

Case Report

Keratoelastoidosis marginalis: a case report and review of literature

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Abstract Marginal keratodermas are broadly classified into acquired and familial forms. Keratoelastoidosis marginalis, also known as digital papular calcific elastosis, is an acquired, marginal, acrokeratoderma that predominantly affects the radial side of the index finger, first web space, and ulnar side of the thumb. It occurs in the middle-aged and elderly. Prolonged UV exposure and repeated trauma to the hands seem to be the etiological factors, although their role has not been well defined in the pathogenesis of keratoelastoidosis marginalis. We report a case of keratoelastoidosis marginalis in a 60-year-old woman.

Key words Keratoelastoidosis marginalis.

Case report

A 60-year-old woman presented for the evaluation of asymptomatic linear plaques on both hands symmetrically distributed along the outer margin of the index fingers and inner margin of the thumbs. The lesions first appeared between the index fingers and thumbs on both hands several years earlier and gradually increased in number, coalescing into linear plaques. The patient's history was remarkable for prolonged sun exposure for several years and repeated trauma to the hands while working in the open fields. None of her other family members were affected.

Physical examination revealed multiple, small, firm, skin-colored, keratotic papules, coalescing into crateriform linear plaques located on the junction between the palmar



Figure 1 Skin-coloured, keratotic papules, coalescing into crateriform linear plaques on lateral margins of both hands.

and dorsal skin on both lateral margins of the thumb and on the radial side of the index finger (**Figure 1**). There was no involvement of the feet. Multiple actinic keratoses were present on the dorsa of the hands, extensor aspect of the forearms and the face. A review of the systems and routine blood and urinalyses were unremarkable. A biopsy taken from the lesions on the patient's left hand revealed marked epidermal orthokeratosis and acanthosis, calcified elastic fibers in degenerated collagen bundles.

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Discussion

Keratoelastoidosis marginalis is a variant of solar elastosis and belongs to a group of disorders known as marginal keratodermas. It is characterized by the development of linear plaques and calcified dermal elastotic masses along the junction of the palms and dorsal surface of the hands. It was first described by Burks, Wise, and Clark in 1960 as degenerative collagenous and elastotic plaques of the hands, then was later termed as keratoelastoidosis marginalis, collagenous and elastotic plaques of the hands, and digital papular calcific elastosis.¹

Keratoelastoidosis marginalis has been reported to occur in people ranging in age from 42 to 78 years, but it remains a disorder mostly affecting the elderly population.² Men are more commonly affected than woman in the white population.³ Repeated traumas to the hands and prolonged sun exposure have been identified to be the inciting factors.⁴ It has also been hypothesized that the repeated episodes of hypoxia of the affected areas resulting from papillary dermal capillary occlusion during periods of pressure, in conjunction with compression from the elastotic material that accumulates in the dermis may cause the hyperkeratosis in Keratoelastoidosis marginalis.² This hypothesis is further augmented by the observation that the dominant hand is more severely affected by the disease process.

Keratoelastoidosis marginalis presents as a slowly progressive, usually asymptomatic bilateral eruption of yellowish white, waxy, and scaly crateriform linear papules that coalesce into plaques at the junction of the dorsal and palmar skin in the first web space

extending along the ulnar surface of the thumb and radial surface of the index finger, and rarely on the palms and dorsa of the hands.⁵

Histopathological examination reveals both epidermal and dermal changes. Epidermal findings consist of prominent orthokeratosis, acanthosis, atrophy, and focal "sawtoothing". Solar elastosis in the reticular dermis may be associated with thickened, fragmented, often calcified elastic fibers between distorted, degenerated collagen bundles. A lymphocytic infiltrate may be present in the papillary dermis.^{4,6,7}

Other marginal keratodermas that may need to be differentiated from Keratoelastoidosis marginalis include acrokeratoelastoidosis of Costa, focal acral hyperkeratosis, acrokeratoelastoidosis of Matthews and Harman, mosaic acral keratosis, hereditary papulotranslucent acrokeratoderma, acrokeratoderma hereditarium punctatum, degenerative collagenous plaques of the hands, and digital papular calcinosis. Acrokeratoelastoidosis is a rare genodermatosis that occurs during childhood and involves both the hands and feet, whereas keratoelastoidosis marginalis affects only the hands and is more closely related to an actinic-related process in older persons. Histopathologically, acrokeratoelastoidosis shows loss of elastic tissue in contrast to keratoelastoidosis marginalis.⁸

Focal acral hyperkeratosis typically occurs on the hands and feet of black children. The histologic changes are confined to the epidermis and spare the dermis. So the absence of elastorrhexis distinguishes focal hyperkeratosis from Keratoelastoidosis

marginalis.⁵ Keratoelastoidosis marginalis is a chronic and progressive disease that is difficult to treat.⁵ Several treatments modalities have been tried with little long-term success. These include laser therapy, high-potency topical corticosteroids, topical tazarotene and tretinoin, oral isotretinoin, and cryotherapy.⁴

References

1. Burks JW, Wise LJ, Clark WH. Degenerative collagenous plaques of the hands. *Arch Dermatol* 1960; **82**: 362-6.
2. Kocsard E. Keratoelastoidosis marginalis of the hands. Synonyms: marginal keratoderma of palms; degenerative collagenous plaques of the hands. *Dermatologica* 1965; **131**: 169-75.
3. Sehgal VN, Singh M, Korrane RV, Nayyar M, Chandra M. Degenerative collagenous plaque of the hand (linear keratoelastoidosis of the hands). A variant of acrokeratoelastosis. *Dermatologica* 1980; **161**: 200-4.
4. Todd D, Al-Aboosi M, Hameed O *et al*. The role of UV light in the pathogenesis of digital papular calcific elastosis. *Arch Dermatol*. 2001; **137**: 379-81.
5. Mengesha YM, Kayal JD, Swerlick RA. Keratoelastoidosis marginalis. *J Cutan Med Surg* 2002; **6**: 23-5.
6. Jordaan HF, Rossouw DJ. Digital papular calcific elastosis: a histopathological, histochemical and ultrastructural study of 20 patients. *J Cutan Pathol* 1990; **17**: 358-70.
7. Mortimore RJ, Conrad RJ. Collagenous and elastotic marginal plaques of the hands. *Australas J Dermatol* 2001; **42**: 211-3.
8. Rahbari H. Acrokeratoelastoidosis and keratoelastoidosis marginalis - any relation? *J Am Acad Dermatol* 1981; **5**: 348-50.

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