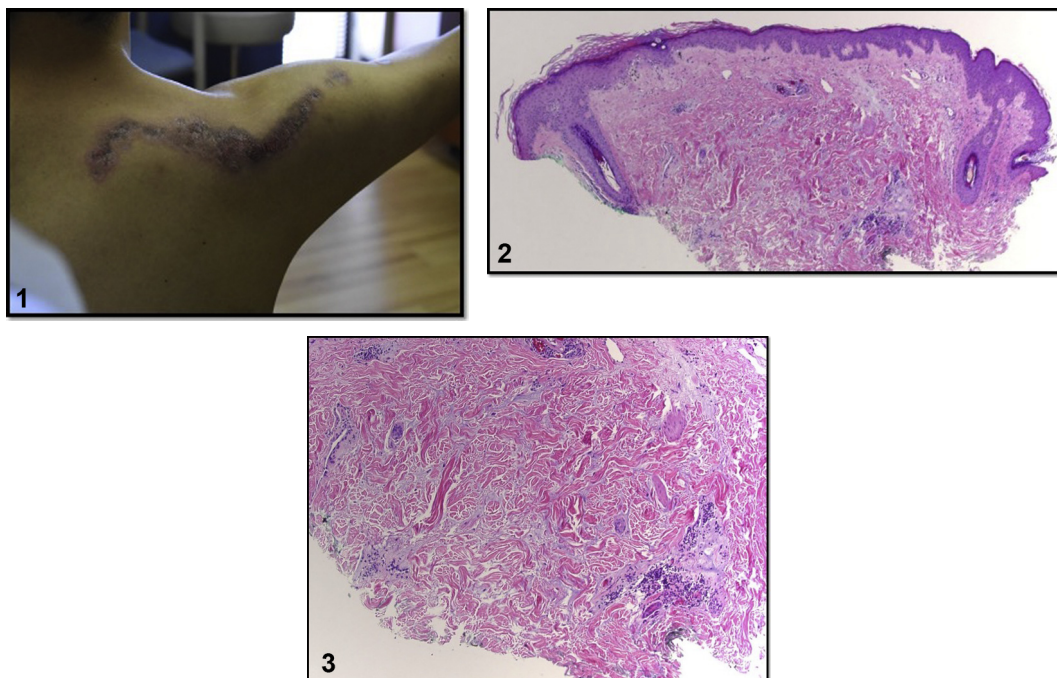


Curvilinear violaceous plaques along Blaschko lines



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A 29-year-old man was referred to dermatology with a 5-year history of violaceous crusted plaques with white central regions of ulcerations and overlying erythema that extended in a curvilinear fashion from the right side of his upper back, shoulder, and upper portion of the right arm along the Blaschko lines (Fig 1). He denied any pain, pruritus, or bleeding. His medical and family histories were unremarkable. The laboratory workup, which included a complete blood count, blood urea nitrogen, urinalysis, liver function test, and antinuclear antibodies, was within normal limits. A punch biopsy of the upper portion of the right arm was obtained (Figs 2 and 3).

Question 1: What is the best diagnosis?

- A. Inflammatory linear verrucous epidermal nevi
- B. Linear psoriasis
- C. Linear cutaneous lupus erythematosus (LCLE)
- D. Linear lichen planus

E. Linear morphea

Answers:

A. Inflammatory linear verrucous epidermal nevi — Incorrect. Inflammatory linear verrucous epidermal nevi, typically, presents early in childhood with

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unilaterally located red scaly papules coalescing into a linear plaque and may be accompanied by significant pruritus. Histology demonstrates elongated rete ridges with broad zones of parakeratosis (underlying loss of granular layer) alternating with orthokeratosis. There is no dermal mucin deposition.

B. Linear psoriasis – Incorrect. Psoriasis is characterized by sharp demarcated red silvery scaly plaques. Histology reveals elongated rete, parakeratosis with diminished stratum granulosum, accumulation of neutrophils in the stratum corneum, and increase of the capillaries in papillary dermis.

