Case 8a
A 60 years old Thai woman from Suphanburi

Chief complaint
Hyperkeratotic plaques at the side of both thumbs and index fingers for 4 months

Present illness
4 months ago, she developed asymptomatic hyperkeratotic plaques on the distal part of lateral side of both index fingers and the medial side of both thumbs. Then the lesions extended to proximal part and 1st web spaces.

Underlying disease
DM, HT, dyslipidemia

Family history
nil

Physical examination
GA
WNL

Skin
Linear hyperkeratotic plaques along lateral side of both index fingers, medial side of both thumbs and 1st web spaces, not tender.

Histopathology (S04-9188A)
There are massive aggregated degenerative elastic fiber intermingle with coarse collagen bundle in the upper dermis.

Compact orthokeratosis in epidermis with mild acanthosis.

Diagnosis
Acrokeratoelastoidosis

Presenter
Suthida Suwanachote

Consultant
Siripen Puavilai

Discussion
Acrokeratoelastoidosis(AKE) is a rare papular palmoplantar keratosis characterized by firm papules or plaques on the sides of the hands and feet. It is a disease with autosomal dominant transmission.

The majority of cases develop before the age of 20 years, unlike this case.

Therefore the differential diagnosis of AKE includes focal acral hyperkeratosis(FAH) and degenerative collagenous plaque of the hand(DCPH), because of their clinical similarities. Histologically there are elastic tissue changes in AKE but not in FAH, in DCPH is predominantly a collagen degeneration.

The effective treatment for AKE has not yet been reported. Since the progression of AKE is slow and the lesion itself is often asymptomatic, no treatment is indicated until an effective and safe therapy can be found.
**TABLE 1. The Differential Diagnosis of AKE**

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Clinical features</th>
<th>Histologic features</th>
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<tbody>
<tr>
<td>Acrokeratoelastoidosis (AKE)</td>
<td>Small round-oval to rhomboid-shaped, yellowish papules on palmar/plantar surfaces of the hands and feet</td>
<td>Hyperkeratosis, epidermal hypertrophy, and decreased elastic fibers in the dermis</td>
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<tr>
<td>Focal acral hyperkeratosis (FAH)</td>
<td>Identical to AKE, except it is more common in blacks</td>
<td>Lack of elastorhexis in the dermis, elastic tissue is intact</td>
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<tr>
<td>Degenerative collagenous plaques (DCP)</td>
<td>More common in older patients (40-60 years), usually associated with chronic sun exposure</td>
<td>Marked degeneration of collagen and elastin fibers</td>
</tr>
</tbody>
</table>


**Reference**

Case 8b
A 67-year-old woman from Petchaboon

Chief complaint: Nodules and tumors on the dorsum of left hand for 30 years

Present illness: She had asymptomatic papules and nodules which gradually enlarge to tumor during the past 30 years

Past history: No underlying disease

Family History: Nil

Physical Examination:
The group of multiple soft, dome-shaped, skin colored papules and nodules and firm tumors on dorsum of left hand
Multiple discrete hyperkeratotic skin color papules on dorsum and lateral aspect of fingers both hands
Café-au-lait macules, axillary freckling and Lisch nodule were absent

Histopathology: (S04-9493)
A. Proliferation of oval and spindle cells tumor with comma shape nuclei in loose stroma
B. Compact hyperkeratosis and swelling epidermis in association with solar elastosis

Diagnosis:
1. Segmental neurofibromatosis
2. Focal acral hyperkeratosis

Presenter: Paan Tan
Consultant: Siripen Puavilai

Comment:
Segmental neurofibromatosis (neurofibromatosis type V) is a rare disorder characterized by cutaneous neurofibromas, with or without café-au-lait macules, that
are limited to a circumscribed body segment. The affected area can vary in size from a narrow strip to an area encompassing half of the body. Distribution may be unilateral or bilateral and symmetric or asymmetric. A median age of onset of is about 28 years. Skin lesions are most commonly found in a unilateral distribution over a cervical or thoracic dermatome. Café-au-lait macules have been reported to occur in 26 percent. Systemic disease is uncommon, and most patients have no family history of neurofibromatosis. Segmental neurofibromatosis is thought to arise from a post-zygotic neurofibromatosis type 1 (NF1) gene mutation. An NF1 microdeletion was identified in a patient with segmental neurofibromatosis using fluorescent in situ hybridization. The mutant allele was present in a mosaic pattern in cultured fibroblasts but was absent from fibroblasts obtained from the patient's normal skin.