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Rowell's syndrome: A Case Report and Review of Literature

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Abstract:

Rowell's syndrome is a rare manifestation of systemic lupus erythematosus characterized by skin lesions resembling erythema multiforme in the background of antinuclear antibody (ANA), anti-La (SS-B)/anti-Ro (SS-A) antibodies and rheumatoid factor (RF) positivity. Here we present the case of a 20 year old Asian girl, known case of SLE for 1 year, presenting with itchy erythematous targetoid lesions and dyspnoea. ANA (with speckled pattern), anti-ds DNA, RF, anti-La and anti-Ro antibodies were positive. Skin biopsy revealed the histological picture of erythema multiforme. Her dyspnoea was attributed to the presence of massive pericardial effusion. Case reports of Rowell's syndrome are very rare in literature and some of the reported cases do not conform to the classical description. This case is close to the classical 'Rowell's Syndrome', which makes it reportable.

Key words: Erythema Multiforme, Systemic Lupus Erythematosus, Antinuclear Antibodies, Pruritus, Rheumatoid Factor.

Introduction

Rowell's syndrome is a rare presentation of lupus erythematosus (LE) with erythema multiforme (EM) like lesions in association with antinuclear antibody (ANA), anti-La (SS-B)/anti-Ro (SS-A) antibodies and rheumatoid factor (RF) positivity [1]. Cases of Rowell's syndrome have been rarely encountered in literature, since its first description which was way back in 1963. Moreover, a recent review demonstrated that most of the reported cases did not fulfil all the diagnostic criteria of Rowell's original description, especially the presence of RF and anti-La antibody [2]. Here we present a case the case of a 20 year old Asian girl, who is a known case of systemic lupus erythematosus (SLE), and presented with targetoid skin lesions and massive pericardial effusion. Her immunological markers and skin histology closely conformed to the classical description of Rowell's syndrome.

Case Report

A 20 year old girl who was a diagnosed case of SLE 1 year back, presented with shortness of breath for 1 month before admission and itchy

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reddish skin rashes over the right thigh for 15 days before admission. Previously, she was diagnosed to be a case of SLE on the basis of fever, joint pain, oral ulcer and immunological markers. She was started on oral steroids, which was gradually tapered off and at the time of current admission she was not receiving any immunosuppression. History revealed that she had chilblain lesions during cold exposure. On physical examination, she had engorged and pulsatile neck veins, tender soft hepatomegaly and bilateral pitting pedal oedema. Cardiovascular examination revealed enlarged heart size and muffled heart sounds. The patient was haemodynamically stable. Few erythematous targetoid lesions were detected over the medial aspect of her right thigh and right upper back.

Laboratory investigations showed mild anemia, leucopenia and an erythrocyte sedimentation rate of 120 mm/h. Serum protein levels, urinalysis, liver and renal function studies were within normal limits. ANA titer was 3(+) with speckled pattern. Anti-ds DNA, RF, anti-La and anti-Ro antibodies were positive and anti-Scl 70, anti-histone, anti-Sm, anti-Jo1 antibodies were negative. Chest skiagram revealed enlargement of heart with bilateral hilar congestion. ECG contained low amplitude QRS complexes without any other obvious abnormalities. Echocardiography confirmed the presence of massive pericardial effusion without any suggestion of tamponade. Abdominal ultrasound did not reveal any obvious abnormality except mild hepatomegaly. Histologic examination of the skin lesions of the thigh reveled vacoular interface dermatitis with lymphocytic infiltration in the dermo-epidermal junction and dyskeratotic cells in epidermis, suggestive of EM.

Discussion

The first described association between SLE and EM was made by Scholtz in 1922 [3]. In 1963, Rowell *et al*. reported a unique syndrome characterized by

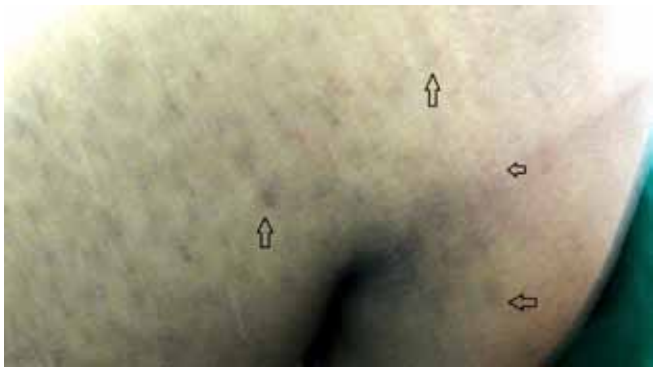

Fig.1: Targetoid skin lesions, resembling erythema multiforme, present in the upper back region.

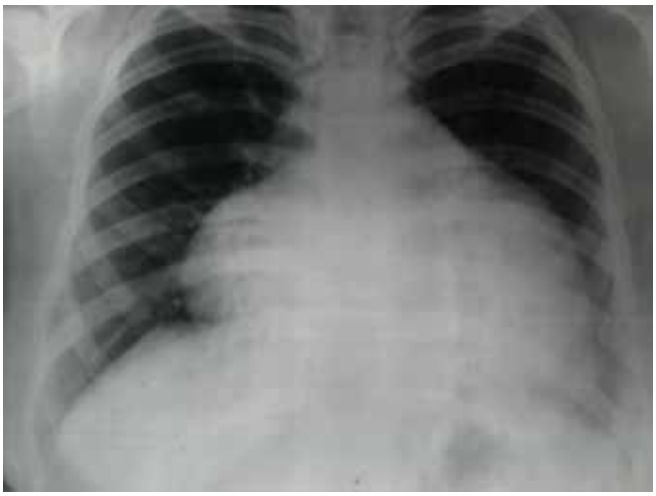

Fig.2: Chest skiagram showing pericardial effusion.

SLE, EM-like lesions, a positive test for RF, speckled ANA and a saline extract of human tissue (anti-SJT) which is now known as similar to Ro (SSA) [1]. Since the first report of Rowell's syndrome very few cases have been reported in the literature in which the presence of EM-like lesions are associated with LE. In 1963, Rowell defined this association as a distinct entity upon discovering different clinical and immunologic findings in four patient during his study including 120 discoid lupus erythematosus (DLE) patients. The original criteria of Rowell's syndrome

comprised of LE, EM 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]. Although this syndrome was originally described in DLE patients, some of these patients developed SLE years after the onset of DLE [3]. In 1995, the observation by Lee *et al*. consolidated the existence of Rowell's syndrome. They suggested chilblains to be included in the diagnostic criteria for Rowell's syndrome. In the year 2000, Zeitouni *et al*. came up with major and minor criteria [4]. Major criteria includes-(i) LE: SLE, DLE, subacute cutaneous lupus erythematosus (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.

Rowell's syndrome has been reported with all subtypes of LE (systemic, acute, subacute or discoid) [1,4,5]. Conversely, all forms of EM (EM minor, EM major) can be associated with Rowell's syndrome [5,6]. Toxic epidermal necrolysis have also been reported in association with this syndrome [7]. Although the proper classification of EM-like lesions occurring in Rowell's syndrome is not clear, these lesions may represent a severe variant of acute cutaneous lupus or, in some cases, subacute cutaneous lupus. Early lesions of annular variety SCLE may resemble erythema multiforme with similar histopathological picture, although direct immunofluorescence usually distinguishes these lesions as those of LE [8].

Cases of the so called "Rowell's syndrome" with fulfilment of all the criteria have very rarely been reported. Here we presented a case of Rowell's syndrome in a diagnosed case of SLE, who presented to medical attention with massive pericardial effusion.

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