ORIGINAL RESEARCH PAPER
Volume - 15 | Issue - 05 | May - 2026 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

RENAL-LIMITED LUPUS NEPHRITIS PRESENTING AS ISOLATED NEPHROTIC SYNDROME WITH SEROLOGICAL DISCORDANCE: A DIAGNOSTIC CHALLENGE

Nephrology

Dr. Kruthi M Gowda Junior Resident, Department of General Medicine, BGS GIMS

Dr. Jalaja B Associate Professor, Department of General Medicine, BGS GIMS

ABSTRACT

Lupus nephritis (LN) is a severe manifestation of Systemic Lupus Erythematosus (SLE), usually associated with systemic clinical features and characteristic serological abnormalities such as anti–double-stranded DNA (anti-dsDNA) positivity and hypocomplementemia involving both C3 and C4. However, atypical presentations with isolated renal involvement and serological discordance may occur, posing a diagnostic challenge. We report a case of a 20-year-old female presenting with bilateral lower limb edema without any systemic features suggestive of SLE. Laboratory evaluation revealed nephrotic-range proteinuria with dyslipidemia and elevated erythrocyte sedimentation rate, while renal function was preserved. Complement analysis showed normal C3 with markedly reduced C4. Autoimmune workup demonstrated negative anti-dsDNA antibodies but strong positivity for anti-Smith (anti-Sm) and U1-snRNP antibodies. Renal biopsy revealed diffuse endocapillary hypercellularity with capillary wall thickening and double contour formation. Immunofluorescence demonstrated a characteristic "full-house" pattern with deposition of IgG, IgA, IgM, C3, and C1q. These findings were consistent with ISN/RPS Class IV-G(A) lupus nephritis with a superimposed Class V component. This case highlights a rare presentation of renal-limited lupus nephritis presenting as isolated nephrotic syndrome with serological discordance, emphasizing the indispensable role of renal biopsy in atypical presentations.

KEYWORDS

Lupus nephritis; Renal-limited lupus; Nephrotic syndrome; Serological discordance; Full-house immunofluorescence

CASE PRESENTATION

A 20-year-old female presented with bilateral lower limb edema of recent onset. There was no history of fever, photosensitivity, oral ulcers, joint pain, alopecia, skin rash, or other features suggestive of systemic autoimmune disease.

On examination, she had bilateral pitting pedal edema. Blood pressure was within normal limits. Systemic examination was unremarkable.

Laboratory investigations revealed nephrotic-range proteinuria (urine protein creatinine ratio of 4.2 g/g) with dyslipidemia (Total Cholesterol 249 mg/dL, Triglycerides 366 mg/dL, LDL 162 mg/dL). Urine analysis did not reveal any casts or red blood cells. Erythrocyte sedimentation rate was elevated (90 mm/hr), while C-reactive protein was normal. Liver function tests revealed hypoalbuminemia (serum albumin 2.2 g/dL). Complete blood count, renal function test and thyroid profile were within normal limits.

Complement levels showed preserved C3 (112 mg/dL) with markedly reduced C4 (<1 mg/dL). Autoimmune profile revealed negative anti-dsDNA antibodies, while anti-Smith (anti-Sm) and U1-snRNP antibodies were strongly positive. Other antibodies including SSA (Ro), SSB (La), and Scl-70 were negative.

Ultrasound abdomen revealed Grade 1 fatty liver and normal renal morphology.

Given unexplained nephrotic syndrome with serological discordance, a renal biopsy was performed. Light microscopy demonstrated diffuse endocapillary hypercellularity, capillary wall thickening, and double-contour formation. Immunofluorescence revealed a "full-house" pattern with granular deposition of IgG (3+), IgA (2+), IgM (2+), C3 (2+), and C1q (3+).

These findings were consistent with ISN/RPS Class IV-G(A) lupus nephritis with a superimposed membranous component (Class V). The activity index was 8/24 and chronicity index was 0/12.

The patient was initiated on corticosteroids, immunosuppressive therapy (Mycophenolate Mofetil) and an Angiotensin Receptor Blocker as per standard lupus nephritis protocols. At follow-up, the patient showed clinical improvement with reduction in edema and proteinuria.

DISCUSSION

Lupus nephritis is typically associated with systemic manifestations of SLE and characteristic serological markers, particularly anti-dsDNA antibodies and hypocomplementemia involving both C3 and C4. Renal-limited lupus nephritis without systemic features is rare, with few cases reported in the literature.

In such cases, serological discordance may further complicate diagnosis. The absence of anti-dsDNA antibodies, as observed in this patient, does not exclude lupus nephritis. Anti-Smith antibodies, although less sensitive, are highly specific for SLE and may provide an important diagnostic clue in such scenarios.

Selective reduction of C4 with preserved C3 suggests activation of the classical complement pathway, which is consistent with immune complex–mediated diseases such as lupus nephritis. This pattern, although less common, has been reported in atypical presentations. The coexistence of anti-Sm positivity with isolated C4 consumption in the absence of anti-dsDNA antibodies makes this case particularly distinctive.

Renal-limited lupus nephritis is a rare entity and may represent an early or incomplete form of systemic lupus erythematosus. These patients may later develop systemic features over time, necessitating close follow-up.

Renal biopsy remains the gold standard for diagnosis. The presence of a "full-house" immunofluorescence pattern is highly suggestive of lupus nephritis and helps differentiate it from other causes of nephrotic syndrome such as primary membranous nephropathy or focal segmental glomerulosclerosis.

This case highlights the limitation of relying solely on serological markers for diagnosis. Similar cases of renal-limited lupus nephritis with negative anti-dsDNA have been rarely reported, emphasizing the heterogeneity of serological presentation. Early recognition of such atypical presentations is crucial, as delayed diagnosis may lead to inappropriate management and progression of renal disease.

CONCLUSIONS

Renal-limited lupus nephritis should be considered in patients with unexplained nephrotic syndrome, even in the absence of classical serological markers. Seronegative or discordant lupus nephritis should not be excluded in nephrotic syndrome without systemic features; renal biopsy remains decisive.

REFERENCES

1. Harrison's Principles of Internal Medicine. 21st ed. New York: McGraw-Hill; 2022.
2. Kidney Disease: Improving Global Outcomes. KDIGO 2021 Clinical Practice Guideline for the Management of Glomerular Diseases. Kidney Int Suppl. 2021;100(4S):S1–S276.
3. Almaani S, Meara A, Rovin BH. Update on lupus nephritis. Clin J Am Soc Nephrol. 2017;12(5):825–835.
4. Weening JJ, D'Agati VD, Schwartz MM, et al. The classification of glomerulonephritis in systemic lupus erythematosus revisited. J Am Soc Nephrol. 2004;15(2):241–250.
5. Anders HJ, Saxena R, Zhao MH, et al. Lupus nephritis. Nat Rev Dis Primers. 2020;6:7.
6. D'Cruz DP, Khamashta MA, Hughes GRV. Systemic lupus erythematosus. Lancet. 2007;369(9561):587–596.