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A case report and differential diagnosis of pruritic pretibial skin lesions

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Abstract

Pretibial pruritic papular dermatitis (PPPD) is a clinical entity first described in 2006. The etiology is uncertain; however, gentle chronic rubbing is likely to be the reason for the skin reaction. Pretibial pruritic lesions may reflect many different systemic diseases and dermatoses. We present a 61-year-old patient with a 2-year history of pruritic pretibial xerosis, keratotic erythematous to brownish papules, and excoriations. Differential diagnosis excluded papular mucinosis, myxoedema, stasis dermatitis, lichen simplex chronicus, prurigo nodularis, lichen amyloidosis, and lichen planus. Regarding clinical-histological correlation, we confirmed a diagnosis of PPPD.

Keywords: pretibial dermatitis, pruritus, rubbing

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Introduction

Pretibial pruritic papular dermatitis (PPPD) is a clinical entity first described in 2006 as an original article posted in *The American Journal of Dermatopathology* (1). The etiology is not well understood; however, gentle chronic rubbing is likely to be the reason for the skin reaction (1). There are no obvious data in the literature for the initial cause of pruritus that starts the rubbing; however, xerosis, psychological stress, and irritants are the most common contributing factors (1). Although it is considered uncommon, PPPD is probably underreported because it is often clinically mistaken for other itchy papular or papulosquamous disorders (1).

Unilateral or bilateral pretibial erythematous to flesh-colored pruritic papules with a smooth surface are present. In later stages, the grouping of papules can create a cobblestone appearance (1, 2).

Histology of PPPD shows mild compact orthokeratosis, flattening of rete ridges, superficial dermal fibrosis, moderate superficial and mid-dermal lymphocytic infiltrate with few eosinophils, and stellate cells (1, 2). These features are an uncommon histopathological pattern and could be a reflection of gentle rubbing and scratching.

Case report

A 61-year-old patient with a 2-year history of pruritic pretibial skin lesions was treated at an outpatient dermatovenerology department in Ljubljana. The skin lesions became worse during the summer, when the patient sweated and wore long pants. She was treated with Euthyrox for 11 years because of hypothyroidism. An allergy for penicillin was suspected. She had no family history of skin disorders.

A clinical exam showed pretibial dry skin with keratotic erythematous to brownish papules and excoriations. In addition, some telangiectasias were seen on the lower extremities (Fig. 1).

She was advised to use compression therapy with long-stretched bandages during the day, topical methylprednisolone aceponate 1 mg / 1 g corticosteroid cream two times per day on pretibial skin lesions, and 10% olive oil in Belobaza® cream as an emollient for skin care. On the second visit, after 5 months, skin lesions with intense pruritus were still present. In the meantime, she was using prescribed corticosteroid cream without the compression therapy and emollient.

Because of treatment inefficiency, a skin biopsy was made. Histology showed orthokeratosis, slight, irregular hyperplasia of the epidermis, and thickened dermal papillae with homogenization of the dermis. There were numerous stellate and spindle cells with superficial to mid-dermal perivascular and interstitial mononuclear infiltrate (Fig. 2). These histological features were typical for PPPD.

The treatment of the patient was continued with hydrocortisone butyrate 1 mg / 1 g corticosteroid cream, oral antihistamine loratadine 10 mg one tablet per day, and 10% olive oil in Belobaza® cream. After 3 months, when she came for her third visit, complete resolution of the skin lesions and pruritus was observed.

