Livedo Reticularis-A Presenting Sign of Neuromyelitis Optica Spectrum Disorder.

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Images in Neurology

Livedo Reticularis—A Presenting Sign of Neuromyelitis Optica Spectrum Disorder

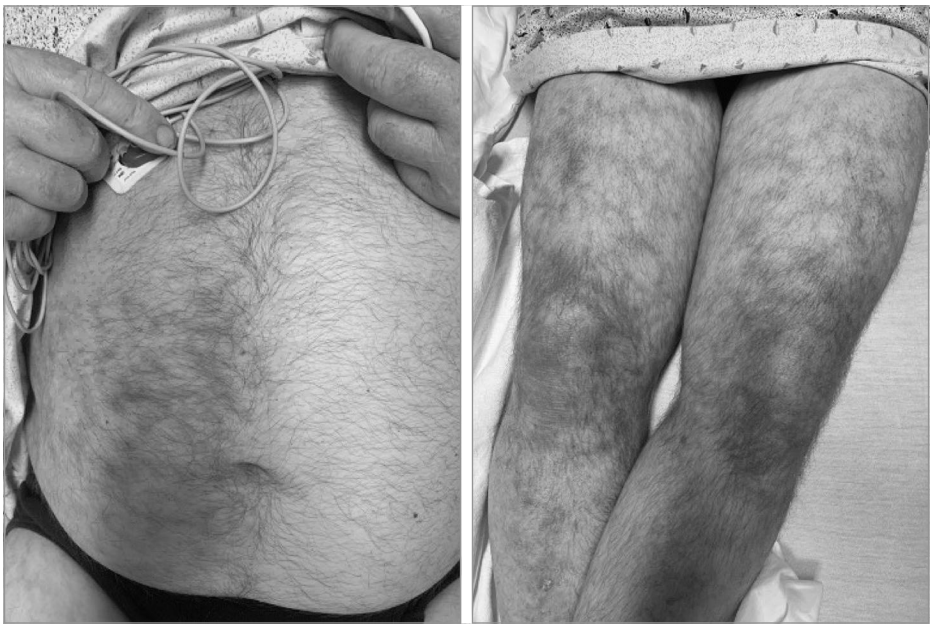
Cleo Zarina Reyes, MD; Negar Moheb, MD; Jay Varrato, DO

A 58-year-old man presented with a 9-day history of intermittent chest and abdominal pain, which was associated with numbness and tingling of the bilateral upper extremities. The patient had a medical history of atrial fibrillation, for which he was taking apixaban, and psoriatic arthritis, which was being managed with ustekinumab. Physical examination revealed a coalescing lacelike, reticular, violaceous, hyperpigmented rash of the bilateral lower extremities, right abdomen, and right flank with a sharp midline delineation consistent with livedo reticularis (LR) (**Figure 1**). He had no further focal neurologic deficits, and results of cardiac, pulmonary, and gastrointestinal workup were negative. Punch biopsy of the right thigh revealed no histologic features supportive of vasculitis. LR spontaneously resolved within 48 hours. The patient had no prior history of LR. Thrombotic panels, cryoglobulin level, antiphospholipid antibodies, antinuclear antibody panel, rheumatoid factor level, and angiotensin-converting enzyme testing were unremarkable.

Four days into admission, the patient developed sudden-onset weakness and numbness of his bilateral lower extremities. Neurologic examination was notable for weakness in the lower extremities both proximally and distally, more on the left side (1/5) than the right side (3/5), and sensory level at T3 to pinprick and temperature. He started experiencing constipation and urinary retention. Emergent magnetic resonance imaging (MRI) of the cervical, thoracic, and lumbar spine with and without contrast revealed a longitudinally extensive transverse myelitis from C3 to T12 with minimal enhancement at C5 to C6 (**Figure 2**). There was no cord compression at any level and no flow voids concerning for dural arteriovenous fistula. MRI of the brain without contrast did not show any significant T2 hyperintensity or pathology. Pertinent workup revealed normal vitamin B_{12} and copper levels, negative Lyme titers and rapid plasma reagin levels, and negative HIV testing. He was treated with 1 g of intravenous methylprednisolone daily for 5 days. On day 4 of initiating steroids, serum aquaporin-4 (AQP4) receptor antibody testing returned positive results (neuromyelitis optica [NMO]/AQP4 fluorescence-activated cell-sorting assay titer 1:100 titer, reference <1:5), and the diagnosis of NMO spectrum disorder (NMOSD) was made. The patient was treated with plasma exchange followed by a short oral steroid taper, resulting in improvement of his lower-extremity weakness.

