An unusual presentation of pigmented purpuric lichenoid dermatitis

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Abstract

Pigmented purpuric lichenoid dermatitis (PPLD) is a rare subtype of pigmented purpuric dermatosis, which classically presents with a mixture of lichenoid papules and patches on the bilateral lower extremities. Herein, we describe an unusual case of a 47-year-old man with PPLD who presented with 1-3mm discrete papules without the presence of larger patches. The diagnosis of PPLD should be considered for patients presenting with bilateral symmetric discrete papules on the legs.

Keywords: Blum, capillaritis, dermatosis, Gougerot, lichenoid, pigmented, purpuric

Introduction

Pigmented purpuric dermatoses (PPD), also known as capillaritis, are a group of chronic skin disorders characterized by the presence of symmetric petechial lesions primarily present on the lower extremities [1,2]. Pathogenesis is believed to be secondary to subtle inflammation of superficial capillaries in the papillary dermis with subsequent erythrocyte extravasation [1,2]. Although clinical presentation varies depending on the subtype of PPD, lesions frequently appear as red-brown macules and patches due to hemosiderin formation from erythrocyte degradation [2]. Five main clinical variants are described and include progressive purpuric pigmented dermatosis, or Schamberg disease (the most common variant), purpura annularis telangiectodes of Majocchi, pigmented purpuric lichenoid dermatitis of Gougerot and Blum, eczematid-like purpura of Doucas and Kapetanakis, and lichen aureus [1,2]. All variants share similar histologic features, including blood vessel dilation, erythrocyte extravasation, hemosiderin deposition, and superficial perivascular lymphohistiocytic infiltrate [1,2].

Pigmented purpuric lichenoid dermatitis of Gougerot and Blum (PPLD) is a rare subtype most often seen in middle-aged to older men [2]. Typical presentation involves a mixture of symmetric Schamberg-like lesions (yellow-brown patches with pinpoint petechiae, resembling cayenne pepper) and red-brown lichenoid papules on the bilateral lower extremities [2]. The disease tends to be chronic and occasionally pruritic [2]. In addition to the common histologic features shared among all PPD, PPLD may demonstrate a lichenoid infiltrate, spongiosis, and focal parakeratosis [2].

Given the rarity of PPLD, limited reports exist, which describe the full spectrum of clinical features seen in this disease. Herein, we describe an unusual case of PPLD which presented with chronic discrete papules without the classic Schamberg-like patches.

Case Synopsis

A 47-year-old man presented with a stable, occasionally pruritic rash on his lower legs and feet that had persisted for the past 10 years. The patient had never sought treatment previously. Skin

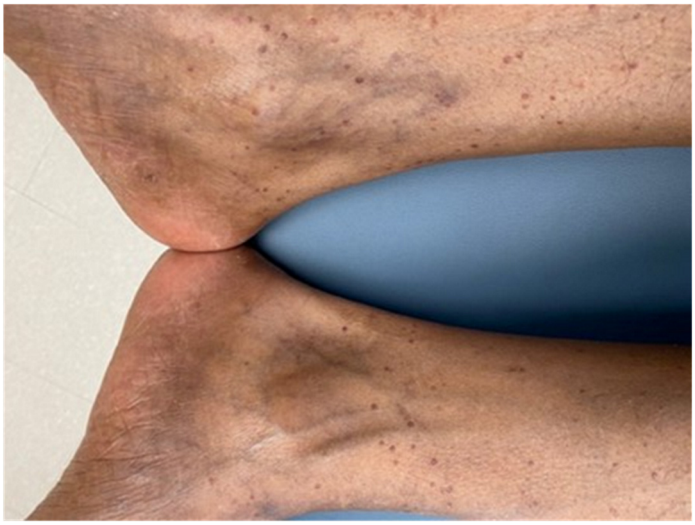

Figure 1. *1-3mm red-brown papules distributed symmetrically on the bilateral lower extremities.*

examination revealed 1-3mm, symmetric red-brown papules on his lower legs and dorsal feet bilaterally (**Figure 1**). Skin biopsies of the left and right shins were performed which showed focal superficial perivascular infiltrate consistent of lymphocytes, histiocytes, and extravasated erythrocytes with overlying subtle interface dermatitis (**Figure 2**). Due to the stable and asymptomatic nature of the lesions, the patient deferred treatment at this time.

Case Discussion

Most published case reports describing patients with PPLD depict the presence of both types of lesions. However, in the patient described here, discrete papules were present chronically without the presence of larger patches.

The optimal treatment regimen for patients with PPD remains uncertain, but current guidelines recommend utilization of non-pharmacological interventions, such as compression stockings and

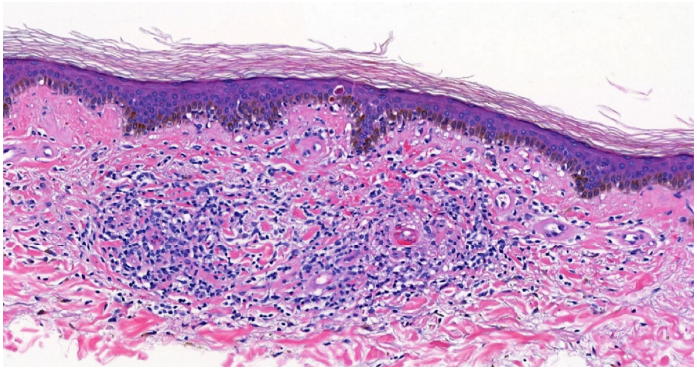

Figure 2. *Histopathology demonstrating focal lymphohistiocytic perivascular infiltrate and extravasated erythrocytes with subtle overlying interface. H&E, 20×.*

topical corticosteroids as first-line therapy options [3-10]. Additional options include but are not limited to topical calcineurin inhibitors [11], phototherapy [6,12-14], pentoxifylline [15], rutoside/ascorbic acid [16], and colchicine [17,18].

There is a lack of data analyzing efficacy of the aforementioned treatment options, specifically in PPLD patients. As such, it remains unclear whether PPLD patients would benefit from a tailored therapeutic regimen which differs from other PPD subtypes.

Conclusion

Pigmented purpuric lichenoid dermatitis classically presents with a combination of cutaneous papules and larger patches. In the patient described here, discrete papules were present without patches. It is important that dermatologists are aware of this uncommon PPLD presentation.

Potential conflicts of interest

The authors declare no conflicts of interest.

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