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Rowell syndrome with recurrence from photoexacerbation: A case report

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Abstract

Rowell syndrome is a controversial entity composed of erythema multiforme-like lesions coexisting with lupus erythematosus. We describe a case of a 61-year-old male with a history of systemic lupus erythematosus who presented with photoexacerbated flaccid bullae and erosive plaques after repetitive sun exposure. Based on his clinical history, biopsy, and laboratory findings, he fulfilled diagnostic criteria for Rowell syndrome as described by Zeitouni et al. With oral prednisone, hydroxychloroquine, mycophenolate mofetil, and local wound care with petrolatum, the patient's number of lesions decreased, as well as his pain and tenderness. He subsequently did not develop any new erosions. This case highlights the diagnostic criteria of this hybrid clinicopathological syndrome and its nature of photosensitivity.

Keywords

Rowell syndrome, lupus erythematosus, erythema multiforme

Introduction

Rowell syndrome (RS) is characterized by manifestations of erythema multiforme (EM)-like lesions in a patient with systemic lupus erythematosus (SLE), discoid lupus erythematosus (DLE), or subacute cutaneous lupus erythematosus (SCLE) with associated serologic abnormalities. The syndrome was first described as a distinct entity by Rowell et al.[1] in 1963, but the simultaneous existence of lupus erythematosus (LE) with EM-like lesions was first described in the literature by Scholtz[2] in 1922. Since 2011, there have been around 80 reported cases in the literature.[3] Although the age range of reported cases of RS is from the second to tenth decades of life,[4,5] the disease seems to exhibit a predilection for middle-aged women.[4] We describe a unique case of RS due to its occurrence in a middle-aged man and its recurrence initiated by ultraviolet (UV) exposure.

Case report

A 61-year-old Hispanic male with a medical history of SLE was admitted for a 5-day duration of a vesiculobullous eruption following 5–6 h of sun exposure. He reported mild skin tenderness exacerbated by movement. On examination, there were over 80 diffuse, heme-crusted erosions with scattered flaccid bullae as well as a few targetoid lesions across his face, chest, and back in a photodistributed pattern with mucosal sparing (Figure 1(a) and (b)). Scattered depigmented patches with perifollicular repigmentation were also noted on his trunk and extremities. Laboratory studies revealed a positive qualitative anti-nuclear antibody (ANA) in a speckled pattern, anti-SS-A (Ro), and anti-SS-B (La). Two punch biopsies were taken of his right medial and lateral chest. The histology revealed vacuolar alteration of the basal layer of the epidermis with a mild superficial perivascular and slightly band-like infiltrate of lymphocytes. The overlying epidermis displayed prominent dyskeratosis at all levels of the epidermis. The deeper dermis displayed a mild increase in mucin content and mild melanin incontinence (Figure 2(a) and (b)). His pathologic diagnosis was interface dermatitis with dyskeratosis consistent with EM. He was started on prednisone 20 mg twice a day as well as topical petrolatum two to three times daily. He was also counseled on the importance of photoprotection including barriers to UV radiation. Our patient was diagnosed with RS based on his medical history of SLE, histopathological diagnosis consistent with EM, and seropositivity for speckled ANA, anti-Ro, and anti-La.

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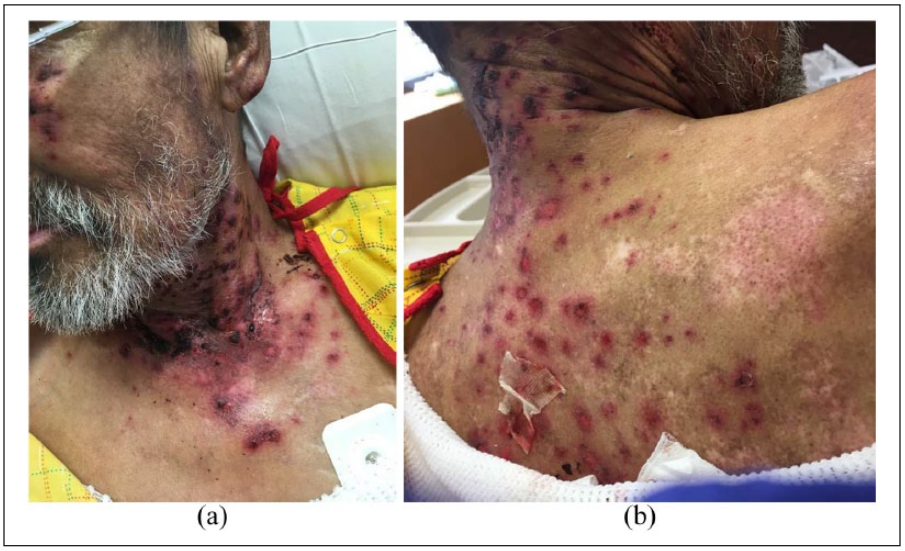


Figure 1. (a) Photodistributed pattern of heme-crusted erosions with scattered flaccid bullae as well as a few targetoid lesions on neck and chest and (b) similar lesions on back.

