Dear Editor,

Bullous systemic lupus erythematosus (BSLE) is an uncommon blistering eruption that can occur in patients with systemic lupus erythematosus (SLE). Although 59 to 85% of SLE patients will have skin manifestations, less than 5% will develop bullous disease, which is even more uncommon (1%) in the pediatric population.1–3

Herein, we report a 16-year-old female, with a recent diagnosis of autoimmune urticaria, characterized by relapsing edema and maculopapular pruritic lesions on the face, lips, abdomen and thighs, refractory to oral prednisolone 40mg/d. Laboratory evaluation showed leukocyte count of 3.300 x 10^9/L, erythrocyte sedimentation rate of 33 mm/hr, normal C3 level, C4 level and C1-inhibitor activity, and positive anti-nuclear (1/640), anti-dsDNA and anti-SSA antibodies. She was referred to our department for further evaluation. Two months later, she developed bilateral knee pain, without swollen and new itchy lesions appeared after sun exposure (Figure 1A, B) despite medication with deflazacort 15 mg/d and hydroxychloroquine 400mg/d. Laboratory evaluation revealed hypocomplementemia. Steroid dose was increased (deflazacort 1 mg/kg/d) and cyclosporine 1.5 mg/kg/d and ebastine 20mg/d were added with transient improvement. Skin lesions became larger and blistered particularly on the limbs. (Figure 1C, D). A skin biopsy revealed sub-epidermal blister full of neutrophils and dermal infiltrate of lymphocytes and neutrophils. Direct immunofluorescence showed linear deposition of IgG, C3 and fibrin at dermo-epidermal junction, compatible with BSLE. Cyclosporine was stopped and dapsone 25 mg/d started, with complete resolution of the skin lesions. Steroids were tapered and stopped and after 2 months. She remains asymptomatic on dapsone 25 mg every other day plus hydroxychloroquine 200 mg/d.

BSLE is characterized by a rapid widespread development of non-cicatricial tense vesicles and bullae over erythematous macules or plaques, usually on photo-exposed areas. There is a predilection for the trunk, upper extremities, supraclavicular region and face. In 31% of cases, urticarial lesions or erythematous plaques are associated, and mucous membrane involvement was seen in 51%.1,4,6

Histology is characterized by subepidermal blister, with predominantly neutrophilic dermal infiltrate and occasional eosinophils. Linear deposition of IgG, IgA, C3, and C1q along the basement membrane zone can be seen on direct immunofluorescence examination.1,4,7

Diagnostic criteria were initially proposed by Camisa and Sharmaa and were revised by Gammon and Briggmanb to include the presence of circulating antibodies to type VII collagen, which is the major component of anchoring fibrils at the dermal-epidermal junction. Therefore, the loss of integrity of the dermal-epidermal junction may cause blistersc.

Differential diagnosis includes bullous pemphigoid, epidermolysis bullosa acquista, IgA bullous dermatosis and dermatitis herpetiformisd1,2.

The activity of the bullous disease can occur isolated or concomitantly with other systemic manifestations of SLE. According to De Risi-Pugliese et al.6, extra-cutaneous SLE manifestations occur in 90% of patients, especially nephritis (50%) and neuropsychiatric manifestations (12%). In our case, arthralgia, raised levels of anti-dsDNA, hypocomplementemia and leukopenia were seen, indicating active SLE.

Dapsone is the treatment of choice for BSLE. Skin lesions may respond even to low dose (25-50 mg/d).3,7 Other treatments for BSLE include corticosteroids, antimalarials and immunosuppressive agents, especially in systemic cases.5,7 Recently, the use of rituximab has been shown to be effective in refractory cases.5 In our patient, the BSLE lesions were resistant to steroids and cyclosporine, and responded dramatically to a low dose of dapsone.

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1 Serviço de Reumatologia, Hospital Garcia de Orta

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Bullous skin lesions can rarely represent the first clinical presentation of pediatric SLE, thus skin biopsy is of extreme relevance for a correct diagnosis of BSLE, proper treatment and to prevent further complications of SLE.

**REFERENCES**


**CORRESPONDENCE TO**

Sandra Sousa  
Serviço de Reumatologia, Hospital Garcia de Orta  
Alameda Dr. Torrado da Silva  
2800 Almada, Portugal  
E-mail: sandrainssousa@gmail.com