C. Linear cutaneous lupus erythematosus (LCLE) – Correct. The dermal mucin deposition is consistent with that of cutaneous lupus erythematosus (CLE). The most common clinical manifestations of CLE are acute CLE, subacute CLE, and discoid lupus erythematosus (DLE). The violaceous nature of the plaques with central crusting in this patient is very reminiscent of DLE. However, LCLE is characterized by a distinct Blaschkoid distribution, and it is less symptomatic than the other forms.¹ The laboratory studies such as the antinuclear antibody can be within normal limits.

D. Linear lichen planus – Incorrect. Lichen planus presents as polygonal, purple, and intensely pruritic lesions. Histopathology shows a classic “sawtooth” appearance of the dermoepidermal junction, stratum basale degeneration with necrosis, and a band-like lymphocytic infiltrate of the dermoepidermal junction.

E. Linear morphea – Although morphea can present along the Blaschko lines, the lesions are generally less violaceous and lack crusting. Histologic characteristics include diffuse interstitial lymphocytic infiltrate, and deeper involvement would show thick hyalinized collagen bundles. Chronic morphea may present histologically with decreased or absent lymphocytic infiltrate, sclerosis, and generalized atrophy.

Question 2: Which of the following aspects of LCLE distinguishes it from other forms of CLE?

- A.** Distinct histopathology
- B.** Younger age of onset
- C.** Higher progression to systemic lupus erythematosus
- D.** Unique symptom triggers
- E.** High photosensitivity

Answers:

A. Distinct histopathology – Incorrect. The histopathologic features of dermal mucin deposition and perivascular infiltrates in LCLE are identical to those seen in DLE.¹ Some of the additional features seen in both LCLE and DLE include follicular plugging, vacuolar interface dermatitis, and periadnexal infiltrates.^{1,2}

B. Younger age of onset – Correct. The mean age of onset is less than 15 years for LCLE,¹ whereas the DLE most commonly occurs in the third, fourth, and fifth decades, and the mean age at presentation is about 40 years in subacute forms of lupus erythematosus.

C. Higher progression to systemic lupus erythematosus – Incorrect. Progression to systemic lupus erythematosus is rare, with very few cases reported in the medical literature. The association with systemic lupus erythematosus is lower for LCLE (6.5%)¹ than that for subacute CLE (18%) and DLE (9.8%–16.7%).³

D. Unique symptom triggers – Incorrect. Although LCLE presents with some differences from other forms of CLE, such as the age of onset, the epigenetic and multifactorial nature of its pathogenesis may lead to similar triggers such as sunlight and trauma for symptomatic lesions.¹ Drugs such as tumor necrosis factor- α inhibitors and heavy metals such as lead that classically trigger both cutaneous and systemic lupus are reasonably expected to trigger LCLE as well.^{1,4}

E. High photosensitivity – Incorrect. Interestingly, LCLE is rarely associated with photosensitivity (11%) when compared with the other cutaneous forms of lupus erythematosus such as DLE (70.9%).¹

Question 3: What is the first-line treatment for acute lesions of LCLE?

- A.** Topical corticosteroids or intralesional corticosteroids
- B.** Tacrolimus
- C.** Hydroxychloroquine
- D.** Oral retinoids
- E.** Dapsone

Answers:

A. Topical corticosteroids – Correct. Topical or intralesional corticosteroids have been the mainstay for LCLE lesions as initial treatment in most cases

results in moderate-to-significant improvement of the lesions.⁵

B. Tacrolimus — Incorrect. Although topical calcineurin inhibitors like tacrolimus are also commonly reported treatments for LCLE, it is more commonly used as a long-term maintenance therapy to avoid the unwanted side effects of chronic corticosteroid therapy.

C. Hydroxychloroquine — Incorrect. Hydroxychloroquine may be effective for long-term therapy in patients for whom topical treatments have failed to reduce symptoms and lesions.

D. Oral retinoids — Incorrect. Oral retinoids are considered second-line therapies. The systemic retinoids have teratogenic effects and long-term risk of hypertriglyceridemia and bone abnormalities.

E. Dapsone — Incorrect. Systemic dapsone is used in cases of refractory lesions; however, it is a less popular alternative because of its side effects.

Abbreviations used:

CLE: cutaneous lupus erythematosus
DLE: discoid lupus erythematosus
LCLE: linear cutaneous lupus erythematosus

Conflicts of interest

None disclosed.

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