DOI: 10.7860/JCDR/2022/57837.16913

Case Report

Dermatology Section

Rowell Syndrome: A True Entity or a Diagnostic Challenge?

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ABSTRACT

Lupus Erythematosus (LE) is an inflammatory condition of the connective tissue which manifests in variable forms. Rowell syndrome is a rare entity, with a female preponderance, where there is the presence of both LE and Erythema Multiforme (EM)-like lesions. Such patients are positive for antinuclear, anti-Ro/La and rheumatoid factor. Subcutaneous LE (SCLE) and EM can often share characteristics. The presence of lesions resembling both the conditions can pose a diagnostic difficulty, especially in the absence of complete clarity on the existence of Rowell syndrome. Present case is of a 47-year-old female, who presented with rashes over the face and trunk, along with photosensitivity. Cutaneous examination revealed multiple erythematous hyperpigmented plaques over the face with atypical targetoid lesions over the trunk and limbs. The diagnosis of Rowell's syndrome was confirmed based on immunology and histopathology. This case has been reported to highlight the importance of delving into the various presentations of LE.

Keywords: Erythema multiforme-like, Hyperpigmented plaques, Limbs, Lupus erythematosus

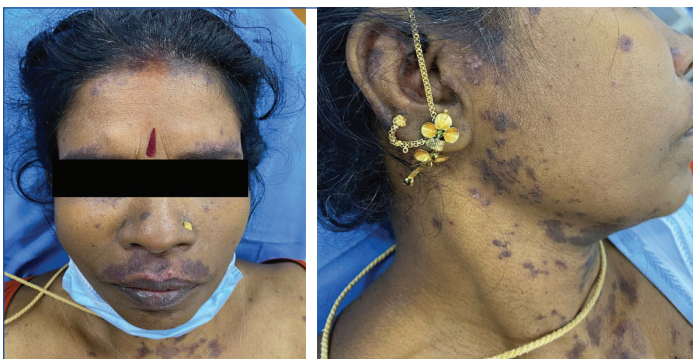
CASE REPORT

A 47-year-old female patient presented to the Dermatology Outpatient with a three month history of rashes over the face and trunk, associated with photosensitivity, which had aggravated in the past 10 days. There were similar lesions on the arms, painless oral ulceration over the hard palate, painful knee and hip joints. There was no such history of similar episodes in the past. She was not taking any medicines and no episodes of fever were reported.

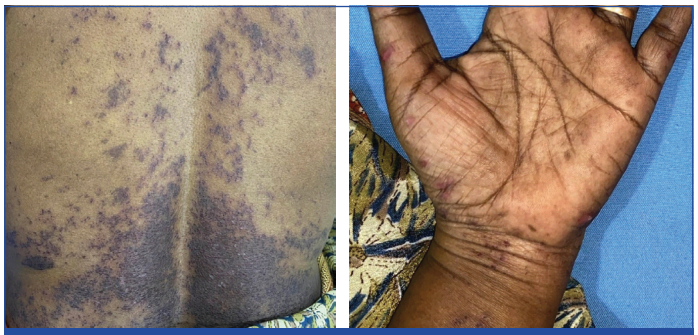
General and systemic examinations were normal. Cutaneous examination showed multiple erythematous and hyperpigmented plaques with ill-defined borders on the forehead, lateral side of face, and upper lip region, resembling photosensitive LE lesions of subacute lupus erythematosus [Table/Fig-1,2]. Raised atypical targetoid lesions (edematous and erythematous to hyperpigmented papules and plaques) were present on sun-exposed areas like the upper chest, back, upper limbs and scalp [Table/Fig-3]. Few, small, erythematous, targetoid lesions (annular patches with rounded borders) were seen over the palms, resembling EM-like lesions [Table/Fig-4]. A few dusky papules and macules were also present over the helix of the ear. The oral cavity also had hyperpigmented patches and an ulcerated, painless erosion was seen on the hard palate [Table/Fig-5].

Dermoscopic examination of the facial lesions showed white structureless areas [characteristic dermoscopic finding in Discoid Lupus Erythematosus (DLE)] with loss of hair follicles, multiple white and yellow dots, and multiple pigment globules, akin to DLE [Table/Fig-6]. A provisional diagnosis of erythema multiforme, lupus erythematosus, and adverse drug reaction was considered and the patient was subjected to further investigations.

