A Case of Bullous Systemic Lupus erythematosus

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Key Words

Systemic lupus erythematosus
Bullous disease
Flares

Abstract

We report a case of bullous systemic lupus erythematosus (SLE). The patient suddenly presented with a widespread blistering eruption on the trunk and neck during the course of treatment of SLE. A skin biopsy specimen showed subepidermal blisters, and direct immunofluorescence (IF) revealed deposition of IgA, IgG and IgM at the dermoepidermal junction. Indirect IF showed linear IgG deposition along the base of 1 M NaCl-split skin. Rapid resolution of the blisters occurred following treatment with dapsone 50 mg daily.

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Most patients with systemic lupus erythematosus (SLE) develop cutaneous manifestations; however, the development of a generalized bullous eruption is a rare condition in SLE [1, 2]. There have been several case reports of established SLE associated with bullous eruptions. These reports included bullous pemphigoid with SLE [3, 4], dermatitis herpetiformis with SLE [5, 6] and an unspecified bullous eruption in SLE [7, 8]. Bullous eruption of SLE or bullous SLE was identified as a distinct clinicopathologic entity by Hall et al. in 1982 [8]. Herein we report an additional patient with bullous SLE; the occurrence of bullae in SLE patients may be an important sign of flare of SLE.

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Case Report

A 9-year-old Japanese female initially developed polyarthritis of the small joints in her hands. On the basis of several examinations, a diagnosis of SLE was made in February 1991. She had been treated with oral prednisolone. In November 1991, she suddenly presented with a widespread blistering eruption on the trunk and neck (fig. 1). A skin biopsy specimen showed a subepidermal blister (fig. 2), and direct immunofluorescence (IF) revealed deposition of IgA, IgG and IgM at the dermoepidermal junction. Indirect IF showed linear IgG deposition along the base of 1 M NaCl-split skin. The serum level of IgG was elevated, the titer of antinuclear antibody was higher than before and hemolytic activity of the complement system was lower (Fig. 1). Blistering eruption on the trunk.

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than the normal level. In spite of the treatment with a high dose (60 mg daily) of prednisolone, she continued to develop blisters. Rapid resolution of the blisters occurred following treatment with dapsone 50 mg daily.

Discussion

Bullous skin lesions are an uncommon finding in patients with SLE. Camisa and Sharma [9] proposed criteria for the diagnosis of bullous SLE, as follows: (a) a diagnosis of SLE by criteria of the American Rheumatism Association, (b) a widespread, non-scarring, vesiculobullous eruption, (c) a subepidermal blister with dermal inflammation characterized by neutrophilic papillary microabscesses like those seen in dermatitis herpetiformis, (d) negative indirect IF for circulating basement membrane zone antibodies (later, they suggested that the criteria be modified to include: negative or positive indirect IF for circulating basement membrane zone antibodies using separated human skin as substrate), (e) direct IF of lesional and non-lesional skin always reveals linear or granular IgG and/or IgM and often basement membrane zone antibodies. Our patient satisfied most of these criteria. Bullous SLE may be confused with bullous pemphigoid, dermatitis herpetiformis or linear IgA dermatitis. Direct IF may be helpful to exclude dermatitis herpetiformis and linear IgA dermatitis. Direct IF of this case showed no granular IgA deposition and linear IgA deposits concomitant with IgG and IgM at the dermoepidermal junction. This finding is in favor of bullous SLE compared with dermatitis herpetiformis and linear IgA dermatitis. By using 1 M NaCl-split skin as a substrate for indirect IF, it is possible to distinguish

Fig. 2. Skin biopsy showing the subepidermal blister.

between bullous pemphigoid and bullous SLE. Bullous pemphigoid antibody binds the roof of split skin, whereas binding is observed at the base of the split skin in bullous SLE [10]. A blistering eruption occurs during flares of SLE, and especially it may be correlated with renal disease [11, 12]. At the time of occurrence of bullae in this patient, general symptoms and the

results of blood examination were under poor control because of insufficient dosage of prednisolone. Bullous eruption may be an important sign of flares in SLE.

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