Intralymphatic Histiocytosis Associated With an Orthopedic Metal Implant

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PRACTICE **POINTS**

- Consider intralymphatic histiocytosis in the differential diagnosis of an asymptomatic skin lesion overlying a joint, particularly in patients with orthopedic metal implants or rheumatoid arthritis.
- Biopsy is essential for the diagnosis of intralymphatic histiocytosis; special stains highlighting dilated lymphatic vessels and intravascular histiocytes may be necessary.
- Intralymphatic histiocytosis is chronic and resistant to therapy; however, patients can be reassured that the condition runs a benign course.

To the Editor:

A 70-year-old white man presented with an asymptomatic patch on the lateral aspect of the right thigh of 15 months' duration. The patient believed the patch correlated with a hip replacement 3 years prior; however, it was 6 inches inferior to the incision site. Physical examination revealed a 30-cm pink and violaceous, asymmetric, reticulated patch (Figure 1). The patch was unresponsive to topical corticosteroids as well as a short course of oral prednisone. The patient's medical history was notable for type 2 diabetes mellitus. Histopathologic examination revealed widely dilated vascular channels containing collections of histiocytes in the superficial dermis. In addition, adjacent features of chronic lymphedema were present, namely interstitial fibroplasia with dilated lymphatic vessels and a lymphoplasmacytic infiltrate (Figure 2). These findings were consistent with intralymphatic histiocytosis, a rare disease most commonly associated with rheumatoid arthritis. Our patient did not have a history or clinical symptoms of rheumatoid arthritis.

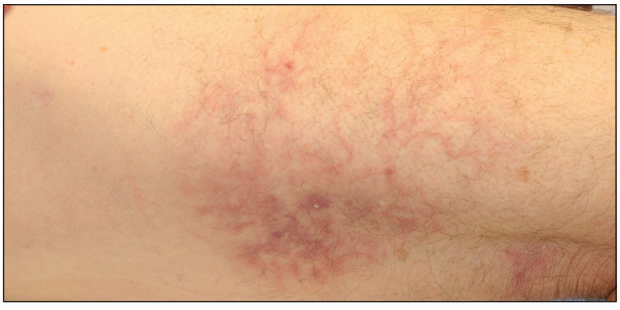

Figure 1. A 30-cm pink and violaceous, asymmetric, reticulated patch on the lateral aspect of the right thigh.

Intralymphatic histiocytosis is a rare cutaneous condition reported by O'Grady et al[1] in 1994. This condition has been most frequently associated with rheumatoid arthritis[2]; however, there has been an emerging association in patients with orthopedic metal implants, with and without a concomitant diagnosis of rheumatoid arthritis. Cases associated with metal implants are rare.[2-7]

The condition presents as asymptomatic red, brown, or violaceous patches, plaques, papules, or

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The authors report no conflict of interest.

