

Rowell Syndrome with Lupus Podocytopathy: A Diagnostic Challenge

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Abstract

Rowell syndrome is a distinct subtype of systemic lupus erythematosus presenting with features of lupus as well as erythema multiforme. It has been well characterized by a different set of systemic signs and symptoms and a unique serological profile. Lupus podocytopathy (LP) is a rare pattern of lupus nephritis with a slightly different clinical and serological profile, natural course of the disease, as well as prognosis. A case of Rowell syndrome is presented, which was later diagnosed with LP on the basis of global effacement of podocytes on electron microscopy.

Keywords: Lupus nephritis, lupus podocytopathy, male, Rowell syndrome

Introduction

Rowell syndrome is characterized by coexistent systemic lupus erythematosus, erythema multiforme, and specific pattern of immunological findings consisting of positive speckled antinuclear antibody, anti-Ro or anti-La antibodies, and rheumatoid factor.¹ In lupus, nephrotic-range proteinuria heralds the findings of lupus nephritis of class III and above. However, in rare cases, patients with massive proteinuria patients on renal biopsy may have normal glomeruli with or without mesangial proliferation.² Further workup by electron microscopy is necessary to look for diffuse foot process effacement for diagnosis of lupus podocytopathy (LP).

Case Presentation

A 20-year-old male with no known comorbidities presented with complaints of sudden-onset erythematous rash on his face along with multiple red-raised lesions over his chest, abdomen, and lower limbs of 2 weeks duration. Ten days after the development of the rash, he developed high-grade intermittent fever associated with chills and joint pains involving the wrists, elbows, and knees. Due to positive serology for dengue and leptospirosis with raised procalcitonin, progressive transaminitis, and leukopenia, he was treated as such with broad-spectrum antibiotics and supportive therapy. However, due to the progressive decline in his general condition, he was transferred to the tertiary care hospital with a diagnosis of sepsis. On initial evaluation, he had fever (101°F), tachycardia, conjunctival congestion, and non-tender, non-matted submandibular, axillary, and inguinal lymphadenopathy. Systemic examination was within normal limits. Dermatological examination revealed 2 distinct types of lesions. Firstly, there was involvement of the face, predominantly the malar region and the bridge of the nose in the form of well-defined erythematous plaque (Figure 1A). Erosions with crusting were noted on the lower lip. Secondly, multiple targetoid erythema multiforme (EM) lesions were observed with symmetrical distribution over forehead, chest, abdomen, and extremities, including the palms and soles (Figure 1B). Investigations revealed hemoglobin levels of 11.9 gm/dL with platelets at 320 000 per mm³. There was leukopenia with a total leucocyte count of 2200 per mm³. A peripheral blood smear was consistent with microscopic hypochromic anemia with leukopenia. Serum creatinine was raised at 6.2 mg/dL, with massive proteinuria of 5.4 gm/24 hours; Serum Glutamic-Oxaloacetic Transaminase(SGOT)-90 IU/mL (normal, 15-37 IU/L) and Serum Glutamic Pyruvic Transaminase(SGPT)-71 IU/mL (normal, 16-63 IU/L). Erythrocyte sedimentation rate, C-Reactive protein and procalcitonin levels were raised. Routine urine examination showed the presence of occasional red blood cells (RBCs) and white blood cells along with a few epithelial cells and casts with *Candida* spp. His tropical fever screen and cultures from blood, urine, and sputum were negative. He was clinically diagnosed as a case of Rowell syndrome. Skin biopsies from the erythematous plaque on the face and the active border of EM on the foot showed epidermal atrophy with basal cell vacuolation, mild-to-moderate lymphomononuclear inflammatory infiltrate with dermal edema and extravasation of RBCs in the dermis. Periodic

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Figure 1. A: Lupus erythematosus skin lesions on the malar area and nose with erythema multiforme lesions on the forehead. The upper lip is spared. B: Erythema multiforme lesions on the sole of foot.

