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Linear Pigmented Purpuric Dermatoses

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Introduction

A 65-year-old woman presented to the dermatology clinic in Montreal with slowly progressive asymptomatic pigmentation in both legs since few years. Patient denied any pigmentation elsewhere. There was no history of chronic use of medications or applications of medicated creams. The eruption was not related to sun exposure. Two episodes of lower extremity superficial thrombophlebitis has occurred at age of 20 and 50. Subsequently, the patient underwent varicose vein ligation and stripping of left lower extremity.

Her medical history included also ulcerative colitis in therapy with mesalazine retention enema and surgically treated renal cell carcinoma. Skin examination revealed linear arrangement of brown muddy non-palpable non-planchable macules and patches located in popliteal fossa bilaterally extending to the thighs. Similar macules were seen on both shins scattered with no particular pattern (cayenne pepper appearance) (Figure 1). Varicose veins and pitting edema were also noted. Peripheral pulses were easily palpated. Differential diagnoses included pigmented purpuric dermatoses (PPD), diabetic dermatopathy, medication-induced pigmentation, purpuric contact dermatitis. Her full

blood count and coagulation profile were normal. Two skin biopsies were obtained and histopathology revealed superficial perivascular polymorphous dermatitis with erythrocyte extravasation and hemosiderin deposits consistent with Schambergs purpura and licheniod lymphocytic infiltrate with grenze zone with hemosiderin deposition more consistent with lichen aureus (Figure 2). There is no evidence of degeneration in the basal layer of the epidermis. Based on clinical and histological characteristics, a diagnosis of linear PPD most likely due to venous hypertension was made.

Comment

PPD is a group of chronic relapsing benign cutaneous entity of unknown etiology that shares similar clinical patterns and histological features. General presentation is red to purple macules that progressively coalesce and evolve to golden-brownish color usually affecting lower extremities. Capillaritis with dilated blood vessels, extravasation of erythrocytes, hemosiderin deposition in papillary dermis and perivascular lymphocytic infiltrate is the histological hallmark of all PPD. There are five major clinical variants Schamberg disease (the commonest), pigmented purpuric lichenoid dermatosis of Gougerot and Blum, purpura annularis telangiectodes of Majocchi, eczematid-like purpura of Doucas and Kapetanakis, and lichen aureus. Most of PPD are idiopathic, however some are associated with medications or systemic diseases. Moreover, predisposing factors that might influence the disease presentation are venous hypertension, exercise and gravitational dependency, capillary fragility, focal infections and chemical ingestion.[1, 2]

Linear PPD has been reported previously as a rare form of PPD and even in a unilateral pattern.[3,4] Linear array could be blaschkolinear, pseudo-dermatomal or following deep venous system.[5,3] In our case it is bilaterally distributed and probably following deep venous system loosely corresponding to small saphenous vein. Interestingly, histopathology shows features of both Schambergs disease and lichen aureus.
There is no standard therapy for PPD. In asymptomatic patients, conservative management is preferred. Other topical and oral therapeutic options can be tried with variable response.[2]

Authors' Contribution

AAK was involved in the data collection, drafting the manuscript, organising article sections, literature review and agreed to be the corresponding author for this article. RB was involved in helping draft the manuscript, interpretation of data, review the content of the article and approved the version to be published.

