Case 10.1

A 59-year-old Thai woman from Nakhon Sawan

Chief complaint: Multiple asymptomatic skin bumps on both dorsum of hands for 2 years

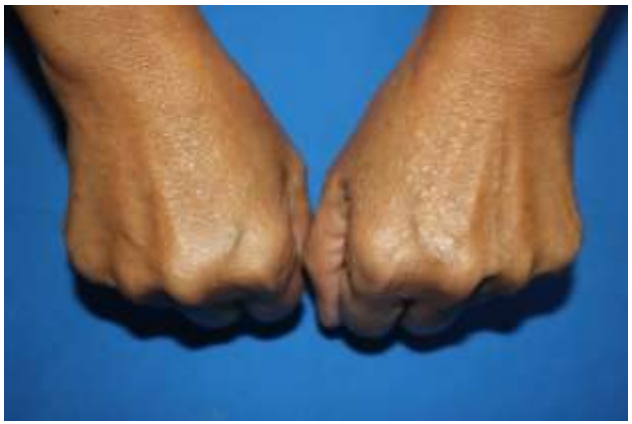

Fig. 10.1.1

Present illness: The patient gradually developed multiple, asymptomatic, skin bumps on both dorsum of hands for 2 years. She had unknown topical treatment without improvement. The lesions progressed to both second and third fingers with pruritus during the past year. She denied relationship with sun exposure. No other family member had been affected.

Past history: allergic rhinitis

Current medication: none

Physical examination: unremarkable

Dermatologic examination: (Fig. 10.1.1)

- Multiple discrete skin-colored papules on both dorsum of hands
- Skin-colored lichenified plaques with minimal scale on both second and third fingers

Histopathology: (S17-036177) (Fig. 10.1.2, 10.1.3)

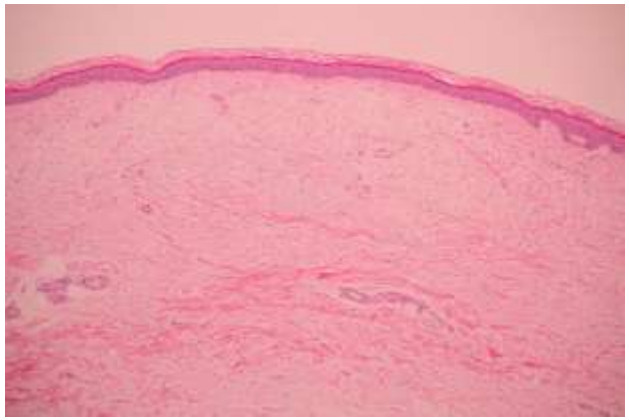

Fig. 10.1.2

- Circumscribed area of mucin deposit with scatter fibroblasts in the upper to mid dermis

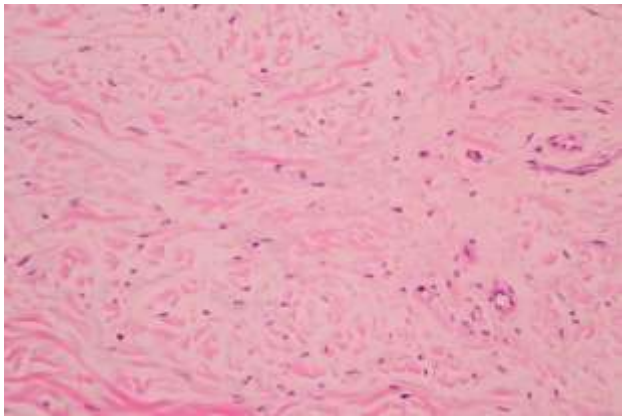

Fig. 10.1.3

- Positive mucin

Investigation:

- Anti-HIV: negative

Diagnosis: Acral persistent papular mucinosis

Treatment:

- 0.1% tacrolimus ointment apply lesions twice a day

Case 10.2

A 61-year-old Thai woman from Bangkok

Chief complaint: Multiple, asymptomatic, skin bumps on both forearms for 10 years

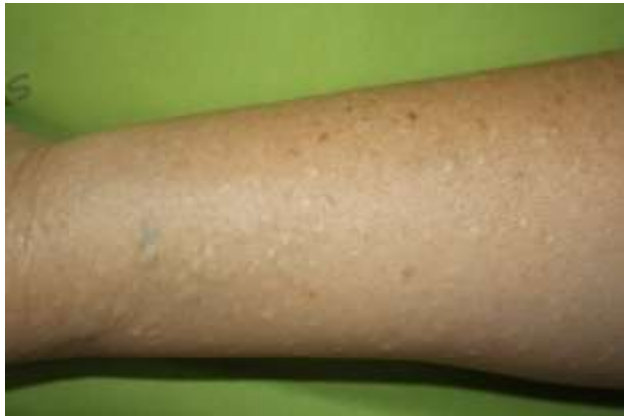

Fig. 10.2.1

Present illness: The patient developed a gradually progressive asymptomatic skin bumps on both forearms for 10 years. She had no previous treatment. No relationship with sun exposure. Her family history was unremarkable.

