Case Report of a Rowell's Syndrome

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Abstract: ***11 year old female presented with history of erythema multiforme like eruption developed lupus erythematous in form of malar rash with unusual laboratory and immunological findings were consistent with Rowell's syndrome. That includes lupus erythematous 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 erythematous, positive rheumatoid factor, positive anti nuclear antibody titre.

1. 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 multiformewas 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(SSA)[1, 2, 7]. However, at the present time there seems to be enough evidence to classify *Rowell*'s syndrome within the subacutecutaneous 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 purpulish rash on sun exposed skin with scarring and depigmenatation which are signature findigs of lupus erythematous. Patient also has ulceration on hard palate, glossitisand gingivitis.

Patient has past history of varicella infection six week back followed by severe erythema multiforme major type I lesion. Patient was given oral steroid and anti histaminics and lesions were healed well. During this admission patient has auto immune screening-Anti nuclear antibody titre(1:80) positive, Anti ds-DNA antibody (+2), Positive rheumatoid factor. Patient also has mild thrombocytopenia without anaemia or leucopenia with normal liver and renal function test and urine analysis. Patient serological marker for Hepatitis B and Hepatitis C and HIV negative. Patient has no other findings suggestive of other system involvement like arthritis, glomerulonephritis and serositis.

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 erythematous with erythemamultiforme.

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. reaffirme the existence of *Rowell''*s syndrome and suggested the inclusion of chilblains to the diagnostic criteria

Table 1: Zeitouniet al. redefined in 2000 Rowell''s syndrome [6]

Sr. no	*Major Criteria*	*Minor Criteria*
1.	LE, SLE, DLE, SCLE	Childblains
2.	Erythema multiform like lesions	Anti-Ro antibody or anti-La antibody
3.	Speckled pattern of ANA	Positive RF

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.

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