

Painful and Pruritic Erosions on the Back

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A 51-year-old black woman presented to the dermatology clinic with painful and pruritic erosions on the back, abdomen, neck, and arms of approximately 2 months' duration. The lesions started on the back and spread in a cephalocaudal manner. The patient denied any new changes in medication. Physical examination revealed large erosions with mild weeping of serosanguineous fluid on the back, abdomen, neck, and upper extremities. A few tense bullae were present on the dorsal aspect of the right hand. She had experienced a similar flare approximately 1.5 years prior to the current presentation. At that time, 2 shave biopsies from vesiculobullous lesions on the right side of the neck were sent for hematoxylin and eosin staining and direct immunofluorescence. Biopsy results showed a subepidermal blister that extended along the course of the hair follicle and was associated with an infiltrate of neutrophilic granulocytes that also extended along the course of the hair follicle. Direct immunofluorescence showed IgG and C3 deposition in the basement membrane zone extending along the floor of the blister where the epidermis was separated from the dermis.

WHAT'S THE DIAGNOSIS?

- bullous pemphigoid
- bullous systemic lupus erythematosus
- dermatitis herpetiformis
- epidermolysis bullosa acquisita
- linear IgA bullous dermatosis

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THE DIAGNOSIS:

Bullous Systemic Lupus Erythematosus

Bullous systemic lupus erythematosus (BSLE) is a rare blistering disease that affects patients with systemic lupus erythematosus (SLE). Our patient had a several-year history of SLE and was being managed by a rheumatologist. She was taking hydroxychloroquine at the time of the flare. Although BSLE tends to present in those with SLE that has already been diagnosed, BSLE has been reported as a possible initial manifestation of SLE.¹

Bullous systemic lupus erythematosus is estimated to occur in less than 5% of patients with SLE and is more common in black women between the second and third decades of life,² though it also can be seen in the pediatric population.³ The lesions of BSLE usually present as subepidermal blisters often located on the face, neck, and arms on an erythematous or possibly urticarial base. Although non-BSLE vesiculobullous eruptions may be seen in patients with SLE, BSLE is differentiated from these other eruptions by its appearance on sun-exposed and non-sun-exposed areas of the body, while other vesiculobullous eruptions associated with SLE typically are limited to sun-exposed sites.⁴

Due to its clinical presentation overlapping with several vesiculobullous conditions, a set of diagnostic criteria have been suggested for BSLE, including the following: (1) fulfillment of the American Rheumatism Association's criteria for SLE⁵; (2) a new-onset vesiculobullous eruption, primarily on sun-exposed skin; (3) histology showing a subepidermal blister with a predominantly neutrophilic infiltrate; (4) presence of IgG, IgA, IgM, and C3 at the basement membrane zone; (5) evidence of antibodies to type VII collagen; and (6) immunoelectron microscopy showing codistribution of immunoglobulin deposits with anchoring fibrils/type VII collagen. To meet the diagnosis of type I BSLE, all 6 criteria must be satisfied. To meet the diagnosis of type II BSLE, only criteria 1 to 4 need to be satisfied.⁶

Patients with BSLE may be presumed to have a different but clinically similar vesiculobullous condition (eg, bullous pemphigoid, cutaneous manifestations of SLE) and may be started on systemic corticosteroids. However, BSLE patients often do not show great improvement while on corticosteroids and may even flare shortly after beginning systemic corticosteroid treatment. The current treatment of choice for BSLE is dapsone, a sulfa drug that is thought to exhibit its anti-inflammatory properties via the inhibition of the alternative pathway of the complement system and through the inhibition

of polymorphonuclear leukocyte functions.⁷ A response to dapsone helps differentiate BSLE from histopathologically and immunopathologically identical conditions such as epidermolysis bullosa acquisita.⁴ Bullous systemic lupus erythematosus can be differentiated from dermatitis herpetiformis with the presence of anti-gladiin and antitissue transglutaminase antibodies, which are found in the latter. Additionally, BSLE may show the presence of IgG and IgM deposition in addition to IgA deposition, as opposed to dermatitis herpetiformis where only IgA is found.⁸ The presence of these additional antibody depositions also help differentiate BSLE from linear IgA bullous dermatosis (LABD), as LABD will only have IgA depositions and often presents with an annular, crown of jewels–like appearance. Finally, there is a well-described phenomenon of LABD being drug induced, particularly after a course of vancomycin,⁹ and such an association with vancomycin has not been documented for BSLE.

Our patient was diagnosed with BSLE following the flare approximately 1.5 years prior to the current presentation. She had been started on dapsone 75 mg daily at that time and was taking 75 mg at the time of presentation. She was admitted and treated as an inpatient with high-dose (1 mg/kg) intravenous prednisone due to the extensive current flare.

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