CASE REPORT

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Rowell Syndrome in a 4-Year-Old Male Child: A Rare Case Report

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ABSTRACT

Rowell syndrome is an unusual disease entity characterized by the occurrence of erythema multiforme(EM) in association with lupus erythematosus(LE). The syndrome occurs mostly in middle aged women. We are reporting this case in a 4 year old child.

KEYWORDS: Rowell syndrome, SLE, Erythema Multiforme, Child

INTRODUCTION

Rowell syndrome is a very uncommon disease entity. It was described back in the year 1963 by Rowell and co-workers.[1] They detailed it as a syndrome characterized by lupus erythematosus with erythema multiforme-like lesions, along with a positive test for rheumatoid factor, speckled antinuclear antibodies, and a serum antibody directed to an extract of human tissues. Later on, major and minor diagnostic criteria were proposed for its diagnosis. Till date, only around 90 cases have been reported. Most of the cases have been described in middle aged women. We are reporting this case in a 4-year-old male child because it is a rare entity as such and occurrence in younger population is quite unusual.

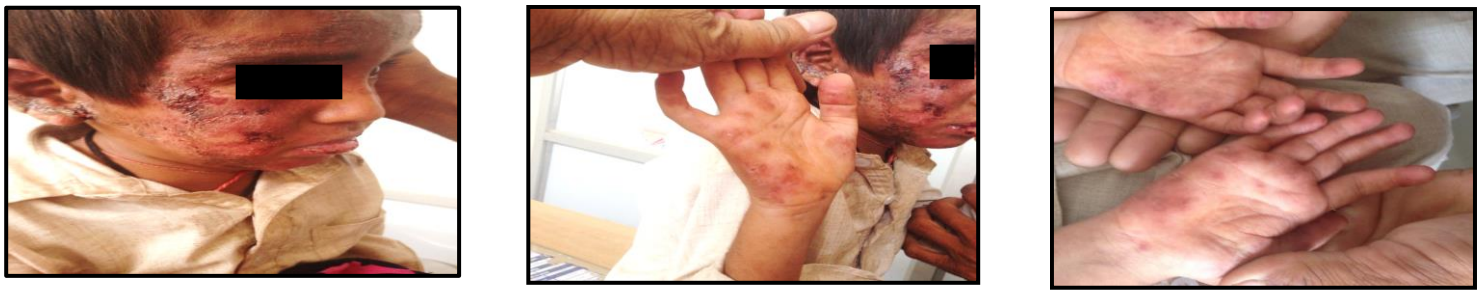
CASE REPORT

A 4-year-old male child presented in our OPD with complaints of on & off fever; erythematous, tender swelling of fingers & toes on exposure to cold; pruritic, erythematous plaques all over the body; multiple oral ulcers; joint pain & myalgia since a duration of 2 years.

The history of Hypertension, Diabetes Mellitus, Tuberculosis, Bronchial Asthma, or seizures in the child were all negative. There was no history of similar complaint in any other family member. Patient was on irregular treatment with topical (clobetasol) and oral steroids (prednisolone 10 mg) as well as emollients. On examination, the sites involved were face, neck, ears, trunk, arms, legs, palms, soles and oral mucosa. (Figures 1 a-c) The involvement of extensor sites was more than flexors and photoexposed sites had more lesions than photoprotected sites. The sites spared were axillae, groins, ocular/ nasal/ genital mucosae. There were multiple erythematous, ill-defined, discrete to confluent papules and plaques on body with superficial erosions and crusting at some sites.

