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Clinical Image
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Unique Cutaneous Manifestation of SLE

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Clinical Image

A 22-year-old Caucasian male with Systemic Lupus Erythematosus (SLE) diagnosed in 2012 presented with a two-day history of rash, shortness of breath, fever, and chest pain. Physical exam showed annular skin plaques studded with vesicles and bullae involving the extremities, chest and back (Figure 1). Laboratory work up revealed a positive antinuclear antibody test by immunofluorescence (1:1280, homogenous pattern), positive SSA and SSB, elevated anti double stranded DNA antibody, low complement C3 (31 mg/dl) and C4 (8 mg/dl) levels, white blood cell count of 2,200 cells/mm^3, platelet count of 70,000 cells/mm^3. Echocardiography of the heart showed reduced ejection fraction of 40% and pericardial effusion. Cardiac MRI was consistent with pericarditis. Histopathology of the bullous lesions revealed sub epidermal neutrophil-rich vesicles while that of the annular polycyclic lesions showed basement membrane vacuolization and dermal mucin deposition. Direct immunofluorescence showed confluent granular deposition of IgG, IgA, IgM and C3 and fibrin along the basement membrane. Kidney biopsy revealed WHO Class III lupus nephritis [Activity index = 10 (0 – 24) and Chronicity index was 3 (0 – 12)]. Patient was diagnosed with bullous lupus erythematosus with pericarditis and new onset lupus nephritis. He was treated with dapsone for his skin lesions with rapid improvement; and steroids, hydroxychloroquine and mycophenolate mofetil for SLE.

Bullous lesions are a rare (<5%) dermatologic manifestation of systemic lupus erythematosus with limited epidemiologic data [1]. They can be the initial presenting feature of SLE, requiring the astute physician to be aware of and consider BSLE when assessing a bullous skin eruption [2]. The lesions typically manifest axially on the trunk, head and neck [2]. As illustrated in Figure 1 however, our patient exhibited bullae primarily on the arms. The most concerning extra cutaneous manifestations of bullous lupus erythematosus include lupus nephritis, hemolytic abnormalities, serositis and neuropsychiatric lupus which can be life threatening [2]. Particular consideration is to be given towards evaluating for lupus nephritis, due to the high risk of concomitant and severe disease [2]. Treatment of skin lesions is with dapsone as a first line agent with a reported efficacy of 90% [2].

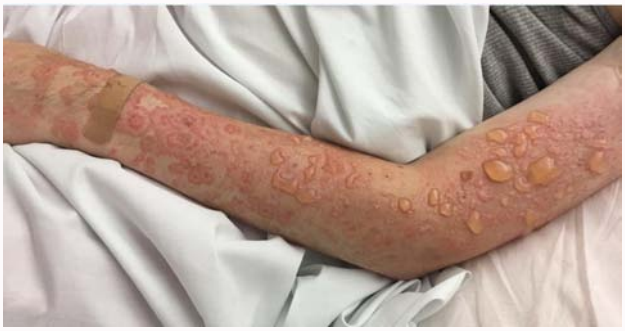

Figure 1: Demonstrates the annular plaques studded with vesicles and bullae both centrally and along the periphery involving the left upper extremity.

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