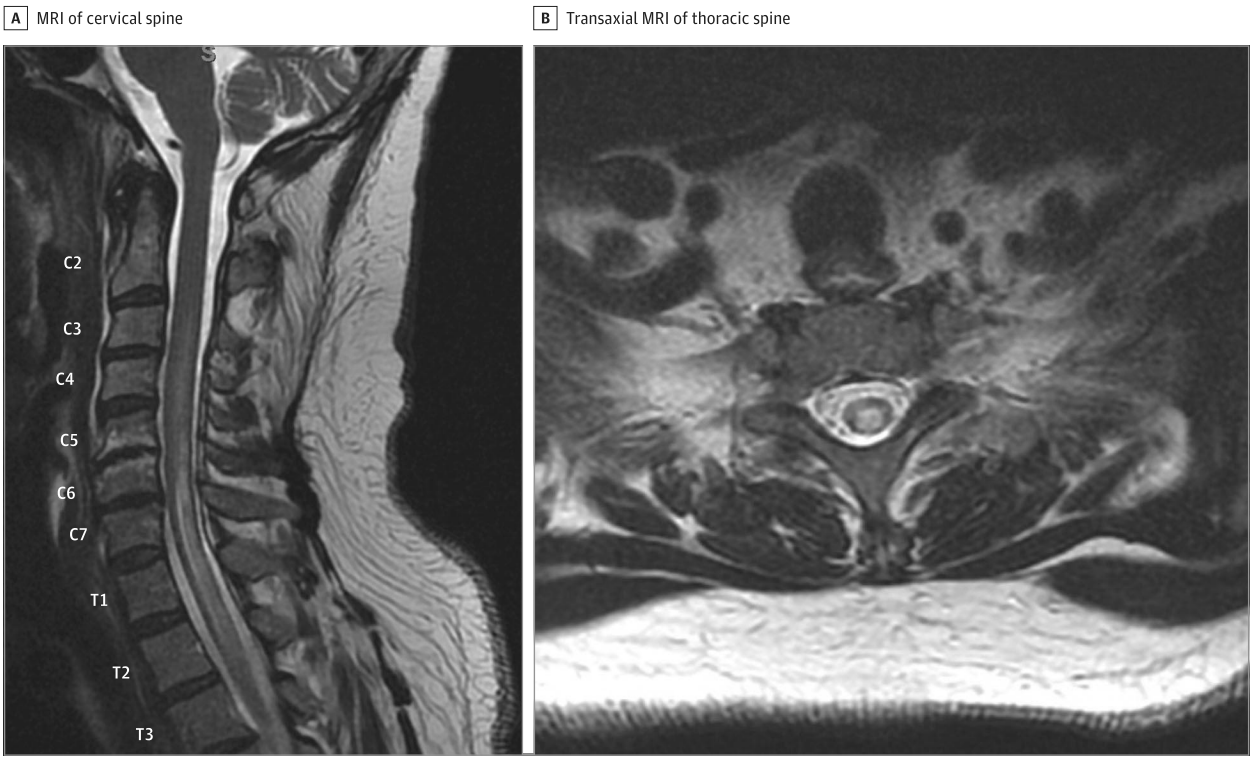
LR is a transient, reddish-blue, netlike pattern of the skin that can be seen in various physiologic and pathologic conditions, including hypothyroidism, reflex sympathetic dystrophy, and amantadine use.[1] The proposed mechanism involves spontaneous cutaneous arteriolar vasospasm, leading to an increased proportion of deoxygenated blood, and any process that reduces blood flow to the skin. Although LR has been rarely reported in patients with multiple sclerosis, to our knowledge, this is the first

Figure 1. Clinical Examination



Coalescing lacelike, reticular, violaceous, hyperpigmented rash of the right abdomen (A), bilateral lower extremities (B), and right flank with a sharp midline delineation consistent with livedo reticularis.

Figure 2. Magnetic Resonance Imaging (MRI) Analysis



A, MRI of the cervical spine showing T2 hyperintensity from the C3 to the T4 levels, associated with faint, ill-defined enhancement at the C5 and C6 levels (not shown). MRI of the thoracic spine also revealed T2 hyperintensity throughout the central thoracic cord without pathologic enhancement or expansion of the cord (not shown). B, Transaxial MRI of the thoracic spine at T1 showing central T2 hyperintensity.

case of LR associated with NMOSD.[2] Additionally, LR has not been described as a known dermatologic finding in patients with psoriatic arthritis.[1,3] This patient's initial presentation of upper-extremity numbness and tingling is likely the first symptom of acute myelitis. This was associated with transient LR, suggesting a temporal correlation between the 2 conditions. Interestingly, this patient's LR followed a dermatomal pattern along where the cord lesion extended. Therefore, cutaneous findings, such as LR, could be considered as a presenting sign for patients with NMOSD in the absence of other known predisposing disorders.

ARTICLE INFORMATION

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