Introduction

Rowell Syndrome was originally described in 1963 as an association or overlap between cutaneous lupus erythematosus and erythema multiforme in the same individual with associated serum immunologic abnormalities. Since its initial description, the syndrome has been controversial as sources question whether it is truly a unique entity or simply part of the spectrum of clinical manifestations of cutaneous lupus erythematosus. We report a case of new-onset SCLE presenting with EM-like and SJS/TEN-like lesions, an entity some describe as Rowell Syndrome.

Case

A 44-year-old female with no significant past medical history presented to our facility in August of 2015 complaining of a rash of 5-6 weeks duration. She reported the rash started on bilateral thighs and subsequently spread to her back, arms, chest and face. She had visited the ER before coming to our facility and was told to follow up with her outpatient dermatologist. He then referred her to our center for further workup and treatment.

Upon arrival the patient exhibited multiple scattered erythematous annular macules and patches with collarette of scale and areas of denudation on bilateral upper and lower extremities as well as abdomen and back (Fig 1). Her chest and face revealed areas of desquamation, her lips were scaly and crusted (Fig 2). She did not display any intraoral or ocular involvement; she did not have any other mucosal lesions. The lesions were painful (7/10 on pain scale) particularly in areas of denudation. On review of systems, she admitted to decreased appetite, cyclic fevers (Tmax 102), and chills.

Antibody work-up revealed positive ANA (homogenous pattern, 1:1280), positive dsDNA, negative Scl-70, negative anticardiolipin, decreased total complement/C3/C4, anti-Smith, and SS-Ro/La negative. Three punch biopsy specimens were obtained; two were submitted for immunofluorescence and one for routine H&E.

Clinical Photos

Fig 1. Extensive area of denudation seen on the back

Fig 2. Lips and neck exhibiting areas of desquamation and crusting

Pathology

Fig 3. Interface dermatitis, vacuolar-type with hyperkeratosis, mild epidermal atrophy and dermal mucin deposition consistent with lupus erythematosus

Direct Immunofluorescence was positive for IgG colloid bodies and C3 granular band at dermal epidermal junction and focal complement deposition in the blood vessels.

Frozen sections revealed full thickness epidermal necrosis consistent with cutaneous lupus erythematosus with a toxic epidermal necrosis like clinical presentation.

Treatment

Our patient was started on IV solumedrol and hydroxychloroquine 200mg PO BID. She was treated topically with hydrocortisone 2.5% ointment and petrolatum jelly twice daily. Her skin lesions improved throughout her stay and she was discharged one week after admission.

Discussion

Many cases of Rowell Syndrome have been reported since Rowell’s original work in 1963. In reviewing the literature, it is evident that a single set of diagnostic criteria does not exist and, moreover, Rowell’s original criteria are not well preserved.

In 2012 Torchia et al. conducted an extensive literature review and suggested new major and minor criteria: Major criteria: chronic cutaneous lupus, EM-like lesions, at least one positivity among speckled ANA, anti-Ro/La antibodies, and negative DIF on EM-like lesions. Minor criteria: absence of triggers, absence of typical EM location, and presence of at least one additional ARA criterion for diagnosis of SLE. They require all 4 major and at least one minor criterion for the diagnosis of Rowell syndrome.

Currently no therapeutic standard for Rowell Syndrome exists. Most patients are treated with therapies directed at their underlying LE. These treatments include corticosteroids, methotrexate, dapsone, hydroxychloroquine, and azathioprine.

While our patient did not meet the proposed diagnostic criteria, we believe this to be a unique and interesting presentation of SCLE that serves to further the discussion regarding the pathogenesis and significance of Rowells.

References


A Rare Case of SCLE presenting with EM-like and SJS/TEN-like lesions: A Review of Rowell Syndrome

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