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Clinical Image

Acrokeratoelastoidosis of Costa: Clinical

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Clinical Image

In this instance, we present a case involving a 42-year-old woman with no significant medical history who came to our dermatological clinic due to chronic, asymptomatic red-violet papules located on the sides of both feet (Fig. 1). These lesions had developed gradually over the past five years. The patient also noted that both of her sisters had similar lesions since childhood. Dermatoscopic examination revealed groups of white to violet papules, some exhibiting umbilication, interspersed with areas lacking distinct structures. A skin biopsy indicated the presence of hyalinized collagen and fragments of elastic fibers, with no signs of granulomatous areas, inflammatory infiltrate, or leukocytoclastic vasculitis. Consequently, we established a diagnosis of acrokeratoelastoidosis of Costa. Treatment involving a 10% salicylic acid cream was attempted for one month but yielded no improvement. Strong corticosteroids were somewhat effective in enhancing the lesions'purplish appearance and slightly reducing their size. However, the patient subsequently discontinued follow-up.

Acrokeratoelastoidosis of Costa is a rare genetic skin disorder inherited in an autosomal dominant manner, first identified by Brazilian dermatologist Oswaldo Costa in 1952 [1]. Typically, the condition presents as flesh-colored papules on the lateral surfaces of the palms, soles, and backs of the hands. It represents a type of marginal keratoderma primarily affecting the lateral aspects of the palmoplantar regions. In addition to inherited cases, sporadic instances of this condition have been documented. Dermatoscopic findings usually include localized clusters of pale to yellowish papules with slight umbilication in some lesions, along with pale yellow structureless areas.

Histologically, the condition is characterized by hyalinized and homogeneous collagen, hyperkeratosis, and a reduction in and fragmentation of elastic fibers (elastorrhexis) [2]. This uncommon form of focal acral keratoderma, whose etiology remains unidentified, typically begins in childhood, although it may be delayed until adolescence. It is essential to differentiate this condition from two groups of disorders: 1) other marginal and focal acral keratodermas, and 2) distinct conditions such as acrokeratosis verruciformis of Hopf. Histopathological examination plays a crucial role in distinguishing acrokeratoelastoidosis from other clinical mimics. No significant morbidity has been reported, and despite some patients expressing cosmetic concerns, the overall prognosis for AKE is favorable [3].

Numerous Treatment Modalities: Topical - emollients and keratolytic agents including salicylic acid, urea, sulfur, coal tar, and tretinoin, as well as topical corticosteroids have been investigated to improve the appearance of the affected acral regions; continuous use may be necessary to sustain any benefits. Oral - corticosteroids, antibiotics, acitretin dapsone, methotrexate, isotretinoin, have shown significant improvement in the condition; however, the risk of relapse upon discontinuation and potential side effects of these medications must be considered.

Physical/surgical - cryotherapy with liquid nitrogen, and surgery with laser erbium have been attempted, yielding generally modest to poor results. Overall, oral retinoids, especially acitretin, have been recognized as the most effective treatment, though relapses are common after treatment cessation [4].

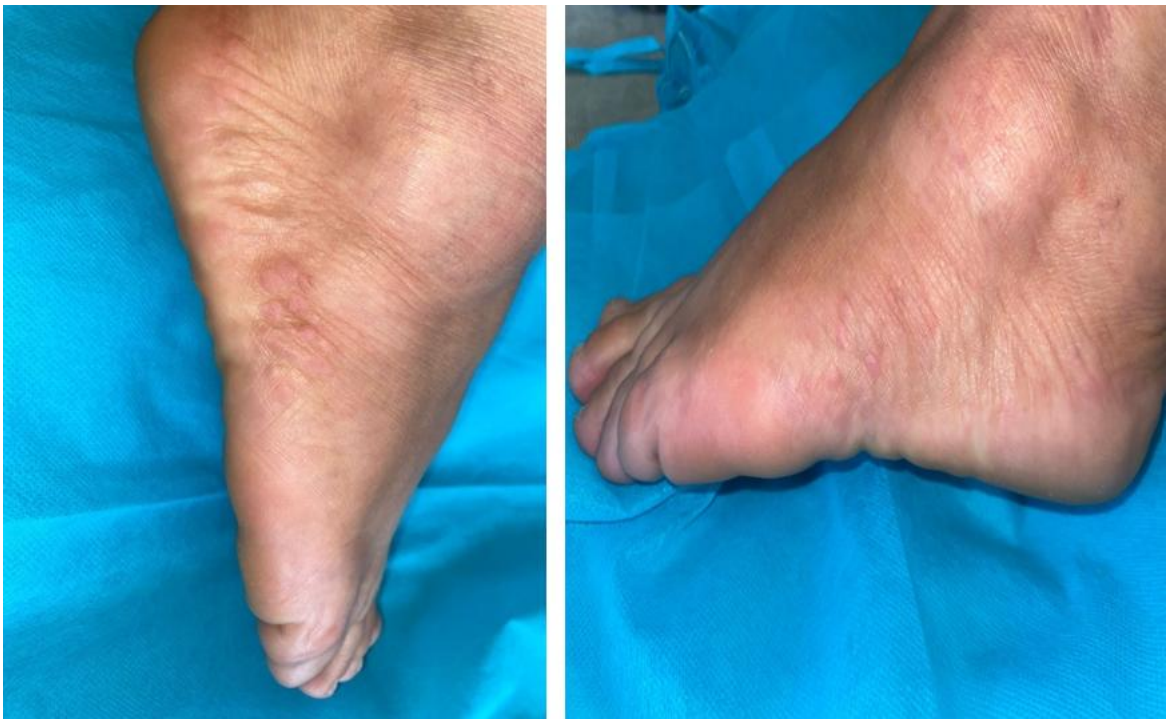

Figure 1: Acrokeratoelastodosis of costa.

Keywords: Dermatoscopic Examination; Biopsy; Salicylic Acid

Conflicts of Interest

The authors declare no conflict of interest in this paper.

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