DERMATOLOGICA SINICA 34 (2016) 92–95

Contents lists available at ScienceDirect

Dermatologica Sinica

journal homepage: http://www.derm-sinica.com

CASE REPORT

Two cases of bullous systemic lupus erythematosus treated successfully with T2 and low-dose corticosteroids

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ARTICLE INFO

Article history:
Received: May 13, 2015
Revised: Jul 29, 2015
Accepted: Aug 14, 2015

Keywords:
bullous lupus erythema
steroid
treatment
Tripterygium wilfordii

ABSTRACT

Two female patients presented with bullous cutaneous lupus on sun-exposed areas and synovitis, and tested positive for antinuclear antibodies, anti-Smith (anti-Sm), anti-dsDNA, and low complement levels. Case 2 also suffered oral ulcers. Thus, diagnoses of bullous systemic lupus erythematosus (BSLE) were made. Both patients responded distinctly to *Tripterygium wilfordii* polyglycosidium (T2; 60 mg/day) combined with oral prednisone (30–40 mg/day), which showed efficacy in 1–2 weeks and produced no side effects. Therefore, T2 combined with low-dose corticosteroids may be an effective and highly tolerable alternative for treating BSLE.

Introduction

Tripterygium wilfordii polyglycosidium (T2) is a traditional Chinese medicine widely used in China in organ transplantation and for the treatment of inflammatory and autoimmune diseases, neurodegenerative diseases, and some tumors. Bullous systemic lupus erythematosus (BSLE) is a rare and severe category of systemic lupus erythematosus (SLE) associated with relative treatment difficulty. Here we report two female BSLE patients treated effectively with T2 and a low dose of corticosteroids.

Case reports

Case 1

A 41-year-old female farmer presented with symmetric bullous and erythematous eruptions on sun-exposed areas. About 6 months prior, light-sensitive and symmetric violaceous erythema had occurred on her face and the dorsum of her hands with no obvious predisposing causes. Facial dermatitis was diagnosed in a local clinic, and intravenous dexamethasone (5 mg/day) was prescribed for a week. As a result, her lesions disappeared, and the treatment was discontinued. However, 3 months later, because she had been working outside for long hours each day, aggravated light-sensitive and symmetric violaceous erythema reoccurred, with several vesicles over it. Arthralgia involving the small joints of the hands, bilateral wrists, and knees was observed, and the symptoms worsened in cold temperatures. Mouth or eye dryness, fever, mucosal ulcer, or Raynaud's phenomenon were not found.

On examination, axillary temperature was 37°C. Diffuse inflammatory violaceous erythema was observed on her face and the dorsum of her hands. Multiple tense, clear vesicles appeared on the upper part of her cheeks and nose, over the erythematous base. Nikolsky's sign was negative. In addition, superficial crusting, postinflammatory hyperpigmentation, and small superficial scars on the dorsum of her hands were noted (Figures 1A and 1B). Tenderness on the small joints of the hands, bilateral wrists, and knees was positive. No further abnormal systemic examination was conducted.

Routine blood and urine tests were normal. Autoimmunity tests were positive for antinuclear antibodies (ANA; 1:640; using HEp-2 cells); anti-dsDNA, anti-Smith (anti-Sm), anti-Sjögren's syndrome-related antigen A (anti-SSA), and anti-SSB; and low levels of C3 [0.56 g/L (0.78–2.1 g/L)], C4 [0.10 g/L (0.17–0.4 g/L)], and hemolytic complement [CH50; 70 g/L (90–180 g/L)]. They were negative for rheumatoid factors and antiphospholipid antibodies. The

Conflicts of interest: The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in this article.

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http://dx.doi.org/10.1016/j.dsi.2015.08.004

