Qual o Seu Diagnóstico?

Múltiplas Pápulas Queratósicas Amareladas nas Eminências Tenar

Calvão J[1], Relvas M[1], Cardoso JC[1], Gonçalo M[1,2]
[1]Dermatology Department of Coimbra University Hospital, Coimbra, Portugal
[2]Faculty of Medicine, University of Coimbra, Coimbra, Portugal

PALAVRAS-CHAVE – Acroqueratoelastoidose; Hiperqueratose acral focal; queratoelastoidose marginal; acroqueratose verruciforme de Hopf; placa colagenosa degenerativa das mãos; queratodermia palmoplantar punctata.

Dermatology Quiz

Multiple Yellowish and Keratotic Papules in the Thenar Eminence

KEYWORDS – *Acrokeratoelastoidosis; focal acral hyperkeratosis; keratoelastoidosis marginalis of the hands; acrokeratosis verruciformis of Hopf; degenerative collagenous plaques of the hands; punctate palmoplantar keratoderma.*

CASE REPORT

A 49-year-old female, with congenital deafness and osteoarthrosis of the first carpo-metacarpal joint related to professional activity (factory worker for 30 years), and no chronic medication, presented with multiple small, monomorphic, round, yellowish keratotic papules, in a linear distribution predominantly in the thenar eminence and along the thumb of both hands in a symmetric way, although more intense in the right hand (Fig. 1 a, b). No lesions were observed on the feet. Lesions were present for 19 years and caused mild local discomfort particularly during heavy manual labour at her work. There were no other similar cases in the family.

Dermoscopy showed 2-6 mm structureless yellowish polygonal globules, mostly in a linear distribution (Fig. 2).

Cutaneous biopsy displayed areas of compact orthokeratotic hyperkeratosis over a slightly depressed but otherwise normal-appearing epidermis (Fig. 3). Irregular fragmentation of the elastic fibres in the reticular dermis was observed with Verhoeff-Van Gieson staining (Fig. 4).

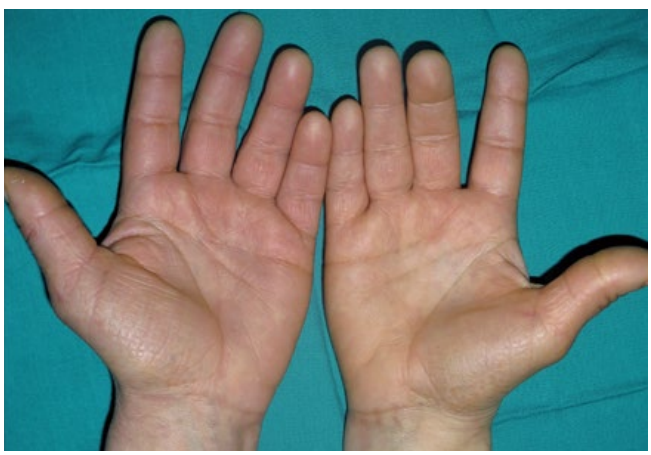

Figure 1a - Multiple small, monomorphic, round, yellowish and keratotic papules, in the thenar eminence and along the thumb of both hands in a symmetric way; b) Skin lesions of the right hand in more detail.

Correspondência: Joana Calvão
Serviço de Dermatovenereologia
Centro Hospitalar e Universitário de Coimbra
Praceta Prof. Mota Pinto
3000-075 Coimbra, Portugal
E-mail: joana.calvao.silva@gmail.com
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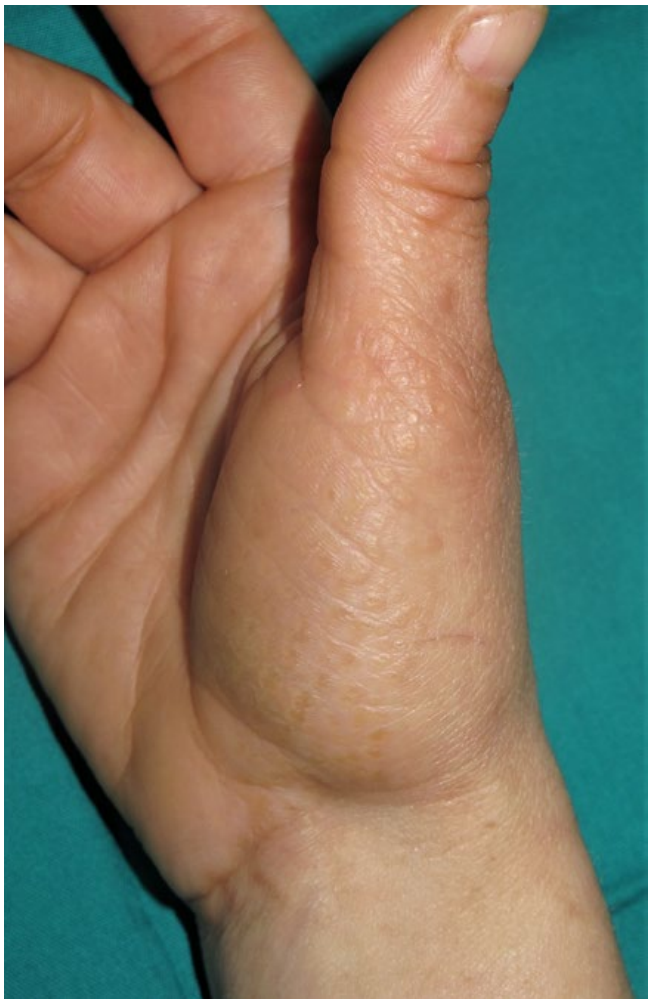

