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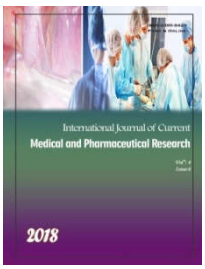


Research Article

# ACROKERATOELASTOIDOSIS: PRESENTATION OF A LOCALIZED LESIONS TO DORSAE OF HANDS AND FEET

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**ABSTRACT**

Acrokeratoelastoidosis is a rare *genodermatosis* that presents clinically as shiny keratotic papules and paques along the junctionn of palms or soles with the dorsae of hands or feet. AKE commonly starts at puberty. Here, we present a case of AKE in a 50-year-old Bangladeshi woman with asymptomatic skin lesion on the dorsal surface of the feet and hands since childhood. She denies any family history of similar condition and her parents are not consanguineous.
Skin examination showed multiple keratotic plaques over the dorsae of hands and feet, some of them are in linear configuration. Histological examination of a 4mm punch skin biopsy specimen revealed orthohyperkeratosis, psoriasiform acanthosis and elastotic plaques with elastorrhexis in the dermis. Elastin stain showed increased elastin fragments with elastorrhexis. On the basis of the above clinicopathological findings, a diagnosis of acrokeratoelastoidosis was made. The patient was reassured.



## INTRODUCTION

Acrokeratoelastoidosis (AKE) is a rare genodermatosis that presents clinically as shiny keratotic papules and paques along the junctionn of palms or soles to the dorsae of hands or feet. Costa defines AKE as frequently located on both thenar and hypothenar eminences, the volar aspect of the wrist, dorsa and sides of the digits, and radial and ulnar margins of the hands; it may also involve the anterior surface of the ankle, tibial, and fibular malleoli areas, the tendo calcaneus, dorsa of the feet and toes, and soles[5]. Sporadic and familial cases following autosomal dominant transmission have been reported.

It is characterized histologically by orthohyperkatosis and elastorrhexis in the reticular dermis.

There are controversy whether focal acral hyperkeratosis and degenerative collagenous plaques of the hands are seperate entities or spectrum of the same condition. AKE and focal acral hyperkeratosis (FAH) are distinguished only on the basis of the absence of elastorrhexis in the later [4]. Another condition with a same clinical appearance is marginal keratoelastoidosis, which is associated with severe sun exposure, marked actinic damage, and genetic inheritance [2].
AKE and degenerative collagenous plaques of the hands (DCPH) are two pathogenically controversial and confusing dermal conditions show acral, symmetric, hyperkeratotic lesions, more frequent on the hands, associated with dermal connective tissue abnormalities. Burks et al. [10] first described DCPH as a chronic, asymptomatic, and slowly progressive condition. The bilateral, symmetric lesions consisted of linear, firm plaques extending from the thumb, around the web, and along the side of the index finger. The surface was keratotic, scaly, or smooth and yellowish in color. Histologically, a rather distinctive deposition of dense collagen and degenerated elastic fibers in the reticular dermis was seen. The affected area was almost a cellular, with an occasional fibroblast but no inflammatory cells. Calcium deposits were common, but not massive.

Differential Diagnosis include, palmoplantar keratoderma punctata and acrokeratosis verruciformis of Hopf[7]. Currently, no effective treatment exists for AKE, which can have a significant cosmetic impact [1].

***Case report***

55- year- old Afghanistani women presented with history of asymptomatic skin lesions on her hands and feet since childhood. She denied any family history of similar condition and her parents are not consanguineous. Review of systems and past medical history were unremarkable. Skin examination showed multiple keratotic plaques over the dorsae of hands and feet, some of them are in linear configuration (Fig. 1)

The differational diagnosis includes, tuberous xanthoma, keloids, lichen ruber moniliformis, and linear morphea.

