lesions among the family members. On cutaneous examination, multiple skin colored, depressed, umbilicated and few crateriform, hyperkeratotic papules were seen on the lateral border of the index finger and medial border of the thumb bilaterally in a symmetrical distribution [Figure 1]. Her soles, nails, hair, teeth, and mucous membranes were normal.

Histopathological examination of a papule revealed orthokeratotic hyperkeratosis, acanthosis, thickened, curved basophilic elastic fibers in the papillary and reticular dermis [Figure 2]. Verhoeff–Van Gieson stain revealed thick, curved, coarse, fragmented elastic fibers (elastorrhexis) in the dermis [Figure 3].

Acrokeratoelastoidosis (AKE) was first described as small, firm, umbilicated skin-colored and keratotic papules along the borders of the hands and feet by Costa.[1] It is a type of palmoplantar marginal papular keratoderma. The marginal papular keratodermas of the palms and soles are a group of disorders which include AKE, focal acral hyperkeratosis (FAH), mosaic acral keratosis, degenerate collagenous plaques of the hands, digital papular calcific elastosis, and keratoelastoidosis marginalis of the hands.[2] The entities described under marginal papular keratodermas are complex and confusing with a considerable overlap.

AKE is a rare disease. Most cases of AKEs that have been reported so far have occurred before the second to third decades of life.[3] The autosomal dominant form of the condition manifests itself in children, adolescents, or young adults. The occurrence of lesions later in life suggests a sporadic form. A history of excessive sun exposure, hyperhidrosis, or repeated trauma may be present.[3]

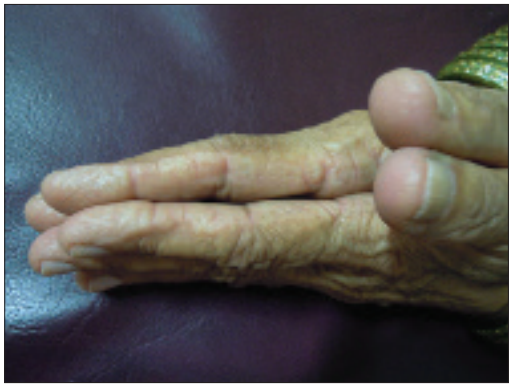

Figure 1: Clustered crateriform papules on the fingers

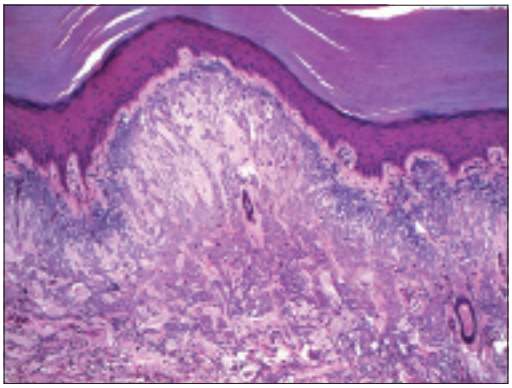

Figure 2: Thickened, curved basophilic elastic fibers in the dermis (hematoxylin and eosin ×40)

The lesions of AKE primarily affect the borders of the palms and soles. Extension to the dorsal and palmar surfaces may occur in some cases. They present clinically as symmetrically located grouped, clustered, small, firm, umbilicated papules with a glossy and translucent surface. Sometimes, the papules may coalesce to form plaques. Clinically, AKE can simulate other

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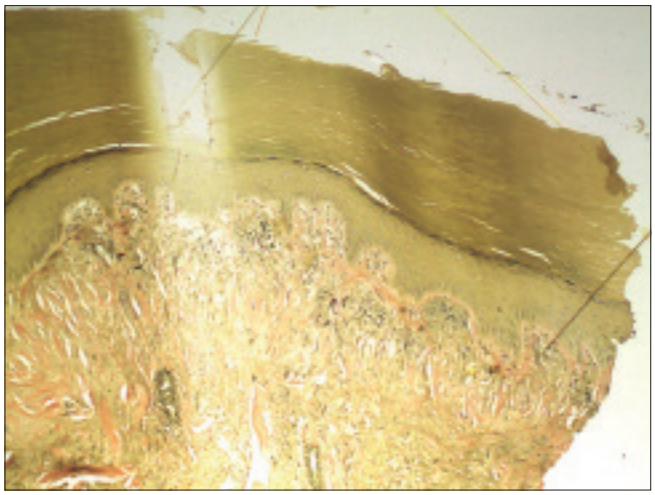

Figure 3: Thick, curved, coarse, fragmented elastic fibers in the dermis. (Verhoeff–Van Gieson stain ×40)

papular acral keratodermas such as focal acral hyperkeratosis and degenerative collagenous plaques of the hands.

Histologically, AKE comprises of epidermal changes such as orthokeratotic hyperkeratosis, acanthosis, hypergranulosis with or without epidermal hyperplasia.[4] The dermis shows reduced number of elastic fibers. The elastic fibers are thickened, curved, fragmented, and appear as basophilic fibers, in the papillary and reticular dermis. Verhoeff–Van Gieson or acid orcien stain may be used to demonstrate the changes in the elastic fibers. The finding of fragmented elastic fibers (elastorrhexis) is absent in FAH, which helps in differentiating AKE from FAH.

Usually, treatment is deferred in these patients as the lesions are asymptomatic.[4] However, the condition is cosmetically unpleasant for patients. Topical treatment includes topical keratolytics such as salicylic acid or tretinoin. Systemic treatment with prednisolone, dapsone, methotrexate, and acitretin has also been tried. Er:YAG laser has been used with some benefit.[5] The patient must be counseled about the nature of the condition and the limited treatment modalities.

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Conflicts of interest

There are no conflicts of interest.

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