Bullous Systemic Lupus Erythematosus

K Parviainen, J Lesher

Citation

K Parviainen, J Lesher. *Bullous Systemic Lupus Erythematosus*. The Internet Journal of Pathology. 2002 Volume 2 Number 2.

Abstract

Bullous systemic lupus erythematosus is a distinct blistering eruption in patients with systemic lupus erythematosus. We present the case of a young, black woman whose bullous eruption led to a diagnosis of systemic lupus erythematosus and lupus nephritis. Skin biopsy showed changes suggestive of dermatitis herpetiformis and direct immunofluorescence showed immune deposits at the basement membrane zone. Bullous lesions responded to dapsone.

CASE REPORT

A 27-year-old African American female presented to the emergency department with a 3-week history of a progressive blistering eruption. She also complained of diffuse hair loss and arthralgias. Her past medical history was unremarkable but her mother had systemic lupus erythematosus. Physical examination demonstrated tense vesicles and crusts on the face, with a predilection for the perioral area (Figure 1). There were also small, tense blisters around the umbilicus (Figure 2) as well as larger blisters of the axillae (Figure 3).

A shave biopsy of a blister edge demonstrated a subepidermal blister containing fibrin, neutrophils and nuclear dust (Figure 4). Direct immunofluorescence of perilesional skin demonstrated deposits of IgG, IgM and C3 at the basement membrane zone. Indirect immunofluorescence was negative. Anti-nuclear antibody was 3+ positive. Complement components were decreased with C3 of 48 (normal 80-200 mg/dl) and C4 of 4 (normal 20-50 mg/dl). Anti-double stranded DNA antibody was elevated at 89% (normal 0-25%) and anti-Smith antibody was markedly elevated with a titer of 1:102,400. She was lymphopenic with a lymphocyte count of 790/mm3. She had proteinuria and hypoalbuminemia so a kidney biopsy was performed which showed changes consistent with class III lupus nephritis.

Treatment with high dose prednisone and pulse cyclophosphamide for lupus nephritis did not halt the blistering. Dapsone was added to her regimen, initially at 25 mg daily. Dapsone was increased to 75 mg a day over a week and the blistering resolved. Her lupus nephritis gradually responded to prednisone and cyclophosphamide, which she was maintained on. She was tapered off dapsone and remained free of blisters for five months. She then developed more extensive blistering along with a flare of her lupus nephritis. Blistering responded to dapsone 100 mg daily but her lupus nephritis has been more difficult to control.

DISCUSSION

Bullous systemic lupus erythematosus (BSLE) is a generalized blistering eruption that uncommonly occurs in patients with systemic lupus erythematosus (SLE). Pedro and Dahl[1] described the first case of BSLE in 1973. Since then, several cases have been reported with similar features.[2,3,4,5,6,7] Many cases have been in young, black women. Blisters may arise on erythematous or normal skin and are non-scarring. Lesions occur on sun-exposed or flexural skin. Skin biopsy shows subepidermal vesicles containing neutrophils with microabscesses, nuclear dust, and fibrin. Patients often respond dramatically to treatment with dapsone. Blistering often parallels flares of SLE involving other organ systems, in particular renal disease.

Camisa and Sharma[8] proposed criteria for this distinct subset of vesiculobullous skin lesions occurring in patients with SLE: (1) a diagnosis of SLE based on American Rheumatism Association criteria; (2) vesicles and bullae arising upon but not limited to sun-exposed skin; (3) histopathology compatible with dermatitis herpetiformis; (4) negative indirect immunofluorescence for circulating basement membrane zone antibodies; (5) direct immunofluorescence positive for IgG and/or IgM and often IgA at the basement membrane zone. Yell et al5 suggested

this classification be revised because of the heterogeneity of clinical and immunohistological presentation. He defined BSLE as an aquired subepidermal blistering disease in a patient with SLE, in which immune reactants are present at the basement membrane zone on direct or indirect immunoflourescence.

Patients with BSLE have demonstrated antibodies against the major anchoring fibril component type VII collagen.[9,10,11] The antigenic domains recognized by BSLE antibodies were within the noncollagenous (NC1), identical to the antigenic domains recognized by antibodies from patients with epidermolysis bullosa acquisita.[12] Although the antigen appears to be identical, BSLE lesions do not leave scars and milia as seen in epidermolysis bullosa acquisita.

Our patient demonstrates a typical case of BSLE. A young, black woman presented with blisters in sun exposed and flexural areas and responded to treatment with dapsone. The blistering eruption was the presenting sign of SLE and led to prompt diagnosis of her rather severe lupus nephritis.

