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Persistent dermatomal eruption on a leg

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Abstract

Unilateral linear capillaritis (ULC) is a rare variant of pigmented purpuric dermatoses (PPD) that is characterized by a linear or pseudo-dermatomal eruption on a single extremity. Although clinically distinct from the other PPD, it shares histopathologic features with this group. Herein, we present a man in his 50s who presented with asymptomatic macules and scaly papules on the left lower extremity in a linear distribution. The eruption persisted despite treatment with topical triamcinolone 0.1% and oral rutocide.

Keywords: pigmented purpuric dermatosis, capillaritis, lichenoid, dermatitis, spongiotic

Introduction

Unilateral linear capillaritis (ULC) is a rare and potentially under-recognized variant of pigmented purpuric dermatoses. It is characterized by an often-striking linear or pseudo-dermatomal distribution on a single extremity and it may clinically mimic several other dermatoses that present with zosteriform patterns [1-3]. Histopathology shows inflammatory changes typical of pigmented purpuric dermatoses and is mandatory in distinguishing it from potential clinical mimics. A case of ULC is presented.

Case Synopsis

A man in his 50s with no past medical history presented with a 4-month history of a persistent, asymptomatic eruption involving the left leg and dorsal foot. Prior to evaluation in our department, the patient was treated with a 21-day course of oral doxycycline for presumed lymphangitis without

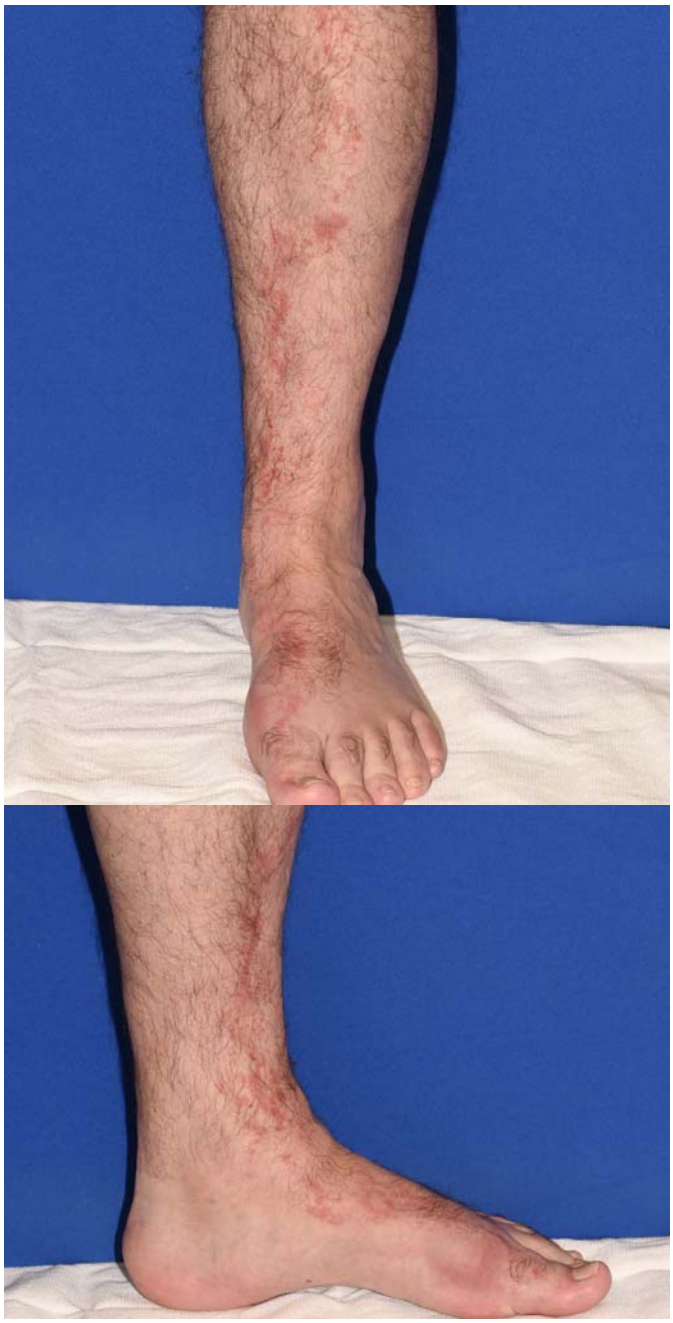

Figure 1. *Non-blanchable red macules admixed with red-orange scaly papules in a zosteriform distribution on the left lower extremity.*

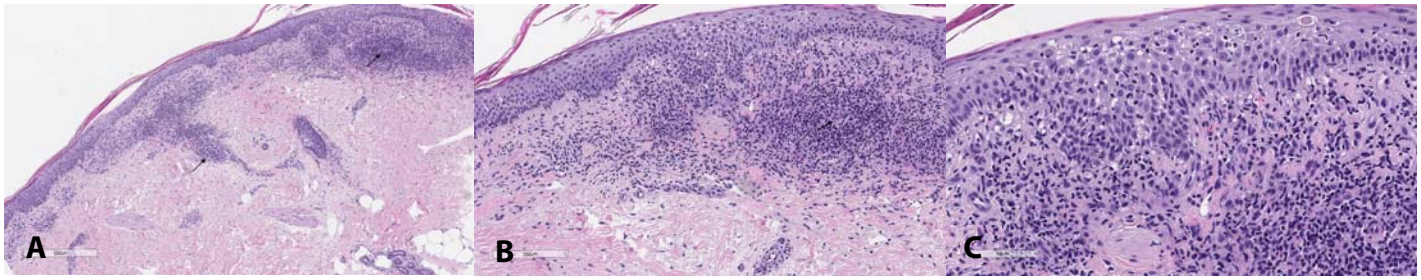

Figure 2 ***A)*** *Superficial perivascular lymphocytic inflammation with a lichenoid infiltrate and spongiosis. Focal lymphocytic vasculitis is indicated by black arrows. H&E, 4×.* ***B)*** *Higher-power view highlighting lymphocytic vasculitis (black arrow). H&E, 10×.* ***C)*** *Erythrocyte extravasation was prominent. H&E, 20×.*

