

SHORT COMMUNICATION

Retiform Plaques: An Additional Diagnostic Consideration

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A 34-year-old female presented for evaluation of a progressive, pruritic rash on the left leg with associated skin-tightening over the knee. Her symptoms began seven years earlier as a patch localized to the proximal anterior thigh. She was then lost to follow-up but later returned because her lesions were expanding down her leg and impacting her range of motion. She was otherwise healthy aside from recurrent genital herpes. Laboratory investigations revealed mild anemia with negative ANA, anti-Smith, RNP, and SCL-70 autoantibodies.

Examination revealed several violaceous-to-hyperpigmented coalescing indurated retiform plaques extending from the medial anterior thigh down the calf in an L2-L3 distribution (**Figure 1A and B**). Biopsy of the left posterior knee revealed a superficial and deep perivascular and interstitial lymphoplasmacytic infiltrate, thickening of deep reticular dermal and subcutaneous septal collagen bundles, and loss of perieccrine adipocytes (**Figure 2A and B**). There was no vasculitis or vasculopathy. The biopsy exhibited typical features of morphea, which, when combined with the clinical findings, was consistent with a diagnosis of linear morphea.

Morphea is an autoimmune condition caused by collagen overproduction that presents with progressive sclerotic skin lesions. An early inflammatory phase is followed by fibrosis, sclerosis, and eventual atrophy. Subtypes include plaque-type, linear, generalized, and mixed.¹

This patient's presentation posed a diagnostic challenge, as retiform lesions have a broad differential diagnosis including vasculitis and vasculopathy. To our knowledge, linear morphea in a retiform pattern has not been previously reported. However, reticulate hyperpigmentation overlying superficial vasculature has been noted in systemic sclerosis.²

Linear morphea typically presents on the face, scalp, or extremities. This patient's left lower extremity lesions have a patterned distribution that is either dermatomal (L2-L3) or blaschkoid. Some authors suggest that linear morphea follows the lines of Blaschko and occurs in susceptible mosaic cells after an environmental trigger.³

Although often idiopathic, morphea is occasionally ascribed to sequelae of infection, trauma, irradiation, or vascular injury. Wolf's isotopic response, in which an unrelated disease (such as morphea) occurs at the site of a previously healed primary

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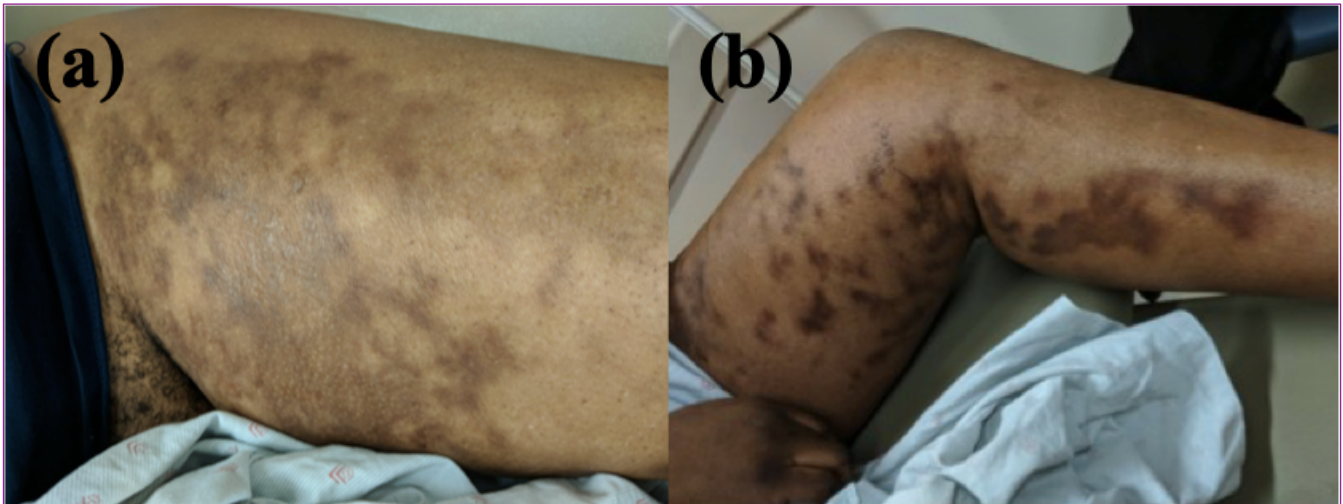


Figure 1. (a) Left leg: stellate-shaped violaceous and hyperpigmented firm plaques of the proximal anterior thigh, (b) Extent of disease from the proximal anterior thigh to the medial calf.