Hemorrhagic crusts were present over lips. There were superficial ulcers with yellowish slough on the oral mucosa. Palms & soles showed presence of targetoid lesions. Other systemic examinations conducted were found to be within normal limits. Due to the concurrent presence of lupus eryththematosus along with erythema multiforme like lesions, we considered the possibility of Rowell syndrome and then subjected the patient to baseline investigations. Complete hemogram was done which showed anemia with a Hb value of 8.7 g/dl, normal TLC at 8800/Cumm and lymphopenia. Urine analysis and serum chemistry were also found to be normal. Immunofluorescent assay (IFA) showed a positive ANA with a speckled pattern. Anti-dsDNA done by IFA was also positive. Anti Ro & anti La Abs performed by enzyme immunoassay (EIA) were raised. Rheumatoid factor was found to be negative. CRP was raised (7.2mg/L). Histologic examination of a plaque from the upper arm revealed mild hyperkeratosis, slight epidermal atrophy and presence of vacuolar degeneration in the basal layer (Figure 2). Dermis showed perivascular and periadnexal infiltrate composed of mainly lymphocytes and melanin incontinence. These findings were consistent with Lupus Erythematosus.

On the basis of clinical, Histopathological & immunological findings, we diagnosed the patient as a case of Rowell's syndrome (SLE+EM like lesions). The child was started on oral prednisolone with a dose of 2mg/kg/day and hydroxychloroquine to which he responded well initially but later his condition deteriorated and he died because of septicemia.



(a) (b) (c)

Figure 1 a-c. Examination of the Patient

DISCUSSION

Rowell et al. described a syndrome characterized by LE and EM-like lesions, RF, speckled ANA, and precipitating antibody to saline extract of human tissue (anti-SjT) in 1963.[1] In 2000, Zeitouni et al. advocated 3 major and minor criteria each to diagnose this condition:[2]

Major criteria:
1) LE(systemic, discoid or subacute)
2) EM like lesions
3) ANA(speckled pattern)

Minor criteria:
1) Chillblains
2) Anti Ro &/or Anti La antibodies
3) +ve Rheumatoid factor

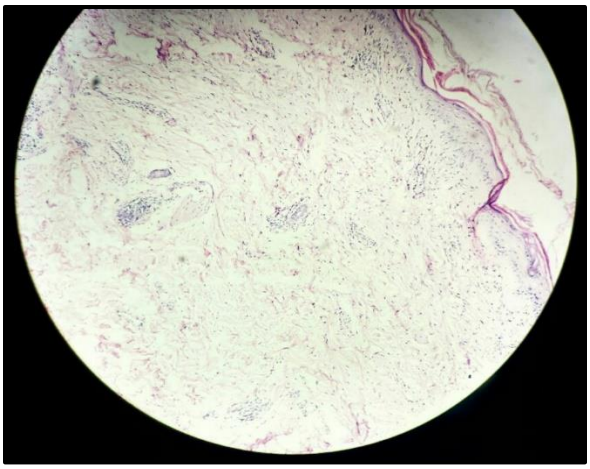


Figure 2. Histologic examination of a plaque from the upper arm

3 major and 1 minor criterion are required to establish the diagnosis. Our patient fulfilled all the major criteria along with chillblains & Anti Ro/La positivity. Occurring in about 88% of the cases, speckled ANA pattern is the most consistent character of Rowell Syndrome, whereas Rheumatoid Factor is the least preserved feature, which is seen only in around 41% of the affected subjects.[3,4] In 2005, Aygodan K et al. proposed Rowell Syndrome to be a sub-entity of subacute lupus erythematosus with erythema multiforme.[5] Some other researchers have variously suggested that Rowell Syndrome is a different variant of cutaneous lupus erythematosus, a subtype of chronic lupus erythematosus or an independent subtype of lupus erythematosus. Moreover in 2012, Torchia concluded that Rowell Syndrome might be incorporated as an autonomous variety of cutaneous LE confined in the spectrum of LE-specific skin conditions.[6] Discovery of new cases like the present one and their addition to the research literature reinforces the belief that Rowell's syndrome is a well-defined clinical entity with definite diagnostic characteristics, although it is not so frequent. In most of the cases, both Rowell Syndrome and SLE respond to a similar therapeutic regimen and strategies. Steroids like prednisolone, antimalarial drugs, dapsone, immunosuppressants like azathioprine and cyclosporine have been used with good efficacy.[7-8]

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