Case Report of a Rowell’s Syndrome

Devang Solanki¹, Ekta Dalal², Nishita Darji³

¹Post graduate student of B. J. Medical College, Ahmadabad, Gujarat, India
²Assistant Professor B. J. Medical College, Ahmadabad, Gujarat, India
³Post graduate student of M. P. Shah Medical College, Jamnagar, Gujarat, India

Abstract: 11 year old female presented with history of erythema multiforme like eruption developed lupus erythematosus in form of malar rash with unusual laboratory and immunological findings were consistent with Rowell’s syndrome. That includes lupus erythematosus with erythema multiforme like skin lesion, positive anti nuclear antibody and positive rheumatoid factor. So we believe our patient meet the criteria for this rarely reported entity.

Keywords: Rowell’s syndrome, erythema multiforme, lupus erythematosus, positive rheumatoid factor, positive anti nuclear antibody titre.

1. Introduction

Rowell’s syndrome is a rare presentation of lupus erythematousus(LE) with erythema multiforme like lesions associated with antinuclear antibody(ANA), anti-La(SSB)/anti-Ro(SS-A) antibodies and rheumatoid factor(RF) positivity[1].The first described association between LE and erythema multiformewas made by Scholtzin 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(SSA)[1, 2, 7]. 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.

2. Case Report

We reported, 11 year female child admitted to our ward with history of skin eruption over the face for two weeks and malena for one week. Physical examination revealed erythematous, well defined and confluent popular rash with central necrosis distributed over the forehead and both cheeks, typically butterfly typed malar flush. Patients has also purpurish rash on sun exposed skin with scarring and depigmennation which are signature findings of lupus erythematosus. Patient also has ulceration on hard palate, glossitis and gingivitis.

Patient was treated with topical steroid ointment and emollients. After two weeks lesions subsided and healed well. Yet patient has not developed any signs of systemic lupus erythematosis (SLE). Hence we believe that our patient met the criteria for this rarely reported entity of Rowell’s syndrome which is distinctive syndrome of cutaneous lupus erythematosus with erythema multiforme.

3. 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].Clinical lesions of RS include LE (systemic, discoid or subacute), ErythemaMultiforme(EM)-likelesions and chilblains. Although this syndrome was originally described in a discoid erythematous lupus (DEL) patient by Rowell [15], further cases with different variants of cutaneous LE such as systemic (SLE) and subacute (SCLE) were reported [1]. At admission a diagnosis of SCLE was considered in our patient due to the presence of elevated anti-DNAds antibody, positive ANAtitre, positive RF and a skin rash.

Classical EM is precipitated by trigger factors such as infectious agents, mainly Mycoplasma pneumonia and HSV or drugs like antibiotics, non steroid anti-inflammatories and anti-convulsants, although other causes including malignant conditions and connective tissue disease have been implicated [1,4,7,9,12,14,15]. EM is not associated with any specific auto-immune serological abnormality [15].The prolonged course (more than six weeks) of erythema multiforme-like lesions observed in our patient does not favor the diagnosis of true erythema multiforme [1, 5, 15] and on the other hand there was no identifiable precipitating factor.

Clinical and histological differentiation of EM from SCLE may be difficult [1, 10, 12]. Early SCLE lesions with annular-polycyclic pattern may resemble EM. Necrotic keratinocytes may be found in SCLE lesions [1] as in EM. In
fact, Herrero and coworkers recently described the presence of necrotic keratinocytes in 6 of 13 (54%) SCLE patients [11]. As some clinical, histological and immunological findings seems to overlap RS and SCLE, it has been suggested by some that lupus erythematosus with EM-like rashes designated as RS represent a subset of SCLE with targetoid lesions, rather than a distinct entity [1, 5, 8].

Although this Syndrome was originally described in DLE patients, some of these patients developed SLE years after the onset of DLE [13]. In1995 Lee et al. reaffirmed the existence of Rowell’s syndrome and suggested the inclusion of chilblains to the diagnostic criteria.

Table 1: Zeitouni et al. redefined in 2000 Rowell’s syndrome [6]

<table>
<thead>
<tr>
<th>Sr. no</th>
<th>Major Criteria</th>
<th>Minor Criteria</th>
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<tbody>
<tr>
<td>1</td>
<td>LE, SLE, DLE, SCLE</td>
<td>Childblains</td>
</tr>
<tr>
<td>2</td>
<td>Erythema multiform like lesions</td>
<td>Anti-Ro antibody</td>
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<tr>
<td>3</td>
<td>Speckled pattern of ANA</td>
<td>Positive RF</td>
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</table>

Three major and one minor criterion required diagnosed Rowell’s syndrome. 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. The immunologic abnormalities described in Rowell’s syndrome may also associate with SCLE[1, 2, 3]. However, patients with these characteristic clinical and immunological features very rarely reported in the literature and we have described a patient whose clinical picture and immunological profile consistent with Rowell’s syndrome.

References