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Neutrophilic urticarial dermatosis with response to dapsone in a patient with lupus

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A 41-year-old female with systemic lupus erythematosus (SLE)/Sjogren's syndrome overlap (positive ANA 1:320 homogenous pattern, positive Ro), celiac disease, Hashimoto's thyroiditis, and facial angioedema presented with a refractory chronic urticarial eruption of the extremities and trunk. She noted associated itching and burning and denied photosensitivity, fevers or arthralgias. Prior diagnosis was chronic idiopathic urticaria but high dose oral antihistamines and montelukast failed to control her symptoms. Physical examination showed edematous urticarial papules on her upper extremities, lower back, chest, palms and angioedema of her upper cutaneous lip. A punch biopsy of the palm and chest revealed superficial and deep perivascular and interstitial neutrophilic infiltrate, without overt evidence of vasculitis. Direct immunofluorescence showed granular deposition of IgA, IgG and IgM in the basement membrane zone. Serologic workup demonstrated elevated ESR, low C4, low-normal C3, and normal dsDNA. Clinical findings and pathology results were most consistent with neutrophilic urticarial dermatosis (NUD). NUD clinically resembles urticaria but presents with neutrophilic dermal infiltrate on pathology and typically affects patients with underlying systemic conditions like SLE or underlying autoinflammatory disorders. Patients with lupus are most commonly Ro positive as in this case.¹ Differential diagnosis includes other neutrophilic dermatosis such as urticarial vasculitis, Sweet syndrome and palisaded neutrophilic and granulomatous dermatosis. NUD is often misdiagnosed as a lupus flare, which can lead to treatment with immunosuppressive drugs. In contrast, NUD usually responds well to colchicine or dapsone.² Our patient received dapsone and experienced rapid and complete resolution of the rash and angioedema.

References:

1. Lee WJ, Kang HJ, Shin HJ, et al. Neutrophilic urticarial dermatosis and Sweet-like neutrophilic dermatosis: under-recognized neutrophilic dermatoses in lupus erythematosus. *Lupus*. 2018;27(4):628-636. doi:10.1177/0961203317736145
2. Gusdorf L, Bessis D, Lipsker D. Lupus Erythematosus and Neutrophilic Urticarial Dermatitis: A Retrospective Study of 7 Patients. *Medicine (Baltimore)*. 2014;93(29):e351. doi:10.1097/MD.0000000000000351

Figures 1-2. Edematous urticarial papules present on the patient's palm and forearm, respectively.

Figure 3. Low magnification of histopathological image showing a superficial and deep perivascular interstitial neutrophilic infiltrate without distinct evidence of vasculitis (hematoxylin-eosin stain).

Figure 4. High magnification showing the deep perivascular interstitial neutrophilic infiltrate (hematoxylin-eosin stain).