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Acral Persistent Papular Mucinosis: is it an under-diagnosed disease?

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Abstract

Acral persistent papular mucinosis is a subtype of localized lichen myxedematosus. It presents as acrally located papules with a benign, but persistent course. It is a scarcely reported disease. We present a female with both the clinical and histopathological described criteria

Introduction

Acral persistent papular mucinosis (APPM) is a type of papular mucinosis. Also known as lichen myxedematosus or scleromyxedema, this is a group of diseases is characterized by mucin deposits in skin. We can classify these diseases into clinicopathological types. The first type is a generalized/sclerodermoid form, also called scleromyxedema, with systemic manifestations. The second is a localized form without systemic involvement. There are five subtypes: discrete papular, nodular, self-healing papular mucinosis, papular mucinosis of infancy, and acral persistent papular mucinosis. Finally, the cases that don't fit any of the groups are considered as atypical forms.

APPM was first described by Rongioletti *et al* in 1986 as a new condition, [1]. Later, in 2001, it was classified as a subtype of localized lichen mixedematosus [2]. It is characterized by acrally located mucinous papules, a benign but persistent course, and an absence of systemic abnormalities [2].

To date, there are 34 reported cases in the literature which fit with the diagnostic criteria (Table 1) [1,3,4,5,6,7]. Another 6 cases have been described, but they do not meet the established criteria for APPM.

Case synopsis

A 53-year old woman presented with a one-year history of a progressive eruption of asymptomatic papules on the back of her hands, wrists, and distal forearms. Physical examination revealed multiple 2-5 mm flesh-colored to translucent papules, which were round and firm with a smooth surface (Figure 1). The skin between lesions appeared to be normal. There were no other similar lesions on the rest of the body.

Routine laboratory tests revealed no abnormalities. Thyroid studies (T3, T4, thyroid-stimulating hormone, anti-thyroid antibodies), serum protein electrophoresis, IgM, IgA, and Ig G serum levels were all normal.

The histologic examination of one of the lesions showed a normal epidermis with a localized deposit of nodular mucin in the papillary and upper reticular dermis, sparing the deep dermis. There was increased spacing between collagen bundles, but the number of fibroblasts was not increased (Figure 2).

Our patient satisfied the diagnostic criteria of APPM (Table 1).

Because the lesions were asymptomatic and not very apparent no treatment was proposed.



Figure 1. Flesh-colored to translucent papules, round and firm, on the back of both hands