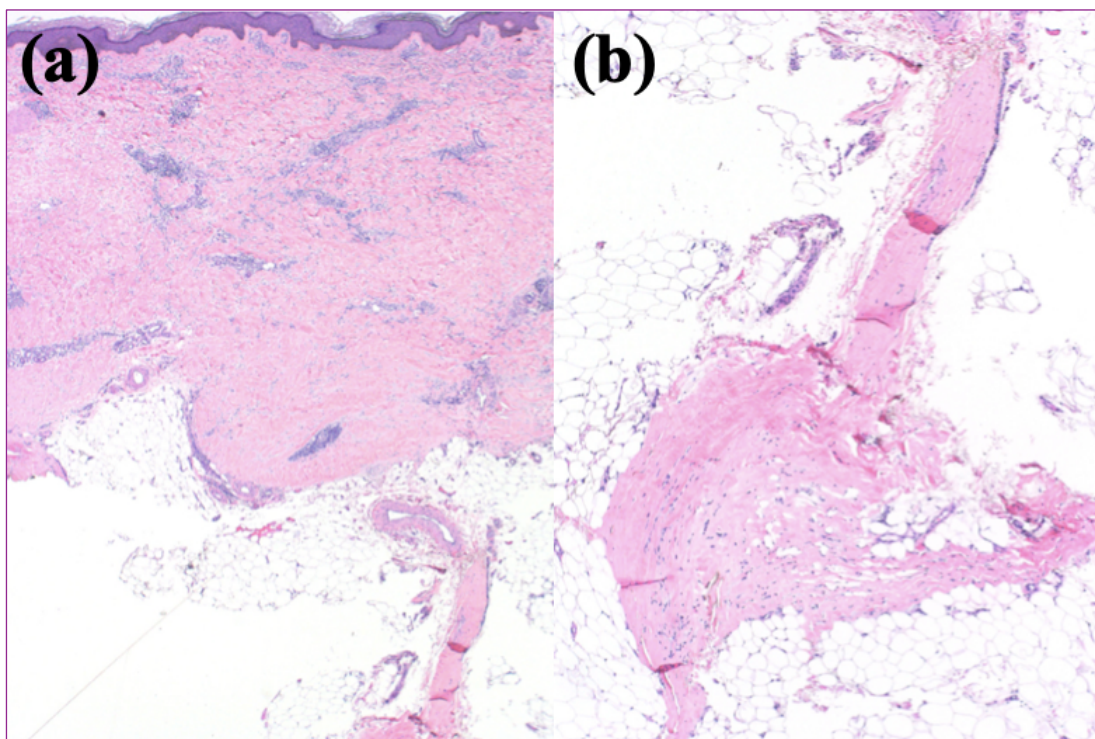


Figure 2. (a) Hematoxylin-eosin staining demonstrates superficial and deep perivascular and interstitial lymphoplasmacytic infiltrate, thickening of deep reticular dermal and subcutaneous septal collagen bundles, and loss of peri-eccrine adipocytes (H&E, 100x), (b) Higher power of the deep portion of the biopsy revealing hyalinization of a subcutaneous septa (H&E, 200x).

disease, could explain the distribution of this patient's lesions as secondary to subclinical dermatomal herpes virus reactivation.⁴ Because viruses may trigger vascular damage and endothelial cell apoptosis has been implicated in the pathogenesis of morphea, this phenomenon could explain both the distribution and morphology of her lesions.⁵

Therapy involves a stepwise approach. For nonfunctional or non-cosmetically sensitive areas, topical agents can be utilized. Phototherapy, particularly UVA1, is effective for patients with more generalized lesions. If disease is deeper or involving critical areas, systemic immunosuppressive agents such as methotrexate or mycophenolate mofetil in combination with systemic corticosteroids are often needed to mitigate inflammation-induced damage.⁵ Physical therapy is paramount for patients at-risk for contractures.⁶

Morphea can significantly impact quality of life. Untreated, linear morphea may cause contractures, disfigurement, and disability. Even with treatment, the five-year recurrence rate is up to 21%.⁶ Thus, patients warrant long-term follow-up. Herein, we provide a rare manifestation of linear morphea presenting with retiform plaques to increase awareness regarding an atypical presentation of this disease.

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