Linear lichenoid pigmented purpura: An unusual histopathologic pattern

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Key words: lichenoid; linear pigmented purpura; linear lichenoid pigmented purpura; pigmented purpuric dermatosis; unilateral linear capillaritis.

INTRODUCTION

Linear pigmented purpura, also known as unilateral linear capillaritis, is a clinical subtype of pigmented purpuric dermatoses (PPDs). Although several variants exist, the PPDs are a benign group of skin eruptions that present with petechiae and hyperpigmented macules, papules, and/or patches. Morphology, clinical examination findings, and histopathology differ across PPD subtypes.[1,2] Thus far, the exact etiology of PPD is unknown.[3]

First described by Riordan et al,[4] linear PPD presents in a strikingly linear distribution, often unilaterally on an upper or lower extremity.[4,5] Linear PPD is more commonly seen in men, with occurrences in young adults and children.[1] Although around 15 cases of linear PPD have been reported in the literature, here we present a rare case of linear pigmented purpura with a concurrent lichenoid inflammatory pattern on histology.[4-10]

Abbreviation used:

PPD: pigmented purpuric dermatoses

CASE REPORT

An otherwise healthy 34-year-old woman with no skin cancer history presented to our academic center for consideration of the systemic treatment of her "lichen planus." She had a 5-year history of very thin papules on her left arm that had spread slightly since the onset. Before evaluation by our institution, she had undergone 2 tangential biopsies of the lesions that were consistent with lichen planus. She had also been treated with tacrolimus ointment 0.1% without any improvement.

On examination, she had discrete red to brown, minimally elevated thin papules on the dorsal aspect of the left hand (Fig 1) and left wrist (Fig 2) in a linear distribution. She had no other areas of involvement on the skin, and her scalp, nails, and mucosa had no inflammatory changes.

A punch biopsy was performed, demonstrating lichenoid granulomatous inflammation and extravasated red blood cells (Fig 3). A Perl stain for iron-highlighted hemosiderin pigment in the papillary dermis (staining blue) (Fig 4).

Both her clinical presentation and histopathologic skin biopsy findings were consistent with a diagnosis of linear lichenoid pigmented purpura, a rare subtype of the PPDs. Considering its benign nature and reports of resolution over time, we recommended clinical observation and no further interventions.

DISCUSSION

Within the literature, 5 distinct variants of PPD have been described, including progressive pigmentary dermatosis (Schamberg disease), lichen aureus, purpura of Majocchi, eczematoid-like purpura of Doucas and Kapetanakis, and pigmented purpuric lichenoid dermatitis of Gougerot and Blum.[1,2] Some rarer forms, including linear and granulomatous pigmented purpura, have been less frequently reported and recognized. PPD most commonly presents bilaterally in the lower extremities. Although some association with venous stasis has been postulated, hemosiderin deposition because of erythrocyte extravasation occurs in the papillary dermis

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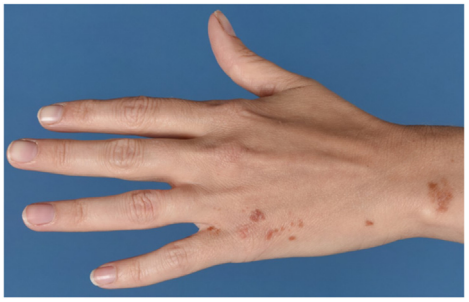

Fig 1. Dorsal aspect of the left hand demonstrating a linear array of thin, red-brown papules consistent with linear pigmented purpura.

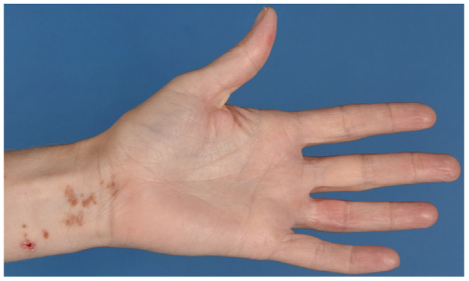

Fig 2. Left wrist demonstrating a linear array of thin, red-brown papules consistent with linear pigmented purpura.