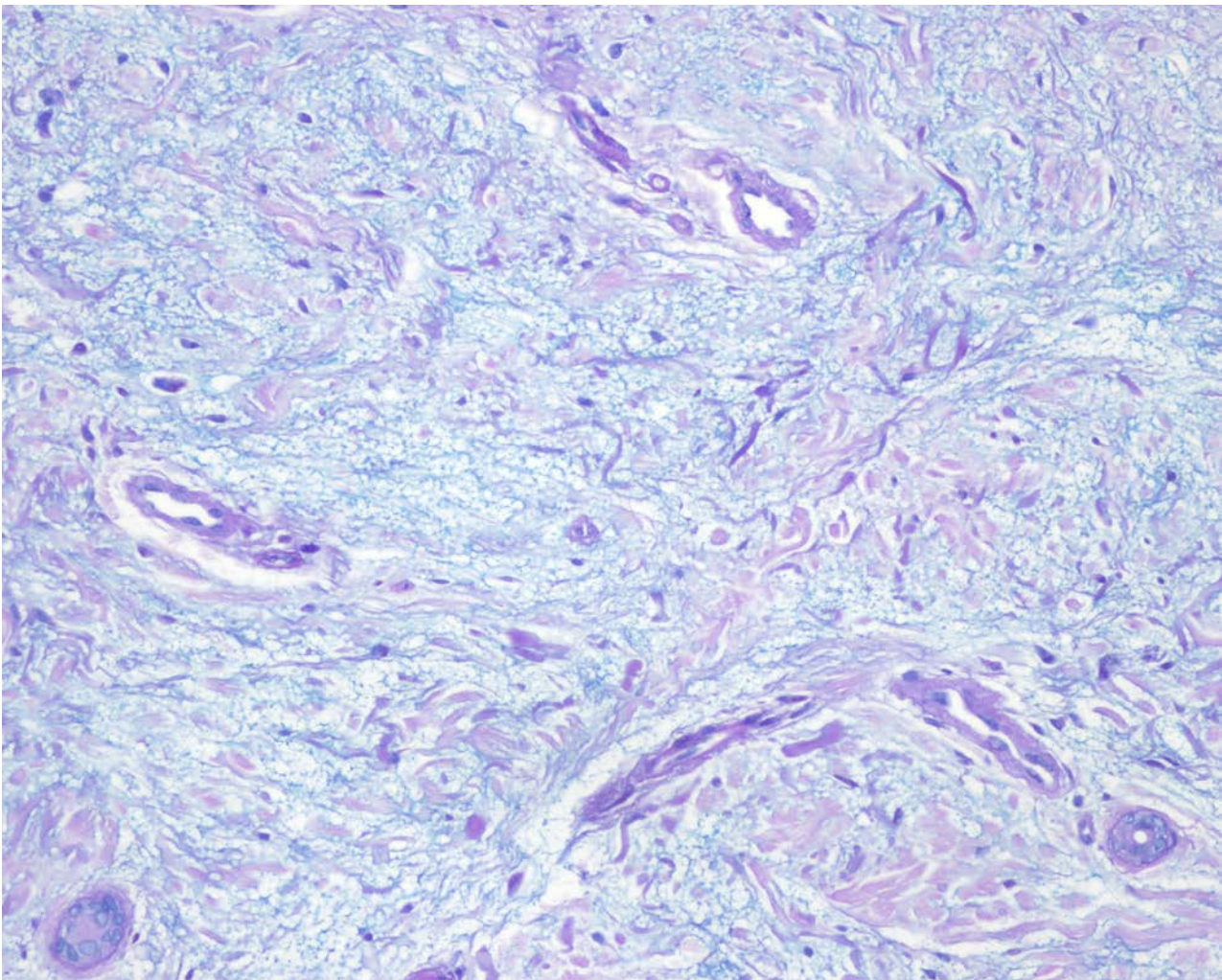


Figure 2. 40x Alcian blue. Normal epidermis with a localized deposit of nodular mucin in papillary and upper reticular dermis, sparing the deep dermis.

Table 1: Clinical and histologic criteria of APPM [5]

Clinical criteria	Histologic criteria
2-5 mm, few to multiple, ivory to flesh-colored papules	Focal, well-circumscribed mucin
	Mucin in papillary and mid dermis, never confined to deep reticular dermis
Persists 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	

Discussion

The etiology of lichen mixedematous remains unknown [3]. An over stimulation of fibroblasts has been implicated in patients with human immunodeficiency virus infection, but not in other cases [4]. In the localized form, the lesions are confined to only a few sites and mainly affects middle-aged women. It is not associated sclerotic features, paraproteinemia, systemic involvement, or thyroid disease [2]. Typically, lesions are asymptomatic, persist, and gradually increase in number [5].

It is important to distinguish APPM from the papular form of lichen myxedematosus. The latter is characterized by the presence of flesh colored translucent papules localized on the extremities and on the trunk; it is usually self-limited [2].

Histologically, it is characterized by well-circumscribed nodular mucin deposits between the collagen bundles, particularly in the superficial dermis, but also in the mid reticular dermis. Only occasionally is there an increased number of fibroblasts present. A Grenz zone is a characteristic finding. The overlying epidermis is normal [5].

Treatment is not required; it is generally a cosmetic concern. Destructive therapies such as liquid nitrogen, carbon dioxide laser, or electrodesiccation [6] are not recommended because they can leave scars [3]. Topical corticosteroids, tacrolimus [7], and pimecrolimus have been used with some success [3,8], although more studies are required to show the benefit of various treatments [5].

Learning points

1. APPM is a localized form of lichen myxedematosus (also called papular mucinosis).
2. It is generally underdiagnosed
3. No association with systemic disease or gammopathy is present.
4. Histologically, it is characterized by the presence of mucin in the papillary and mid dermis and by variable fibroblast proliferation.
5. It is persistent and asymptomatic.
6. Treatment is not required.

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