Collagenous and Elastotic Marginal Plaques of the Hands
-A Case Report
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Collagenous and elastotic marginal plaques of the hands is a rare acquired slowly progressive and asymptomatic dermal connective tissue abnormality. Herein, we present a 72-year-old man with a 2-3 year history of asymptomatic symmetrical sclerotic linear plaques extending along the junction of the dorsal and palmar skin of the hands from the lateral aspect of the thumb to the medial aspect of the index finger. He was a farmer. No other family members had similar symptoms. A skin biopsy showed a dermal acellular zone composed of thickened bundles of collagen haphazardly arranged, some perpendicular to the epidermis, admixed with elastic fibers and amorphous basophilic elastotic materials. Granular calcium deposits were identified, particularly within degenerated collagen bundles. According to the clinical and histological features, collagenous and elastotic marginal plaques of the hands was diagnosed. Actinic degeneration and chronic repetitive pressure have been implicated in its pathogenesis, and our patient reported marked actinic exposure with chronic occupational friction. Topical treatment with urea and salicylic acid was applied, and avoiding sunlight and wearing glove were also advised. No progression about skin lesions or limitation about hand ambidexterity was mentioned after treatment. (Dermatol Sinica 26: 107-111, 2008)

Key words: Collagenous and elastotic marginal plaques of the hands, Collagen, Elastic fiber, Calcification, Pathogenesis

INTRODUCTION
Collagenous and elastotic marginal plaques of the hands (CEMPH) is an acquired slowly progressive and asymptomatic sclerotic skin disorder. The first case series in 1960 describe degenerative alternation of collagen and elastic fibers in the dermis, confined to the junction of the palmar and dorsal surfaces of hands. The main clinical manifestation is linear sclerotic plaques along the junction of the palmar and dorsal surfaces of hands in elder males. The medial aspect of the thumb and radial aspect of the index fingers are usually involved. Here, we present a typical case living in Hualien, Taiwan.

CASE REPORT
A 72-year-old man presented with symmetrical, slowly progressive sclerotic plaques on the radial side of both hands, which he had for approximately 2-3 years. There was no painful or pruritic sensation. No arthralgia or
Fig. 1
(A) Linear sclerotic plaques along the junction of the dorsal and palmar skin of bilateral hands from medial aspect of the thumbs to the lateral aspect of the index fingers.
(B) Slight yellow hue and glistening indurative plaques with adhesive scalings and central depression.

Fig. 2
(A) Thinning or hyperplastic epidermis with hyperkeratosis (H&E, original magnification x100)
(B) There is diffuse acellular zone composed of thickened collagen bundles in haphazard array or perpendicular to the epidermal surface (H&E, original magnification x200).

Fig. 3
(A) Masson trichome stain (original magnification x100) identified blue-color broad collagen bundles perpendicular to epidermis.
(B) Pinkus acid orcein stain (original magnification x200) identified intervening elastotic materials between broad collagen bundles.
Raynaud’s phenomenon was reported. He was a farmer, and had a history of occupational solar exposure with chronic pressure and friction over the hands. There was no family history of similar lesions. No significant past medical or drug history was noted. The dermatologic examination revealed linear sclerotic plaques along the junction of the dorsal and palmar skin of the bilateral hands from the medial aspect of the thumbs to the lateral aspect of the index fingers (Fig. 1A). These hyperkeratotic plaques with adhesive scaling and a central depression had a slightly yellow hue and glistered (Fig. 1B). The initial clinical differential diagnosis included localized sclerosis, solar elastosis, focal acral hyperkeratosis, and acrokeratoelastoidosis. The skin biopsy revealed a thinning epidermis with hyperkeratosis and a large acellular zone in the dermis composed of thickened collagen bundles in a haphazard array or perpendicular to the epidermal surface (Fig. 2A). The thickened collagen bundles had basophilic changes, and amorphous basophilic materials deposited in the dermo-epidermal junction, inter-collagen space, and lower part of the acellular zone (Fig. 2B). Masson trichrome stain identified broad collagen bundles (Fig. 3A), and Pinkus acid orcein stain showed intervening elastotic materials between the broad collagen bundles (Fig. 3B). A von Kossa stain showed several areas of granular or clumped calcification over the dermoepidermal junctions and the lower part of the acellular zone. CEMPH was diagnosed according to the clinical and histological findings. Topical treatment with urea and salicylic acid was applied, and avoiding sunlight and wearing glove were also advised. No progression about skin lesions or limitation about hand ambidexterity was mentioned after treatment.

