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Lupus Erythematosus with Erythema Multiforme like lesions: Rowell Syndrome; a Clinical or Histological Diagnosis?

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Abstract

Observation: Rowell syndrome (RS) is a rare and as such very controversial condition. Though defining diagnosing criteria exist, yet many previously published reports lack one or more of these criteria. Erythema multiforme has multiple etiological factors but coexistence of cutaneous lupus erythematosus (CLE) and erythema multiforme (EM) like eruption in a single individual has been defined under Rowell syndrome (RS) along with other criteria. We report a case of a 25 year old female who had lesions of lupus erythematosus along with erythema multiforme-like lesions and laboratory findings were consistent with Rowell syndrome. Here we suggest that histopathological criteria should also be included in defining criteria of Rowell syndrome.

Introduction

Rowell's Syndrome (RS) was described by Professor *Neville Rowell* and colleagues in 1963. Patients with the syndrome have discoid or systemic lupus erythematosus (LE), annular targetoid skin lesions like erythema multiforme (EM) associated with a characteristic pattern of immunological abnormalities [1]. It is a rare condition with worldwide distribution.

In 2000, *Zeitouni* et al redefined these major and minor diagnostic criteria for Rowell syndrome [2]. The diagnosis of this syndrome requires the presence of all the major criteria and at least one of the minor. On search of literature, till 2012 only 71 cases of Rowell Syndrome have been reported [3] here we describe a similar case in 25 year old female.

Case Report

A 25 year old female presented with two localized, erythematous, pruritic plaques of seven months duration present each on nose and lips along with itchy, erythematous plaques over dorsal aspect of index fingers of both hands since ten days. There was history of joint pains also. History dates back to seven months when patient noticed erythematous plaque over root of nose which was associated with mild to moderate itching. Lesion increased in size over the next few months along with development of mild scaling over them. One month ago, she developed a similar lesion on the lip and a swelling in neck region. Few days later, patient developed erythematous papules over dorsal aspect of index fingers of both hands along with fever and joint pains in the knees, elbows, wrists. It was associated with photosensitivity, Raynaud's phenomenon and chilblains. There was no associated history of oral ulcerations or infections like Herpes Simplex virus, Mycoplasma, Orf. She was a diag-

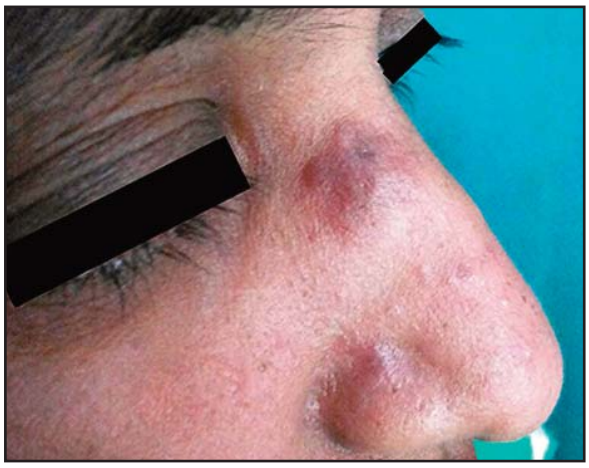

Figure 1. Showing erythematous plaque over the root of the nose

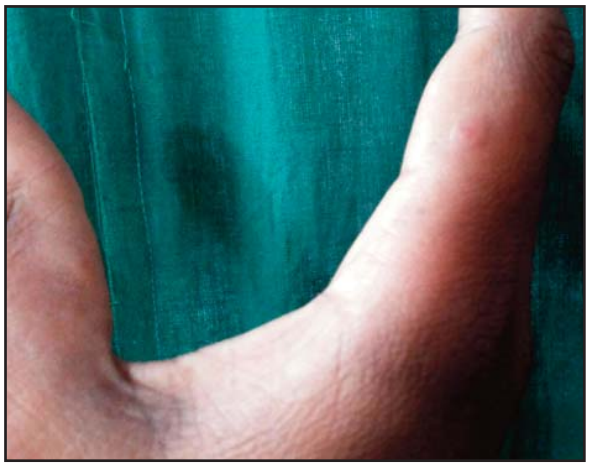

Figure 2. Showing targeted lesion on the side of the index finger

nosed case of hypothyroidism on treatment with thyroxin (100 microgram/day). Family history was not significant.

