Hyperpigmented Patch on the Leg

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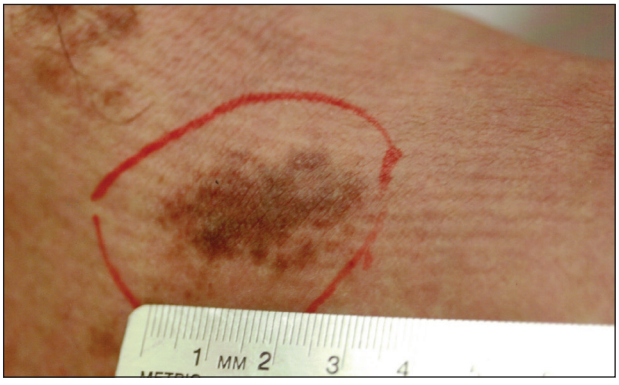

A 32-year-old man presented with an asymptomatic pigmented lesion on the left foot that developed over the course of 4 months. Physical examination revealed a 4-cm asymmetrical, deeply pigmented macule on the left foot. A shave biopsy of the lesion was performed.

WHAT'S THE **DIAGNOSIS?**

a. lentigo maligna
b. lichen aureus
c. purpura annularis telangiectodes of Majocchi
d. Schamberg disease
e. solar lentigo

PLEASE TURN TO PAGE E2 FOR THE DIAGNOSIS

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The authors report no conflict of interest.
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THE **DIAGNOSIS:** Lichen Aureus

The clinicopathological findings were diagnostic of lichen aureus (LA). Microscopic examination revealed a relatively sparse, superficial, perivascular and interstitial lymphohistiocytic infiltrate with scattered siderophages in the upper dermis. Extravasation of red blood cells also was noted (Figure 1). An immunohistochemical stain for Melan-A highlighted a normal number and distribution of single melanocytes at the dermoepidermal junction with no evidence of pagetoid scatter. A Perls Prussian blue stain for iron demonstrated abundant hemosiderin in the dermis (Figure 2).

Pigmented purpuric dermatosis (PPD) describes a group of cutaneous lesions that are characterized by petechiae and pigmentary changes. These lesions most commonly present on the lower limbs; however, other sites have been reported.[1] This group includes several major clinical forms such as Schamberg disease, LA, purpura annularis telangiectodes of Majocchi, eczematidlike purpura of Doucas and Kapetanakis, and lichenoid PPD of Gougerot and Blum. Lesions typically demonstrate a striking golden brown color clinically and by definition occur in the absence of platelet defects or vasculitis.[1]

Factors implicated in the pathogenesis of pigmented purpura include gravitational dependency, venous stasis, infection, and drugs.[2] It is suggested that cellular immunity may play a role in the development of the disease based on the presence of $CD4^+$ T lymphocytes in the infiltrate and the expression of HLA-DR by these lymphocytes and the keratinocytes.[3] Lichen aureus differs in that it relates to increased intravascular pressure from an incompetent valve in an underlying perforating vein.[4]

Lichen aureus, also referred to as lichen purpuricus, is one major variant of PPD. The name reflects both the characteristic golden brown color and the histopathologic pattern of inflammation.[1] Lichen aureus usually presents as a unilateral, asymptomatic, confined single lesion located mainly on the leg,[1] though it can develop at other sites or as a localized group of lesions. Extensive lesions have been reported[5] and cases with a segmental distribution have been described.[6] In contrast, Schamberg disease demonstrates pinhead-sized reddish lesions giving the characteristic cayenne pepper pigmentation. These lesions coalesce to form thumbprint patches that progress proximally.[1] Majocchi purpura is annular and telangiectatic, while lichenoid purpura of Gougerot and Blum presents with flat-topped, polygonal, violaceous papules that turn brown over time.

