Case Report

A Case of Rowell Syndrome in a Filipino Adolescent: A Diagnostic and Therapeutic Challenge

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Abstract

In 1963, Rowell *et al*. described a syndrome combining lupus erythematosus (LE) and erythema multiforme (EM)-like lesions. In this report, we present a 15-year-old female who presented with both systemic LE and EM-like skin lesions meeting all of the major and one of the minor criteria for a diagnosis of Rowell syndrome. Her condition improved with administration of systemic and topical corticosteroids, and hydroxychloroquine. Rowell Syndrome, a rare entity, is often debated as a coincidental overlap of other conditions according to Bonciolini, *et al*.[1] In light of Rowell Syndrome's infrequency and the paucity of available literature, we emphasize the clinical significance of recognizing this challenging condition.

Keywords: Erythema multiforme, lupus erythematosus, Rowell syndrome

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Introduction

Rowell syndrome as a distinct entity was formally defined in 1963 by Rowell *et al*., who identified unique serological markers in patients with this combined condition.[2] Consequently, there was a revision of the diagnostic criteria in 2000 by Zeitouni *et al*. to address these inconsistencies. As redefined, the major criteria encompass the simultaneous presence of lupus erythematosus (LE), erythema multiforme (EM)-like lesions (with or without mucosal involvement), and the speckled pattern in antinuclear antibody (ANA) tests. Minor criteria include the clinical observation of chilblains, the presence of positive anti-Ro or anti-La antibodies, and a positive RF result.[3,4]

We report the case of a 15-year-old female diagnosed as LE, with EM-like lesions, chilblains, and positive ANA with a speckled pattern fulfilling three major + 1 minor criteria consistent with the diagnosis of Rowell syndrome.

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Case Report

A 15-year-old Filipino female presented with multiple erythematous papules and plaques on the face described as nonpruritic and nontender. The lesions gradually increased in size and number, affecting the trunk, upper, and lower extremities. The patient also presented with oral ulcers, easy fatigability, and episodes of epistaxis. There was no history of upper respiratory infection, fever, or recent drug intake.

Physical examination showed generalized erythematous targetoid plaques with dusky central discs, some with central bullae, pale peripheral ring with erythematous halo

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and erosions, crusts, and areas of desquamation. There were chilblain-like lesions on both palms and soles and alopecic patches on the bilateral temporal areas [Figure 1]. Diagnostic examinations, such as ANA and anti-dsDNA, were requested, which revealed positive results. A 4-mm skin punch biopsy revealed vacuolar interface changes with few necrotic keratinocytes, superficial to mid perivascular infiltrate composed of lymphocytes, neutrophils, and melanophages. Few infiltrates are seen around adnexal structures [Figure 2]. These findings are suggestive of the diagnosis of erythema multiforme (EM). Alcian blue stain revealed increased mucin deposition in the dermis, and PAS staining showed thickened basement membrane consistent with LE. Direct immunofluorescence was not performed due to resource-limited settings. Following comanagement with rheumatology, hydroxychloroquine 100 mg/tablet thrice daily, hydrocortisone 100 mg TIV every 8 h (4 mkd), clobetasol propionate 0.05% lotion twice daily, antihistamines, and emollients led to marked lesion improvement within 2 weeks. Residual hyperpigmentations were noted during outpatient follow-up, with no recurrence thereafter.

Discussion

Rowell syndrome is an autoimmune condition characterized by a triad of systemic LE (SLE), EM, and a positive rheumatoid factor. This syndrome exhibits skin manifestations resembling EM, including target-like lesions, alongside the systemic effects observed in SLE. The precise cause remains uncertain, and there are not more than 71 cases that have been reported in the international literature[5] of which 18 cases are from India.[6] No formal publications documenting this disease entity exist within the local context.

Diagnosis can be challenging, and treatment entails managing the underlying SLE while addressing the associated skin symptoms. The existence of RS is still debated whether as a separate distinct clinical entity or an independent chronic cutaneous LE (CLE) subtype.[7,8] Given its infrequency, additional research will enhance our understanding of the pathogenesis and establish optimal therapeutic approaches.[3]

The Systemic Lupus International Collaborating Clinics criteria provide a framework aiding clinicians in identifying the systemic involvement associated with Rowell syndrome.[9]

Following the criteria set by Zeitouni *et al*., the patient fit the three major + 1 minor criteria to diagnose a case of Rowell syndrome.[3] According to Torchia *et al*., there is no significant histological differences between CLE and EM