On muco-cutaneous examination, two round well-defined erythematous and indurated plaques of size 2 x 3 cm were present over root of the nose and centre of the upper lip. Both had slight central atrophy and mild scaling (**Figure 1**). Erythematous plaques with halo and central hyperpigmented targetoid lesion were present over the volar aspect of index finger of both the hands (**Figure 2**). Skin biopsy was done from lesion on the right finger. All mucosae were spared.

Autoimmune screening revealed positivity for Rheumatoid Arthritis factor (RF), Anti-Ro, and Anti-La antibodies while Anti Nuclear Antibodies (ANA) was negative. Histopathological examination revealed laminated hyperkeratosis, acanthosis, hypergranulosis and lymphocytic infiltrate around tips of rete ridges but without any evidence of necrotic keratinocytes. It was found to be consistent with lupus erythematosus.

A short course of tapering oral and topical steroids for a few months showed complete resolution of cutaneous lesions without any scarring. Since then there has been no recurrence during the follow-up of many months.

Discussion

The association between Lupus Erythematosus (LE) and Erythema multiforme (EM) was originally described by *Scholtz* in 1922 [**4**]. During a study of 120 patients with chronic discoid LE, *Rowell* et al in 1963 reported this entity in four female patients characterized by discoid LE, EM-like lesions, a positive test for Rheumatoid Factor, speckled Anti Nuclear Antibodies (ANA) and a saline extract of human tissue (anti-SJT) [**1**].

In 2000, *Zeitouni* et al redefined Rowell's syndrome with major and minor criteria [**2**].

Major criteria included

i) Lupus erythematosus: Systemic LE, Discoid LE or Subacute Cutaneous LE

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

iii) Speckled pattern of ANA.

Minor criteria:

i) Chilblains

ii) Anti-Ro antibody or anti-La antibody

iii) Positive Rheumatoid Factor.

The diagnosis of this syndrome requires the presence of all the major criteria and at least one of the minor.

Since the diagnosis of RS is based on clinical and immunological criteria, the case presented here fulfilled most of the criteria described by *Zeitouni* other than ANA factor. As analyzed by *Antiga* et al speckled pattern of ANA was found in only 60.4% of the reported cases of RS [**3**].

The histopathology of EM like lesion in our case was consistent with LE which made us probe the histopathological findings of previously reported cases of RS. In the original case series of four patients by *Rowell*, only one patient had the histopathological description consistent with EM like lesion [**1**]. So the other three cases could have been Subacute

cutaneous Lupus Erythematosus (SCLE) patients misdiagnosed as RS.

Similarly, *Lyon* et al reported two cases initially diagnosed as Rowell syndrome with EM like lesions associated with positive Ro/SSA antibodies and RF but on histopathological analysis findings were suggested of Subacute cutaneous Lupus Erythematosus(SCLE) [**5**]. Again *Modi* et al reported two cases of RS in which the histopathological examination of clinically typical EM lesions revealed the presence of LE features while in other case reports by *Pandhi* et al well as *Duarte* et al histopathological findings were consistent with EM though the lesions were clinically suggestive for SCLE [**6,7,8**].

Thus there is a difficulty of differentiating clinically between the annular polycyclic type of SCLE and EM. Furthermore, histologically also the presence of necrotic keratinocytes is not exclusive to EM as it has been reported by *Herrero* et al in six out of thirteen patients of SCLE [**9**].

The dilemma is whether Rowell's syndrome is a separate entity in the spectrum of LE or it is a subset of SCLE. The arguments in favour of RS as a type of SCLE is early lesions of annular-polycyclic pattern of SCLE may resemble erythema multiforme with similar histopathological findings. In addition, the immunologic abnormalities described in Rowell's syndrome may also be associated with SCLE. But the response to treatment is good as per previous reports and progression to SLE is seen in few cases although recurrences are common [**2,10**]. The response to treatment was good in present case also. Thus the prognosis of the condition favors its existence as separate entity.

To classify Rowell Syndrome as a discrete entity, diagnostic criteria need to be further defined so as to eliminate the possibility of SCLE wrongly diagnosed as RS. Hence it is suggested that histopathological criteria or more precise tests like direct immunoflouresscence may be added to the diagnostic criteria.

To conclude Rowell syndrome is a rare condition. As more and more cases are reported, the existence of this entity will be further clarified. For now, it should be considered a distinct disorder, suspected and screened in all patients with LE and EM-like lesions.

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