Collagenous and Elastotic Marginal Plaques of the Hands

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PRACTICE **POINTS**

- The etiology of collagenous and elastotic marginal plaques of the hands (CEMPHs) is attributed to collagen and elastin degeneration by chronic actinic damage, pressure, or trauma.
- It is important to keep CEMPH in mind when dealing with occupational cases of repeated long-term trauma or pressure to the hands as well as excessive sun exposure. It should be separated from other diseases and avoid being misdiagnosed as a malignant lesion.

To the Editor:

Collagenous and elastotic marginal plaques of the hands (CEMPHs) has several names including degenerative collagenous plaques of the hands, keratoelastoidosis marginalis, and digital papular calcific elastosis. This rare disorder is an acquired, slowly progressive, asymptomatic, dermal connective tissue abnormality that is underrecognized and underdiagnosed. Clinical presentation includes hyperkeratotic translucent papules arranged linearly on the radial aspect of the hands.

A 74-year-old woman described having "rough hands" of more than 20 years' duration. She presented with 4-cm wide longitudinal, erythematous, firm, depressed plaques along the lateral edge of the second finger and extending to the medial thumb in both hands (Figure 1). She had attempted multiple treatments by her primary care physician, including topical and oral medications unknown to the patient and light therapy, all without benefit over a period of several years. We have attempted salicylic acid 40%, clobetasol cream 0.05%, and emollient creams containing α-hydroxy acid. At best the condition fluctuated between a subtle raised scale at the edge to smooth and occasionally more red-pink, seemingly unrelated to any treatments.

The patient did not have plaques elsewhere on the body, and notably, the feet were clear. She did not have a history of repeated trauma to the hands and did not engage in manual labor. She denied excessive sun exposure, though she had Fitzpatrick skin type III and a history of multiple precancers and nonmelanoma skin cancers 7 years prior to presentation.

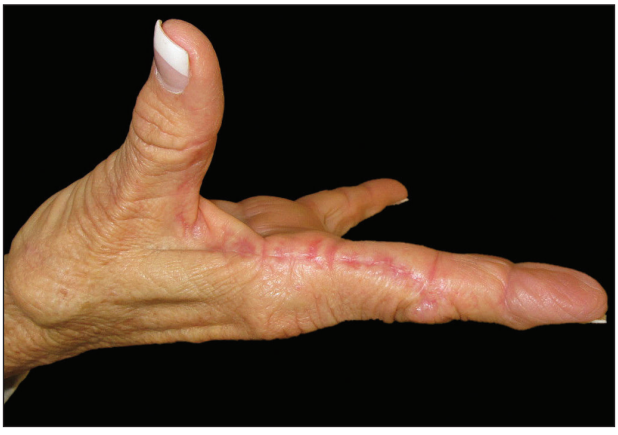

Figure 1. Longitudinal, erythematous, firm, depressed plaques (4-cm wide) along the lateral edge of the second finger and extending to the medial thumb on the right hand.

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Histology of CEMPH reveals a hyperkeratotic epidermis with an avascular and acellular replacement of the superficial reticular dermis by haphazardly arranged, thickened collagen fibers (Figure 2A–2C). Collagen fibers were oriented perpendicularly to the epidermal surface. Intervening amorphous basophilic elastotic masses were present in the upper dermis with occasional calcification and degenerative elastic fibers (Figure 2D).

Collagenous and elastotic marginal plaques of the hands is a chronic, asymptomatic, sclerotic skin disorder described in a 1960 case series of 5 patients reported by Burks et al.[1] Although it has many names, the most common is CEMPH. Collagenous and elastotic marginal plaques of the hands most often presents in white men aged 50 to 60 years.[2] Patients typically are asymptomatic with plaques limited to the junction of the palmar and dorsal surfaces of the hands with only minimal intermittent stiffness around the flexor creases. Lesions begin as discrete yellow papules that coalesce to form hyperkeratotic linear plaques with occasional telangiectasia.[3]

The etiology of CEMPH is attributed to collagen and elastin degeneration by chronic actinic damage, pressure, or trauma.[4,5] The 3 stages of degeneration include an initial linear padded stage, an intermediate padded plaque stage, and an advanced padded hyperkeratotic plaque stage.[4] Vascular compromise is seen from the enlarged and fused thickened collagen and elastic fibers that in turn lead to ischemic changes, hyperkeratosis with epidermal atrophy, and papillary dermis telangiectasia. Absence or weak expression of keratins 14 and 10 and strong expression of keratin 16 have been reported in the epidermis of CEMPH patients.[4]

