SKIMages

Diagnostic Challenges of Hypertrophic Lupus Erythematosus

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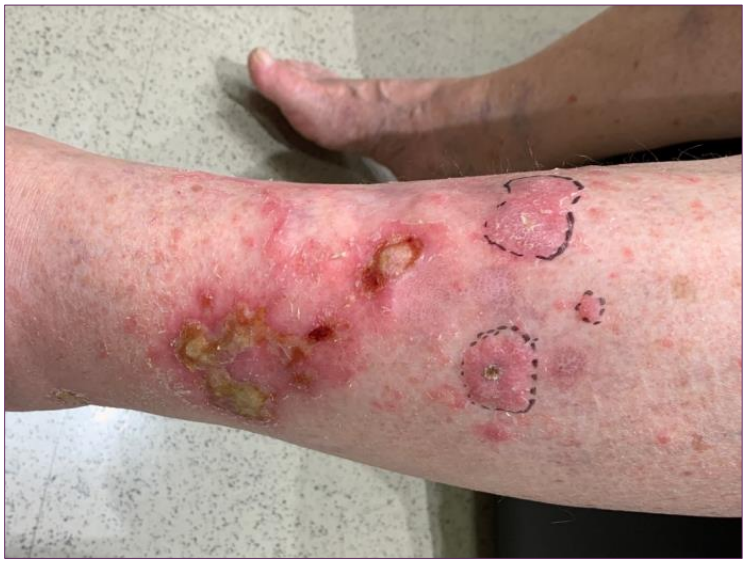
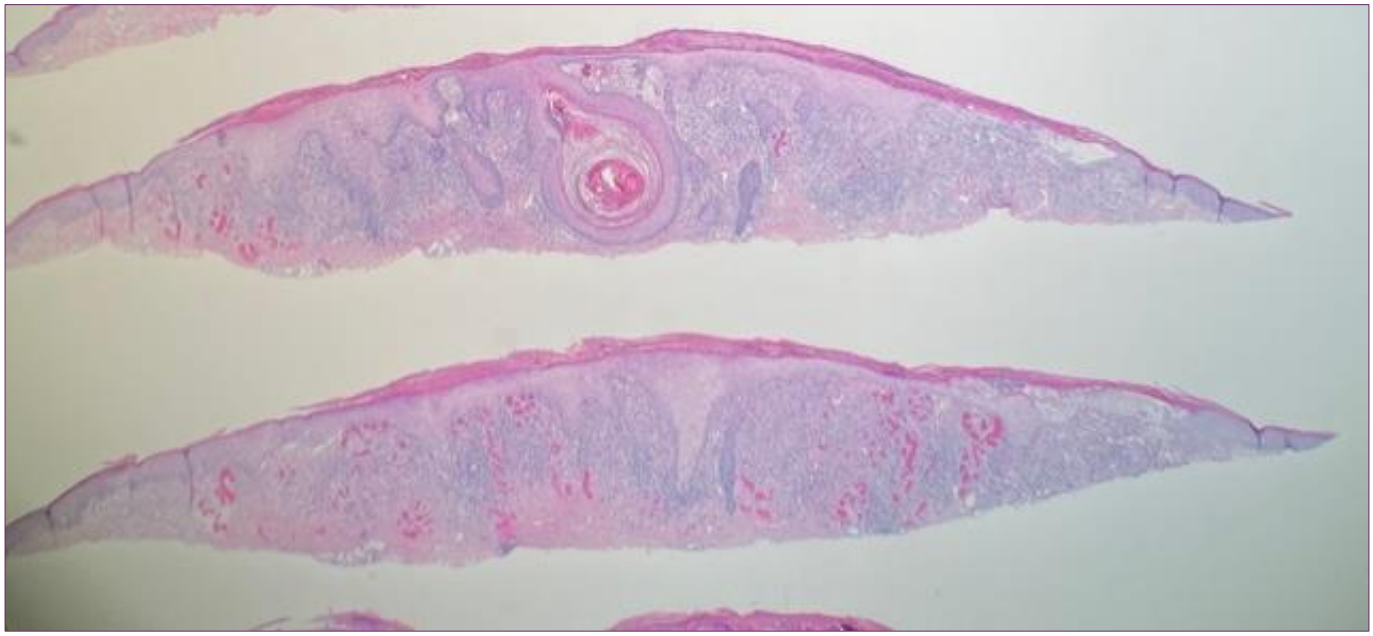


