Acrokeratoelastoidosis of Costa Mimicking Dyshidrosis: A Case Report

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ABSTRACT:

Acrokeratoelastoidosis is a rare genodermatosis with an autosomal dominant pattern of inheritance. The clinical presentation is usually asymptomatic keratotic papules confluent to plaques on the margin of hands and feet. The histopathological findings are fragmentation and diminishing of elastic fibers within the dermis. We identified a case of 40-year-old female presented with multiple papules on both sites of hands and feet. Initially, she was diagnosed and treated as dyshidrosis. The skin biopsy showed fragmented elastic fibers in the dermis, compatible with acrokeratoelastoidosis. She has been treated with topical tretinoin. The lesions are slightly improved.

Key words: Acrokeratoelastoidosis, Acrokeratoelastoidosis of Costa, Palmoplantar keratoderma, Marginal papular acrokeratodermas

Introduction

Acrokeratoelastoidosis (AKE) is considered as one condition of the marginal papular acrokeratodermas group[1]. It is a rare condition and usually observed in childhood or adolescence[2]. The pathogenesis is not well understood. The disease appears to be an autosomal dominant inherited disease, but can also be sporadic[3]. The typical skin characteristics are asymptomatic multiple yellow keratotic papules with an umbilicated surface. AKE lesions are commonly distributed bilaterally on the border of hands and feet. The histopathological findings are focal parakeratosis, acanthosis, and elastorrhexis of elastic fibers in the reticular dermis. The treatment options include topical and systemic medications, such as corticosteroids and retinoids. To date, there is no curative therapeutic regimen for AKE[2].

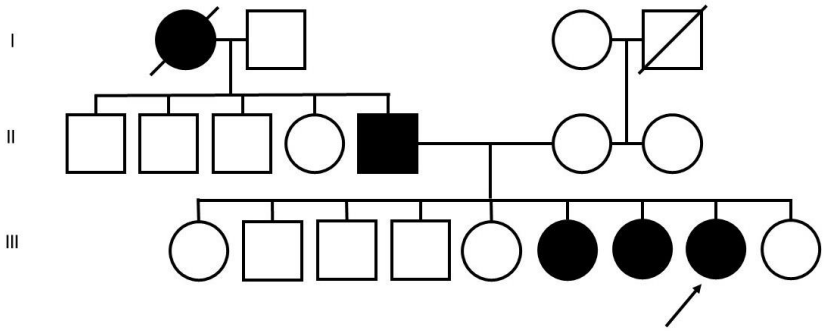

Figure 1 Pedigree chart of the case's family

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Received: 16 January 2023 Revised: 10 February 2023 Accepted: 28 March 2023

Case report

A 40-year-old Thai female presented with asymptomatic papular lesions scattered on both hands and feet for a few years. The rashes had gradually increased in number and size. She declined itching or painful sensation. She had no history of hyperhidrosis, repetitive trauma, friction, or excessive sun exposure. Her father and sisters had similar skin lesions, as shown in the pedigree (Figure 1). During the first visit, she was diagnosed as dyshidrosis and treated with topical superpotent corticosteroids. But her symptoms did not get alleviated.

On dermatologic examination, multiple well-defined flat-topped shiny skin-colored hyperkeratotic papules were noted along the knuckles and margins of both hands and feet (Figure 2). No puffy finger, digital pitting scar or abnormality of nailfold were detected. A skin biopsy taken from her papular lesion of right hand (Figure 3), revealed insignificant change in epidermis. Minimal superficial perivascular lymphocytic infiltration was observed in the dermis. However, Verhoeff-Van Gieson (VVG) special stain showed fragmentation and reduction of dermal elastic fibers. Masson trichrome stain revealed normal collagen. From clinical and histological findings, the diagnosis of AKE was made and she had been treated with topical 0.025% tretinoin plus 10% urea cream. Trivial flattening of the papular rash was reported after 3-month of therapy.