Acid-Schiff stain highlighted an increase in dermal mucin (Figure 2A). Immunological testing revealed antinuclear antibody (ANA) positivity by indirect immunofluorescence (IIF) at 1:320. The rheumatoid factor was positive at 32 IU/mL (normal range - less than 20 IU/mL), and further testing revealed positive U1nRNP (0.34), anti-Sm (0.16), and anti-dsDNA

(0.24) antibodies by Enzyme-Linked immunosorbent assay with low C3 (26 IU/mL) and C4 (8 IU/mL) levels (normal range C3 - 88-201 IU/mL; and C4 - 15-45 IU/mL). Anti-phospholipid antibody profile was negative. A kidney biopsy was performed due to high suspicion of lupus nephritis, which showed tubular atrophy accompanied interstitial fibrosis in <5%

of cortical parenchyma without interstitial inflammation suggestive of diffuse podocytopathy (Figure 2B). Immunofluorescence on kidney biopsy showed trace deposits of IgG in the glomeruli with negative IgA, IgM, C3, C1q, kappa, and lambda (Figure 3A). To further confirm the findings of light microscopy, the renal tissue was subjected to electron microscopy, which showed diffuse effacement of foot processes diagnostic of LP (Figure 3B). On the basis of clinical, immunological, and histopathological findings, he was diagnosed as a case of Rowell syndrome and was started on intravenous immunoglobulin 2 g/kg in a divided dose over 5 days, tablet prednisolone 60 mg once a day, tablet hydroxychloroquine 200 mg twice a day, topical mometasone cream, and broad-spectrum sunscreen. There was a dramatic response, with the patient turning afebrile within 48 hours. Leukopenia and transaminitis settled down over the next 10 days. At discharge, serum creatinine returned to baseline, and urine protein creatinine ratio decreased to 12 mg/mmol. At 1-month follow-up, the patient had near-complete resolution of skin lesions and normal liver and renal function. The patient was started simultaneously on tablet mycophenolate mofetil 1 gram twice-daily and is presently under clinical remission. Prednisolone was tapered by 10 mg every 15 days till 20 mg and then by 5 mg every month till 5 mg once-daily maintenance dosing.

Main Points

- Rowell syndrome is a distinct subtype of systemic lupus erythematosus presenting with features of lupus as well as erythema multiforme with target lesions on acral areas.
- Rowell syndrome is characterized by positive speckled antinuclear antibody, anti-Ro or anti-La antibodies, and rheumatoid factor.
- Rarely in lupus, the histopathological findings on renal biopsy may be scant and may not correlate with nephrotic-range proteinuria.
- In such cases, further workup by electron microscopy is necessary to look for diffuse foot process effacement for diagnosis of lupus podocytopathy.
- Lupus podocytopathy is a distinct clinicopathological entity with 2 subtypes, namely minimal change disease and focal segmental glomerulosclerosis, with the latter having a worse prognosis.