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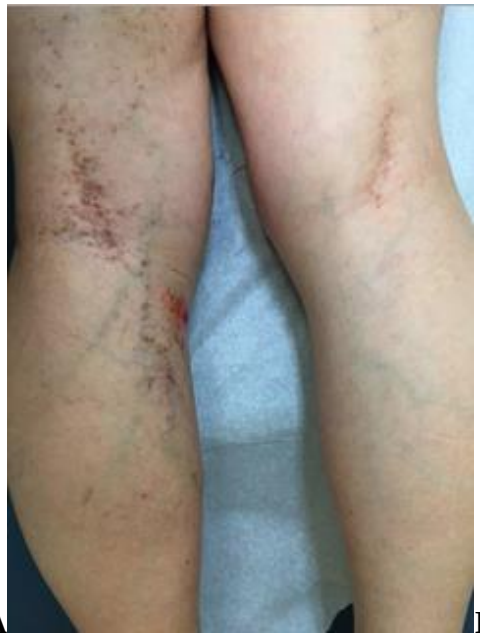
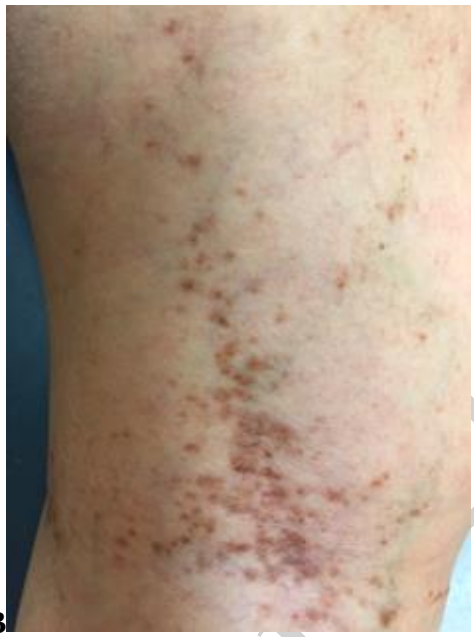
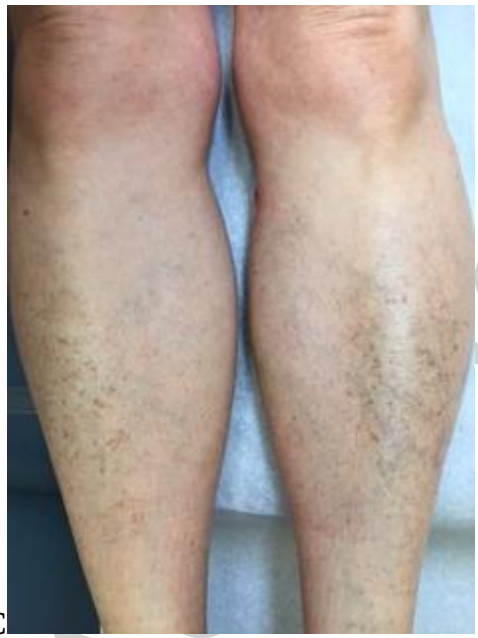
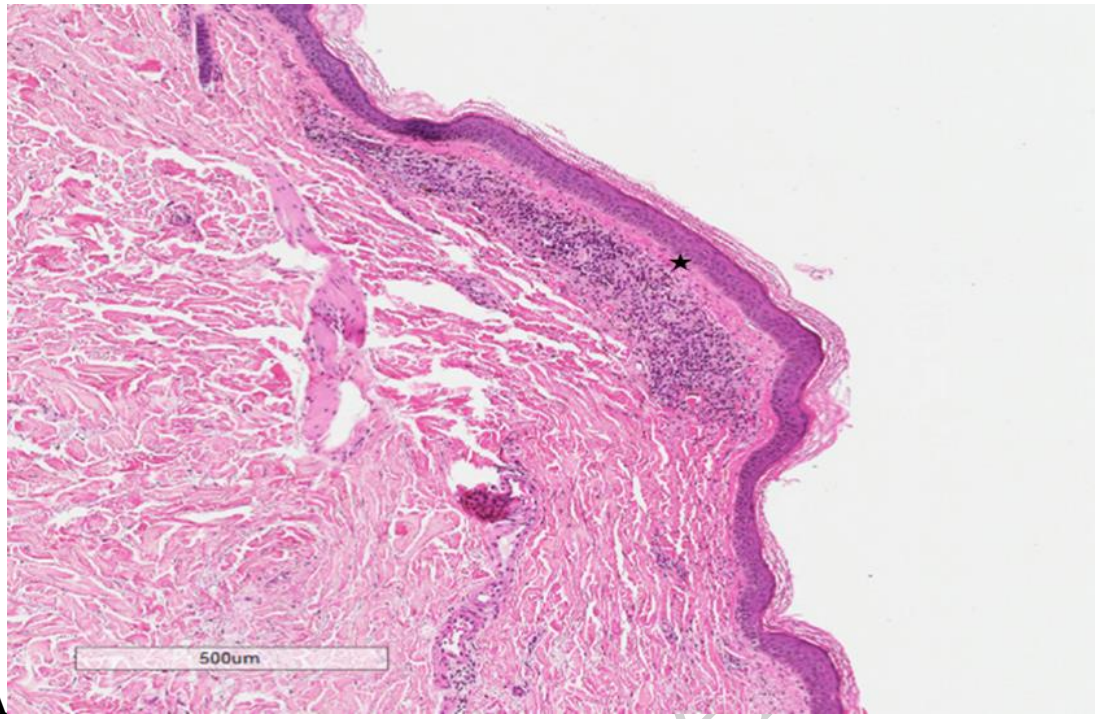
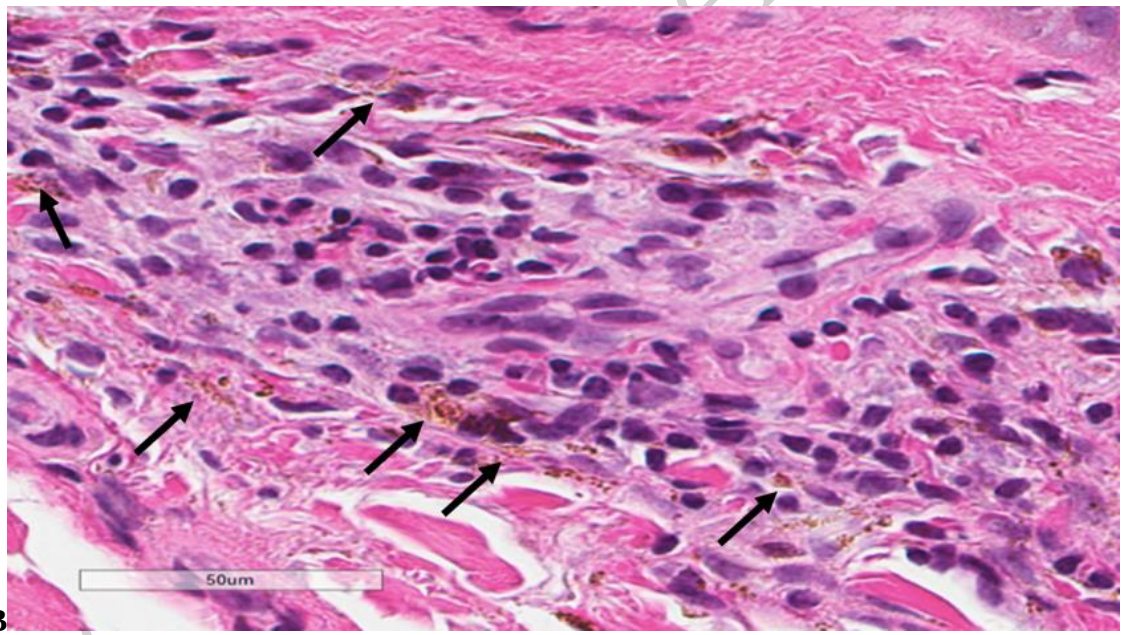




Figure 1: A: Brownish non palpable macules and patches arranged in linear distribution on the back of the legs bilaterally. The left one is more involved than the right since it extends to the posterior thighs. Prominent varicose veins are noted. **B:** Closer image showing the linear distribution of the eruption affecting the left leg probably following deep venous system. **C:** Similar scattered macules are distributed on both shins.



A



B

Figure 2: A: Heamatoxylin and eosin stain at x 40 magnification showing dermal lichenoid lymphocytic infiltrate with grenze zone (asterisk) with hemosiderin deposition. There is no evidence of degeneration in the basal layer of the epidermis. **B:** Heamatoxylin and eosin stain at x 200 magnification showing polymorphous perivascular infiltrate with prominent hemosiderin deposition (black arrows).