Figure 1b - Skin lesions of the right hand in more detail.

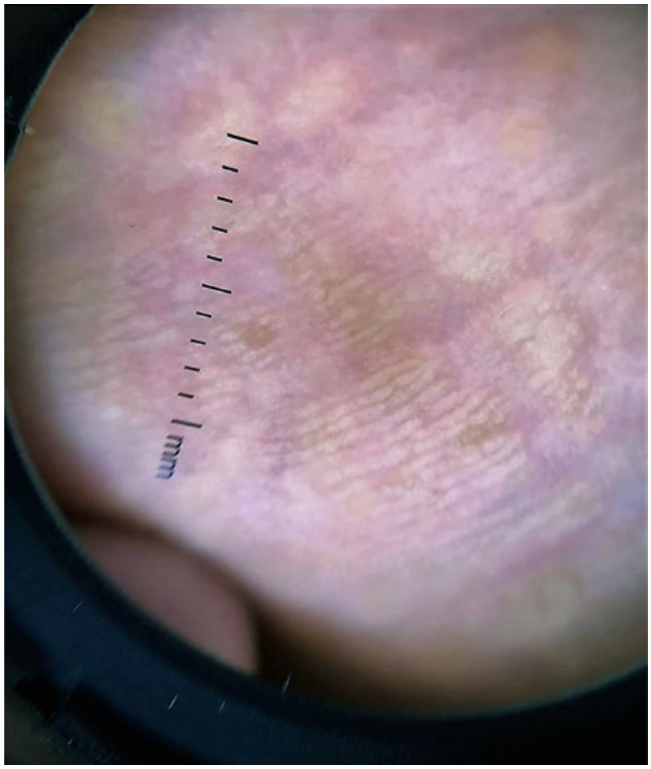

Figure 2 - Dermoscopy showing structureless yellowish polygonal globules in a linear distribution.

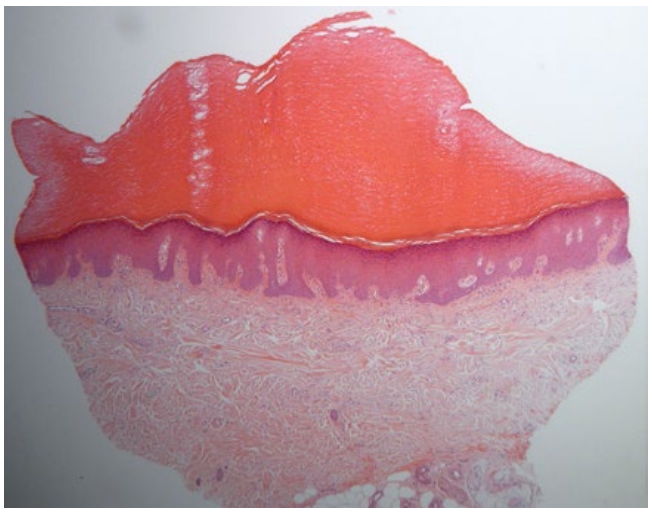

Figure 3 - Compact orthokeratotic hyperkeratosis (H&E stain ,x40).

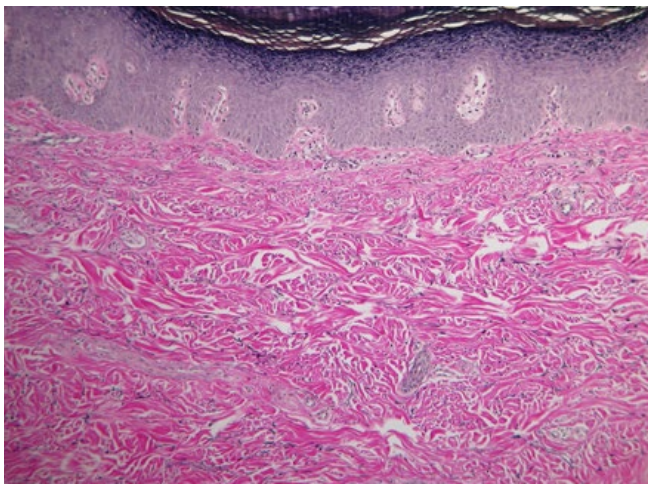

Figure 4 - Irregular fragmentation of the elastic fibers in the reticular dermis (Verhoeff–Van Gieson stain a) - x100; b) - x200).