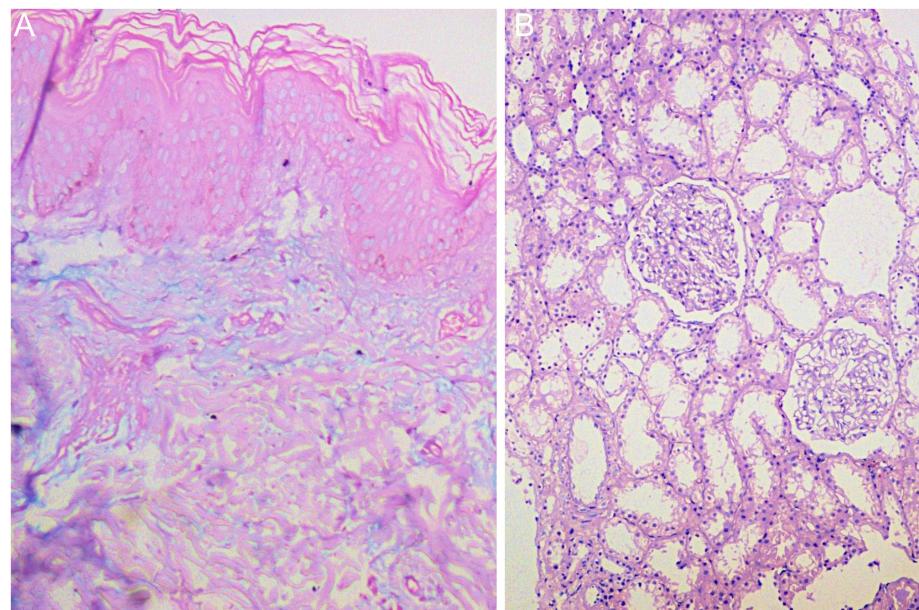


Figure 2. A: Histopathology of skin biopsy showing focal mucin deposition (bluish stained areas) in dermis on PAS staining (PAS; x40). B: Histopathological Examination(HPE) of kidney biopsy showing tubular atrophy accompanied interstitial fibrosis in <5% of cortical parenchyma without interstitial inflammation. There was a noticeable absence of mesangial proliferation in spite of significant proteinuria (Hematoxylin and Eosin(H&E); x40).

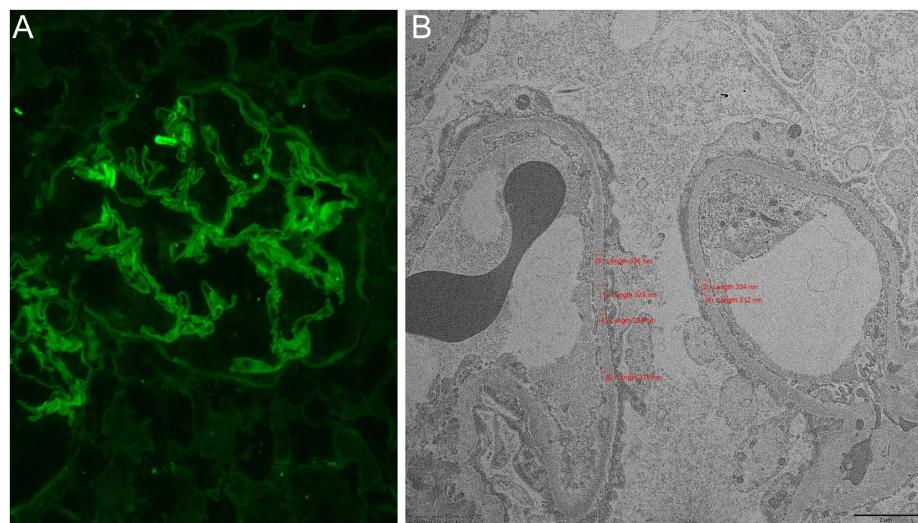


Figure 3. A: Immunofluorescence on kidney biopsy showing trace deposits of IgG in the glomeruli with negative Immunoglobulin A(IgA), Immunoglobulin M(IgM), Complement Component 3(C3), Complement Component 1q(C1q), kappa, and lambda (x100; Direct Immunofluorescence(DIF)). B: Electron microscopy on kidney biopsy showing global effacement of podocytes diagnostic of lupus podocytopathy.

Discussion

Rowell syndrome (RS) is an uncommon presentation of LE with erythema multiforme-like lesions associated with specific serological changes. The association between LE and EM was first observed by Shlotz in 1922.¹ In 1963, Neville Rowell et al¹ described this as a distinct entity while studying 120 patients with chronic cutaneous lupus erythematosus (CCLE). Revised diagnostic criteria were formulated by Zeitouni et al² in 2000. Major criteria include (i) LE: discoid lupus erythematosus or subacute cutaneous lupus erythematosus or systemic lupus erythematosus, (ii) erythema multiforme-like skin lesions with or without involvement of the mucous membranes, and (iii) speckled pattern ANA. Minor criteria include (i) chilblains, (ii) positive anti-Ro antibody or anti-La antibody, and (iii) positive rheumatoid factor. All 3 major criteria and at least 1 minor criterion are required for the diagnosis of RS. This case met the clinical diagnostic criteria but had a slightly unusual immunological profile. Skin biopsy