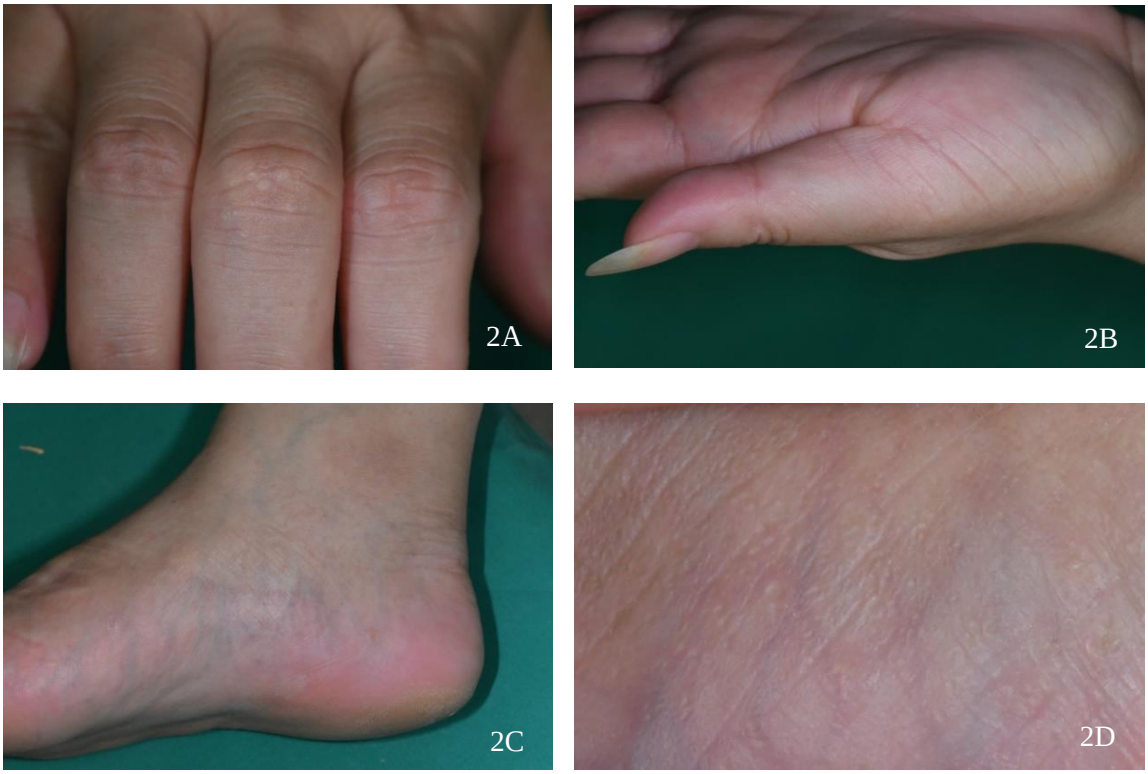

Figure 2 Multiple flat-topped shiny skin-colored papules at the knuckles (2A) and palmodorsal junction of both hands and feet (2B, 2C). Some of the lesions show umbilicated appearance (2D)

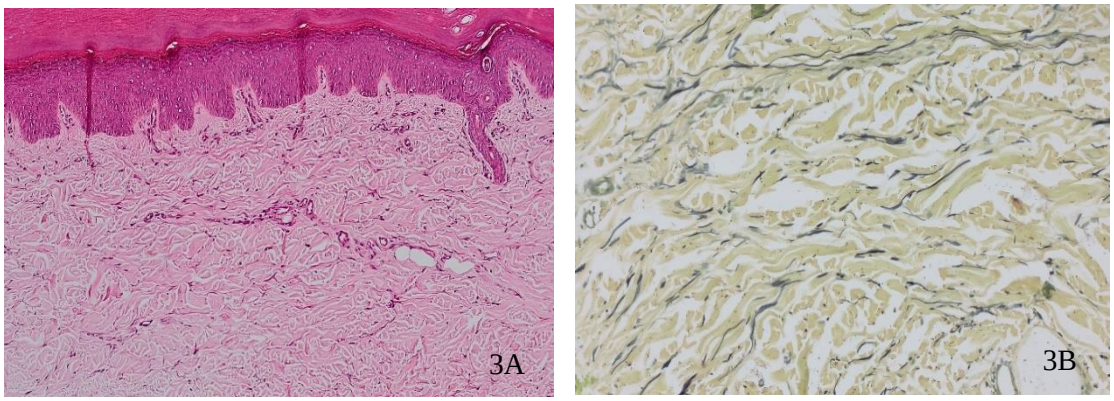

Figure 3 Compact orthokeratosis, hypergranulosis, mild regular acanthosis with mild superficial perivascular lymphocytic infiltration were noticed from Hematoxylin & Eosin stain (X100, 3A). Verhoeff-Van Gieson stain showed fragmentation and reduction of elastic fibers in the dermis (X200, 3B).