Hypertrophic lupus erythematosus (HLE), a variant of cutaneous LE, often occurs in the absence of systemic symptoms and can be misdiagnosed as squamous cell carcinoma (SCC) as both appear clinically as erythematous scaling plaques in sun exposed areas and histologically show significant epithelial hyperplasia.[1] Distinguishing HLE from SCC can be difficult, thus a detailed clinical history and adjunct testing to rule out inflammatory conditions are necessary to direct treatment. Multiple clinical factors including a history of discoid lupus erythematosus, positive serologies, direct immunofluorescence studies, and anatomical location of the lesions on the face, arms, or chest, coupled with histological findings can lead pathologists to confidently diagnose HLE; however, the absence of systemic symptoms in the clinical history often leads to misdiagnosis as squamous neoplasia.[3] CD123 immunostains are helpful in diagnosing HLE and should be considered for locally recurring presumed SCC.[2]

A 76-year-old woman presented for evaluation of multiple well-differentiated SCCs (wdSCC) of the left lower leg. Previous treatments included curettage, multiple Mohs micrographic surgeries (MMS), and topical fluorouracil resulting in more inflammation and pain. Despite



surgical clearance, she developed new biopsy-proven wdSCC near treated sites. She presented for a second opinion due to non-healing wounds and surgical fatigue. A trial of intralesional Kenalog (ILK) injections significantly decreased pain and size of plaques. The patient had a negative past medical history of personal or familial autoimmune diseases, rash elsewhere, joint pain, and immunosuppression.

Examination showed erythematous, violaceous round plaques and smaller satellite papules with thin overlying scale coalescing on the left lower shin and erosions with yellow-brown crust on the lateral and medial aspect of the left lower leg. Labs were negative for ANA, SSA, and SSB. Urinalysis was negative for blood and protein. Ten prior left lower leg outside biopsies reported wdSCC. A second-read revealed lichenoid dermatitis with pseudoepitheliomatous hyperplasia.

Due to her clinical course and differential including HLE, immunostains were performed revealing CD123 positivity and increased peripheral epidermal plasmacytoid cells. CD123 tends to be more strongly positive in HLE. At the bases of the rete pegs, there are interface changes throughout, especially at the base of the elongation. This tends to occur less frequently with SCC. In some biopsies, follicular plugging was identified along with a subtle increase in dermal mucin. Oftentimes, this will lead to a differential of HLE vs HLP. In addition, eosinophils were identified which is often present in hypertrophic lupus and not hypertrophic lichen and planus. Intralesional Kenalog (0.5 cc of Kenalog 20) was injected into 4 sites on the left lower leg. She was also given betamethasone 0.05% cream to apply twice daily to the affected areas of the left lower leg for 3 weeks. At 3-week follow-up, patient noted improvement in lesions evidenced by decrease in plaque size, erythema, and swelling. An additional 2.0 cc of Kenalog 20 were injected, and she was started on daily triamcinolone 0.1% ointment. She was referred to Rheumatology-Dermatology clinic and noted continued improvement with physical exam notable for atrophic pink plaques on the left lower leg, with no evidence of active disease. She was continued on triamcinolone 0.1% ointment

daily to the left lower leg as needed. These histological findings with improvement after ILK injections and topical steroids favored the diagnosis of HLE.

Given the excellent cure rate of wdSCC when treated appropriately, apparent recurrent local metastasis should prompt clinicians to rule out inflammatory processes, such as HLE or hypertrophic lichen planus. Diagnosing HLE is difficult due to a lack of systemic symptoms and clinical and microscopic similarity to other cutaneous diseases. No single criterion confidently differentiates HLE from SCC histologically, postulating the need for CD123 immunostaining in evaluating locally recurring, supposed SCCs.[1] CD123 is highly expressed on the surface of plasmacytoid dendrocytes, seen in HLE lesions densely at the epidermal-dermal junction, while SCC lacks this positive band staining. Recurrent lesions unresponsive to MMS and positive CD123 immunostaining are highly suggestive of HLE.

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