histopathology in this case showed a mixture of features of LE and EM in the form of basal cell vacuolation, dermal edema, extravasation of RBCs, and dermal mucin deposition with negative IIF. According to Torchia et al³ there are no significant histological differences between CCLE and EM lesions, and this was seen in the current case too. Kidney biopsy in the patient showed minimal change on light microscopy and IIF but showed diffuse podocytopathy (cells lining the glomeruli) on electron microscopy. Lupus podocytopathy is a newly emerging entity of non-immune complex-mediated lupus nephropathy being increasingly reported in the literature. The diagnostic criteria for LP are mentioned in Table 1.⁴ Lupus podocytopathy is sub-classified as minimal change disease (MCD) or focal segmental glomerulosclerosis types, with the latter having a worse outcome due to higher rates of acute kidney injury and hypertension at diagnosis, more severe tubulo-interstitial involvement on kidney biopsy, and low steroid responsiveness

as compared with MCD/mesangial proliferative lesions.⁵ The patient belonged to the MCD type. In LP, the proteinuria is out of proportion to the degree of mesangial deposits or hyperproliferation as seen by light microscopy, but podocytopathy observed under electron microscopy correlates with that of proteinuria. This suggests an alternative pathophysiological pathway that is unrelated to immune complex deposits as seen in classical SLE.⁶ In LP, it is believed that abnormal functioning of T cells leads to the production of cytokines and chemokines that cause podocyte effacement.⁷ Malar rash is the most common extra-renal clinical manifestation of SLE with LP and is seen in approximately half of the patients.⁵ Hematological abnormalities are observed frequently in LP, consisting of leukopenia (44%), anemia (26%), and thrombocytopenia (20%).⁶ Other manifestations of SLE, like oral ulcers and arthritis, have been reported along with the prevalence of anti-dsDNA antibody, anti-Smith antibody, and positive anticardiolipin antibody with varying frequencies in different case series. RS/LP should be managed similarly to that of SLE, often aggressively if the situation demands. Therapeutic options include oral or injectable steroids, antimalarials like hydroxychloroquine, and other immunosuppressive agents like azathioprine, cyclosporine, and cyclophosphamide. Response to treatment is unpredictable, and the patient may experience frequent recurrences.⁸ Most of the cases of RS or LP reported are middle-aged women. This case had many rare and unusual features in the form of fulminant picture in a young male mimicking sepsis and rapid progression to renal involvement with minimal change on histopathology and podocytopathy on electron microscopy. To the best of the authors' knowledge, this case is the first in the literature of LP presenting as Rowell's syndrome. Positive antigen/serology for dengue and leptospira led the clinicians astray before focus on the characteristic SLE and EM skin lesions co-existing led to the correct diagnosis. A high index of suspicion is required, especially in those with no prior diagnosis of LE, as early diagnosis and prompt aggressive treatment of RS may be required to prevent irreversible complications.

Data Availability Statement: The data that support the findings of this study are available on request from the corresponding author.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

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Table 1. Proposed Criteria for Diagnosis of Lupus Podocytopathy

Parameter	Features
Clinical	Diagnosis of SLE by ACR criteria, full nephrotic syndrome (i.e., nephrotic-range proteinuria, hypoalbuminemia, and edema).
Light microscopy	Normal glomeruli or FSGS, mesangial proliferation permitted, endocapillary proliferation, necrosis and/or crescents not permitted.
Immunofluorescence microscopy	Deposits absent or confined to mesangium.
Electron microscopy	Diffuse and severe foot process effacement (typically >70%), deposits absent or confined to mesangium.

ACR, American College of Rheumatology; FSGS, focal segmental glomerulosclerosis; SLE, systemic lupus erythematosus.

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