Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Case Report

So-called Rowell's Syndrome: Report of a Case

Selda Pelin Kartal Durmazlar MD,* Bilgen Oktay MD, Cemile Eren MD, Fatma Eskioğlu MD

From the Department of Dermatology, Ministry of Health Ankara, Dışkapı Yıldırım Beyazıt Education and Research Hospital, Dışkapı, Ankara, 06110, Turkey

E-mail: pelin@dr.com

* *Corresponding author*: Selda Pelin Kartal Durmazlar MD, Ministry of Health Ankara Dışkapı Yıldırım Beyazıt Education and Research Hospital, Dışkapı, Ankara, 06110, Turkey

Published:
J Turk Acad Dermatol 2009; **3 (2)**: 93201c
This article is available from: http://www.jtad.org/2009/2/jtad93201c.pdf
Key Words: *Rowell*'s syndrome, lupus erythematosus, erythema multiforme

Abstract

Observations: A 30 year-old woman with a 6-year history of systemic lupus erythematosus presented with pruritic, erythematous skin rashes on the face, targetoid lesions on the hands and multiple erosions of buccal mucosa. Laboratory investigations showed speckled antinuclear antibody with a titer of 1:320, anti-La (SS-B), anti-Ro (SS-A) antibodies and rheumatoid factor positivity. Histologic examination of lesional skin of the hand was consistent with erythema multiforme. Complete clearing of skin lesions was achieved with oral 80 mg/d prednisolone and 100 mg/d azathioprine within four weeks.

Introduction

Rowell's syndrome is a rare presentation of lupus erythematosus (LE) with erythema multiforme like lesions associated with antinuclear antibody (ANA), anti-La (SS-B)/anti -Ro (SS-A) antibodies and rheumatoid factor (RF) positivity [1].

The first described association between LE and erythema multiforme was made by *Scholtz* in 1922 [2]. In 1963, *Rowell* et al. reported a new syndrome characterized by LE, erythema multiforme-like lesions, a positive test for RF, speckled ANA and a saline extract of human tissue (anti-SJT) which is now regarded as similar to Ro (SS-A) [1, 2, 3]. However, at the present time there seems to be enough evidence to classify *Rowell*'s syndrome within the subacute cutaneous lupus erythematosus (SCLE) subset [1].

Nevertheless, we describe a patient whose clinical picture is consistent with so-called *Rowell*'s syndrome.

Case Report

A 30 year-old woman with a 6-year history of systemic lupus erythematosus (SLE) presented with pruritic, erythematous skin rashes on the face and hands. Her daily medication included

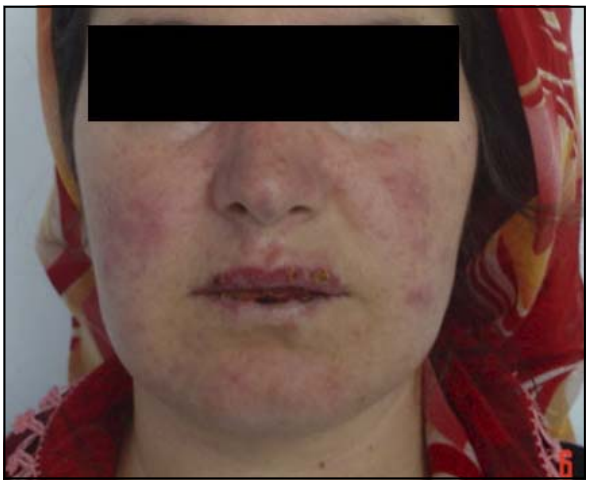

Figure 1. The patient on presentation; numerous erythematous annular plaques that coalesced on the face, hemorrhagic crusting on the lips