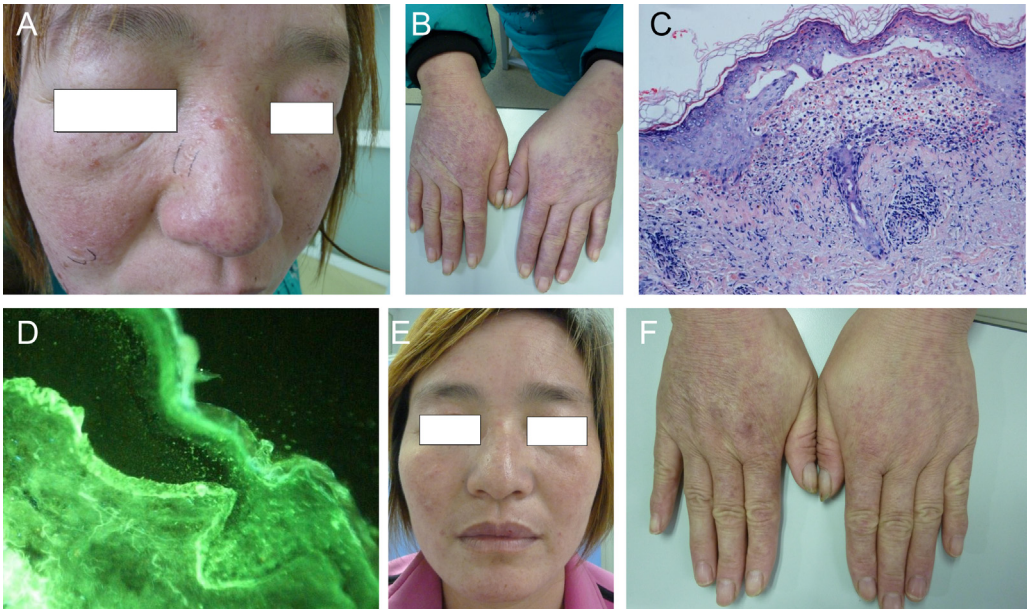

Figure 1 Patient 1: (A, B) A 41-year-old female with violaceous erythema diffused on her face and the dorsum of her hands. Multiple tense and clear vesicles dispersed on the upper part of her cheeks and nose. Nikolsky's sign was negative. (C) Histopathology of the facial vesicles revealed a subepidermal split, liquefaction of basal cells, a neutrophilic microabscess in the dermal papillae, and mild infiltration of neutrophils and histocytes around vessels and cutaneous appendages (hematoxylin–eosin; original magnification ×100). (D) Direct immunofluorescence revealed granular immunoglobulin G deposition along the basement membrane zone (original magnification ×400). (E, F) After the combined treatment of *Tripterygium wilfordii* polyglycosidium and low-dose prednisone for 6 months, hyperpigmentation was left on her face and the dorsum of her hands.

erythrocyte sedimentation rate (ESR) was 40 mm/hour (0–5 mm/hour). A biopsy of the facial vesicles revealed a subepidermal split, liquefaction of basal cells, a neutrophilic microabscess in the dermal papillae, and mild infiltration of neutrophils and histocytes around vessels and cutaneous appendages (Figure 1C). Direct immunofluorescence (DIF) revealed immunoglobulin G and C3 deposition along the basement membrane zone (Figure 1D).

A diagnosis of BSLE was considered. Resting, keeping warm, and avoiding sunshine were advised. Oral prednisone (30 mg/day), T2 (60 mg/day), and calcium supplements were prescribed. Two weeks into the treatment, the inflammatory violaceous erythema on sun-exposed areas darkened, and arthralgia dissipated. The skin eruptions improved and arthralgia of all involved joints disappeared within 2 months. Oral prednisone and T2 were reduced to 20 mg/day and 40 mg/day, respectively. By the 4th month, her skin eruptions had almost vanished (Figures 1E and 1F), and arthralgia did not relapse. T2 was discontinued, and prednisone was gradually reduced to 10 mg/day. Serological ANA decreased to 1:80, and C3, C4, and CH50 recovered, although anti-dsDNA, anti-Sm, anti-SSA, and anti-SSB were still positive. No side effects were reported.

Case 2

A 42-year-old female farmer presented with violaceous erythema and vesicles on sun-exposed areas. Like Case 1, she had erythematous and vesicular rashes on her face, neck, and extremities for about 2 weeks because of much sun-exposed farming. She also complained of fever, fatigue, pruritus, painful oral ulcer, and arthralgia involving the knees and ankles. Other systemic symptoms were not found.

On examination, axillary temperature was 37.8°C. Sporadic violaceous vesiculopapules, erythema, discoid plaques, and tense bullae based on the erythema or plaques were seen on her face, neck, and distal extremities. Some vesicles were ruptured and crusted. Nikolsky's sign was negative (Figures 2A and 2B). Oral ulcers could be seen, while other mucosae were unaffected. Tenderness on the bilateral knees and ankles was found, but additional abnormal systemic problems were not.

The blood cell count was normal. Urine protein was 2+. The ESR was 60 mm/hour. Autoimmunity indices were positive for ANA (1:1280); anti-dsDNA and anti-Sm; and low levels of C3 (0.46 g/L), C4 (0.12 g/L), and CH50 (50 g/L) were noted. A biopsy of a vesicle on the left arm and DIF showed similar results to those of Case 1. A renal biopsy was rejected.

BSLE was diagnosed. Oral prednisone (40 mg/day), T2 (60 mg/day), calcium supplements, and ethacridine lactate paste for the eruptions were prescribed. After 1 week, the vesicles on sun-exposed areas decreased, and arthralgia began to dissipate. Urine protein returned to normal. Oral prednisone was then reduced to 30 mg/day. By the end of the 1st month, the skin eruptions and arthralgia of all involved joints greatly improved. The drugs were further downregulated—prednisone to 20 mg/day and T2 to 40 mg/day. The skin eruptions and arthralgia resolved. After 2 months, T2 was discontinued, and prednisone was gradually decreased to 10 mg/day. By the 6th month, hyperpigmentation was left on the original eruptions (Figures 2C and 2D), and no more arthralgia was found. Urine protein was negative and ANA 1:40, C3, C4, and CH50 returned to normal, but anti-dsDNA, anti-Sm, anti-SSA, and anti-SSB were still positive. No side effects were reported.

Discussion

BSLE is a form of subepidermal autoimmune bullous dermatosis, a rare entity that accounts for less than 5% of SLE. Females in their 20s to 40s have high chances of being diagnosed with BSLE.[1] Etiology is unclear. Genetics and autoimmunity for VII collagen in the basement membrane zone may take part in the separation of the epidermis and dermis. The most common presentation of BSLE is asymptomatic tense blisters confined to sun-exposed areas or sometimes present in a more widespread distribution. Multisystem