improvement. He denied history of trauma to the involved areas. Physical examination revealed scattered red, non-blanchable macules admixed with scaly, thin red-orange papules extending from the left upper shin to dorsal foot in a dermatomal distribution (**Figure 1**). The right lower extremity was unaffected. Histopathologic examination revealed superficial perivascular lymphocytic inflammation with a lichenoid infiltrate, spongiosis, and focal lymphocytic vasculitis (**Figure 2**). Erythrocyte extravasation was also seen. Peri-eccrine inflammation was absent. The patient was treated with triamcinolone 0.1% cream twice daily and oral rutocide 100mg daily without improvement at follow-up three months later.

Case Discussion

Unilateral linear capillaritis (ULC) is a rare variant of pigmented purpuric dermatoses (PPD). Approximately 10 cases of ULC have been reported in the literature since its original

description by Riordan et al. in 1992 [1-5]. As a group, PPD is characterized by progressive petechiae, purpura, and pigmentary changes of the skin [6]. Although significant overlap may occur between sub-groups, five clinical variants are classically recognized [7]. Schamberg disease

generally presents with "cayenne pepper" petechiae distributed symmetrically over the shins and dorsal feet. Purpura annularis telangiectoides (Majocchi disease) is characterized by annular, red, telangiectatic patches on the legs. Lichenoid pigmented purpura of Gougerot and Blum involves flat-topped, coalescent, thin papules and plaques on the legs, whereas eczematid-like purpura of Doucas-Kapetanakis is typified by diffuse, eczematous, purpuric patches. The fifth variant, lichen aureus, presents with localized papules/plaques with a distinct golden color.

Unilateral linear capillaritis represents a rare and potentially under-recognized sixth variant of PPD [3]. Clinically, it is characterized by an asymptomatic, progressive, unilateral eruption of purpuric macules/patches, with the vast majority of cases occurring on a lower extremity [1-5]. As cases become chronic, ezematous patches and lichenoid papules may develop. The most distinctive feature of ULC is its striking linear or pseudo-dermatomal distribution. This distinguishes it from the more common PPD variants, in which symmetrical lower extremity involvement is characteristic. Unilateral linear capillaritis is five times more common in males than in females and most cases have occurred between the first and fourth decades of life [1-5].

As with other PPD variants, the etiology of ULC is unknown. Proposed causative or exacerbating factors include venous hypertension, capillary fragility, contact allergy to dyes, or systemic medications [4, 7]. Given its frequent blaschkolinear distribution, ULC may also represent a manifestation of somatic mosaicism wherein keratinocytes along embryologic migration lines are either prone to or incite inflammation in those areas [8]. Similar pathophysiology has been proposed for other blaschkolinear inflammatory disorders including the linear variants of lichen planus (LP) and discoid lupus erythematosus. Alternatively, ULC may be a primarily dermatomal process, although it is unclear what role cutaneous nerves might have in its pathogenesis [3].

The histopathologic findings of ULC are similar to those of other PPD variants [6, 7]. There is perivascular lymphocytic inflammation involving the small vessels of the papillary dermis. Erythrocyte extravasation with hemosiderin deposition is typical. As in our patient, a lymphocytic vasculitis characterized by vascular endothelial swelling may occur, but fibrinoid necrosis is rare [7]. Varying degrees of epidermal spongiosis and lymphocytic lichenoid inflammation may also be seen, both of which were observed in our case. Notably, a granulomatous histopathologic variant of PPD was recently described, but not in association with the ULC sub-type [9].

Unilateral linear capillaritis must be distinguished from other inflammatory disorders occurring in linear, blaschkoid, or dermatomal distributions. Linear LP may clinically resembles ULC, but is differentiated by its intense pruritus, violaceous color, and prominent post-inflammatory hyperpigmentation. LP would also be expected to show hypergranulosis and lacks the erythrocyte extravasation that is typical in ULC. Lichen striatus may be considered in pediatric cases of ULC, but is distinguished by its hypopigmented papules and lymphocytic peri-eccrine inflammation. Inflammatory linear verrucous epidermal nevus is easily differentiated by its intense pruritus and papillomatosis on histopathological examination.

Like other PPD variants, ULC is resistant to treatment. Topical corticosteroids may be helpful if pruritus is present, but are otherwise of marginal benefit [3]. Psoralens with UVA, rutocide, ascorbic acid, and griseofulvin have been used with variable success [7, 10] Spontaneous resolution occurs after several months to years in untreated cases.

Conclusion

This case highlights ULC as a rare and distinct clinical variant of PPD. Histopathology showing changes typical of PPD may be necessary to distinguish it from other dermatoses characterized by linear or zosteriform patterns.

Potential conflicts of interest

The authors declare no conflicts of interests.

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