A Case of Rowell Syndrome: Excellent Response to Oral Cyclosporine

Dear Editor,

Lupus erythematosus is a multisystem disease which frequently involves the skin. There are several variants of cutaneous lupus, which are defined and classified by the location and the depth of the inflammatory infiltrate, adnexal involvement, presence or absence of interphase dermatitis, and chronology (1). The most common clinical subtypes are acute, subacute and chronic cutaneous lupus erythematosus; however, other rare specific and non-specific cutaneous involvements also exist (2). Rowell syndrome is one of these rare specific variants and was originally described as the association of lupus erythematosus, erythema multiforme-like lesions without any known precipitating factors, and immunological abnormalities such as a speckled pattern of antinuclear antibody (ANA) staining, positive Anti-La antibody, and reactive rheumatoid factor (3). Subsequently, in order to enhance diagnostic specificity, the criteria were redefined as major (lupus erythematosus, erythema multiforme-like lesions, speckled pattern of ANA staining) and minor (chilblains, positive Anti-La or Anti-Ro antibodies, reactive rheumatoid factor); patients should present all three major criteria plus at least one minor criterion to be diagnosed with Rowell syndrome (4). First line treatment options for cutaneous lupus as well for Rowell syndrome comprise topical corticosteroids and calcineurin inhibitors, systemic anti-malarial therapy, and systemic corticosteroids (for active disease). In anti-malarial resistant disease, retinoids, dapsone, methotrexate, and other systemic immunosuppressive agents can be considered,

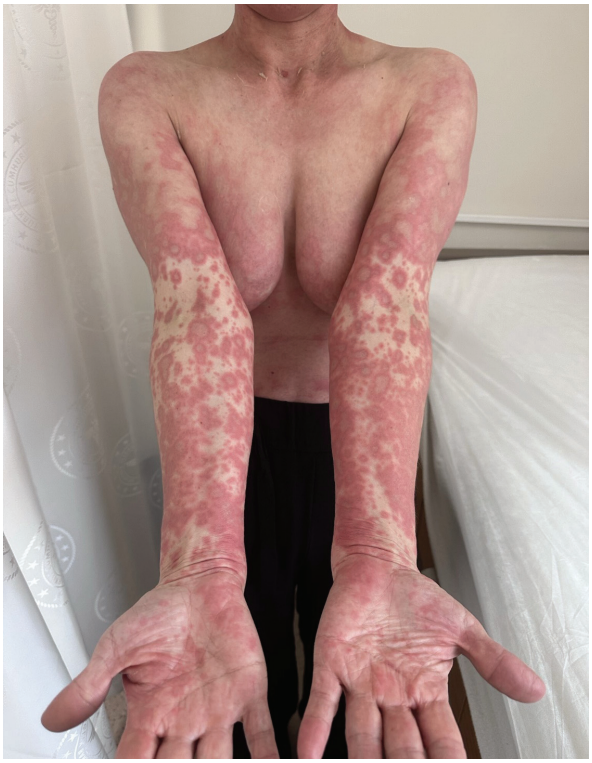

Figure 1. Violaceous-dark erythematous plaques with a prominent arcuate/targetoid shape at the periphery.

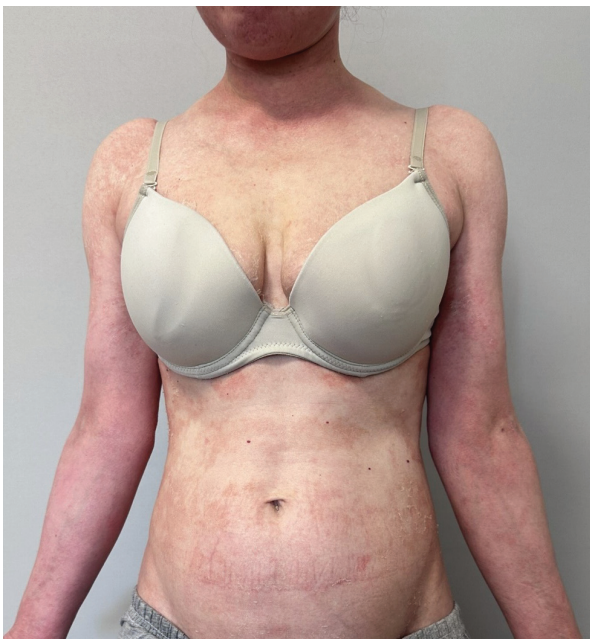

Figure 2. Complete clinical response at the fourth week of cyclosporine treatment.

though with a lower level of evidence (5). Herein, we present the case of a patient with Rowell syndrome with a therapeutic approach that is rarely included in the literature. Informed consent was obtained and signed from the patient regarding the use of the patient's information for the purposes of writing a case report publication.

A 38-year-old woman who had been examined by the Rheumatology Department for connective tissue disease (CTD) because of her morning stiffness and peripheral arthritis was referred to us for consultation due to the new onset of a mild, itchy rash. The patient's lesions first appeared on her face, neck and upper trunk, subsequently becoming generalized. There was no previous history of recent infection or medication. The patient underwent follow-up under hydroxychloroquine therapy (400 mg/day) for CTD for 2 months. Dermatological physical examination showed violaceous-dark erythematous plaques with a prominent arcuate/targetoid shape at the periphery were present on her whole body, with oral mucosal erosions (Figure 1). In previous laboratory studies, ANA positivity with a speckled pattern and Anti-Ro positivity were observed. Rheumatoid factor was non-reactive. A punch biopsy was performed. Histopathological examination showed prominent interface dermatitis with basal vacuolar degeneration and apoptotic keratinocytes, which corresponds to subacute cutaneous lupus erythematosus. Due to the presence of three major criteria and one minor criterion, the patient was diagnosed with Rowell syndrome. She was given additional treatment comprising 1 mg/kg/day oral methylprednisolone. However cutaneous involvement progressed rapidly, and the methylprednisolone dose was increased to 500 mg/day intravenously for three consecutive days as pulse steroid therapy. Thereafter, upon clinical irresponsiveness, oral cyclosporine was started at 3.5 mg/kg/day, and systemic corticosteroid therapy was gradually ceased. An excellent and complete clinical response was achieved at the fourth week of the cyclosporine treatment (Figure 2).

Cyclosporine is a rapid-acting immunosuppressive agent which inhibits calcineurin and blocks T-lymphocyte response (5). Just one Rowell syndrome case treated with cyclosporine has been reported so far in the literature (6). Even though cyclosporine therapy is not suggested for cutaneous lupus erythematosus without systemic involvement (4), it can considered as a treatment method due to its rapid and dramatic effectiveness, especially in cases that do not respond to steroids while presenting with rapid progression.

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