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CASE REPORT

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A rare case of acral persistent papular mucinosis

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Abstract

In patients with asymptomatic papules of hands and feet, a clinical differential of acral persistent papular mucinosis should be thought of.

KEYWORDS

dermatology, mucinosis, myxedematosus, scleromyxedema

1 | INTRODUCTION

Acral persistent papular mucinosis is a very rare subtype of localized lichen myxedematosus. Based on the diagnostic criteria proposed by Rongioletti and Rebora in 2001,[1] there are only 35 cases of APPM reported so far.[2,3] We present a 66-year-old Chinese man with asymptomatic papules on his hands and wrists, which adequately meet clinical and pathological criteria for acral persistent papular mucinosis.

2 | CASE HISTORY/ EXAMINATION

Our patient complained of 9 months of persistent and asymptomatic skin lesions on his upper limbs, especially on hands and wrist. The lesions appeared spontaneously and progressively enlarged. He denied previous insect bites, injuries, or trauma to the affected sites. There was no relationship with sun exposure. No other family member was affected. He had no past medical history apart from longstanding hyperlipidaemia which was controlled with simvastatin. Physical examination revealed multiple small 3 to 4 mm firm round skin-colored papules located exclusively on the dorsum of the hands, wrists, and forearms (Figure 1).

3 | DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS, AND TREATMENT

Differential diagnoses included papular granuloma annulare, molluscum contagiosum, lichen nitidus, lichen amyloidosus, papular elastorrhexis, and eruptive collagenomas. Punch biopsy specimen from a papule was performed. Histology showed focal spongiosis, lymphocytic exocytosis, and mild perifollicular inflammatory infiltrate with no increased fibroblasts (Figure 2). The upper and middle dermis showed increased mucin deposits which stained positively with Alcian blue. Laboratory studies were normal, showing no evidence of plasma cell dyscrasia, thyroid dysfunction, connective tissue disorders, and human immunodeficiency virus infection. Hepatitis C infection screen was not performed. The patient was diagnosed with acral persistent papular mucinosis. He declined treatment as he had no symptoms.

4 | OUTCOME AND FOLLOW-UP

The chronic but benign disease course of acral persistent papular mucinosis was explained to the patient. Given the absence of any systemic involvement and active symptoms, the patient chose to observe his skin for spontaneous resolution. A future follow-up appointment was offered.

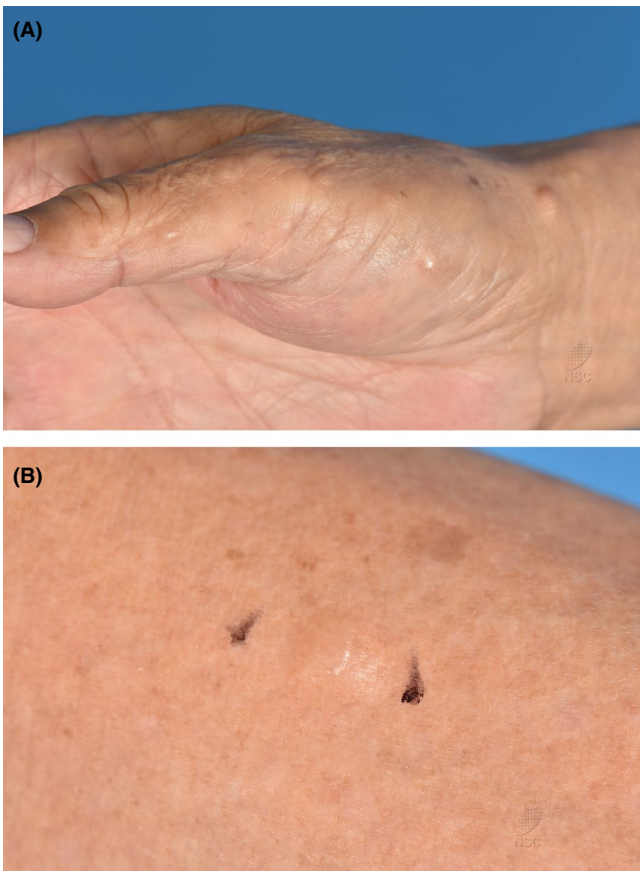

FIGURE 1 (A) Clinical photographs of skin-colored papules on hands and forearms, (B) Close-up image of papule