Figure 1

Figure 1: Vesicles and crusting around the lips

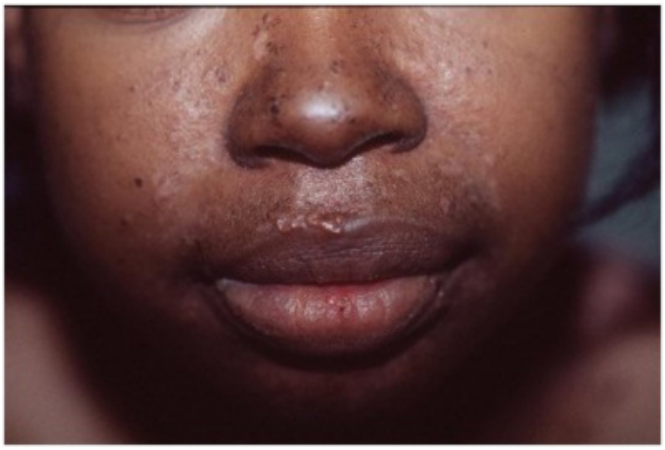


Figure 2

Figure 2: Blisters on the abdomen, concentrated around the umbilicus

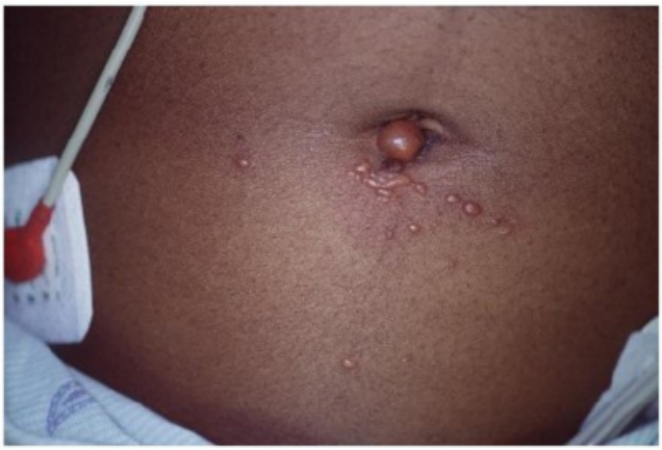


Figure 3

Figure 3: Tense bullae of the axilla

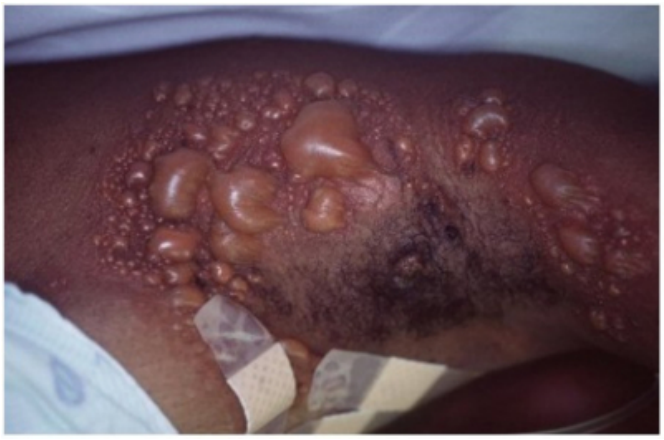
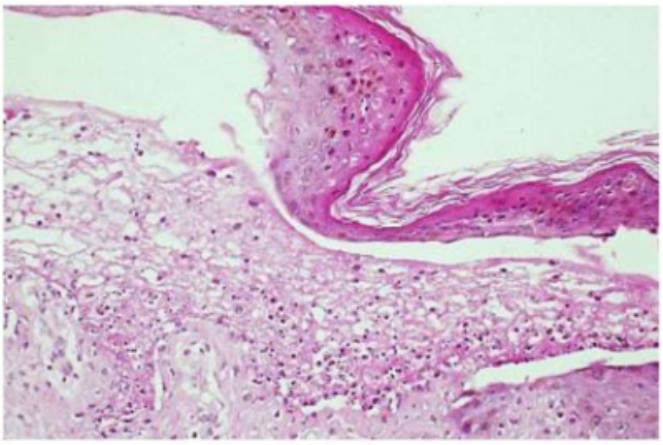


Figure 4

Figure 4: Subepidermal blister containing fibrin, neutrophils and nuclear dust



References

1. Pedro SD, Dahl MV. Direct immunoflourescence of bullous lupus erythematosus. Arch Dermatol 1973;107:118-120.
2. Burrows NP et al. Bullous eruption of systemic lupus erythematosus: a clinicopathological study of four cases. Brit J Dermatol 1993;128:332-338.
3. Camsia C. Vesiculobullous systemic lupus erythematosus. A report of four cases. J Am Acad Dermatol 1988;18:93-100.
4. Rappersberger K, Tschachler E, Tani M, Wolff K. Bullous disease in systemic lupus erythematosus. J am Acad Dermatol 1989;21:745-52.
5. Yell et al. Bullous systemic lupus erythematosus: revised criteria for diagnosis. Brit J Dermatol 1995;132:921-928.
6. Olansky AJ, Briggaman RA, Gammon WR, Kelly TF, Sams WM. Bullous systemic lupus erythematosus. J Am Acad Dermatol 1982;7:511-520.
7. Kettler AH, Bean SF, Duffy JO, Gammon WR. Systemic lupus erythematosus presenting as a bullous eruption in a child. Arch Dermatol 1988;124:1083-1087.
8. Camisa C, Sharma HM. Vesiculobullous systemic lupus erythematosus: Report of two cases and review of the literature. J Am Acad Dermatol 1983;9:924-933.
9. Gammon WR, Woodley DT, Dole KC, Briggaman RA. Evidence that anti-basement membrane zone antibodies in bullous lupus erythematosus recognize epidermolysis bullosa acquisita antigen. J Invest Dermatol 1985;84:472-476.
10. Woodley DT, Briggaman RA, O'Keefe EJ, Inman AO, Queen LL, Gammon WR. Identification of the skin basement membrane autoantigen in epidermolysis bullosa aquisita. N Engl J Med 1984;310:1007-1013.
11. Barton DD, Fine J-D, Gammon WR, Sams WM. Bullous systemic lupus erythematosus recognize epidermolysis bullosa acquisita antigen. J Am Acad Dermatol 1986;15:369-373.
12. Lapiere J-C, Woodley DT, Parente MG, et al. Epitope mapping of type VII collagen: identification of discrete peptide sequences recognized by sera from patients with acquired epidermolysis bullosa. J Clin Invest 1993;92:1831-1839.

Author Information

Karen C. Parviainen, MD
Resident, Section of Dermatology, Medical College of Georgia

Jack L. Lesher, Jr., MD.
Professor and Chief, Section of Dermatology, Medical College of Georgia