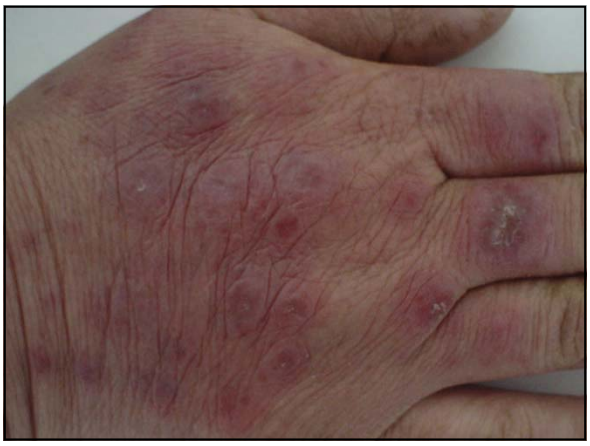

Figure 2. The patient on presentation; erythematous targetoid lesions on hands

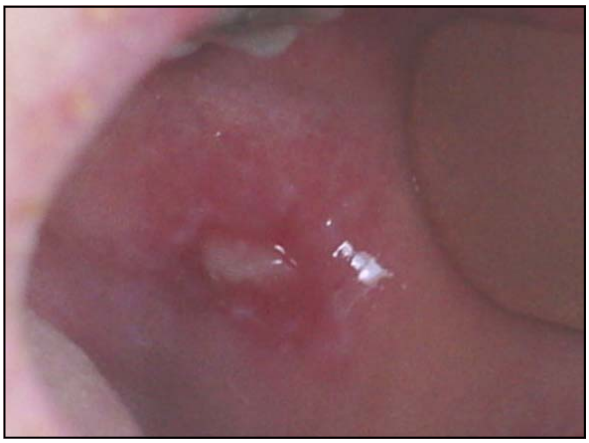

Figure 3. The patient on presentation; erosion on the buccal mucosa

prednisolone 15 mg and azathioprine 50 mg. It was learned that she was not taking any medication except prescribed ones for SLE. The patient's past medical history was otherwise unremarkable. She did not have a history of upper respiratory tract and herpes virus infection or any infection associated with fever at the onset of the lesions. On physical examination, she had numerous erythematous annular plaques that coalesced on the face, hemorrhagic crusting on the lips, erythematous targetoid lesions on the hands and multiple erosions on the buccal mucosa (**Figure 1, 2, 3**).

History revealed that she had chilblain lesions during cold exposure.

Histologic examination of lesional skin of the hand revealed hyperkeratosis, epidermal necrosis, vacuolar degeneration of the dermal-epidermal junction, and papillary dermal edema consistent with erythema multiforme and direct immunoflourescence study was negative. Laboratory investigations showed mild anemia, slight leucopenia and an erythrocyte sedimentation rate of 40 mm/h. Serum protein levels, urinalysis, liver and renal function studies were within normal limits. ANA titer was 1:320 speckled, RF, anti-La and anti-Ro antibodies were positive and anti-Scl70, anti-histon, anti-Sm, anti-dsDNA, anti-Jo1 antibodies were negative. Abdominal ultrasonography and posterior-anterior chest radiography showed no pathology. Minimal pericardial fluid was detected on echocardiography.

Complete clearing of skin lesions was achieved with oral 80mg/d prednisolone and 100mg/d azathioprine within four weeks (**Figure 4, 5**).

Discussion

Since the first report of *Rowell*'s syndrome not more than 35 cases have been reported in the English literature in which the presence of erythema multiforme-like lesions associated with LE. However, a recent review demonstrated that most of the reported cases did not fulfill all the diagnostic criteria of *Rowell*'s original description, especially the presence of RF and anti-La antibody [1]. In 1963 *Rowell* defined this association as a distinct entity upon discovering