WHAT IS YOUR DIAGNOSIS?

ACROKERATOELASTOIDOSIS

Based on these characteristic clinical and histological manifestations the diagnosis of acrokeratoelastoidosis was established.

Acrokeratoelastoidosis (AKE) is a rare type of palmoplantar keratosis.[1] First described by Oswaldo Costa in 1953, AKE presents as asymptomatic, firm, shiny papules with occasional keratosis or umbilication, typically located on the peripheral margins of the palms and soles.[2] The origin of AKE is not fully elucidated, but it may result from abnormal formation of elastic material by dermal fibroblasts.[1] There are familial cases, generally with an autosomal dominant pattern of transmission probably related to a chromosome 2 mutation. They have their onset in childhood and young adulthood.[1]

Sporadic cases have their onset later in life and are usually related with a history of excessive sun exposure, hyperhidrosis[1,3] or repeated trauma.[4] Association with systemic[5] and localized scleroderma[6] has been described, raising the question of an autoimmune process, and a recent case report suggested an association with immunosuppression.[7] In our case, both late onset and absence of family history suggest a sporadic aetiology, probably related to recurrent trauma in the occupational context, which can also explain more intense lesions in the right hand, the dominant hand.

The most common histopathology findings are hyperkeratosis, mild acanthosis and abnormal dermal elastic fibres, which are fewer and fragmented (elastorrhexis), as observed with Verhoeff-Van Gieson stain.[1,3]

In what regards the differential diagnosis, focal acral hyperkeratosis, described for the first time by Dowd *et al,*[8] is clinically identical to AKE, but elastorrhexis is absent on histology.[9] Keratoelastoidosis *marginalis* of the hands (KEMH), acrokeratosis verruciformis of Hopf, degenerative collagenous plaques of the hands and punctate palmoplantar keratoderma should also be considered (Table 1).[1,2,10]

Although KEMH can also be related to heavy manual work and repeated trauma to the hands, it is more closely related to actinic damage and predominantly affects the radial side of the index finger, first web space, and ulnar side of the thumb. Elastic fibres are often thickened, fragmented and calcified.[11]

Acrokeratosis verruciformis of Hopf is a rare genodermatosis characterized by keratotic lesions and verrucous plaques on the dorsum of the hands and feet that are usually present from birth or early childhood. Histopathology includes papillomatosis (circumscribed epidermal elevations known as "church spires"), acanthosis, hyperkeratosis, and hypergranulosis without parakeratosis.[12]

The term "degenerative collagenous plaques of the hands" refers to a rare acquired skin disorder localized to the hands, morphologically resembling those of AKE, but with no involvement of the feet and no familial predisposition. Histologically, there is a distinctive deposition of dense collagen and degenerated elastic fibres in the reticular dermis.[13]

Punctate palmoplantar keratoderma is an is an autosomal dominant genodermatosis that affects both palms and soles; histology shows marked hyperkeratosis, parakeratosis, and mild acanthosis without significant dermal changes.[14]

Usually no treatment is required for AKE because lesions are benign and mostly asymptomatic, however topical 10% salicylic acid[15], urea, calcipotriol and corticosteroids, systemic or topical retinoids, Nd-YAG and Er:YAG lasers[16], cryosurgery and iontophoresis[1,7,17] have been tried with transient and minor benefits. In the case described, we first tried 8% salicylic acid, with no significant improvement, so we opted for an emollient cream with 5% urea.

Table 1 - Main differential diagnosis of acrokeratoelastoidosis.

DISEASE	CLINICAL PRESENTATION	HISTOPATHOLOGY
Acrokeratoelastoidosis (AKE)	- Asymptomatic, firm, shiny papules with occasional keratosis or umbilication - Peripheral margins of the palms and soles	- Hyperkeratosis, mild acanthosis - Elastorrhexis
Focal acral hyperkeratosis	Hands and feet (similar to AKE)	- Hiperkeratosis, acanthosis - No dermal changes (no elastorrhexis)
Keratoelastoidosis marginalis of the hands	Exclusively hands, more related to actinic damage	- Elastic fibres are often thickened, fragmented and calcified
Acrokeratosis verruciformis of Hopf	Keratotic lesions and verrucous plaques on the dorsum of the hands and feet	- Papillomatosis ("church spires"), acanthosis, hyperkeratosis, and hypergranulosis without parakeratosis
Degenerative collagenous plaques of the hands	Similar to AKE but exclusively in the hands	- Deposition of dense collagen and degenerated elastic fibres in the reticular dermis
Punctate palmoplantar keratoderma	Palms and soles	- Marked hyperkeratosis, parakeratosis, and mild acanthosis - No significant dermal changes

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Conflitos de interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.
Suporte financeiro: Não existiram fontes externas de financiamento para a realização deste artigo.
Confidencialidade dos dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.
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