Targetoid Clinical Morphology as a Diagnostic Clue of the Lichenoid Histopathologic Subtype of Pigmented Purpuric Dermatosis

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INTRODUCTION

Lichenoid pigmented purpuric dermatosis (PPD) is a histopathologic subtype of PPD characterized by a dense band-like lymphohistiocytic infiltrate in the upper portion of the dermis.1 There are several histopathologic variants of PPD, but common features across subtypes include hemosiderin deposition, red blood cell extravasation, and papillary dermal fibrosis.2

The diagnosis of lichenoid PPD can be difficult, as it can present with histopathologic and clinical features resembling mycosis fungoides (MF). Additionally, the histopathologic and clinical patterns of PPD may not correlate. Herein, we describe 3 cases of lichenoid PPD with targetoid clinical morphology, for which the clinical differential diagnosis did not initially include PPD. In all 3 cases, the initial histopathologic differential diagnosis included MF, prompting immunohistochemical evaluation. Although the lichenoid pattern, fibrosis, and exocytosis can raise concern for MF, targetoid clinical morphology may prompt inclusion of lichenoid PPD in the differential diagnosis.

CASE SERIES

Case 1

A 34-year-old man presented with a 3-year history of a pruritic and progressive rash on the lower extremities. Physical examination showed erythematous targetoid plaques with violaceous centers on the lower extremities (Fig 1). The clinical differential diagnosis included lichen simplex chronicus and fixed drug eruption (FDE). Microscopic examination revealed a band-like infiltrate composed of lymphocytes and histiocytes. Fibrosis was present in the superficial dermis, consistent with chronicity. There was no interface tissue reaction (Fig 2, A to C). Owing to lymphocyte exocytosis and evidence of chronicity, immunohistochemical stains for CD3, CD4, CD8, and CD7 were performed and reflected a polyclonal, reactive T cell predominant infiltrate. Correlation and scrutiny for extravasated erythrocytes as well as hemosiderin deposition (Fig 2, C and

Fig 1. Erythematous targetoid plaques with violaceous centers on the lower extremity.
established the diagnosis of lichenoid PPD. Treatment with mometasone 0.1% ointment twice a day for 3 weeks followed by 3 times a week as maintenance resulted in improvement in physical findings and resolution of pruritus.

Case 2
An 83-year-old man presented with a 1-year history of an eruption limited to the lower extremities. Examination revealed erythematous targetoid plaques and patches with violaceous centers (Fig 3). The clinical differential diagnosis included erythema annulare centrifugum, nummular dermatitis, and FDE. Microscopic examination revealed a patchy lichenoid infiltrate composed predominantly of lymphocytes. No interface tissue reaction was observed, but lymphocyte exocytosis was noted. Chronicity was indicated by the presence of fibrosis. Additionally, there were scattered extravasated red blood cells and siderophages (Fig 4). Immunohistochemical stains for CD3, CD4, CD8, and CD7 were performed and reflected a reactive T cell predominant infiltrate.

Case 3
A 25-year-old man presented with a 5-year history of a pruritic rash on the thighs and buttocks. Physical examination demonstrated well-demarcated erythematous-to-violaceous targetoid plaques on the buttocks and thighs, some with lichenification (Fig 5). The clinical differential diagnosis included erythema annulare centrifugum, MF, and FDE. A lichenoid infiltrate without interface tissue reaction or cytologic atypia was observed. There were extravasated red blood cells and siderophages, along with fibrosis and exocytosis (Fig 6). Similar to the previous cases, the T cell infiltrate was considered reactive based on the ratio of CD8:CD4 expression and retention of CD7 expression. Treatment with
triamcinolone 0.1% ointment twice daily for 6 weeks was ineffective, but tacrolimus 0.1% ointment daily for 8 weeks resulted in some improvement.

**DISCUSSION**

PPD refers to a group of dermatoses characterized by petechiae, purpura, and red-brown pigmented macules, patches, or plaques that most frequently involve the lower extremities.\(^3\,^4\)

The exact etiology of PPD is not known, but diet, infection, diabetes, venous insufficiency, exercise, autoimmune conditions, and medication hypersensitivity have been described as possible causes.\(^3\)

PPD can be classified into clinical and histopathologic subtypes. Clinical variants include Gougerot-Blum purpura, purpura annularis telangiectodes (Majocchi disease), lichen aureus, Schaumburg disease, and eczematoid-like purpura of Doucas and Kapetanakis.\(^1\) There are also clinical variants of PPD that do not fit into the previously described categories, which can make diagnosis difficult. In the cases presented here, there are overlapping clinical features of purpura annularis telangiectodes and eczematoid-like purpura.

While there are several histopathologic variants of PPD, common features include a superficial perivascular lymphocytic infiltrate and extravasated red blood cells with frequent extension to the epidermis, with or without hemosiderin deposition.\(^1\)

Histopathologic subtypes include lichenoid, interface, perivascular, granulomatous, and spongiotic.\(^1\) The interface pattern has basal vacuolization, dyskeratotic keratinocytes, and lymphocyte exocytosis. The perivascular pattern is characterized by only superficial perivascular inflammation, while granulomatous PPD shows non-caseating granulomas.
admixed with a lymphocytic infiltrate, and spongiotic PPD demonstrates spongiosis without interface, lichenoid, or granulomatous inflammation.1,5

The histopathologic pattern of a lichenoid infiltrate without interface tissue reaction and with findings of chronicity and lymphocyte exocytosis may prompt consideration of MF. Differentiating between MF and PPD can be challenging, and these groups of disorders can demonstrate overlapping histopathologic, immunohistochemical, and even molecular findings. Although PPD typically demonstrates a CD8 predominant and polyclonal T cell infiltrate, CD4 predominance and monoclonality may be observed.6 Additionally, PPD with progression to MF has been described, and pigmented purpura may serve as an early manifestation of MF or rarely support the coexistence of both conditions.1

The association between lichenoid PPD and targetoid lesions has not been described in prior case series on PPD.1,2,6,7 However, this is likely because a targetoid clinical appearance has not been previously recognized in PPD. In the 3 cases presented here, PPD was not included in the initial clinical differential diagnosis; instead, MF, FDE, and erythema annulare centrifugum were considered. While clinical and histopathologic subtypes of PPD often fail to align, targetoid clinical morphology may serve as a diagnostic clue to aid in clinicopathologic correlation and in accurate histopathologic diagnosis of lichenoid PPD.

Conflicts of interest
None disclosed.

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