A Case of Bullous Systemic Lupus Erythematosus

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Introduction: Bullous systemic lupus erythematosus (BSLE) is a rare manifestation of systemic lupus erythematosus (SLE) with an incidence of 3.4 cases per million per year. (1) It primarily affects females, and adults between 20 and 40. BSLE manifests with vesicles and bullae that affect the trunk, head, neck, arms, legs, and mucosal membranes (2). Most lesions resolve with hyper/hypo-pigmentation without scarring or milia, but milia occurs in 21% of cases and scarring occurs in 16% of cases. The criteria for diagnosis of BSLE:

1. Acute onset of vesicles and/or bullae on normal or erythematous skin

2. Histopathology of subepidermal blistering with neutrophil-dominant infiltrate in superficial dermis

3. Direct immunofluorescence of linear or granular immunoglobulin at basement membrane

4. Elevated antinuclear antibody

5. Exclusion of other causes (3)

Prior diagnosis of SLE is supportive of but not required for diagnosis of BSLE since it may be the first manifestation of SLE. First-line treatment for BSLE is dapsone which has an efficacy of 90%. Second-line treatment for patients unresponsive or who cannot tolerate dapsone include: glucocorticoids, cyclophosphamide, azathioprine, methotrexate, mycophenolate mofetil, methotrexate, and rituximab (2).

Case Presentation: A 19-year-old female with a history of systemic lupus erythematosus, lupus nephritis, anemia of chronic disease, chronic impetigo, and history of septic shock secondary to Streptococcus pyogenes two months ago presents with bilateral lower extremity swelling with bullous lesions. The patient's symptoms started one week ago and have progressively worsened. The patient reports her bullous lesions begin as fluid-filled blisters that leave scars when they reduce. The patient was on prednisone, mycophenolate mofetil, and hydroxychloroquine for systemic lupus erythematosus

prior to admission. The patient's SLE has manifested with diffuse joint pain and skin rash. The patient is positive for antinuclear antibody, anti-chromatin IgG, anti-dsDNA, anti-RNP, and anti-Smith with low complement levels. Skin punch biopsies were performed on the left anterior thigh. Histopathology shows vacuolar interface dermatitis with subepidermal splitting with lymphocyte infiltrate in superficial dermis. Direct immunofluorescence was negative for IgG, IgG4, IgM, and IgA and showed discontinuous weak granular deposits of C3 at the basement membrane and non-specific deposits of fibrinogen in connective tissue. Patient was treated with dapsone, which provided significant improvements in skin lesions.

Conclusion: Bullous systemic lupus erythematosus is a rare manifestation of SLE that should be considered as a differential diagnosis for vesiculobullous lesions in patients with SLE. BSLE must be

differentiated from other cutaneous bullous skin lesions including bullous pemphigoid, linear IgA dermatosis, pemphigoid gestationis, and epidermolysis bullosa acquisita. While scarring is not a sequela for most cases of BSLE, it should not be considered a cause for ruling out. In this case presentation, a diagnosis of BSLE was made based on patient history, clinical presentation, antibodies, and histopathology in the setting of negative direct immunofluorescence. Criteria for diagnosis of BSLE should consider reassessment for patients with this presentation.

This may be the first manifestation of SLE in patients who have not been diagnosed.

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