Answer to Dermacase *continued from* **page 563**

2. Unilateral linear capillaritis

First described by Riordan and colleagues in 1992,[1] unilateral linear capillaritis (ULC), also known as *unilateral pigmented purpuric eruption* or *quadrantic capillaropathy,* is a rare variant of progressive pigmented purpuric dermatoses (PPPD); only around 10 cases have been reported in the literature.[1-4] The condition usually affects the lower extremities, and to a lesser extent the upper extremities, of boys and young men.[2] Clinically, it is characterized by a progressive unilateral arrangement of hyperpigmented macules and patches that can become purpuric and extensive. Similar to PPPD, the histology of ULC shows a mild to moderate perivascular lymphocytic infiltrate, both superficial and mid-dermal, with extravasation of erythrocytes and hemosiderin deposition.[2] The diagnosis in our case was confirmed after results of a biopsy showed the typical histologic findings of ULC.

Like other PPPD variants, the underlying cause of ULC is unknown.[1-5] Suggested causative factors include abnormalities in the cellular immune system, drugs, infection, capillary fragility, or phlebostasis.[2] The unilateral or linear distribution of ULC along Blaschko lines might also be explained by an underlying genetic abnormality, such as mosaicism.

Differential diagnosis

Clinical differential diagnosis of ULC should include conditions that might present in a linear distribution, such as linear psoriasis, inflammatory linear verrucous epidermal nevus, lichen striatus, and linear lichen planus (LP).[6-8] Linear psoriasis, an extremely rare form of generalized psoriasis, presents with asymptomatic, sharply demarcated plaques that are erythematous and scaly; the lesions are arranged in a linear distribution.[6,7] Inflammatory linear verrucous epidermal nevus presents as a pruritic, erythematous, and scaly plaque that affects children and, less commonly, adults. The eruption progresses over months to years and rarely does it resolve spontaneously.[6] Lichen striatus usually presents as small flat-topped skin-coloured to hypopigmented papules, arranged in a linear distribution, that primarily affect the extremities of children.[6,8] These asymptomatic lesions appear over days to weeks and tend to spontaneously resolve over a period of months to a few years. Linear LP shows the typical violaceous flat-topped papules of LP, arranged in a linear distribution; these might be followed by postinflammatory hyperpigmentation.[6]

The differential diagnosis should also include other PPPD variants.[9] Progressive pigmented dermatosis is an asymptomatic seasonal eruption most commonly occurring in the spring and summer; it commonly fades spontaneously over a period of several months to years.[9] Clinically, this subtype is characterized by pinpoint erythematous macules and patches with a mild overlying scale, usually involving the legs of adult women. Schamberg disease usually presents as asymptomatic and persistent purpuric lesions, discrete or confluent, on the lower legs of adult patients; the lesions are non-blanching and nonpalpable.[5,9] Purpura annularis telangiectodes is a PPPD subtype, characterized by annular patches of punctate, purpuric, reddish-brown macules with telangiectasias; it commonly presents on the bilateral lower extremities of adolescent girls.[9] Pigmented purpuric lichenoid dermatosis (Gougerot and Blum disease) is a chronic variant of PPPD that also involves the lower extremities of adult men. It takes the form of polygonal or round reddish-brown lichenoid papules that sometimes coalesce into plaques, with or without an overlying scale; it is usually associated with purpura or telangiectasia.[9] Lastly, lichen aureus, although rarely occurring in a segmental distribution, has a distinct golden colour, papular component, chronic course, and band-like dermal inflammatory infiltrate that makes its differentiation from ULC possible.[5,8,9]

Management

The prognosis for ULC is usually favourable, as the eruption tends to resolve spontaneously within 2 years.[2] There is no evidence of any effective treatment, but recently psoralen-UVA treatment has been reported to improve the condition.[2] Our patient preferred to take no treatment, and within 6 months the eruption had partially resolved.

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Competing interests
None declared

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