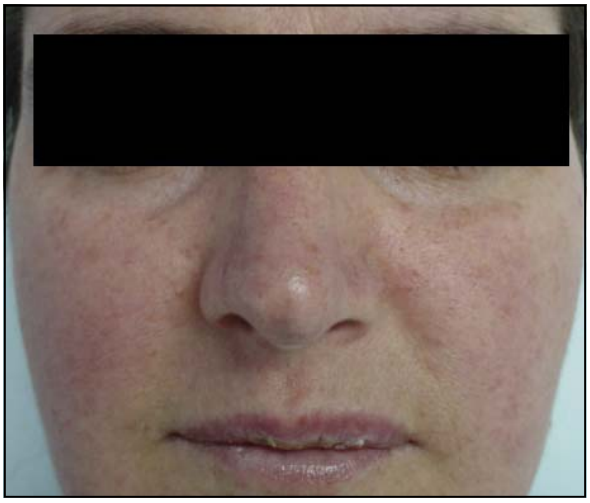

Figure 4. Lesions responded to the treatment

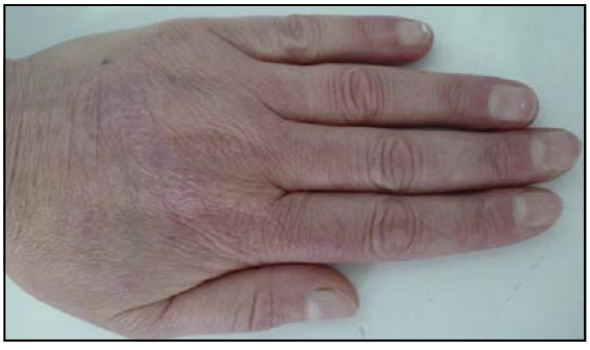

Figure 5. Lesions responded to the treatment

different clinical and immunologic findings in four patient during his study including 120 discoid lupus erythematosus (DLE) patients [**3**]. The original criteria of Rowell's syndrome consist of LE, erythema multiforme like lesions and immunological abnormalities such as speckled pattern of ANA, RF and saline extract of human tissue (anti-SJT) positivity which is now regarded as similar to anti-Ro [**1, 2, 3**]. Although this syndrome was originally described in DLE patients, some of these patients developed SLE years after the onset of DLE [**4**]. In 1995 *Lee* et al. reaffirmed the existence of *Rowell*'s syndrome and suggested the inclusion of chilblains to the diagnostic criteria. In 2000 *Zeitouni* et al. redefined *Rowell*'s syndrome with major and minor criteria [**2**].

Major criteria included

i) LE: SLE, DLE, SCLE,

ii) Erythema multiforme like lesions (with/ without involvement of the mucous membranes),

iii) Speckled pattern of ANA.

Minor criteria were

i) chilblains,

ii) anti-Ro antibody or anti-La antibody,

iii) positive RF.

However, at the present time there seems to be enough evidence to classify Rowell's syndrome within SCLE subset rather than accepting it as a separate entity, because early lesions of annular-policyclic pattern of SCLE may resemble erythema multiforme with similar histopathological findings [**1, 5**]. In addition, the immunologic abnormalities described in *Rowell*'s syndrome may also associate with SCLE [**1, 5, 6**]. However, patients with these characteristic clinical and immunological features very rarely reported in the literature and we have described a patient whose clinical picture was consistent with so-called *Rowell*'s syndrome.

References

1. Aydoğan K, Karadoğan SK, Balaban Adım S, Tunalı Ş. Lupus erythematosus associated with erythema multiforme: report of two cases and review of the literature. J Eur Acad Dermatol Venereol 2005; 19: 621-627. PMID: 16164723

2. Zeitouni NC, Funaro D, Cloutier RA, Gagne E, Claveau J. Redefining Rowell's syndrome. Br J Dermatol 2000; 142: 343-346. PMID: 10730772

3. Rowell NR, Beck JS, Anderson JR. Lupus erythematosus and erythema multiforme-like lesions. Arch Dermatol 1963; 88: 176-180. PMID: 14043605

4. Millard LG, Rowell NR. Chilblain lupus erythematosus (Hutchinson). A clinical and laboratory study of 17 patients. Br J Dermatol 1978; 98: 497-506. PMID: 656324

5. Shteyngarts AR, Warner MR, Camisa C. Lupus erythematosus associated with erythema multiforme: does Rowell's syndrome exists? J Am Acad Dermatol 1999; 40: 773-777. PMID: 10321612

6. Sontheimer RD. Subacute cutaneous lupus erythematosus: 25-year evolution of a prototypic subset (subphenotype) of lupus erythematosus defined by characteristic cutaneous, pathological, immunological, and genetic findings. Autoimmun Rev 2005; 4: 253-263. PMID: 15990071