

Received: 2016.11.13
Accepted: 2016.12.27
Published: 2017.03.29

ISSN 1941-5923
© Am J Case Rep, 2017; 18: 317-319
DOI: 10.12659/AJCR.902351

Systemic Lupus Erythematosus and Bullous Pemphigoid with Dramatic Response to Dapsone

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 11
Final Diagnosis: Bullous pemphigoid in systemic lupus erythematosus
Symptoms: Bullous lupus • photosensitive rash • synovitis
Medication: —
Clinical Procedure: Pharmacological treatment
Specialty: Rheumatology

Objective: Unusual clinical course
Background: Bullous pemphigoid is an autoimmune blistering disease, with relapses, isolated or associated with other autoimmune diseases such as systemic lupus erythematosus (SLE). Joint manifestations rapidly respond to small or moderate doses of corticosteroids, whereas skin manifestations usually respond to antimalarial drugs.
Case Report: We describe the clinical case of an 11-year-old girl with SLE. She showed bullous skin lesions with arthralgia, mild proteinuria, resolved after steroid treatment. At the tapering of her prednisone dose, the patient had new skin lesions requiring an increased dose of prednisone. She started dapsone at the dosage of 1 mg/kg/day, maintaining low dose prednisone; this treatment was successfully followed by the dramatic disappearance of skin lesions and limb pain.
Conclusions: Bullous skin lesions can represent the first clinical presentation of pediatric SLE and could influence the treatment and the outcome of these patients. This case showed an atypical course as both skin manifestations and arthritis promptly and persistently resolved with dapsone without the use of high-dose glucocorticoids. Only a few cases of patients with SLE associated with bullous pemphigoid have been reported in the literature, and very few in the pediatric population.

MeSH Keywords: Dapsone • Lupus Erythematosus, Systemic • Pemphigoid, Bullous

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/902351>



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Background

Bullous pemphigoid (BP) is an autoimmune blistering disease. Isolated BP exhibits a relatively benign prognosis with relapses, but may be associated with other autoimmune diseases in the context of complex clinical syndromes [1,2].

Skin and joint involvement are frequent manifestations of systemic lupus erythematosus (SLE). There are three forms of cutaneous SLE: chronic cutaneous (discoid) lupus, subacute cutaneous lupus, and acute cutaneous lupus. Joint involvement is typically not erosive. Anti-Ro is closely associated with a photosensitive rash and with subacute lupus. Joint manifestations rapidly respond to small or moderate doses of corticosteroids [3], whereas skin manifestations usually respond to antimalarial drugs [2,4,5].

Only few cases of patients with SLE associated with BP have been reported in the literature [6,7], and very few in the pediatric population [8–15].

Case Report

We describe the case of an 11-year-old female with SLE. She had a history of periodic fevers, recurrent joint swelling and tenderness (knees and ankles), back pain, abdominal pain, and a butterfly rash. She developed mild proteinuria, which was successfully treated with steroids. Since the age of 10 years, she had developed a vesiculobullous eruption with clinical and histological features of BP, with typical skin features (Figure 1A). Biopsy of a bulla revealed subepidermal blister, and perivascular neutrophils and eosinophil infiltration. She showed a persistent high titer of anti-dsDNA antibodies (345 IU/mL by ELISA, $\text{nv} < 100$), and anti-nuclear antibodies (1: 160 by IFI). Her ENA (anti-Ro, LA, RNP), anti-desmoglein 1, 3 antibodies, anti-BP 180, anti-BP 230, anti-skin-AMB, and anti-skin-ASI antibodies were negative. The genetic study of auto-inflammatory syndromes (FMF, TRAPS; MVK, NLRP3, NLRP12) was negative. For

the skin lesions relapses, she received corticosteroids (prednisone 1 mg/kg/day) resulting in the remission of the clinical manifestations (Figure 1B) and the normalization of serum amyloid A (SAA), erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP). At the tapering of prednisone dose, the patient had new skin lesions requiring an increased dose. She showed striae rubrae, hypertrichosis, and growth velocity reduction. Due to the frequent relapses, she started oral administration of dapsone at the dosage of 1 mg/kg/day, which resulted in dramatic disappearance of the lesions and allowed gradual dose tapering of prednisone down to 2.5 mg/day. She subsequently showed an increase of growth velocity, a pubertal onset, and menarche, which occurred seven months after the start of dapsone. During the first 18 months of follow-up she manifested four relapses of the skin lesions associated with knee and back pain. However dermatological lesions after dapsone treatment were mild, and promptly resolved with short courses of low-dose oral prednisone.

Discussion

We described an unusual presentation of pediatric SLE with bullous skin lesions. The association between SLE and pemphigoid is uncommon in the pediatric population, with only 10 cases reported in the literature [8–15]. Moreover, while cutaneous lupus responds to hydroxychloroquine, a satisfactory outcome with dapsone treatment has only sporadically been reported [4,5]. In the described case, dapsone treatment showed efficacy on skin lesions and the treatment was followed also by the complete remission of the other manifestations of the disease, without the employment of other immunosuppressive drugs [16].

Conclusions

In our patient, this therapy allowed a good control of clinical outcome with a low dose of steroids in a phase of her life characterized by growth spurt and pubertal development.

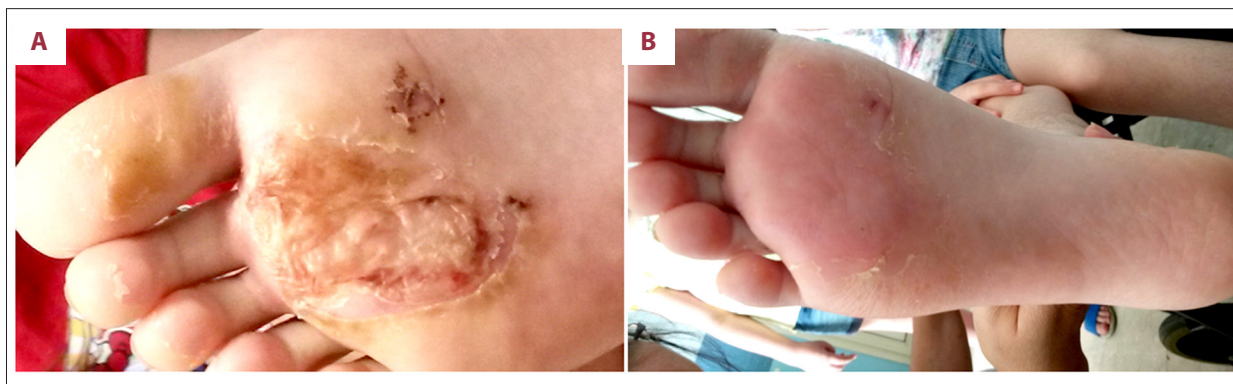


Figure 1. (A) Severe skin lesions before dapsone treatment. (B) Skin lesions after start of steroids treatment (prednisone 1 mg/kg/day).

For patients at the adolescent age, in fact, steroid doses should be low and only given for brief periods. This care prevents delay in bone maturation and impaired pubertal development.

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Conflicts of interest

Authors have no conflicts of interest.