Lichen Aureus

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Lichen aureus is a localized variant of pigmented purpuric dermatitis (PPD) and is rarely reported. A case of lichen aureus in a 28-year-old woman and a 13-year-old boy are presented. Biopsy results revealed the characteristic pattern of lichen aureus, including diffuse deposition of hemosiderin and dermal infiltrate of lymphocytes and histiocytes, some of which had Birbeck granules in the cytoplasm. The former case is unusual for its zosteriform distribution and localization (lower abdomen).

igmented purpuric dermatitis (PPD) encompasses several subgroups including purpura annularis telangiectodes (Majocchi disease), progressive pigmentary dermatosis (Schamberg's disease), pigmented purpuric lichenoid dermatitis (Gougerot-Blum disease), eczematoidlike purpura, and lichen aureus.1 Lichen aureus, originally reported in 1958 by Martin,² is a rarely reported subset of PPD. We describe 2 cases of lichen aureus, one of which showed a zosteriform distribution on the lower abdomen.

Case Reports

Patient 1—A 28-year-old Japanese woman presented with a 1-year history of a slowly spreading rash on her right lower abdomen without history of antecedent trauma. At initial presentation, she told a history of being treated with minocycline and clarithromycin for pelvic peritonitis due to Chlamydia.

The lesions on the right half of her lower abdomen consisted of closely aggregated, superficial, orange-brown flat papules (Figure 1). Each patch had petechiae at the center and a yellow or golden hue at the periphery. The distribution was asymmetric and zosteriform. Results of a Rumpel-Leede test (capillary fragility test) were negative.

Biopsy results revealed mild orthohyperkeratosis and partial flattening of the rete ridges in the epi-



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Figure 1. Closely aggregated, superficial, orange-brown patches distributed in zosteriform on the lower abdomen.

dermis. A bandlike infiltrate of lymphocytes and histiocytes beneath a narrow grenz zone of normal tissue was observed in the upper dermis, interspersed with extravasated erythrocytes (Figure 2). The histiocytes had convoluted or bean-shaped nuclei and abundant eosinophilic cytoplasm. With electron microscopy, large histiocytic cells containing Birbeck granules in their cytoplasm were seen among the lymphocytes in the upper dermis (Figure 3), indicating that these histiocytic cells were Langerhans cells. Pigment-laden macrophages stained positive for iron.

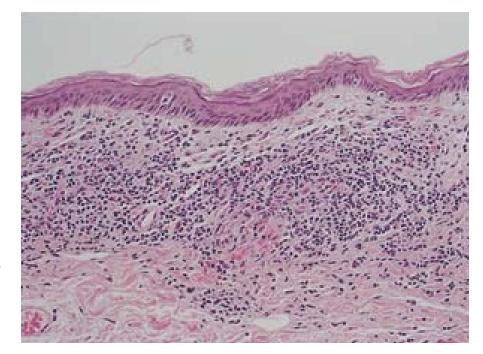


Figure 2. Dense, bandlike infiltrate beneath a narrow grenz zone of normal tissue in the upper dermis. The infiltrate is composed of lymphocytes and histiocytes that had convoluted nuclei. Extravasated erythrocytes also were evident.

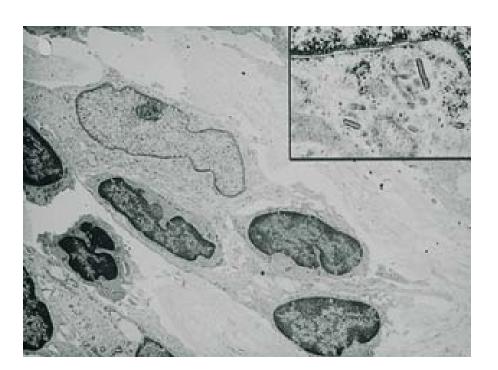


Figure 3. Large histiocytes showing Birbeck granules (inset) in the cytoplasm seen adjacent to the lymphocytes. (H&E, original magnification ×3000 and ×12,000).

The lesion persisted unchanged for 3 weeks of treatment with betamethasone dipropionate ointment twice a day. She underwent an operation for appendicitis while being treated for lichen aureus, and neither the surgical procedure nor the systemic administration of antibiotics changed the skin lesion. She then was treated with Chinese herbal therapy

(granules of Unsei-in extract) for 2 months, and most of the golden and purplish color disappeared.

Patient 2—A 13-year-old boy, otherwise in good health, developed a nonpruritic erythematous patch on his left thigh that gradually enlarged and increased in number over 10 months. The lesion had since persisted unchanged.

Dermatologic examination at his first presentation revealed a group of lesions on his inner left thigh. The lesions consisted of flat-topped papules 1 to 2 mm in diameter with a macular halo of rust-colored pigmentation (Figure 4). Very fine petechial elements could be seen in the papules. Skin biopsy results revealed a normal epidermis and dermal changes similar to those in the previous case, though the histiocytic infiltration was less dense and the bandlike pattern was less distinct.

Comment

Lichen aureus, also known as lichen purpuricus, is classified as a localized uncommon variant of PPD. The eruption is usually asymptomatic, highly chronic, and therapy resistant. The lesions are clinically characterized by a distinctive gold, rust, or orange-brown color and show a predilection for men and lower extremities, most commonly the lower legs. Histopathologically, the major finding is a dense, bandlike lymphohistiocytic infiltration with hemosiderin deposits separated from the epidermis by a narrow grenz zone in the upper dermis.³

Patient 1 is an unusual case because the lesions showed zosteriform distribution on the lower abdomen. In rare instances, the lesions may be in a linear or zosteriform pattern, and several authors have described zosteriform or segmental lichen aureus of the extremities. ⁴⁻⁶ In these cases, the lesions corresponded neither to a dermatome nor to Blaschko lines, but did follow the course of veins. ^{6.7} More recently, Palleschi et al⁸ reported bilateral acral lichen aureus that showed linear disposition. In patient 1, the lesions loosely followed the dermatome but not the course of vessels or Blaschko lines.

The etiology of lichen aureus is unknown. Several explanations concerning the pathogenic mechanism of PPD have been postulated. One possible explanation is the involvement of cellular immunity. Most of the infiltrating cells in the lesions were CD4⁺ T cells, and these cells, as well as keratinocytes, expressed HLA-DR antigens.⁹ As in patient 1, remarkable accumulation of Langerhans cells in the upper dermis also was observed.¹⁰

An alternative explanation is venous insufficiency. Shelly et al¹¹ reported an obvious varicose perforator vein in patients with lichen aureus. This hypothesis, however, cannot explain the appearance of the eruption on the trunk and upper extremities as seen in patient 1 nor the eruptions appearing in a young age group, as in patient 2. We believe that venous insufficiency is only one of the many possible causes of endothelial damage.



Figure 4. Rust-colored patches with purpuric elements on the inner left thigh.

Other investigators have suggested drugs,¹² capillary fragility,¹³⁻¹⁵ or infection¹⁵ as causative factors of PPD and lichen aureus; however, no conclusive evidence has been obtained. In patient 1 in this report, treatment of pelvic peritonitis and appendicitis with antibiotics had no effect on the skin lesions. It seems likely that the eruption disappeared spontaneously in patient 1 because the antiallergenic and anti-inflammatory efficacy of Chinese herbal therapy is still dubious.

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