Rowell Syndrome: A Rare Presentation of Systemic Erythematous Lupus

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Abstract
A 27-year-old woman presented to hospital with a 4-week history of a progressive, painful, and pruritic erythematous-violaceous mucocutaneous rash involving the face, lips, genitals, palms, and feet. She was previously healthy and reported no medications around the time of rash onset. She denied any preceding viral or bacterial infections. No blisters or bullae were seen on exam. Targetoid lesions were identified on her palms and erythema multiforme (EM) major was diagnosed.

Résumé
Une femme de 27 ans s’est présentée à l’hôpital après quatre semaines d’une éruption cutanéo-muqueuse érythémateuse-violacée progressive, douloureuse et prurigineuse touchant le visage, les lèvres, les organes génitaux, la paume des mains et les pieds. Elle était auparavant en bonne santé et n’avait pas pris de médicaments au moment de l’apparition de l’éruption. Elle a nié toute infection virale ou bactérienne antérieure. L’examen n’a révélé aucune cloque ou bulle. Des lésions ciblées ont été identifiées sur ses paumes et un érythème polymorphe (EM) majeur a été diagnostiqué.

Keywords: Rowell syndrome; lupus; erythema multiforme

Introduction
Laboratory investigations revealed bicytopenia (hemoglobin 103 g/L, WBC 1.6E9/L) and hypocomplementemia (C3 0.26 g/L, C4 0.05 g/L). ANA was positive and ENA-profile demonstrated positive anti-chromatin (>8.0), anti-ribosomal P (>8.0), anti-SSA/Ro (5.2), anti-Sm (>8.0) and anti-RNP (>8.0). Renal and cardiac function were normal. Infectious work-up for HIV, hepatitis A, B and C, enterovirus PCR, CMV, syphilis, HSV1/2, VZV and parvovirus B19 were negative. EBV IgG was positive, indicative of past infection. Based on positive serology, cytopenias, and clinical findings, systemic erythematous lupus (SLE) was diagnosed. Biopsy of the skin lesions demonstrated chronic inflammation and patchy epithelial necrosis involving the epidermis, consistent with interface dermatitis. Given the presence of SLE and EM, a diagnosis of Rowell syndrome was made. She was started on daily prednisone with taper and had subsequent...
improvement in her lesions. Hydroxychloroquine and mycophenolate mofetil were added prior to discharge.

Rowell syndrome (RS) is a rare entity characterized by EM-like lesions in those with lupus erythematosus, a positive ANA in a speckled pattern, and positive anti-SSA/Ro or anti-SSB/La.¹ A majority of cases occur in middle-aged women with a pre-existing diagnosis of lupus, unlike this case where clinical manifestations presented simultaneously.² RS has a similar prognosis and management to SLE.² While debate exists on whether RS is a separate form of cutaneous lupus versus an overlap syndrome, this case highlights why clinicians should consider lupus on the differential for EM when an underlying infectious, drug or malignant trigger does not exist.³ The authors confirm that informed, written patient consent was obtained for this work (Figure 1).

Consent

Informed, written patient consent was obtained for this work.

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Conflict of interest

None of the authors have any conflicts of interest to declare.

Authorship statement

All authors contributed to the preparation of this manuscript.

References