DISCUSSION

Since the first case was reported in 1960, different names have been given to this skin disorder, including degenerative collagenous plaques of the hands, keratoelastoidosis marginalis, digital papular calcific elastosis, and collagenous and elastotic marginal plaques of the hands (CEMPH). CEMPH is the commonly accepted name, because it describes the collagenous and elastotic changes in this skin disorder.

CEMPH develops in the sixth to seventh decade of life. Men are more predominantly affected than women. A high proportion of cases are reported in Caucasians. However, there is no familial predisposition. The clinical manifestations are bilateral with symmetrical linear plaques at the junction of the dorsal and palmar skin from the medial aspect of the thumb distally onto the lateral aspect of the index finger. The lesions extend to the distal interphalangeal joint of the index finger. The ulnar side of the hand and other sites on the fingers are rarely involved. The course of this disease is slowly progressive and asymptomatic.

The pathological findings of CEMPH reveal an acellular zone of haphazardly arranged collagen with some bundles running perpendicular to the epidermis. The bundles of collagen are admixed with fragmented elastic fibers and distinctive angulated amorphous basophilic elastotic masses in the upper dermis. These masses are composed of degenerative elastic fibers and calcium. Chronic actinic damage and repetitive long-term trauma and pressure inducing degenerative collagenous and elastotic processes are implicated in the pathogenesis of CEMPH. Three different evolutional stages are involved in the pathogenesis: an initial linear padded stage, an intermediate padded plaque stage, and an advanced padded hyperkeratotic plaque stage. Enlarged and coalescent thickened collagen and elastic fibers cause vascular compression and ischemic changes, marked 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 recorded in CEMPH patients. The effects of vari-
The age of the patients and distribution of the skin lesions in AKE are different from CEMPH. AKE is a rare skin disorder which occurs at an early age, usually in childhood or adolescence. The skin lesions develop on the hands as well as the feet. No predisposing factors such as trauma or sun exposure have been elicited in AKE. Sporadic as well as familial cases have been discovered. The histologic findings in AKE are hyperkeratosis in the epidermis and fragmentation of elastic fibers in the reticular dermis. Focal acral hyperkeratosis (FAH) is another similar clinical entity occurring in African Americans below the age of ten years. The lesions in FAH are also distributed on the hands and feet. The most characteristic histologic features are orthohyperkeratosis, moderate acanthosis and slight hypergranulosis of the epidermis without dermal changes. Other keratinization disorders that present keratotic papules along the borders of the hands and feet in the differential diagnosis of CEMPH include verruca plana, acrokeratosis verruciformis of Hopf, punctate porokeratosis and punctate palmoplantar keratoderma.

No specific treatment has been proposed in past studies. Because the clinical course is slowly progressive and asymptomatic, no definite remedy is indicated. Sun-protection and avoidance of repetitive pressure or friction should be suggested to prevent disease progression.

REFERENCES

手部膠原與彈力纖維板塊
-病例報告

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手部膠原性與彈力纖維板塊是一種罕見後天性皮膚真皮結締組織的異常，病程緩慢，且沒有明顯的臨床症狀。我們提出一位72歲的男性病人在左右手掌心與掌背的交界處，產生無症狀性對稱性的直線狀硬皮變化的斑塊。病灶的進展是由大拇指的外側往食指的內側進行，病灶的產生約有2-3年。職業是農夫，家族其它的成員並沒有相同的症狀。皮膚切片下可見沒有細胞組成的區域在真皮層，增厚的膠原蛋白凌亂地排列，有些膠原蛋白的排列會與表皮走向垂直，夾雜有嗜鹼性的鈣質沈積與退化的彈力纖維。經由臨床症狀與組織切片，診斷為手部膠原與彈力纖維板塊。陽光傷害與慢性重複性壓力摩擦是形成手部膠原與彈力纖維板塊的可能致病機轉。透過尿素與水楊酸的局部治療，並建議病人防曬與穿戴手套，發現皮膚的症狀並沒有持續進行或是手指的靈活度有受影響。（中華皮誌：26: 107-111, 2008）