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Case Report

Clinical presentation of bullous systemic lupus erythematosus in a pediatric patient: a case report

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ABSTRACT

Bullous systemic lupus erythematosus (BSLE) is a rare blistering eruption that can occur in patients with systemic lupus erythematosus. Although 59 to 85% of patients will have skin manifestations, less than 5% will develop the bullous disease, which is even rarer (1%) in the pediatric population. We present the case of a 16-year-old female patient in whom a diagnosis of BSLE was made through clinical presentation, histological study by biopsy, and direct immunofluorescence. Due to the low incidence of cases reported in pediatric patients, it is possible that the diagnosis may not be made immediately, which is why we emphasize the importance of obtaining an accurate diagnosis and providing timely treatment.

Keywords: Dermatology, Dermatosis, Skin diseases

INTRODUCTION

Bullous systemic lupus erythematosus (BSLE) can describe as a type III autoimmune disease that presents its pathogenesis due to autoantibodies against collagen VII located in the basement membrane. The recruitment of neutrophils secondary to the presence of these autoantibodies explains the proteolysis and cell separation characteristic of this pathology. Systemic lupus erythematosus has different variables, the one reported in this case presents a low frequency within the pediatric population.[1] It has an acute clinical presentation characterized by tense vesicles or bullae on erythematous skin and/or mucous membranes. Histologically BSLE is characterized by a subepithelial detachment accompanied by a superior dermal neutrophilic infiltrate and papillary microabscesses.[2]

CASE REPORT

A 16-year-old female was hospitalized due to painful and pruritic vesicles and bullae that displayed sudden and rapid growth on the oral mucosa, external genital organs, thighs, and legs over the last 3 weeks. She also presented headache, fever, weight loss and myalgia over the last 2 months. The clinical examination revealed a prostrated underweight patient, with hypotension of 90/60 mmHg.

Skin findings showed post-inflammatory macules along with multiple small tense vesicles and bullae varying from 1 mm - 1 cm over slightly erythematous skin, that was predominant in the neck, trunk, and extremities including palms. Oral mucosa showed two erosive lesions over the soft palate and multiple erosions were present in the vaginal mucosa. No lymphadenopathy and signs of

arthritis were detected; the remaining physical examination revealed no alterations.

Findings from laboratory studies showed a leukocyte count of 7.630×109 /L, erythrocyte sedimentation rate of 70 mm/hr, Urinalysis revealed the presence of proteinuria 2+100 mg/dl, and leukocyturia, with no casts. Renal function was preserved, 24-hour urine protein of 1.34 g. Ac IgM herpes II 2.20, IgG herpes II 0.20, IgG/IgM herpes I negative.

Additional studies were notable for low complement levels, C3=49 mg/dL and C4=7 mg/dL; antinuclear antibody was positive (1:2/560) with a fine speckled pattern, anti-double-stranded DNA and anti-histone were positive. Pregnancy test and lumbar puncture culture were negative.

Intravenous antibiotics and steroids were administered initially, but the patient showed no improvement during hospitalization. She displayed further extensive lesions to the trunk, axillae, and vulva as well as disruption of the bullous lesions, which remained as hyperemic scars.

Two 4 mm punch biopsies were performed, histological findings showed focal necrosis, subepidermal blister full of neutrophils, and dermal infiltrate of lymphocytes and neutrophils. Direct immunofluorescence for IgA, IgM, C3c, C1q, Kappa, and lambda showed a granular pattern with pseudo linear deposition in the basement membrane zone, and IgG with a perivascular granular pattern with pseudo linear deposition.

Clinical and laboratory findings confirmed the diagnosis of BSLE. As an initial treatment, we administered prednisone (40 mg/day) and hydroxychloroquine (200 mg/day). Although this resulted in an improvement of most lesions, some of the plaques on the trunk and arms persisted as well as psychiatric manifestations, therefore we decided to add six doses of intravenous cyclophosphamide (500 mg each), achieving symptomatic improvement after 2 weeks.