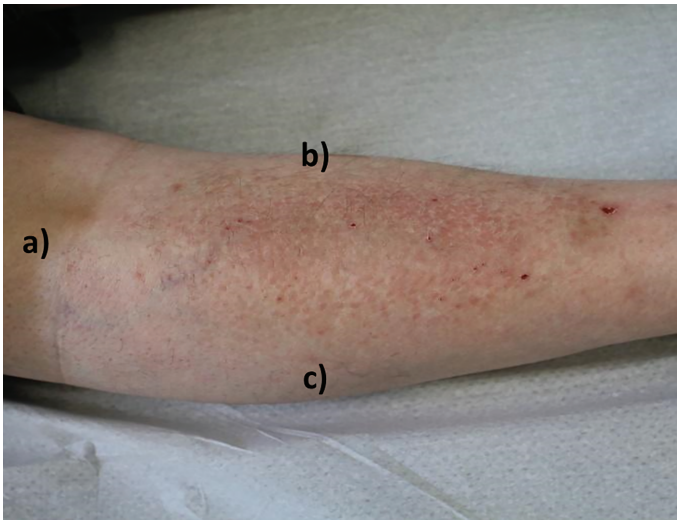

Figure 1 | Pretibial dry skin with keratotic erythematous to brownish papules grouping into plaque with a cobblestone appearance, with some excoriations and telangiectasia; a) knee, b) lateral part of the calf, and c) medial part of the calf.

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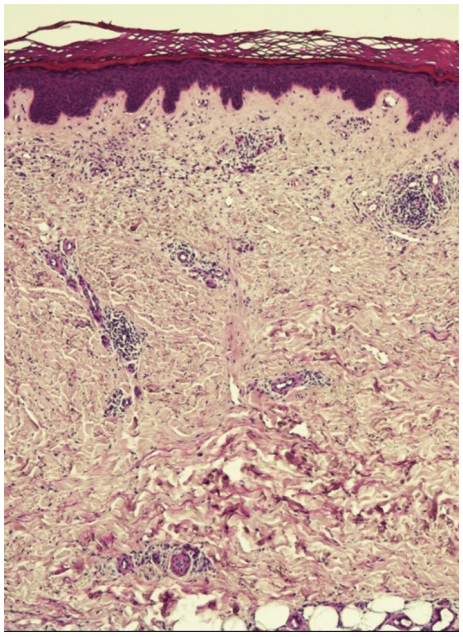

Figure 2 | Histological features typical of pretibial pruritic papular dermatitis (PPPD) include orthokeratosis and slight, irregular hyperplasia of the epidermis and thickened dermal papillae with homogenization of the dermis. There are numerous stellate and spindle cells along with superficial to mid-dermal perivascular and interstitial mononuclear infiltrate.

Discussion

Due to a medical history of hypothyroidism and clinical presentation of venous disease, differential diagnosis was expanded to papular mucinosis, myxedema, and stasis dermatitis. Papular mucinosis, also known as localized lichen myxedematosus, presents with pretibial waxy papules and plaques, mostly without pruritus, systemic involvement, or thyroid dysfunction (3, 4). Myxedema occurs in patients with Graves' disease with diffuse pretibial non-pitting edema and skin thickening with nodules (5). Only varicosities with mild perimalleolar pitting edema were seen in our case, without other features of venous insufficiency (6).

According to the differential diagnosis mentioned above, one of the following histological reports was expected, but not found. In papular mucinosis, there is accumulation of mucin, there is edema in the upper and mid-dermis, and fibroblast proliferation is seen histologically (3). In pretibial myxedema, there are depositions of mucin in reticular dermis, separation of collagen bundles with the subepidermal grenz zone of normal collagen, and thickening of the dermis with perivascular and periadnexal lymphocytic infiltrate, mast cells, and large stellate fibroblasts (7). In stasis dermatitis, there are features of acute or chronic eczema with dilated capillaries, surrounded by cuffs of fibrin, hemosiderin deposits, and hyperplastic venules (8).

In differential diagnosis of pretibial skin lesions, the development of dermatoses with a similar clinical course due to persistent physical stimulus, such as lichen simplex chronicus (LSC) or prurigo nodularis (PN), or with a similar clinical appearance, such as lichen amyloidosis (LA) or lichen planus (LP), are expected (1, 9).