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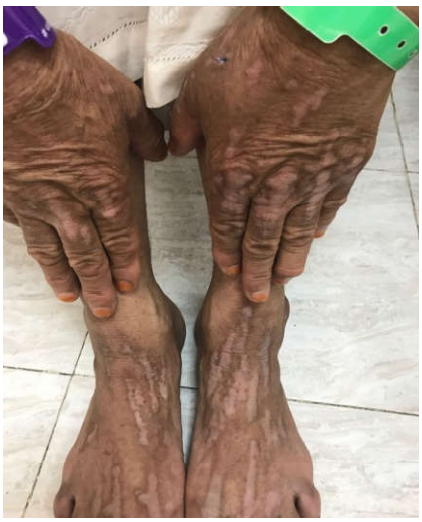

**Figure 1** Multiple keratotic plaques on dorsae of hands and feet

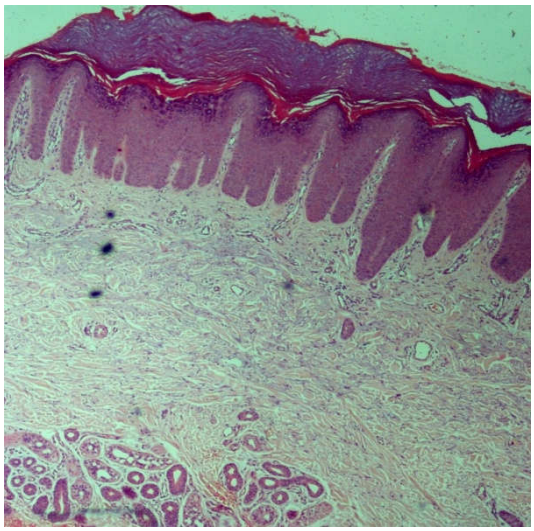

**Figure 2** Histological examination of the skin lesion (Hematoxylin and eosin, original magnification × 40) showing orthohyperkeratosis, psoriasiform acanthosis and elastotic plaques with elastorrhexis ( basophilic material) in the dermis**.**

Histological examination of a 4 mm punch skin biopsy specimen revealed orthohyperkeratosis, psoriasiform acanthosis and elastotic plaques with elastorrhexis in the dermis (figure 2). Elastin stain showed increased elastin fragments and elastorrhexis. On the basis of the above clinicopathological findings, a diagnosis of acrokeratoelastoidosis was made. The patient was reassured.

## DISCUSSION

AKE is a rare *genodermatosis*. AKE commonly starts at puberty, and rarely later on. Women appear to be affected more frequently than men.

Clinically, AKE is characterized by multiple symmetrical soft, shiny, *translucent,* firm, and yellow papules on the lateral margins of the palms and soles, dorsal surfaces of the hands and feet, the frontal surface of the lower legs, and across the joints, o*ccasionally similar to knuckle pads.* [5] The lesions are sometimes umbilicated. Our case is unusual in that the lesions were localized only to the dorsal surfaces of the hands and feet. To our Knowledge this is the first report of such condition.

The etiology for AKE is not fully explained. It appears to be a genotype with a potentially hereditary genome and a possible chromosome 2 association. . Keratotic papules may result from an overproduction of filaggrin, which may accumulate as a diffuse basophilic band above the granular layer before being incorporated into the dense horny cell protein matrix. Although moderately thickened elastic fibers were demonstrated, collagen, oxytalan, and elaunin fibers were normal.[6] A report has been published showing that elastorrhexis is a feature of the human skin as well as the apparent normal skin in a patient with AKE, indicating that this disorder can be considered a general defect of elastic tissue that is somewhat limited in situ. [6] A relationship was suggested between AKE and systemic scleroderma.[7]

Elastic fibers in AKE may be thinned, thickened, or normal and may represent genetic variants. Consequently, FAH, PTAK, MAK, and PPK should not be considered a separate entities but clinico-pathologic variants of AKE.

AKE may not require treatment because the lesions are symptomless and without adverse effects. [6] However, the condition may be cosmetically unappealing to patients, Various treatment options have been tried with little or no help. [3, 6]

The various treatments reported include topical medications (corticosteroids, salicylic acid, and tretinoin), immunosuppressant therapies using prednisone or methotrexate, antibiotic therapy, and destructive therapy with plastic surgery. All of them were few or no interest[6] Retinoic acid (50 mg / d) was reported to exhibit some response, although the results were still unsatisfactory. [6] Er:YAG laser has limited effect.

## CONCLUSION

Due to the scarcity of AKE, little is known about its occurrence, etiology, and pathophysiology. Genetic researches may discover more about this disease and may therefore be useful in identifying better treatment options.

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