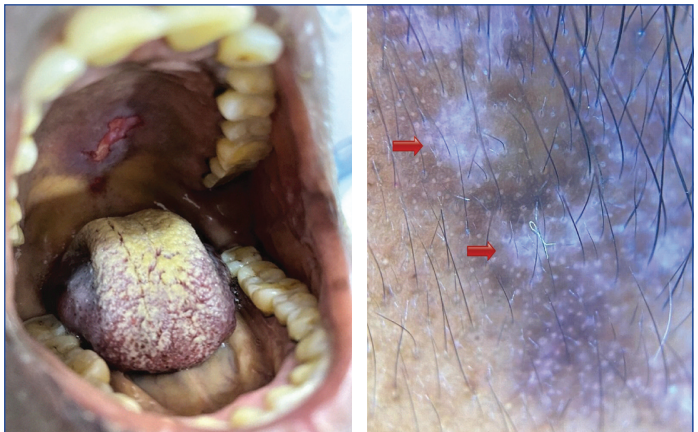
Complete haemogram showed signs of anaemia (hemoglobin – 8.2 gm/dL), reduced total counts (3600 cells/cu.mm) and elevated Erythrocyte Sedimentation Rate (ESR) (30 mm/hour). Antinuclear



[Table/Fig-1]: Clinical image of the face showing erythematous, violaceous and pigmented plaques predominantly over the perioral area, eyebrows and hairline. **[Table/Fig-2]:** Clinical image of neck showing erythematous, violaceous and pigmented plaques. (Images from left to right).

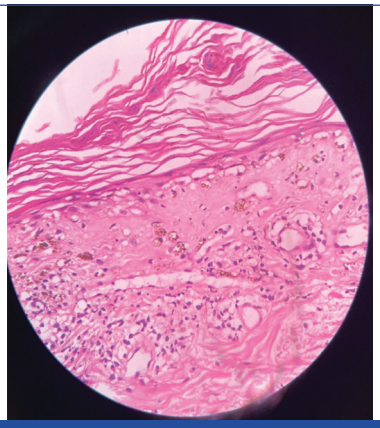


[Table/Fig-3]: Clinical image of the back showing erythematous, violaceous and pigmented plaques. **[Table/Fig-4]:** Clinical image of the palm with erythematous and targetoid lesions. (Images from left to right).



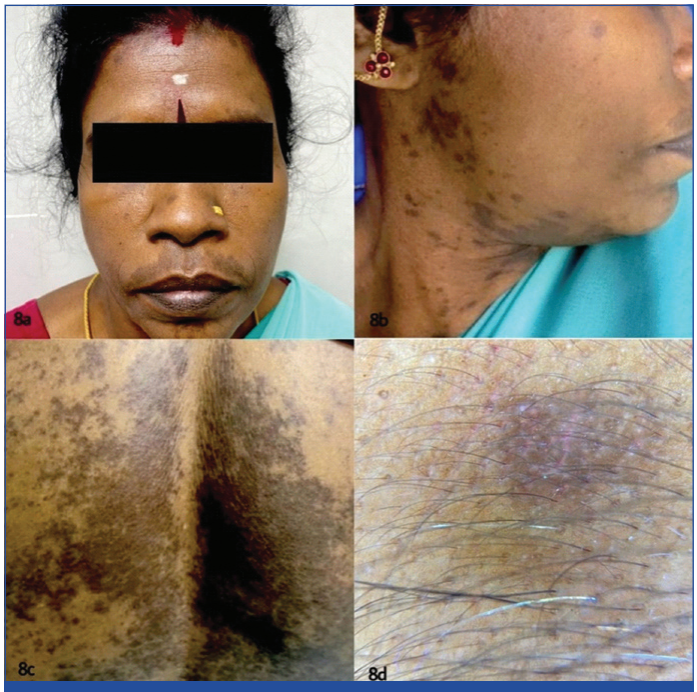
[Table/Fig-5]: Clinical image of the oral cavity showing a well-defined erosion over the hard palate and candidiasis over the tongue. **[Table/Fig-6]:** Dermoscopy (non contact, polarised) of the lesion on the face showing white structureless areas indicating fibrosis. (Images from left to right).

Antibody (ANA) profile, Anti-ds DNA, HBsAg, Anti-HCV, CRP, anti-Ro and anti-La antibodies were requested. Among these, ANA (1:100 dilution) and Anti-Ro antibody were found to be positive. Fine, speckled, nuclear pattern of ANA on Indirect Immunofluorescence (IIF) was seen. Histopathological study of a 3.5 mm punch biopsy, taken from a lesion on the lower back, showed necrotic keratinocytes with basal cell degeneration, pigment incontinence, dilated dermal capillaries with surrounding lympho-histiocytic mononuclear infiltration [Table/Fig-7]. Based on the aforementioned clinical history, and histological and laboratory findings, a differential diagnosis of Subacute Cutaneous Lupus Erythematosus (SCLE) was proposed, but with the existence of EM-like lesions. Thus, given the simultaneous presence of both LE and EM-like lesions, along with a supporting ANA profile, a diagnosis of Rowell syndrome was ascertained.



[Table/Fig-7]: Lesion showing necrotic keratinocytes, basal cell degeneration, pigment incontinence, dilated capillaries with perivascular lymphocytic infiltration (Haematoxylin & Eosin staining, 40X).