Past history: allergic rhinitis, dyslipidemia

Current medication: simvastatin 20 mg/day, cetirizine 10 mg/day

Physical examination: unremarkable

Dermatologic examination: (Fig. 10.2.1)

- Multiple discrete, skin-colored papules on extensor surface of both forearms

Histopathology: (S18-012201) (Fig. 10.2.2, 10.2.3)

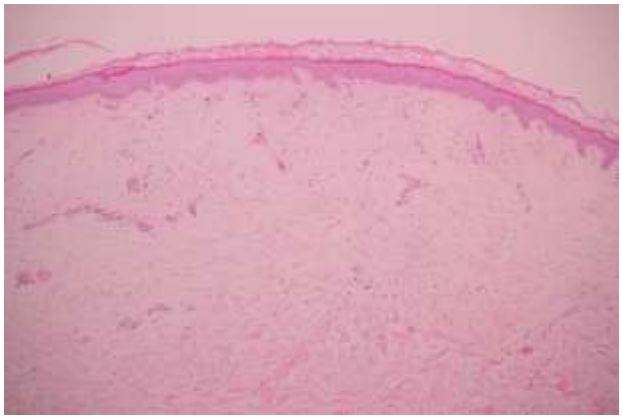

Fig. 10.2.2

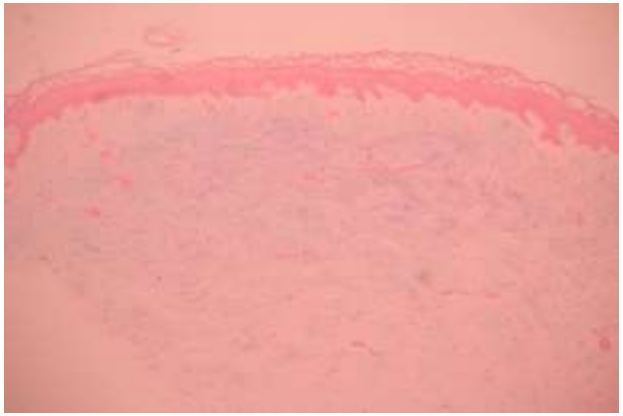

Fig. 10.2.3

- Diffuse mucinous deposit with slightly increased number of blood vessels with plump fibroblasts in the upper and mid dermis
- Positive mucin

Investigation:

- Anti-HIV: negative

Diagnosis: Acral persistent papular mucinosis

Treatment:

- 0.05% clobetasol propionate cream apply lesions twice a day

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Consultant: Vasanop Vachiramon, MD

Discussion:

Acral persistent papular mucinosis (APPM) is a rare subtype of localized lichen myxedematosus with an unknown etiology. The condition was firstly described by Rongioletti et al in 1986 as a distinct clinical and histological form of dermal mucinoses.[1] Later, in 2001, Rongioletti et al established a classification of lichen myxedematosus into 2 main groups: the generalized form and localized form. The generalized form or scleromyxedema, which has an association with systemic disorders and, almost constantly, with paraproteinemia. The localized form or localized lichen myxedematosus, which criteria are as follows: [1] papular or nodular/plaque eruption; [2] mucin deposition with variable fibroblast proliferation; and [3] the absence of both monoclonal

gammopathy and thyroid disease. APPM was classified as a subtype of localized lichen myxedematosus.[2] HIV infection has been anecdotally reported in patients with APPM.[3]

The clinical manifestation of APPM is few to multiple ivory to flesh-colored papules 2-5 mm in diameter located on the dorsum of wrists and hands and extensor surface of distal forearms. The lesions may gradually progress in number, but do not spontaneously resolve.[4] Although lesions are generally asymptomatic, pruritus also has been reported.[5, 6] APPM occurs more common in female and onset at adulthood.[4, 7-9]

Dermoscopic appearance of APPM revealed a reddish-orange background color with multiple red dotted and globular vessels.[10]

Histopathological features from hematoxylin and eosin staining show a focal deposition of mucin in the papillary and upper reticular dermis, typically sparing a subepidermal zone. The number of fibroblast was variable. A well-circumscribed deposition of mucin is positively stained when stained with alcian blue.[2, 4, 9]

The differential diagnosis includes granuloma annulare, molluscum contagiosum, acrokeratoelastoidosis, keratoelastoidosis marginalis of the hands, hereditary papulotranslucent acrokeratoderma, focal acral hyperkeratosis, degenerative collagenous plaques of the hands, lichen amyloidosis, eruptive syringoma and syringotropic mycosis fungoides as well as other forms of mucinosis.[9] Diagnosis of APPM can relies on clinical and histologic criteria in Table 1

This condition is benign in nature and the treatment is rarely necessary due to the absence of symptom. Topical corticosteroids may show some benefit but recurrent is possible if the medication is discontinued.[11] A partial response has been reported with 0.1% tacrolimus ointment.[12] Others destructive modalities such as electrofulguration[9], electrocoagulation[13], Erbium-YAG laser[14] can be used but scarring can occurred as a sequel. Spontaneous resolution may occur in localized lichen myxedematosus, but no tendency to spontaneous resolution was previously reported in APPM.[9]

Table 1: Diagnostic criteria of APPM

Clinical criteria	Histologic criteria
• 2-5 mm, few to multiple, ivory to flesh-colored papules	• Focal, well-circumscribed mucin
• Exclusively located on back of hands, wrists, occasionally distal aspect of forearms	• Mucin in papillary and mid dermis, never confined to deep reticular dermis
• Persist without spontaneous resolution, may increase in number	• Spared grenz zone
• Predominately female patients	• Variable fibroblast proliferation, usually absent
• No systemic disease overlap	
• No associated gammopathy	

Adapted from Harris JE et al[4]

References:

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