Case Report

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

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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
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 immunofluorescence 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
different clinical and immunologic findings in four patients 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