The patient was started on sunscreen, topical mid-potent corticosteroid (mometasone furoate 0.1%) cream, once-daily application, intravenous dexamethasone 4 mg twice a day for a week, and oral hydroxychloroquine 200 mg twice a day. The patient showed gradual resolution of skin lesions and the steroid was then gradually tapered to 30 mg of oral prednisolone daily for one week, followed by 20 mg of oral prednisolone daily for one week and a maintenance dose of oral prednisolone 15 mg once daily was achieved. The patient was asked to continue hydroxychloroquine 200 mg twice daily along with topical agents. The patient is currently on follow-up on a fortnightly basis and is responding to treatment [Table/Fig-8].



[Table/Fig-8]: Follow-up image of patient 10 weeks after initiation of therapy showing improvement clinically and dermoscopically.

DISCUSSION

Rowell Syndrome is an uncommon condition characterised by the presence both LE and EM-like lesions in patients with a typical immunologic pattern. EM is characterised by the presence of target lesions which derive their name from the appearance of rings of different colours that is most commonly seen on the hands and extensor aspect of limbs. Classic EM occurs due to certain infectious causes like *Mycoplasma pneumoniae*, drugs like non steroidal anti-inflammatory drugs, antibiotics, anticonvulsants, or malignancies [1]. LE is an autoimmune condition with variable presentation. Lesions of SCLE are widespread, asymmetrical in distribution without any scarring. It frequently involves the extensor surface of the arms, upper chest, upper back, shoulders and neck, where systemic involvement is usually absent but anti-Ro antibodies are frequently present [2]. Psoriasiform and annular polycyclic are the predominant forms known, but other presentations have also been described. The histological similarity in both these conditions is the presence of lymphocytic exocytosis into the epidermis, inflammation at the dermoepidermal junction, vacuolar degeneration of basal epidermal cell layer along with oedema of the upper dermis [3].

In concordance with the case reports by Yachoui R and Cronin PM, [2] and Gallo L et al., [4], the index patient also had characteristic features – middle-aged woman with clinical features suggestive of both SCLE and EM. Till date, very few cases of Rowell syndrome have been reported. Although the disease is common in young adults, and the median age reported is 32, with instances of paediatric and elderly cases [4]. It was first identified by Rowell in 1963 in four female patients with DLE, speckled immunofluorescence pattern, who were positive for Rheumatoid factor and anti-Ro/La antibodies.

The definition of RS remains a matter of debate, as a few scholars have opined that LE associated with EM may be an overlapping syndrome, a subtype of LE or merely a co-incidence [5]. There was no identifiable precipitating factor for EM in the index patient.

The criteria by Zeitouni NC et al., to diagnose Rowell syndrome comprise major as well as minor criteria [6]. Major criteria include SLE, DLE, or SCLE; EM-like lesions; and a speckled pattern of ANA positivity. Minor criteria consist of anti-Ro/anti-La antibody, a positive RF and chilblains [7]. To diagnose RS, all three major criteria and atleast one minor criterion must be present [2]. In this case, all three major criteria along and one minor criterion (positive anti-Ro antibody) were fulfilled, which clinched the diagnosis [Table/Fig-9].

Criteria for the diagnosis of Rowell syndrome [6]	Present case
Major criteria:	
LE, DLE, or SCLE EM-likelesions (with/without involvement of mucous membranes) Speckled pattern of ANA	Present Present Present
Minor criteria:	
Chilblains Anti-Ro/SSA or anti-La/SSB Positive rheumatoid factor	Absent Present Absent

[Table/Fig-9]: This table compares the findings of the index patient against the Zeitouni NC et al., criteria [6].

Speckled ANA is considered to be a consistent finding of Rowell syndrome, which was positive in the index patient. Both LE and RS follow a similar line of management. Patients with Rowell syndrome generally have a favourable prognosis and show complete resolution of lesions within one year of starting therapy [7]. Antimalarials, steroids, dapsone, azathioprine, and cyclosporine have shown to be beneficial [8]. The index patient responded well to intravenous dexamethasone 4 mg twice a day and oral hydroxychloroquine 200 mg twice a day.

CONCLUSION(S)

SCLE and Rowell syndrome have overlapping clinical, histological, and immunological findings. Index case also highlights the features shared by SCLE and EM, often making it difficult to diagnose. There have been very few reports of Rowell syndrome in literature and not much is known about this rare entity. Hence, this case has been reported to highlight the various features of Rowell syndrome and to bring into question of whether this condition is a possible designation of subacute lupus erythematosus or a true entity on its own.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: May 28, 2022
- Manual Googling: Jul 14, 2022
- iThenticate Software: Aug 30, 2022 (8%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **May 17, 2022**
Date of Peer Review: **Jun 17, 2022**
Date of Acceptance: **Jul 21, 2022**
Date of Publishing: **Sep 01, 2022**