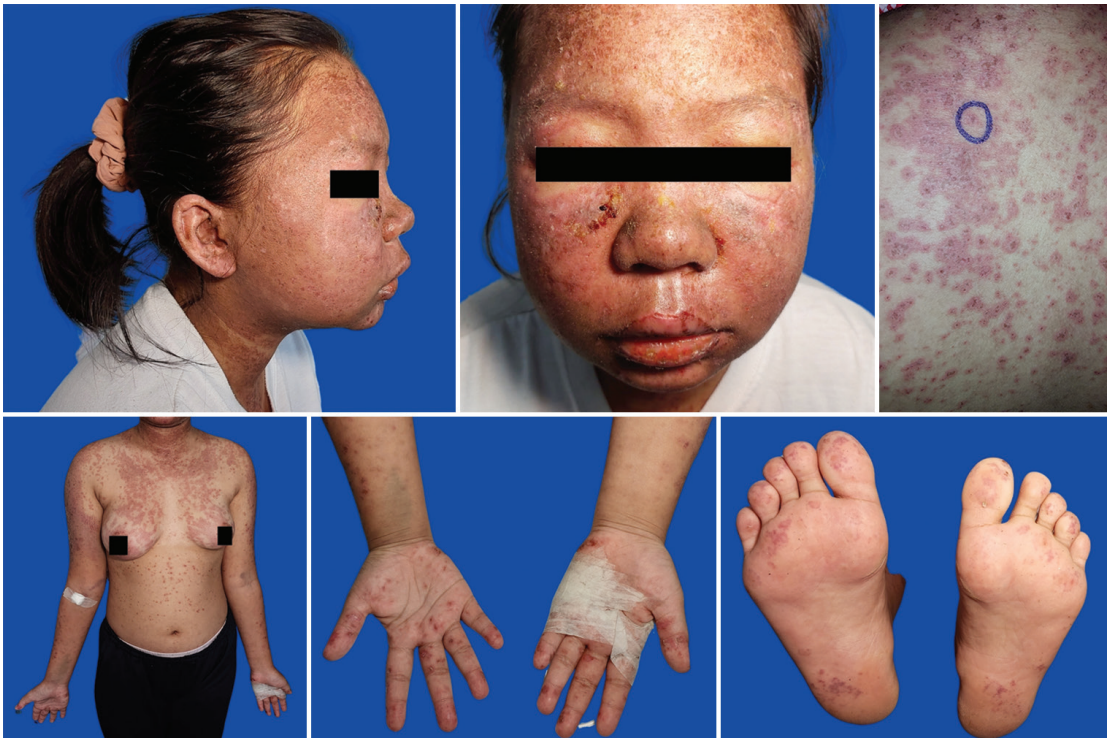

Figure 1: Physical examination showed generalized erythematous targetoid plaques with dusky central discs, some with central bullae, pale peripheral ring with erythematous halo and erosions, crusts, and areas of desquamation. There were chilblain-like lesions on both palms and soles and alopecic patches on the bilateral temporal areas

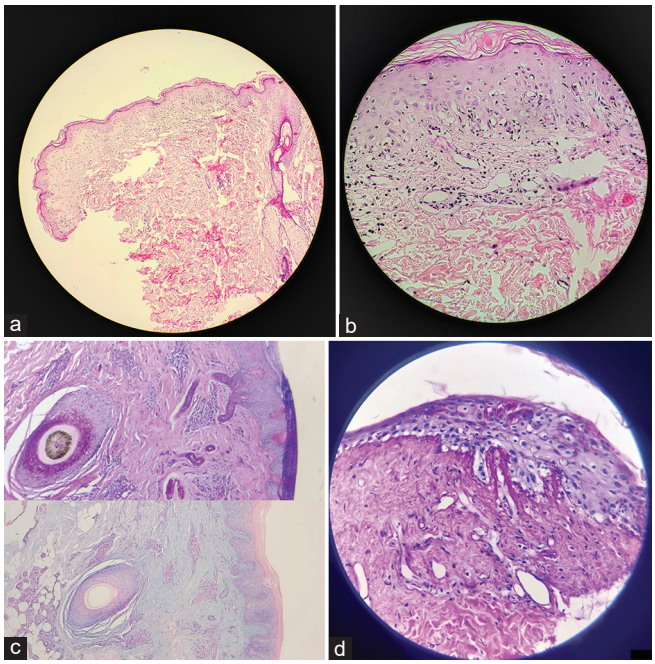

Figure 2: Following views show (a) ×10 magnification and (b) ×40 magnification. Histopathological examination revealed vacuolar interface changes with few necrotic keratinocytes, superficial to mid perivascular infiltrate composed of lymphocytes, neutrophils, and melanophages, few infiltrates seen around adnexal structures. (c) Alcian blue stain was performed and revealed increased mucin deposition in the dermis, (d) PAS stain showed thickened basement membrane

lesions, and the presence of necrotic keratinocytes is not specific for EM, as it may also be found in subacute CLE lesions.[7] Early lesions of an annular-polycyclic pattern of LE may resemble EM.[8] Such overlap between LE and EM has been reported by Mendonca, in which repeated biopsies of lesions previously read as EM showed LE instead.[10] Unifying histologic features of Rowell syndrome and lupus are parameters such as periadnexal lymphocytic infiltrates, absence of dermal eosinophils, and CD123 positivity of ≥10% of the inflammatory infiltrate.[6]

Immunosuppressive agents such as corticosteroids, antimalarials, and immunomodulatory medications control the autoimmune response and reduce inflammation.[11] Given resource constraints, the patient was treated with steroids and hydroxychloroquine. Providing supportive care is also essential in the management.[11] Patients with Rowell syndrome face complications that may involve vital organs, including the kidneys, heart, and lungs, necessitating close monitoring.[12] Skin lesions in Rowell syndrome can be prone to infection, demanding careful wound care and infection prevention measures.[13] Collaborative care involving rheumatologists, dermatologists, and other specialists is essential to promptly mitigate potential complications.[14]

Conclusion

This case highlights the importance of meticulous clinical and serological evaluation in patients with overlapping LE and EM-like lesions, facilitating timely diagnosis, appropriate management, and improved patient outcomes. Increased recognition of this condition can lead to better understanding of its prevalence and clinical spectrum within diverse populations and improve patient care. Furthermore, given the paucity of documented cases within the Filipino population and the potential for severe systemic complications, this report is a vital contribution to local and international medical literature.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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