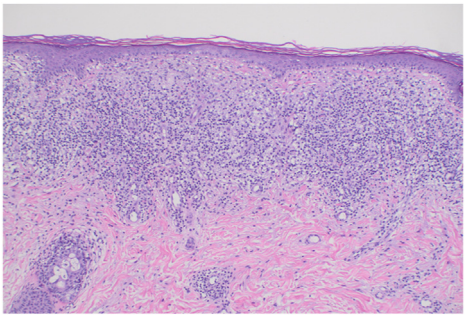

Fig 3. Punch biopsy, hematoxylin and eosin stain demonstrating lichenoid and granulomatous infiltrate, and extravasated red blood cells. (Hematoxylin-eosin stain; original magnification: ×100.)

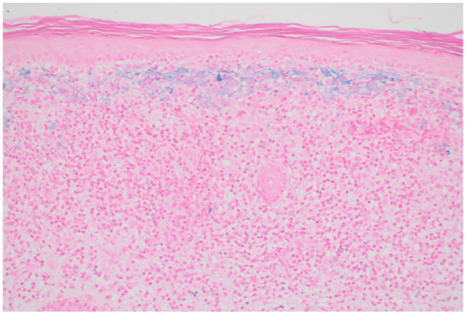

Fig 4. Perl stain highlighting hemosiderin pigment in the superficial dermis (*blue*). (Perl stain; original magnification: ×200.)

from small cutaneous vessels rather than deeper blood vessels.[1,2] The development of PPD has not been significantly associated with any underlying diseases or hematologic abnormalities; however, some medications, including acetaminophen, aspirin, glipizide, lorazepam, chlordiazepoxide, hydralazine, among others have been identified as triggers.[1-3]

Regardless of subtype, histopathology of PPD shows perivascular lymphocytic inflammation in the papillary and mid-dermis, dilated cutaneous blood vessels with endothelial cell swelling, and erythrocyte extravasation with associated hemosiderin deposition, responsible for the red-to-brown lesion pigmentation observed on examination. Lymphocytic inflammatory patterns visible on histopathology may be further classified into perivascular, spongiotic, lichenoid, or granulomatous.[2] Across PPD variants, lymphocytic infiltrate in a lichenoid pattern is classically associated with lichen aureus or pigmented purpuric lichenoid dermatitis of Gougerot and Blum.[1,2] We report another, rare case of linear pigmented purpura with a lichenoid infiltrate.[9]

Linear pigmented purpura is a rare, often overlooked subtype of PPDs given its frequent unilaterally. Cases have been reported in various locations, including the legs, arms, and torso.[4-10] Given their similar presentations, linear PPD can often be mistaken for other dermatologic disorders with linear, blaschkoid, or dermatomal-like distributions, including lichen aureus or other PPD subtypes, linear epidermal nevi, linear morphea, lichen striatus, or linear lichen planus among others. Unlike linear PPD, lichen aureus is usually bilateral with lichenoid, golden-brown papules, whereas pigmented purpuric lichenoid dermatitis of Gougerot and Blum tends to exhibit the "cayenne pepper" petechiae of Schamberg disease with lichenoid, red-brown papules, and frequent pruritus.[1-3] Linear epidermal nevi demonstrate hyperkeratosis, acanthosis, and papillomatosis on histopathology and are often present from a young age.[5] Linear morphea and lichen striatus are most often seen in children and can often be differentiated on physical examination. In more challenging cases, a skin biopsy can be done showing evidence of sclerosis in linear morphea and peri-eccrine lymphocytic inflammation in lichen striatus.[9,10]

In our case, linear PPD was initially mistaken for linear lichen planus. Linear PPD lacks much of the significant pruritus, deep purple color, and hyperpigmentation of linear lichen planus on clinical examination.[7,9,10] Further, skin biopsy of lichen

planus usually reveals a lichenoid band of inflammation in the papillary dermis with epidermal hyperplasia and hypergranulosis. Although a tangential biopsy sampling the epidermis and papillary dermis may appear similar for both linear lichen planus and linear pigmented purpura, a punch biopsy can aid in the differentiation of the 2 diagnoses.

