A Case of Rowell's Syndrome Masquerading as Erythema Multiforme

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Abstract
Background: Rowell's Syndrome is a rare entity which usually manifests as Lupus Erythematosus with erythema multiforme like lesions. All major criteria as well as minor criteria are to be met in order to diagnose the patient as Rowell's syndrome. Methods: In this article we are discussing a case of a female patient aged 40 years with polyarthralgia and targetoid lesions and positive rheumatoid factor and ANA showing speckled pattern and positive anti – Ro antibodies. Results: Patient received treatment with methyl prednisolone, hydroxychloroquine and topical steroids and other supportive medication. Patient responded and improved well with the treatment. Conclusion: In all individuals with systemic lupus erythematosus and lesions resembling erythema multiforme, it is advisable to rule out Rowell's syndrome.
Keywords: Rowell's Syndrome, Systemic lupus erythematosus, Discoid Lupus Erythematosus, Erythema Multiforme, Rheumatoid Factor.

1. Introduction

Professor Neville and his colleagues first described Rowell's Syndrome in 1963 (1). Patients suffering from Rowell's syndrome have Systemic Lupus Erythematosus or Discoid Lupus Erythematosus, annular skin lesions like erythema multiforme and immunological abnormalities with a characteristic pattern (2). Rowell's syndrome is not common but is seen worldwide and it occurs in almost all subtypes of lupus erythematosus like acute, subacute, systemic, discoid types (4). Major criteria for Rowell's Syndrome include positive ANA report with speckled pattern, erythematous multiforme and lupus erythematosus like lesions. Minor criteria for Rowell's Syndrome include positive Rheumatoid factor, positive anti – Ro antibody (SS-A), anti – La antibody (SS-B) (6). For diagnosing a patient with Rowell's syndrome, all major and minor criteria are to be fulfilled (5).

2. Case Report

A 40-year-old post-menopausal female, a known case of hypothyroidism for 6 years on regular treatment with tablet levothyroxine 75 mcg & also a known case of Rheumatoid Arthritis for 5 years on irregular medication presented with complaints of polyarthralgia, rashes over upper chest, lips & genital area associated with itching for 3 months. Patient also has history of photosensitivity, painless oral ulcers, difficulty in swallowing, decreased appetite, disturbed sleep & weight loss of around 20kg for past 3 months along with symptoms like nausea, vomiting, burning sensation over chest, throat & abdomen and severe hair fall. Patient was a known case of steven Johnson syndrome for which she took treatment 2 years ago. She also had folliculitis & oral candidiasis which was treated. No other comorbidities are present. Her obstetric history was good. On local examination, erythematous maculopapular rashes over upper chest, axilla, lips & genitalia were seen associated with itching along with erosive lesions over hard palate & oral candidiasis. Vitals were stable and systemic examination revealed no abnormalities.

All necessary investigations were done. Patient RA factor and ANA was positive. ANA showed speckled pattern on immunofluorescence. with SS-A native & RO-52 strongly positive while JO-1 weak positive & borderline Sm antibody. Other antibodies came negative. Erythrocyte Sedimentation Rate, C-Reactive protein & Lactate dehydrogenase levels were elevated. Her complete blood counts showed the picture of pancytopenia and peripheral smear showed picture of dimorphic anemia with pancytopenia with severe anisopoikolocytosis showing pencil shaped cells & tear drop cells, decreased platelets & WBC with hyper segmented neutrophils. Retic count, RFT, LFT, electrolytes, urine routine, USG pelvis study are all normal. Based on the clinical picture of erythematous rash as well as investigations, patient was diagnosed to be having Overlap Syndrome (Rhupus) as well as subacute cutaneous lupus erythematosus which overall gives the picture of Rowell's syndrome. Patient was stated on Hydroxychloroquine 6 mg/kg/day, methyl prednisolone 1mg/kg/day, topical mometasone furoate (0.1%), broad spectrum sunscreen, Tablet fluconazole & candid-B oral mouth wash for candidiasis, Tablet Ferrous sulphate for anemia & other symptomatic treatment was given. Rashes

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disappeared gradually and significant improvement is observed in the patient. Patient is being followed up regularly and there are no further symptoms.

Table 1: Investigations	
INVESTIGATIONS	VALUES
RA FACTOR	POSITIVE
ANA	
SS-A NATIVE	STRONG POSITIVE
Ro- 52	
Jo-1	WEAK POSITIVE
WBC COUNTS	3120 / CUMM
RBC / HB	1.62 mill/ 5.4g
Platelet count	73,000

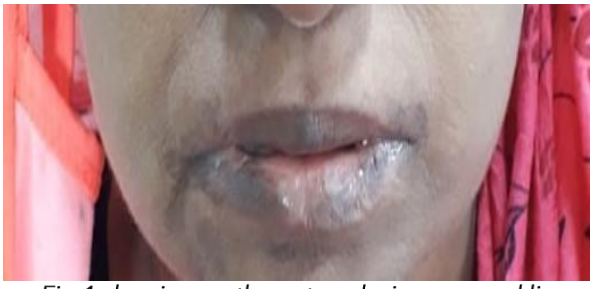

Fig 1 showing erythematous lesions around lip

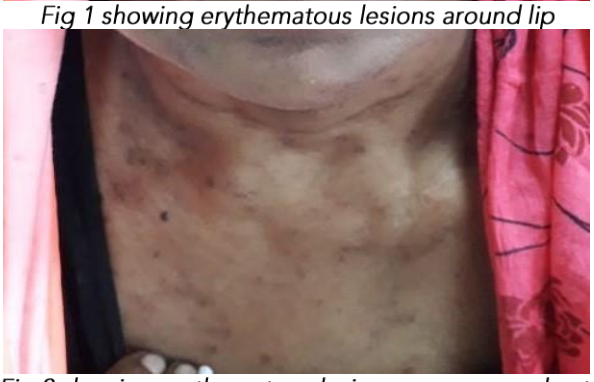

Fig 2 showing erythematous lesions over upper chest.

3. Discussion

Rowell's syndrome seen mostly in women of 20 – 70 years is a rare entity (3). It consists of lesions like erythema multiforme coexisting with lupus erythematosus and immunological findings which are characteristic (7). Rowell's syndrome first reported in patients with positive RA factor, erythema multiforme like lesions over face, chest, arms etc, chronic discoid lupus erythematosus, positive ANA report with speckled pattern and positive anti-Si-T, an antibody to saline extract from human tissue (8). Currently Rowell's Syndrome is classified under subtype of cutaneous lupus erythematosus (8). Genetical and environmental factors include the syndrome's pathogenesis. This syndrome's higher incidence is attributed to estrogen signaling, drugs like terbinafine and sun exposure (7). Azathioprine, hydroxychloroquine, prednisolone, dapsone, and cyclosporine are some drugs which have been used in treatment of patients with good results (5). In this case, our patient responded well to prednisolone & HCQ's.

4. Conclusion

Recent literature debated whether Rowell's syndrome is an overlap syndrome with true association or just a coincidence of erythema multiforme and discoid type of lupus erythematosus despite the refined criteria. Hence although Rowell's syndrome is rare, there is a need to rule out the syndrome in all the individuals with lupus erythematosus and lesions resembling erythema multiforme without any signs of causative factor.

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Ethical Consent: Informed consent was taken from the patient included in the study.

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Conflict Of Interest: The authors declare no conflict of interest.

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