Targetoid-like Lesions in the Setting of Systemic Lupus Erythematosus: A Case of Rowell Syndrome

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Excessive sun exposure in the context of tapered doses of hydroxychloroquine (HCQ) and methotrexate (MTX), erythema multiforme–like (EM-like) skin lesions, and speckled patterned antibodies led to a diagnosis of Rowell syndrome (RS) in a patient with systemic lupus erythematosus (SLE).

A 39-year-old woman with longstanding SLE, Raynaud phenomenon, and Sjögren syndrome reported 6 days of worsening rash following a picnic. Six months prior, her HCQ dose was halved and MTX dose decreased. She denied common triggers of EM or Stevens-Johnson syndrome (SJS), specifically herpes infections and new medications.

Examination revealed dusky, necrotic papules, plaques, vesicles, and bullae on erythematous bases, many targetoid in appearance, involving face, trunk, and extremities (Figure 1). Histopathology demonstrated basket-weave stratum corneum, underlying broad epidermal necrosis, and vacuolar interface changes of EM (Figure 2). Anti-Ro, anti-dsDNA, and anti-Sm antibodies were positive; rheumatoid factor (RF) was negative.

This presentation is typical of RS: EM-like lesions complicating lupus erythematosus (LE). The literature outlines 3 major criteria [SLE, discoid LE, or subacute cutaneous LE; EM-like lesions; and speckled pattern antinuclear antibody (ANA)] and 3 minor criteria (chilblains, anti-Ro antibodies, anti-La antibodies, and positive RF), with all 3 major criteria and 1 minor required for diagnosis[1,2]. However, because anti-Ro/La antibodies are speckled patterned on indirect immunofluorescence, some criteria are redundant[3]. In this patient with SLE, EM-like lesions, and speckled patterned antibodies including anti-Ro and anti-Sm, RS was diagnosed. ANA was not obtained, because once positive in ANA-associated autoimmune disease, repeat ANA is not indicated[4]. Differential diagnosis of RS includes bullous lupus, EM, SJS, fixed drug eruption, bullous pemphigoid, and leukocytoclastic vasculitis[5].

RS was attributed to excessive sun exposure in the setting of tapered HCQ and MTX. Management included systemic corticosteroids, HCQ dosage increase, and sun avoidance.

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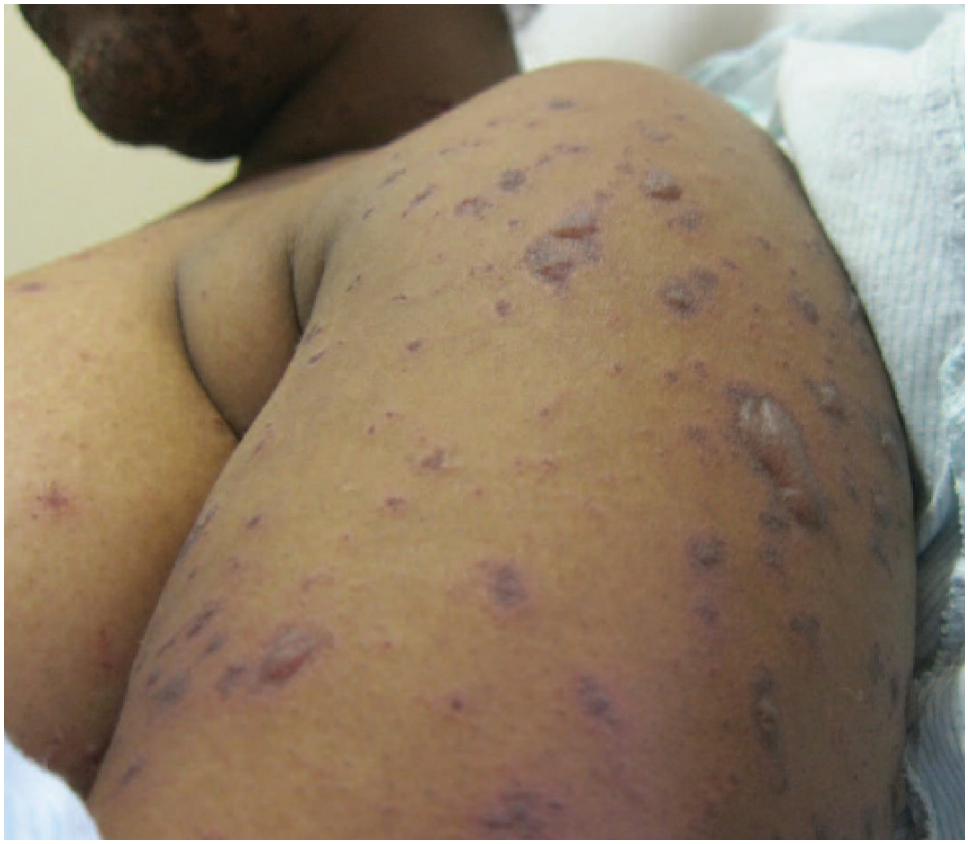

Figure 1. Necrotic vesicles and bullae on a dusky, erythematous base, many targetoid in appearance, are scattered over the arm.

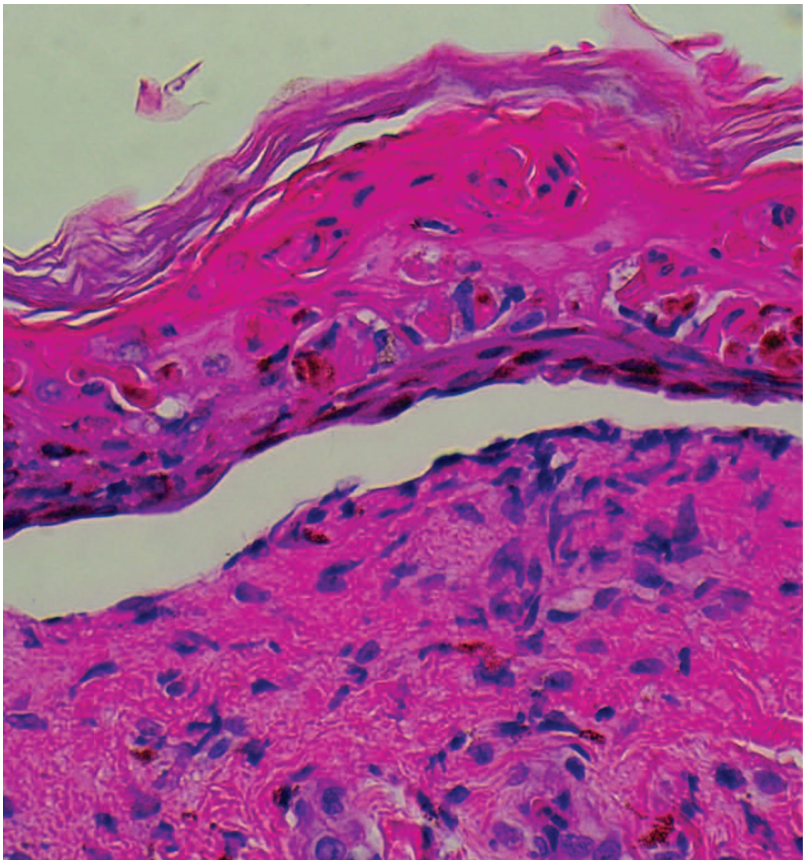

Figure 2. Basket-weave stratum corneum overlies broad epidermal necrosis with separation between the necrotic epidermis and the underlying dermis. Dyskeratotic keratinocytes and vacuolar interface changes extend into the surrounding epidermis and adnexal structures.