LSC is a well-described, common pruritic disorder resulting from repeated rubbing and scratching. Typically, only one site is involved (10). In LSC, papules aggregate into irregular, indistinctly bordered scaly plaques with lichenification and peripheral hyperpigmentation, which are absent in PPPD. In addition, excoriations are frequently present in LSC, but not as prominent as in PPPD. The histology of skin biopsy in LSC reveals acanthosis, hypergranulosis, patchy hyperkeratosis with parakeratosis, and vertically arranged collagen fibers in dermal papilla (1, 2). In fact, among the 44 patients included in the original article, 18 cases were initially misdiagnosed as LSC (1).

PN lesions are symmetrically distributed hyperkeratotic pruritic brownish-red nodules and papules on the extensor surfaces of the extremities. In histology, we observe thick compact orthohyperkeratosis, irregular epidermal hyperplasia, focal parakeratosis, hypergranulosis, and necrotic keratinocytes in the epidermis. There is fibrosis of the papillary and reticular dermis with vertically arranged collagen fibers, increased numbers of fibroblasts and capillaries, and lymphocytic infiltrate with macrophages (11).

Like PPPD, LA presents with pretibial pruritic, scaly, flesh-colored to hyperpigmented papules (10). However, histological features such as dermal amyloid appearing as eosinophilic globules, which under polarized light show green birefringence on Congo red staining, distinguish LA from PPPD (1). Amyloid in LA is not derived from immunoglobulins or serum proteins, but from keratin peptides of necrotic keratinocytes damaged by chronic scratching (11). Pruritus seems to be the cause and not a symptom of the papular skin lesions. Consequently, treatment of LA should not be directed at removing amyloid, but at improving the pruritus (12).

LP presents as violaceous, pruritic papules and plaques with shiny surfaces and a network of fine white lines known as Wickham striae. The most common sites of involvement are skin, mucous membranes, and nails (10). Typical histological features of LP are hyperkeratosis without parakeratosis, focal increases in the granular layer, irregular acanthosis, vacuolar degeneration of the basal cell layer, a band-like lymphocytic infiltrate at the dermal-epidermal junction, and the presence of Civatte bodies (13).

Pathologically, PPPD resembles pigmented purpuric dermatosis, but in our case hemosiderin was absent and there was more papillary dermal fibrosis (14).

The latest comprehensive clinical and pathologic review focuses on lymphocyte atypia and CD30 expression in tissue biopsy to distinguish PPPD from lymphomatoid papulosis. The authors concluded that a relatively low number of atypical lymphocytes with weak or light CD30 staining intensity, as well as characteristic pruritus and pretibial distribution, favor PPPD over lymphomatoid papulosis (15).

Dermoscopy is recommended as a non-invasive tool for improving the recognition of PPPD. The main features are dotted or globular vessels on a pinkish-white background, with or without peripheral whitish collarette scaling with a petaloid appearance (9).

Conclusions

Pretibial pruritic lesions may reflect many different systemic diseases and dermatoses. One must consider a group of dermatoses due to psychological stress, such as rubbing and scratching, sometimes without a clear initial cause for pruritus. Furthermore, persistent rubbing can cause different clinical and histological patterns. The reason for this phenomenon is not fully understood, but some authors suggest it could be a reflection of the nature of rubbing. For example, LSC is thought to be the result of long-standing rubbing in a back-and-forth manner, PN may be a result of vigorous scratching or rubbing lasting for months or years, and LA is thought to follow prolonged scratching or friction with subsequent deposits of amyloid in dermal papillae (1). Histopathological signs of chronic scratching such as epithelial hyperplasia

with hypergranulosis and compact orthokeratosis, and coarse collagen in vertical streaks in the stratum papillare are present in PN, LSC, and LA, but absent in PPPD. Thus, PPPD, although less well known, is another distinct reaction pattern to chronic rubbing (2). For correct diagnosis, one must consider clinical-histological correlation as well as possible contributing factors. If patients are observed and examined carefully, rare entities or even new diagnoses will be found.

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