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Costa's acrokeratoelastoidosis: Two sporadic case reports

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Abstract
Acrokeratoelastoidosis is a rare genodermatosis of unclear cause. It usually begins in childhood or adolescence and is clinically characterized by multiple small, firm, yellowish papules on the lateral borders of the hands and feet. There are several differential diagnoses that histology excludes by showing elastorrhexis associated with hyperkeratosis and acanthosis. The aesthetic prejudice caused by this condition can be important in some patients, justifying a treatment but to date none is satisfactory.

Keywords: Acrokeratoelastoidosis, Costa, Palmoplantar keratoderma, Genodermatosis, Elastic fibres

Introduction
Costa's acrokeratoelastoidosis (AKE) is a rare form of palmoplantar keratoderma type III, characterized by keratin and elastic tissue abnormalities. We report 2 cases seen in dermatological consultation.

Clinical Presentation
Case 1: A 25-year-old female patient, phototype IV, presents for 9 years whitish itching papules with smooth surface, located on the lateral edges of both feet (Figure 1A). At dermoscopy, yellowish structureless patterns were found.
Case 2: 22-year-old female patient, phototype III, who has been suffering from pruritic confluent erythematous papules on the lateral surfaces of both feet for 6 years (Figure 1B).
In our 2 patients there was no notion of sun exposure nor of hyperhidrosis, but we found the wearing of ill-fitting shoes. Skin biopsy confirmed the diagnosis of AKE, showing hyperkeratosis with hyperplasia of the granular layer (Figure 2) and a decrease in the elastic fibers of the reticular dermis when stained with Orcein (Figure 3). Local retinoids 0.025% under occlusion were initiated, without clear improvement after 6 months.

Discussion
First described in 1952 by the Brazilian dermatologist Oswaldo Costa, acrokeratoelastoidosis (AKE) or inverse papular acrokeratosis is a rare genodermatosis transmitted mostly in an autosomal dominant mode, most probably related to chromosome 2, although sporadic and recessives forms have been reported [1].
Its pathogenesis is still unclear. Besides the hereditary factor, it is assumed that the keratotic papules that are clinically seen in AKE may be the result of a defect in the secretion or excretion of elastic material by dermal fibroblasts [2]. For sporadic forms, chronic trauma and excessive sun exposure have been suggested as potential causal factors [3]. Moreover, the association with systemic and localized scleroderma has been reported by Yoshinaga and Tanaka, suggesting an autoimmune process [4, 5].
This rare condition usually begins in childhood, although its onset may be delayed until adolescence as in our two patients. There doesn't seem to be a predilection for gender or race based on the limited data available in the literature on this issue [6].
Clinically, AKE manifests as multiple asymptomatic, yellowish, sometimes shiny, keratotic papules, distributed symmetrically along the marginal borders of the palms and soles or in the pre-tibial region [2]. Hyperhidrosis and pruritus is also commonly noted [7]. Dermoscopy reveals focal clusters of yellowish and structureless areas [8].

Histology can rule out other differential diagnoses, such as focal acral hyperkeratosis where there is no elastorrhexis and the alterations are limited to the epidermis (hyperkeratosis and acanthosis). Marginal keratoelastoidosis also has the same clinical appearance, is usually associated with intense sun exposure and marked actinic lesions with a significant genetic predisposition. This condition includes: focal acral hyperkeratoses, AKE, papulotranslucent acrokeratoderma, mosaic acral keratosis, and punctuate palmoplantar keratoderma. Other conditions to consider in the differential diagnosis of AKE include Hopf's acrokeratosis verruciformis, degenerative collagenous plaques, palmoplantar punctate keratoderma, papular calcinosis, primary cutaneous amyloidosis and lichen nitidus [3].

Typical histopathological features of AKE are hyperkeratosis, hypergranulose, mild acanthosis, collagen homogenization, and a decrease in the number of elastic fibers that become mainly fragmented in the superficial and reticular dermis, best seen by acid Orcein stain or Elastic–van Gieson [3].

Except for the cosmetic damage that this condition may cause in some patients, the overall prognosis is good [6]. Generally, no treatment is needed. But for some bothered patients, various local or systemic therapeutics have been tried, but without any real improvement [9]. Among these therapeutics are keratolytics, dermocorticoids, liquid nitrogen, and more recently the Erbium Yag laser has been suggested by Erbil *et al*. [10].