Treatment of linear PPD is difficult and usually ineffective, similar to the treatment of other PPD variants. Across case reports of PPD, oral antihistamines, pentoxifylline, topical steroids, psoralen plus UV-A, rutoside, ascorbic acid, and other agents have been trialed with no evidence suggesting anyone's superior treatment strategy.[1,2] In 2 cases of linear PPD, Mar et al[7] reported symptomatic improvement of pruritus with topical steroids; however, time to lesion resolution did not seem to vary across patients based on steroid usage.[6,8-10]

The clinical examination focused on the color and morphology of the lesions as well as punch biopsy may prevent misdiagnosis of linear lichenoid pigmented purpura as linear lichen planus or other dermatologic disorders. Because most cases of linear PPD partially or completely spontaneously remit within 24 months of diagnosis, accurate diagnosis can prevent unnecessary, potentially harmful treatments.[4-10]

Clinical photographs were taken by Cindy Doles, the medical photographer affiliated with Department of Plastic Surgery at the University of Texas Southwestern Medical Center.

Conflict of interest

None disclosed.

REFERENCES

1. Sardana K, Sarkar R, Sehgal VN. Pigmented purpuric dermatoses: an overview. *Int J Dermatol*. 2004;43(7):482-488. https://doi.org/10.1111/j.1365-4632.2004.02213.x
2. Spigariolo CB, Giacalone S, Nazzaro G. Pigmented purpuric dermatoses: a complete narrative review. *J Clin Med*. 2021;10(11):2283. https://doi.org/10.3390/jcm10112283
3. Kim DH, Seo SH, Ahn HH, Kye YC, Choi JE. Characteristics and clinical manifestations of pigmented purpuric dermatosis. *Ann Dermatol*. 2015;27(4):404-410. https://doi.org/10.5021/ad.2015.27.4.404
4. Riordan CA, Darley C, Markey AC, Murphy G, Wilkinson JD. Unilateral linear capillaritis. *Clin Exp Dermatol*. 1992;17(3):182-185. https://doi.org/10.1111/j.1365-2230.1992.tb00201.x
5. Kim JC, Kim YC. Zosteriform unilateral linear capillaritis. *Ann Dermatol*. 2021;33(3):298-300. https://doi.org/10.5021/ad.2021.33.3.298
6. Elshimy N, Walker B. Unilateral linear capillaritis: a case and summary of the literature 25 years on from its original description. *Clin Case Rep Rev*. 2017;3:1-3.
7. Mar A, Fergin P, Hogan P. Unilateral pigmented purpuric eruption. *Australas J Dermatol*. 1999;40(4):211-214. https://doi.org/10.1046/j.1440-0960.1999.00364.x
8. Ma HJ, Zhao G, Liu W, Dang YP, Li DG. Unilateral linear capillaritis: two unusual Chinese cases. *Eur J Dermatol*. 2007;17(2):160-163. https://doi.org/10.1684/ejd.2007.0132
9. Hoesly PM, Cappel MA, Sluzevich JC. Persistent dermatomal eruption on a leg. *Dermatol Online J*. 2019;25(10):13030/qt2k49w08d.
10. Rodriguez-Garijo N, Tomas-Velazquez A, Estenaga A, Antoñanzas J, Idoate MA, Espana A. A case of unilateral linear capillaritis. *Pediatr Dermatol*. 2020;37(4):750-751. https://doi.org/10.1111/pde.14151