Some authors have championed a role for dermoscopy in diagnosis of LA.[7] By dermoscopy, LA demonstrates a diffuse copper background reflecting the lymphohistiocytic dermal infiltrate, red dots and globules representing the extravasated red blood cells and the dilated swollen

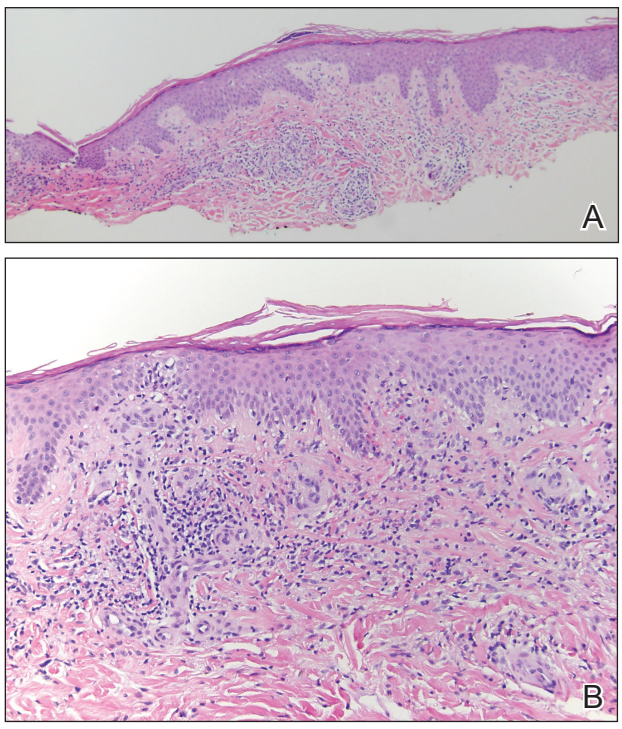

FIGURE 1. Lichen aureus histopathology revealed a superficial lymphohistiocytic infiltrate (A)(H&E, original magnification ×40) with scattered siderophages and extravasated red blood cells (B)(H&E, original magnification ×200).

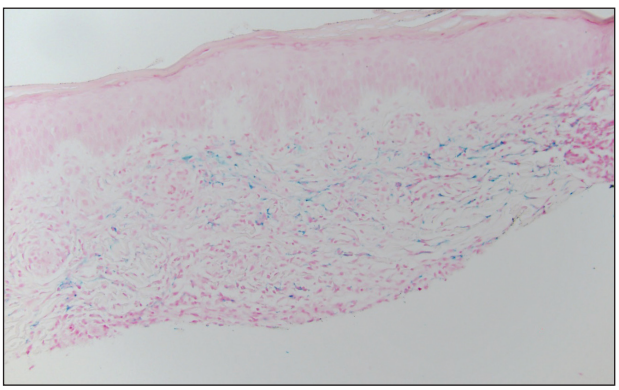

FIGURE 2. Lichen aureus histopathology using the Perls Prussian blue stain for iron demonstrated abundant hemosiderin in the dermis (original magnification ×40).

vessels, and grey dots that reflect the hemosiderin present in the dermis.[8]

Histologically, LA demonstrates a superficial perivascular infiltrate composed mainly of $CD4^+$ lymphocytes surrounding the superficial capillaries. Over time, red cell extravasation leads to the formation of hemosiderin-laden macrophages, which can be highlighted with Perls Prussian blue stain. A bandlike infiltrate with thin strands of collagen separating it from the epidermis also may be noted.[9]

An important consideration in the differential diagnosis of PPD is mycosis fungoides (MF). Mycosis fungoides is a cutaneous T-cell lymphoma that clinically presents as a single or multiple hypopigmented or hyperpigmented patches or as erythematous scaly lesions in the patch or plaque stage. These lesions eventually may evolve into tumor stage.[10] Mycosis fungoides may mimic PPD clinically and/or histopathologically, and rarely PPD also may precede MF.[11] Involvement of the trunk, especially the lower abdomen and buttock region, favors a diagnosis of MF. Typically, histopathologic examination of MF demonstrates an epidermotropic lymphocytic infiltrate composed of atypical cerebriform lymphocytes overlying papillary dermal fibrosis. Although classic MF would be difficult to confuse with PPD, the atrophic lichenoid pattern of MF may show remarkable overlap with PPD.[12] Such cases require clinicopathologic correlation, immunophenotyping of the epidermotropic lymphocytes, and occasionally T-cell clonality studies.

Lichen aureus is a chronic persistent disease unless the underlying incompetent perforator vessel is ligated. Various treatments have been used for other forms of pigmented purpura including topical corticosteroids, topical tacrolimus, systemic vasodilators such as prostacyclin and pentoxifylline, and phototherapy.[1] Clinical follow-up is recommended for lesions that show some clinical or histopathological overlap with MF. Additional biopsies also may prove useful in establishing a definitive diagnosis in ambiguous cases.

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