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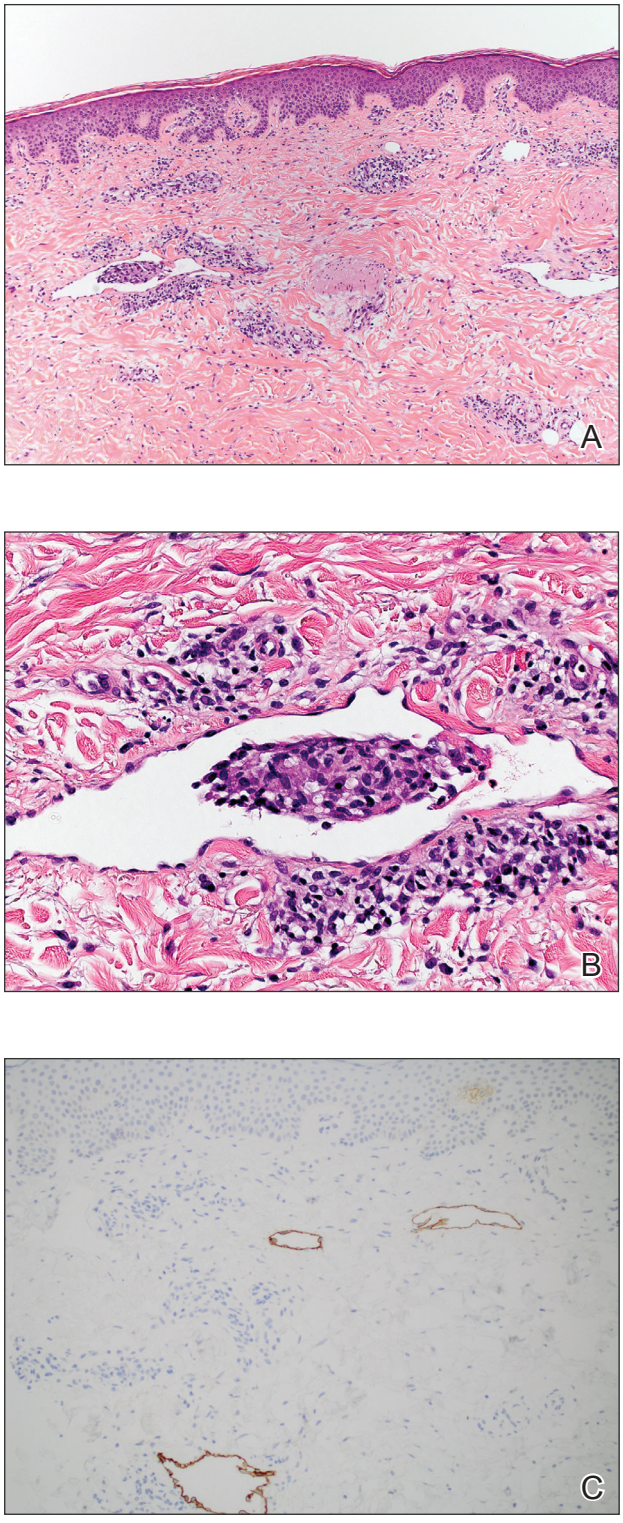

Figure 2. Histopathology revealed widely dilated vascular channels containing collections of histiocytes in the superficial dermis with adjacent features of chronic lymphedema (A)(H&E, original magnification ×10) as well as a collection of histiocytes in a dilated lymphatic channel (B)(H&E, original magnification ×40). D2-40 staining demonstrated ectatic lymphatic vessels in the upper dermis (C)(original magnification ×20).

nodules that are ill defined and tend to demonstrate a livedo reticularis–like pattern. The lesions typically are overlying or in close proximity to a joint. Histopathologic findings include dilated vascular structures in the reticular dermis, some with empty lumina and others containing collections of mononuclear histiocytes. There also may be an inflammatory infiltrate in the adjacent dermis composed of a mix of lymphocytes, plasma cells, and/or histiocytes. Endothelial cells lining the dilated lumina express immunoreactivity for CD31, CD34, D2-40, Lyve-1, and Prox-1. Intravascular histiocytes are positive for CD68 and CD31.[6]

The pathogenesis of intralymphatic histiocytosis remains undefined. Some hypothesize that intralymphatic histiocytosis could be the early stage of reactive angioendotheliomatosis, as these conditions share clinical and histological features.[8] Reactive angioendotheliomatosis also is a rare condition that may present as erythematous to violaceous patches or plaques. The lesions are commonly found on the limbs and may be associated with constitutional symptoms. Histologic findings of reactive angioendotheliomatosis include a proliferation of epithelioid, round, or spindle-shaped cells within the lumina of dermal blood vessels, which show positivity for CD31 and CD34.[9] Others suggest the lesions of intralymphatic histiocytosis arise from lymphangiectasia; obstruction of lymphatic drainage due to congenital abnormalities; or acquired damage from infection, trauma, surgery, or radiation.[2] Due to the common association with rheumatoid arthritis and orthopedic implants, it is likely that lymphatic stasis secondary to chronic inflammation plays a notable role.

Therapies such as topical and systemic corticosteroids, local radiotherapy, cyclophosphamide, pentoxifylline, and arthrocentesis have been attempted without evidence of efficacy.[2] Although intralymphatic histiocytosis is chronic and resistant to therapy, patients can be reassured that the condition runs a benign course.

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