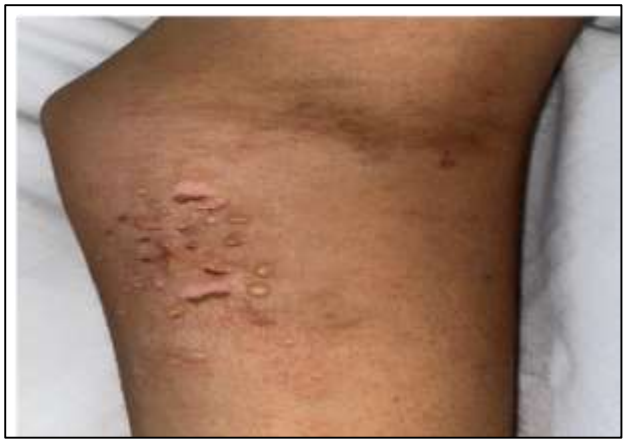

Figure 1: Post-inflammatory macules, multiple small tense vesicles and bullae.

Figure 2: Tense vesicles and bullae.

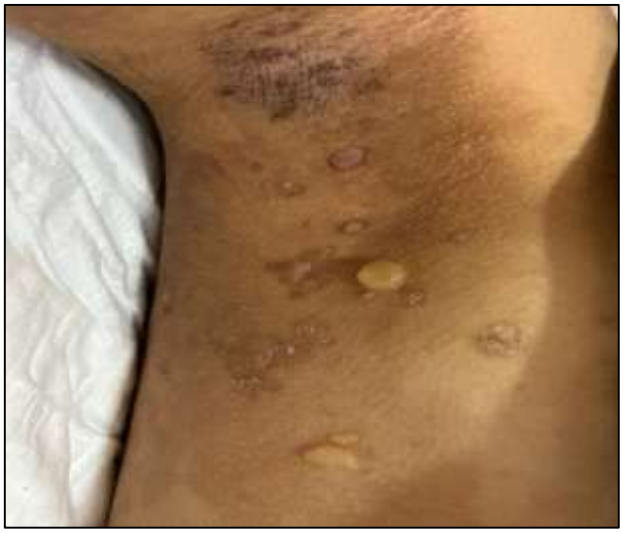

Figure 3. Multiple erosions.

DISCUSSION

BSLE is a rare disorder for which epidemiologic data are also limited.[1] The estimated incidence is 0.2 to 0.05 cases per million inhabitants/year. The reported incidence of BSLE is approximately 2-3% of all subepidermal autoimmune bullous skin diseases.[3] Affects less than 5% of patients with systemic lupus erythematosus.[4] The prevalence of BSLE in the childhood-onset lupus population estimates to be lower than 1%.[1]

Is more frequent in females than in males, by the epidemiology of systemic lupus erythematosus. BSLE usually occurs in adults in the age range between 20 and 40 years, but also can affect older adults or children like in the case of the patient whose presentation occurred at the age of 16 years, with an atypical form of presentation.[5] It has been reported predominance of African descent in most of the cases, discording with this case. However, it can be presented in all races, ages, and gender.[6]

The clinical presentation, in this case, is interesting since the dermatological lesions could be misdiagnosed with other blistering pathologies, however, the systemic manifestation that the patient presented was key to guiding the diagnosis.

Regarding systemic alterations, it should be noted that the patient did not present renal involvement (lupus nephritis), which is the most frequent systemic association, reported in case series in up to 50%.[7]

Histologically, BSLE is characterized by a subepithelial detachment accompanied by a superior dermal neutrophilic infiltrate and papillary micro-abscesses, as presented in this case.[2] Due to the biopsy taken from the patient, it was possible to reach an accurate diagnosis and provide the appropriate treatment, so we emphasize this methodology to achieve better management. Likewise, in the direct immunofluorescence a linear deposition of IgG was found in the area of the basal membrane that was useful to differentiate the diagnosis from DH and LABD.

The therapeutic response to dapsone is satisfactory, therefore is considered the treatment of choice with rapid cessation and healing of the lesions in a course of days to weeks.[7] However in this case due to the systemic manifestations, dapsone was not initially prescribed and lesions appeared despite high doses of corticosteroids and antimalarial drugs (chloroquine and hydroxychloroquine) which supports the hypothesis of the relative resistance reported in the literature.[8]

CONCLUSION

Due to the low incidence of cases reported in pediatric patients, it is possible that the diagnosis may not be made immediately, which is why we emphasize the importance of obtaining an accurate diagnosis and providing timely treatment.

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