Figures legend

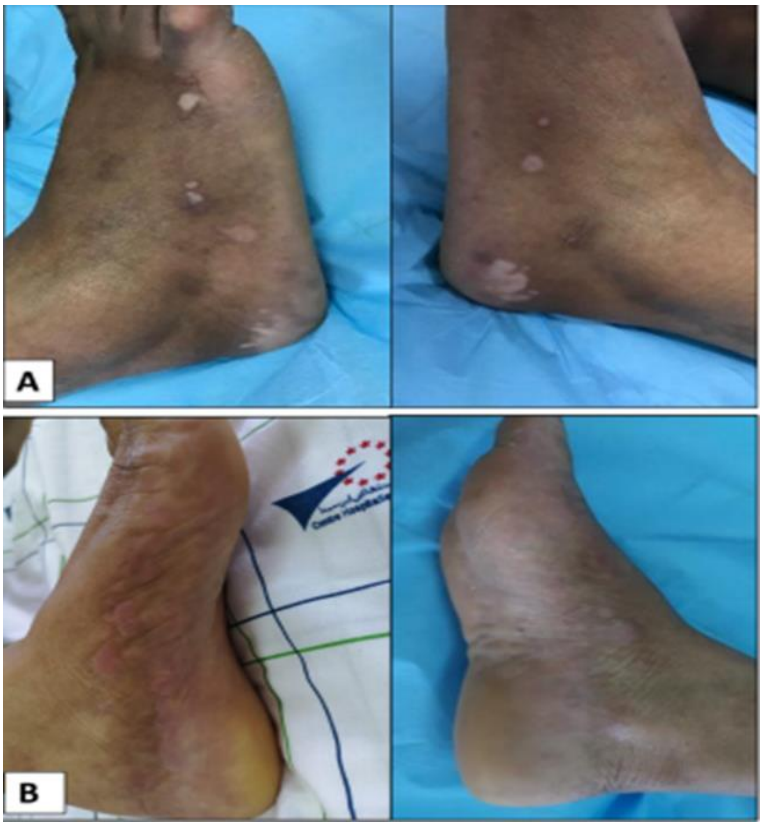

Fig 1: (A) Whitish papules with smooth surface, confluent in some places, located on the lateral edges of both feet. (B) Confluent erythematous papules on the lateral sides of both feet

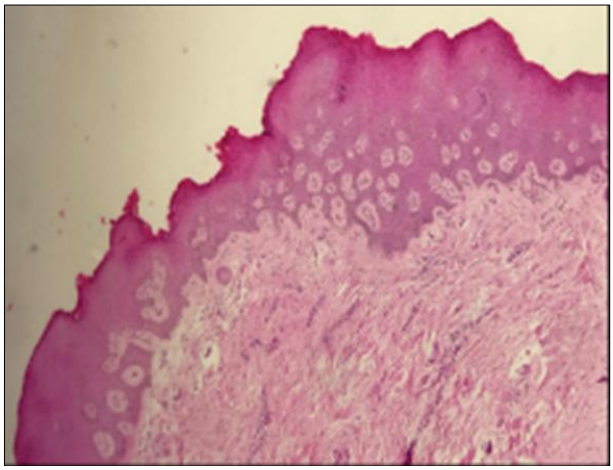

Fig 2: Histological image showing hyperkeratosis with acanthosis (HE)

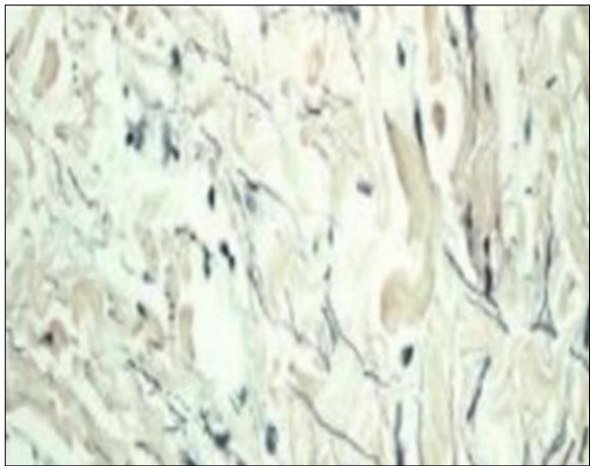

Fig 3: Histological image after staining with Orcein showing a decrease of elastic fibers in the reticular dermis

Conclusion

Costa's AKE raises a problem of aesthetic management. There is no codified therapeutic consensus. Various treatments have been proposed to reduce the hyperkeratotic aspect and to limit its extension. But all of them are still disappointing.

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