Juvenile bullous systemic lupus erythematosus – case report

Sousa S[1], Santos MJ[1]

ACTA REUMATOL PORT. 2020;45:74-75

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.

1 Serviço de Reumatologia, Hospital Garcia de Orta

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 lgG, lgA, C3, and C1q along the basement membrane zone can be seen on direct immunofluorescence examination[2,4,7].

Diagnostic criteria were initially proposed by Camisa and Sharma[8] and were revised by Gammon and Briggaman[9] 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 blisters[4].

Differential diagnosis includes bullous pemphigoid, epidermolysis bullosa acquisita, IgA bullous dermatosis and dermatitis herpetiformis[1,2].

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)[3,5,7]. Other treatments for BSLE include corticosteroids, antimalarials and immunosuppressive agents, especially in systemic cases[4,7]. Recently, the use of rituximab has been shown to be effective in refractory cases[2]. In our patient, the BSLE lesions were resistant to steroids and cyclosporine, and responded dramatically to a low dose of dapsone.

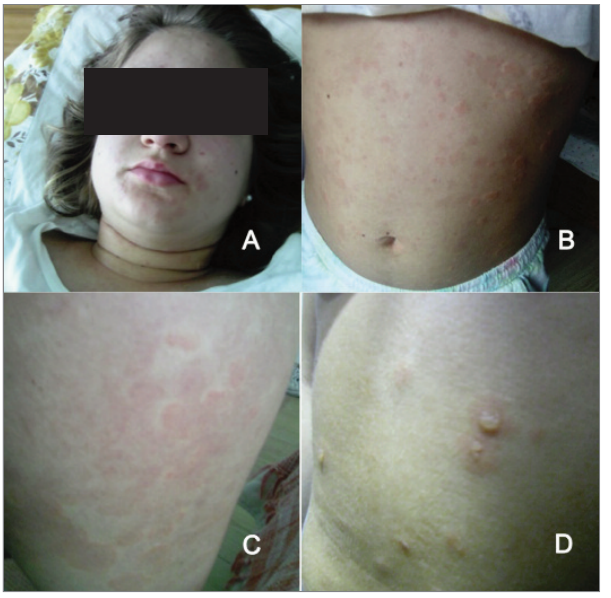

FIGURE 1. A) B) and C) Coalescing erythematous plaques and papules on face, abdomen and left leg; D) Tense bullae overlying an erythematous base on the knee

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.

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

REFERENCES

1. Padrão EMH, Teixeira LF, Maruta CW, et al. Bullous systemic lupus erythematosus – a case report. Autops Case Rep. 2019;9(1):e2018069.
2. Contestable JJ, Edhegard KD, Meyerle JH. Bullous Systemic Lupus Erythematosus: A Review and Update to Diagnosis and Treatment. Am J Clin Dermatol. 2014;15(6):517-24.
3. Liu KL, Shen JL, Yang CS, Chen YJ. Bullous Systemic Lupus Erythematosus in a Child Responding to Dapsone. Pediatr Dermatol. 2014;31(4):104-6
4. Lourenço DM, Gomes RC, Aikawa NE, Campos LM, Romiti R, Silva CA. Childhood-onset bullous systemic lupus erythematosus. Lupus. 2014;23(13):1422-5.
5. Hans-Bittner NR, Bittner G, Filho GH, Takita LC. Bullous systemic lupus erythematosus in a 10-year-old child. An Bras Dermatol. 2017; 92(5 Suppl 1): 37–39.
6. De Risi-Pugliese T, Cohen Aubart F, Haroche J, et al. Clinical, histological, immunological presentations and outcomes of bullous systemic lupus erythematosus: 10 New cases and a literature review of 118 cases. Semin Arthritis Rheum. 2018;48(1):83-89.
7. Duan L, Chen L, Zhong S, et al. Treatment of Bullous Systemic Lupus Erythematosus. J Immunol Res. 2015;2015:167064.
8. Camisa C, Sharma HM. Vesiculobullous systemic lupus erythematosus: Report of two cases and a review of the literature. 1983; 9(6):924—933
9. Gammon WR, Briggaman RA. Epidermolysis bullosa acquisita and bullous systemic lupus erythematosus: disease of autoimmunity to type VII collagen. Dermatologic Clinics. 1993;11(3):535–547