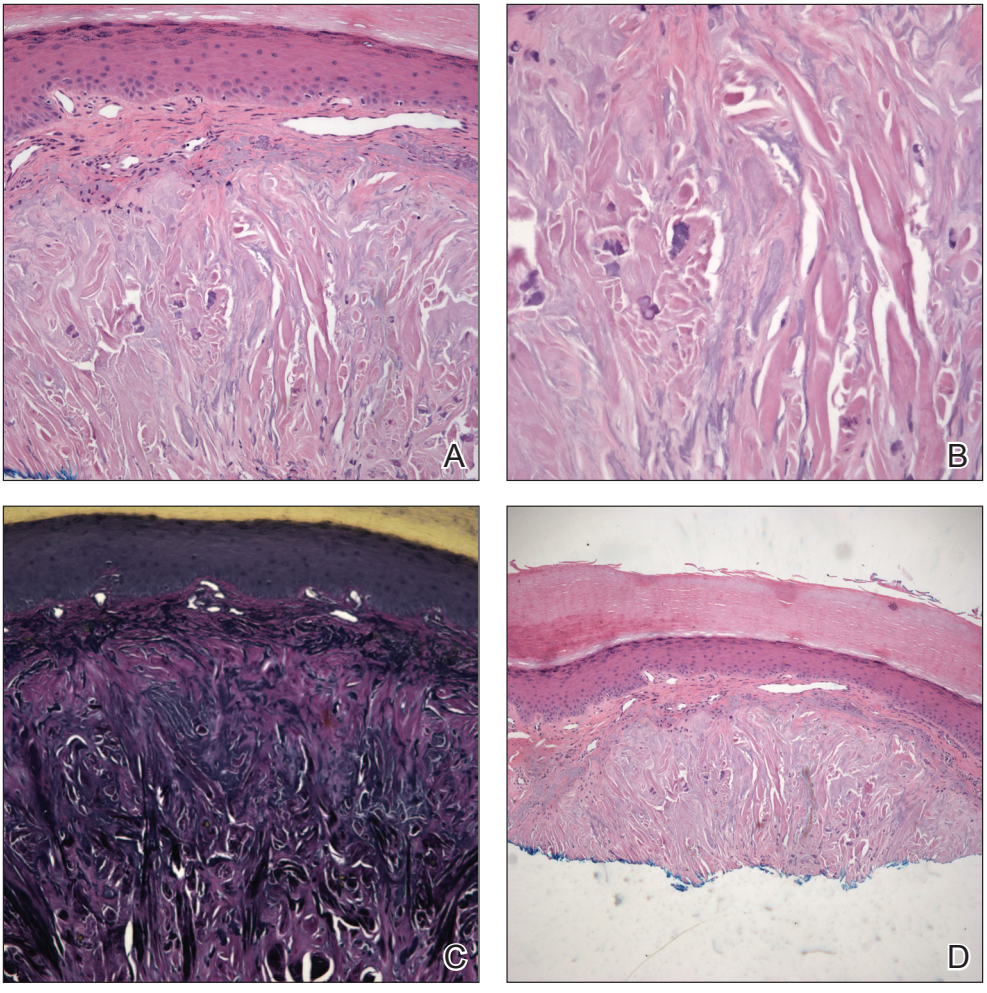

Figure 2. Histopathology shows vertically oriented, thickened collagen bundles with mixed elastin (A)(H&E, original magnification ×10). The collagen bundles are arranged haphazardly (B and C)(H&E, original magnification ×20[B]; Verhoeff-van Gieson, original magnification ×10[C]). Altered elastic fibers are present in the upper dermis (D)(H&E, original magnification ×4).