Discussion

Acrokeratoelastoidosis (AKE) or Acrokeratoelastoidosis of costa was first described by Brazilian dermatologist, Oswaldo Costa, in the 1950s[4]. It is a rare autosomal dominant genetic disorder involving on 2p25-p12 locus located by genetic mapping which possibly related to chromosome 2[5]. The AKE is also found as a sporadic case[2,3]. The pathogenesis of AKE remains unclear, but there is some evidence from electron microscopic studies showing that the fibroblasts in reticular dermis are reduced in number but contain dense granules in the ectoplasm. These granules are thought to be the precursor of abnormal elastic fibers. Moreover, the finding of a dense band over the granular layer is related to the overproduction of filaggrin, which is corresponding with the formation of keratotic papules of the disease[2]. AKE is usually presented in children or adolescences. The disease has no significant race predominance[2]. The classic clinical presentations are multiple small, yellow or white, oval or polygonal, keratotic papules, sometimes umbilicated, located symmetrically bilateral on the margin of hands and feet. Most of the patients declined history of hyperhidrosis or any associated systemic disease[2].

The differential diagnoses of AKE are hereditary (focal acral hyperkeratosis, FAH, punctate palmoplantar keratoderma, PPK, and hereditary papulotranslucent keratoderma, PTAK) and acquired types (degenerative collagenous plaques of the hands, DCPH) of marginal papular acrokeratodermas group. FAH has very similar cutaneous lesions as AKE and is also presented in childhood to young adults, but more common in black persons. Punctate PPK has skin manifestations undistinguished from AKE[2]. PTAK presents with translucent papules that can be involved in the dorsal or volar of hands and feet. It is precipitated by exposure to water[6]. But the histopathologic finding of FAH, punctate PPK and PTAK reveals no elastorrhexis[2,6]. DCPH is caused by repetitive minor trauma and UV exposure. It usually affects middle-aged and elderly patients. DCPH is different from AKE by histological findings of dense collagen and calcium deposition. On the other hand, degeneration of elastic fiber and collagen can be observed in the reticular dermis of DCPH[7]. Furthermore, verruca vulgaris and Acrokeratosis verruciformis of Hopf should be

taken into consideration of AKE differential diagnosis. The distinct dermoscopic findings such as dotted or linear vessels, verrucous growth, and bleeding suggest the diagnosis of common warts[8]. Acrokeratosis verruciformis of Hopf lesions look similar to flat warts which are mostly distributed on the dorsal of hands and feet. The histopathology finding of church spire appearance in the epidermis is its characteristic[9].

Histological examination frequently shows some minor epidermal changes, such as focal hyperkeratosis, acanthosis, and hypergranulosis. VVG staining reveals diminished and fragmented elastic fibers (elastorrhexis), predominantly in the reticular dermis[2].

The treatment of AKE is still challenging. Several modalities include topical retinoids, topical steroids, salicylic acid, cryosurgery, systemic corticosteroids, methotrexate, dapsone, and oral retinoids have been described with limited outcomes[2]. Erbium-doped yttrium aluminium garnet (Er:YAG) laser is considered as another option for recalcitrant case[10].

In our case, the patient was presented with multiple skin-color papules on the knuckles and margin of both hands and feet. Similar cutaneous findings were observed in her father and sisters. The diagnosis of dyshidrosis was initially made by the first in-house dermatologist. This may be due to its shiny appearance and hand-foot distribution, which can be mimicking pompholyx. Nevertheless, the skin biopsy was finally done and showed unremarkable change from Hematoxylin and eosin (H&E) stain. VVG special stain demonstrated reduction in number and fragmentation of elastic fibers. The clinicopathological data supported the diagnosis of AKE. Slight clinical improvement has been noticed after 3-month course of topical retinoid and 10% urea cream application. After the patient was informed about benign nature of the disease, she denied trying other options of treatment.

In conclusion, we reported a case of AKE mimicking dyshidrosis. The disorder seems to be underrecognized by physicians. Because there are many cutaneous conditions that appear to be similar to AKE, in case of doubt, histological examination should be performed to confirm the diagnosis. Although AKE is usually benign and asymptomatic, it can cause cosmetic concerns. Due to the rarity of this condition, the therapeutic recommendations are mostly reported as a single case. Further study regarding effective treatment for AKE should be carried on.

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