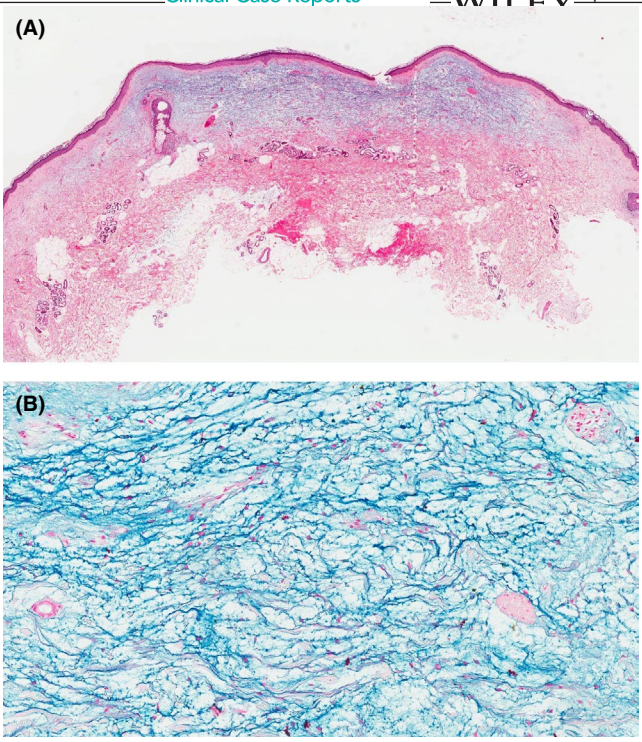

FIGURE 2 (A) Increased upper and mid-dermal mucin deposits are seen (H&E, magnification x20), (B) mucin deposits stain positively with Alcian blue (Alcian blue, magnification x200)

5 | DISCUSSION

Cutaneous mucinoses are heterogeneous disorders in which mucin deposits abnormally in skin. The etiology is unclear. They are classically divided into two groups; (a) primary forms, in which mucin accumulation is the major histologic feature and leads to clinically distinctive lesions and (b) secondary forms, in which mucin accumulation is only an associated finding.

Primary cutaneous mucinoses can be divided into degenerative or inflammatory forms and hamartomatous or neoplastic forms. Degenerative or inflammatory primary mucinosis is further subclassified into dermal or follicular types. In 2001, Rongioletti and Rebora proposed a classification for dermal mucinosis.[4] It is differentiated into two main groups: generalized form (or scleromyxedema) and localized form (or lichen myxedematosus). The former is associated with systemic disorders which include paraproteinemia, endocrinopathies, autoimmune connective tissue diseases, and hematologic malignancies. The latter has no associations with systemic diseases. The diagnostic criteria of scleromyxedema and lichen myxedematosus are summarized in Table 1. Cases that do not meet criteria for either forms are classified as atypical.

The localized variants of lichen myxedematosus are further subdivided into four distinct subtypes, including (a) a discrete papular form, (b) acral persistent papular mucinosis, (c), cutaneous mucinosis of infancy, and (d) a pure nodular form. The first case of acral persistent papular mucinosis was reported by Rongioletti et al as a subtype of lichen myxedematosus in 1986.[5] In acral persistent papular mucinosis, small papules are localized exclusively on dorsum of hands, wrists, and extensor aspects of distal forearms with no other clinical or laboratory manifestations. Lesions may persist and increase in number slowly with time. Histologically, Alcian blue staining demonstrates upper dermal mucin accumulation with collagen fibers separation from hyaluronic acid deposition.

To date, only 35 cases of acral persistent papular mucinosis have been reported.[2,3] It is reported to have a female predominance, and mean age at onset was 42.9 ± 15.9 years.[2]

TABLE 1 Diagnostic criteria of scleromyxedema versus lichen myxedematosus

Scleromyxedema	Lichen myxedematosus
Generalized papular eruption and sclerodermoid features	Papular eruption (or nodules and/or plaques due to confluence of papules)
Microscopic triad (mucin deposition, fibroblast proliferation, fibrosis)	Mucin deposition with variable fibroblast proliferation
Monoclonal gammopathy	Absence of monoclonal gammopathy
Absence of thyroid disorder	Absence of thyroid disorder

The possible association with solid organ or hematological malignancies remains unclear. As patients with acral persistent papular mucinosis are generally asymptomatic, treatment is rarely necessary. For patients who request treatment due to cosmetic concerns, topical corticosteroids, calcineurin inhibitors, liquid nitrogen, electrocoagulation, laser ablation, and surgical excision have been reported as successful therapeutic options.[2,3,6]

We report another case of acral persistent papular mucinosis. This diagnosis should be considered in patients with acral papules. It is appropriate to exclude other systemic involvement when mucin accumulation is confirmed.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

JJHT and DYW: prepared manuscript. NG: described dermatopathology input.

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