Clinical and Histological Findings of Differential Diagnoses

	Clinical Findings	Histological Findings
Collagenous and elastotic marginal plaques of the hands	6th and 7th decades, asymptomatic, junction of the palmar and dorsal surfaces of hands, discrete yellow papules coalesce to form hyperkeratotic linear plaques	Epidermis: hyperkeratotic; superficial reticular dermis: avascular and acellular replacement by thickened haphazard deposits of collagen fibers; intervening amorphous basophilic elastotic masses in upper dermis with occasional calcification and degenerative elastic fibers
Genodermatoses		
Acrokeratoelastoidosis of Costa	Autosomal dominant; occurs in children and young adults; small, firm, keratotic papules with central umbilication along the margins of the hands and feet	Epidermis: orthokeratotic hyperkeratosis from overproduction of filaggrin in the granular layer; reticular dermis: basophilic, thick, curled and fragmented elastic fibers with dilated capillaries
Focal acral hyperkeratosis	Predominately occurs in black patients aged <10 y, hands and feet are involved	Orthohyperkeratosis, moderate acanthosis, and slight hypergranulosis of the epidermis with no dermal involvement
Hyperkeratotic palmoplantar psoriasis	Thickening and scaling of the palms and soles, associated with the formation of deep painful fissures	Epidermal hyperplasia; loss of the granular skin layer with prominent dermal capillaries; mixed dermal infiltrate of lymphocytes, macrophages, and neutrophils
Hyperkeratotic palmoplantar lichen planus	Papules or yellow-colored plaques with pruritus, erythema, and compact hyperkeratosis	Epidermis: hyperkeratosis, acanthosis, and wedge-shaped hypergranulosis; degeneration in the basal layer and a lichenoid pattern of lymphocytic infiltration at the dermoepidermal junction
Palmoplantar Keratodermas		
Callosities	Repeated trauma or friction over bony prominences causing thickened and hardened skin	Prominent hyperkeratosis and acanthosis with moderate papillomatosis
Arsenic keratoses	Round, verrucous, or acuminate keratotic papules most commonly on palms and soles; hyperpigmentation changes and often hyperkeratotic lesions	Atypical keratinocytes in the epidermis with thick hyperkeratosis and vacuolated cells without solar elastosis; hyperpigmentation due to dermal arsenic deposition and melanosis

Collagenous and elastotic marginal plaques of the hands do not have a specific treatment, as it is a benign, slowly progressive condition. Several treatments such as laser therapy, high-potency topical corticosteroids, topical tazarotene and tretinoin, oral isotretinoin, and cryotherapy have been tried with little long-term success.[4] Moisturizing may help reduce fissuring, and patients are advised to avoid the sun and repeated trauma to the hands.

The differential diagnosis of CEMPH is summarized in the Table. Two genodermatoses—acrokeratoelastoidosis of Costa and focal acral hyperkeratosis—clinically resemble CEMPH. Acrokeratoelastoidosis of Costa is an autosomal-dominant condition that occurs without trauma in children and young adults. Histopathology shows orthokeratotic hyperkeratosis due to an overproduction of filaggrin in the granular layer of the epidermis. The reticular dermis shows basophilic, thick, curled and fragmented elastic fibers with dilated capillaries that can be seen with Weigert elastic, Verhoeff-van Gieson, or orcein stains. Focal acral hyperkeratosis occurs on the hands and feet, predominantly in black patients. On histology, the epidermis shows a characteristic orthohyperkeratosis, moderate acanthosis, and slight hypergranulosis with no dermal involvment.[6]

Chronic hyperkeratotic eczematous dermatitis is another common entity in the differential characterized by hyperkeratotic plaques that scale and fissure. Biopsy demonstrates a spongiotic acanthotic epidermis.[7,8]

Psoriasis of the hands, specifically hyperkeratotic palmoplantar psoriasis, is associated with manual labor, similar to CEMPH. Histology shows epidermal hyperplasia; regular acanthosis; loss of the granular skin layer with prominent dermal capillaries; and a mixed dermal infiltrate of lymphocytes, macrophages, and neutrophils.[9] Hyperkeratotic palmoplantar lichen planus presents with pruritic papules in the third and fifth decades of life. Histologically, hyperkeratosis, acanthosis, and wedge-shaped hypergranulosis with a lichenoid lymphocytic infiltration at the dermoepidermal junction is seen.[10]

Palmoplantar keratodermas due to inflammatory reactive dermatoses include callosities that develop in response to repeated trauma or friction on the skin. On histology, there is prominent hyperkeratosis and acanthosis with moderate papillomatosis.[11] Drug-related palmoplantar keratodermas such as those from arsenic exposure can lead to multiple, irregular, verrucous, keratotic, and pigmented lesions on the palms and soles. Histologically, atypical keratinocytes are seen in the epidermis with thick hyperkeratosis and vacuolated cells without solar elastosis.[12]

In conclusion, CEMPH is an underdiagnosed and underrecognized condition characterized by asymptomatic hyperkeratotic linear plaques along the medial aspect of the thumb and radial aspect of the index finger. It is important to keep CEMPH in mind when dealing with occupational cases of repeated long-term trauma or pressure to the hands as well as excessive sun exposure. It also is imperative to separate it from other diseases and avoid misdiagnosing this degenerative collagenous and elastotic disease as a malignant lesion.

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