AN ATYPICAL PRESENTATION OF BULLOUS SYSTEMIC LUPUS ERYTHEMATOSUS

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Introduction:
Bullous systemic lupus erythematosus (BSLE) is a particularly uncommon manifestation of noncuring subepidermal bullous eruption of systemic lupus erythematosus (SLE) with an incidence of 0.26 per million persons per year in adults. BSLE presents with rapid onset of vesicles and bullae distributed throughout the body, concentrated on the face, neck, extremities, and trunk. The simultaneous occurrence of a primary bullous disease in a patient with SLE can be distinguished from BSLE, as bullous pemphigoid (BP), dermatitis herpetiformis (DH), linear IgA dermatosis, and epidermolysis bullosa acquisita (EBA) have all been reported in association with SLE. Typically, patients with BSLE will meet criteria for Systemic Lupus Erythematosus (SLE) as defined by the American College of Rheumatology (ACR) in addition to a widespread vesiculobullous eruption that is commonly unrelated to the severity of the SLE. BSLE is a rare and often difficult to diagnose manifestation of SLE, especially when it is the initial presentation. Here we report such a case of a 57-year-old Caucasian male with BSLE.

Case Report:
A 57-year-old, Caucasian male presented with a 1-month history of a diffuse vesiculobullous, pruritic, and painful eruption. He admitted to an abrupt onset of lesions. His medical history was significant for stage IV chronic kidney disease secondary to local segmental glomerulosclerosis (FSGS), anemia of chronic disease, and hyperparathyroidism. Physical examination revealed innumerable tense bullae on his trunk as well as upper and lower extremities bilaterally, with sparing of both knees and elbows. (Figures 1-2) Skin fragility and apparent scarring were absent. Upon oral examination, multiple erosions were observed. A biopsy specimen of perilesional skin demonstrated neutrophil-rich subepidermal bullae with clumps of neutrophils filling the dermal papillae and lining up along the dermo-epidermal junction (DEJ) with strong discontinuous IgG and C3 lining the basement membrane (Figure 3).

Routine laboratory investigations revealed the following results: hemoglobin, 8.6 g/dL (reference range, 13.0–16.0 g/dL); total white blood cell count, 5,200/mL (reference range, 4,000–10,500/mL); platelet count, 487,000/mL (reference range, 100,000–400,000/mL); serum creatinine, 3.06 mg/dL (reference range, 0.5–1.4 mg/dL); urea nitrogen, 59 mg/dL (reference range, 6–20 mg/dL); serum total protein, 6.8 g/dL (reference range, 6.0–8.3 g/dL); albumin, 4.3 g/dL (reference range, 3.2–5.5 g/dL); and total globulin, 2.5 g/dL (reference range, 1.7–3.9 g/dL). A urinalysis revealed a urine protein value of 100 mg/dL (reference, 0). Liver function tests revealed no abnormalities. Additional investigations revealed an antinuclear antibody of 1:40 with a positive indirect fluorescent antibody as well as an elevated anti-dsDNA at 32 (reference, ≤4). Smith antigen and Sjögren syndrome A and B antigens were negatives. Tests for antibodies to type VII collagen were not able to be performed.

A glucose-6-phosphate dehydrogenase level was found to be unremarkable and treatment with dapsone 25mg PO daily was initiated due to his longstanding history of anemia. There was no improvement noted after a 2-week period. He was then switched to a regimen of oral prednisone tapered over 3 weeks before starting hydroxychloroquine 200mg PO twice daily which was later increased to 400mg PO daily. Accurate resolution of bullous lesions was observed within 3 weeks and he admits to continued clearance of all lesions.

Discussion:
BSLE is believed to be triggered by circulating autoantibodies to type VII collagen. This eruption may represent the concurrence of lupus with an autoimmune blistering disease due to the autoantibodies to a component of the basement membrane zone. Most patients have been young, black women, but all ages, races, and both sexes are affected.

The clinical presentation of BSLE is generally that of an acutely generalized vesiculobullous eruption in patients who meet the American Rheumatism Association revised criteria for SLE. Lesions may involve flexural or extensor skin and mucosal surfaces of the mouth and pharynx. Blisters may form on erythematous skin and may be preceded by erythematous macules and plaques. Lesions may be large and tense and resemble those of BP or small and grouped and resemble those of DH. BSLE does not exhibit mechanical fragility of the skin with tense bullae that subsequently heal with scars and milia contrasting with EBA. Patients may complain of itching, which may sometimes be severe. However, pruritus is not prominent. Additionally, the primary lesions seen in both SLE and discoid lupus are not commonly associated with BSLE. There are conflicting reports that BSLE bullous eruptions coincide with other systemic manifestations of SLE. Certain studies report that BSLE clinical signs have no relationship with SLE features and that BSLE can be the initial manifestation of SLE.

Table 1. Differential Diagnosis of Subepidermal Bullous Eruption

<table>
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<tr>
<th>Type</th>
<th>Clinical Features</th>
<th>Histological Features</th>
<th>Direct Immunofluorescence</th>
<th>Target Antigens</th>
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<td>SLE</td>
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