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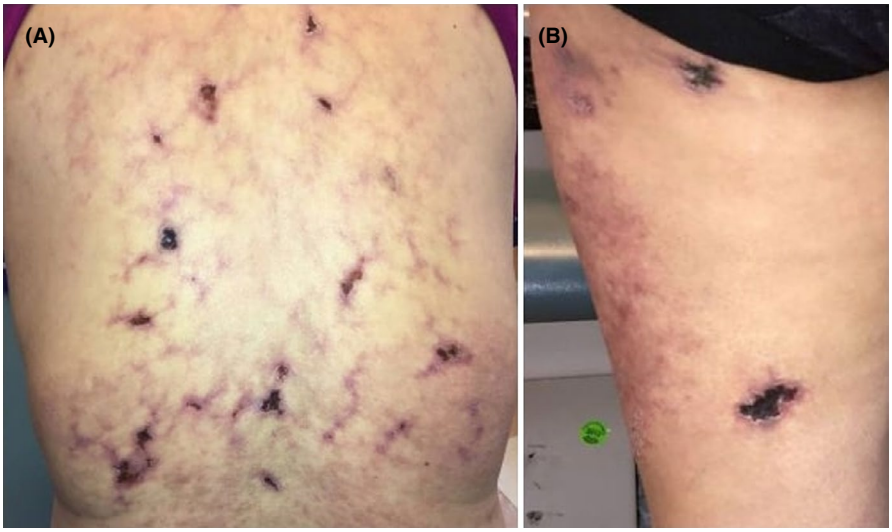
Clinical Images: A not so common overlap presentation in systemic lupus erythematosus



Melanoma differentiation–associated gene 5 (MDA-5) is associated with dermatomyositis and often presents with a distinct cutaneous presentation and progressive interstitial lung disease (ILD) (1–3). MDA-5 signaling often leads to increased interferon signaling, which contributes to atypical vasculopathy, leading to atypical, and at times ulcerative or necrotic, cutaneous manifestations, which are challenging to diagnose and treat (2). Rarely, patients with lupus present with necrotic skin lesions as an overlapping presentation from MDA-5 dermatomyositis. We report a 38-year-old woman who presented with malar rash, oral ulcers, nonscarring alopecia, and arthritis. Laboratory tests revealed high titers of antinuclear antibodies and low levels of complements. She was diagnosed with lupus and started on steroids, hydroxychloroquine, and azathioprine. After 3 months, she presented with new painful necrotic skin lesions on her back (**A**), buttocks/legs (**B**), elbows, and hips. We noted elevated liver function test (LFT) results, normal muscle enzyme levels, low positive titers of anticardiolipin and β2-glycoprotein immunoglobulin M antibodies, whereas other antibodies were negative. A concern for antiphospholipid syndrome or vasculitis was raised. Her imaging did not reveal any evidence of thrombosis or vasculitis, a chest computed tomography scan revealed ILD, and her skin biopsy did not reveal any vasculitis or thrombotic vasculopathy. Interestingly, the skin biopsy revealed interface dermatitis with parakeratotic scaling and negative immunofluorescence staining results, raising a concern for other interface processes. On the basis of evidence from previous literature describing such necrotic skin lesions in patients with MDA-5 dermatomyositis and lack of response to high-dose steroids, MDA-5 levels were checked, which were highly positive (3). Therefore, intravenous immunoglobulin (IVIG) was added to her medication regimen, which led to marked improvement in skin lesions, LFT results, and ILD. On retesting, all antiphospholipid antibodies were negative. Therefore, consistent with previous reports, a skin biopsy lacking evidence of vasculitis or thrombotic vasculopathy, and a great response to IVIG, we report a not so common overlap presentation of necrotic skin lesions driven by MDA-5 in lupus (2).

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