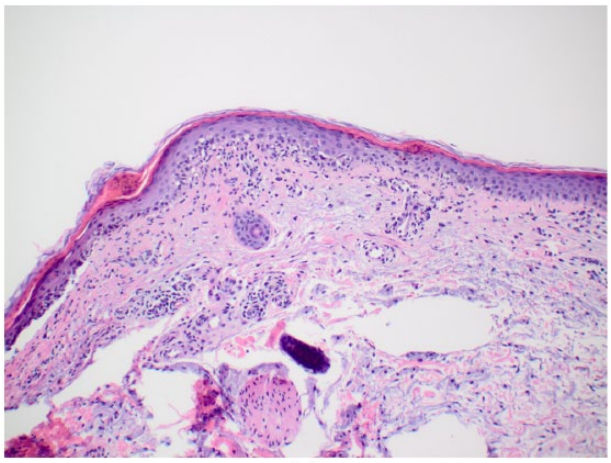
Although compliant with his prednisone regimen, the patient was readmitted after 1 month for a similar cutaneous manifestation with burning and pruritic sensations. The patient had been avoiding direct sun exposure but had spent several hours in the shade thinking it would not be problematic. He had new erosions on his face and left shin but no new lesions on the chest or back. He was started on hydroxychloroquine 200 mg twice a day and mycophenolate mofetil 500 mg twice a day. The patient's prednisone dose was increased to 30 mg twice a day, with plans to taper in an outpatient setting. Petrolatum was applied to the affected areas two to three times daily while hospitalized. He was again advised to avoid UV exposure, found even in indirect sunlight. His symptoms and lesions improved continuously during his hospital course, and, by the time of discharge, he had less than 30 lesions. Unfortunately, he was lost to follow-up after discharge.

Discussion

Rowell et al.[1] first proposed the diagnostic criteria of RS in 1963, which Zeitouni et al.[6] subsequently modified and expanded in 2000. The current standard requires all three major and one of four minor criteria for diagnosis. The major criteria include the following: the presence of systemic, discoid, or subacute LE, EM-like lesions in the absence of any known precipitating factors with or without mucous membrane involvement, and a speckled pattern of ANA. The minor criteria include the following: chilblains, anti-Ro and/or anti-La antibodies, and rheumatoid factor (RF). Most of the cases of RS in the literature do not adhere to Rowell's original criteria.[7] A positive speckled ANA has been the most consistent finding in the reported cases, while anti-Ro and RF have not been as well preserved. Because of these observations, Zeitouni et al. proposed the addition of anti-Ro to the diagnostic criteria and removal of the RF requirement for diagnosis.

There is controversy surrounding the viability of RS as a distinct diagnosis. Shteyngarts et al.[8] suggested that the coexistence of LE with EM might be coincidental, and the combination imparts no unique characteristics. In their review, they argue that this new entity with features of both LE and EM did not change the course, therapy, or prognosis of either disease. Yachoui and Cronin[9] described RS as merely a subset of SCLE with targetoid lesions due to their clinical and histological overlap. EM itself is classically a brief and self-limited reaction to a drug or pathogen that is characterized by acral and/or mucosal targetoid lesions with a central dusky necrotic zone, a middle skin-colored zone, and a peripheral erythematous zone.[10,11] It is rarely recurrent or persistent unless associated with inflammatory bowel disease or malignancy.[11] EM patients do not generally have the positive serologic markers included in the RS criteria. In contrast, RS is distinguished by idiopathic, recurrent, and diffuse EM-like lesions with positive serology and clinical evidence suggestive of a systemic process.

Abnormal reactivity to UV light is an important factor in the pathogenesis of both cutaneous and systemic manifestations of LE.[12] UV radiation induces keratinocyte apoptosis, which has been identified as a contributing factor to the initiation of the autoimmune reaction cascade.[12] In primary and UV-induced skin lesions of LE, there is histological evidence of a significantly increased number of apoptotic keratinocytes compared to normal skin.[12] The accumulation of these immunogenic cells due to a defect in clearance may trigger the autoimmune response.[12] As evidenced in our patient's



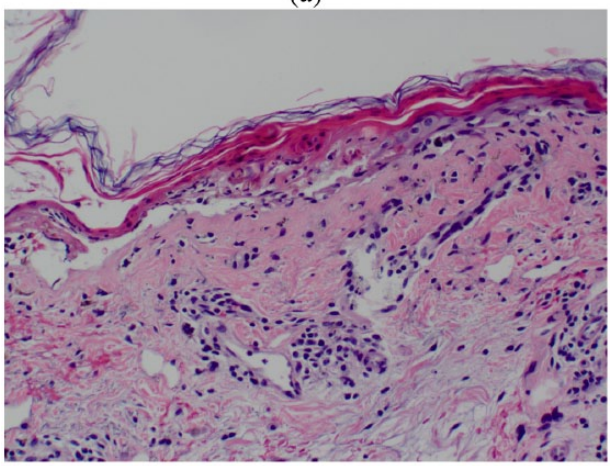


Figure 2. (a) Vacuolar alteration of the basal layer of the epidermis with a mild superficial perivascular and slightly band-like infiltrate of lymphocytes. (b) Prominent dyskeratosis at all levels of the epidermis with a mild increase in mucin content and mild melanin incontinence of deeper dermis.
Hematoxylin and eosin stain, original magnifications at (a) 100× and (b) 200×.

clinical history, RS may be initiated and exacerbated by UV radiation. In summary, we report a case of RS that manifested and recurred after prolonged UV exposure, thus highlighting its photosensitive nature and the importance of photoprotection in the management of RS.

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Informed consent

Written informed consent for patient information and images to be published was provided by the patient.

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