10.5152/turkjnephrol.2023.23546



Turkish Journal of Nephrology

# Lupus Nephritis, Enteritis with Cystitis Masquerading as Bilateral Hydroureteronephrosis

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**Received:** March 25, 2023 **Accepted:** June 08, 2023

**Cite this article as:** George Mathew G, Babu D. Lupus nephritis, enteritis with cystitis masquerading as bilateral hydroureteronephrosis. *Turk J Nephrol.* 2023;32(4):367-369.

## INTRODUCTION

Lupus nephritis with cystitis and nonspecific enteritis is a rare clinical manifestation of systemic lupus erythematosus (SLE) with abdominal symptoms like diarrhea, nausea, and abdominal pain.[1,2] Lupus cystitis is estimated to involve 0.01%-2% SLE patients with a female preponderance.[3] The prevalence of gastrointestinal involvement inclusive of enteritis, colitis, pancreatitis, protein-losing enteropathy, ascites, and hepatic dysfunction constitutes around 40%-60% of the SLE cases.[4] Here we present a rare case of lupus nephritis with enteritis and cystitis in a 29-year-old male treated with intravenous cyclophosphamide (Euro-Lupus regimen).

## CASE DESCRIPTION

A 29-year-old male presented o the hospital with complaints of pedal edema, increased urinary frequency, and alternating episodes of diarrhea and constipation. He was a karate trainer, nonsmoker, teetotaler without a family history of kidney disease. He has not been on any regular medications in the recent past. On examination, his pulse was 98/min and blood pressure was 126/80 mmHg, and no significant abdominal findings were noted. Serum creatinine was 1.6 mg/dL, serum albumin was 2.6 g/dL, hemoglobin was 9.4 g/dL, and 24-hour urine protein was 3.9 g/day. Urine analysis revealed 3+ albumin, microscopic hematuria with plenty of leukocytes. Antinuclear antibodies (ANA) and double-stranded deoxyribonucleic acid (dsDNA) were positive with low C3 (63 mg/dL) and low C4 (8 mg/dL) with negative anti-neutrophilic cytoplasmic antibodies. His ultrasound revealed mild ascites, thickened bladder wall with bilateral hydroureteronephrosis (HUN). A Magnetic resonance(MR) urogram revealed bilateral hydroureteronephrosis with bladder wall thickening (Figure 1).

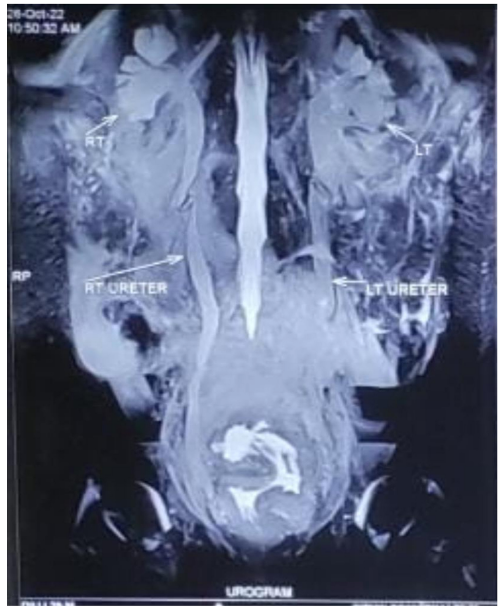


**Figure 1.** MR urogram showing bilateral hydroureteronephrosis with bladder wall thickening. MR, Magnetic resonance.

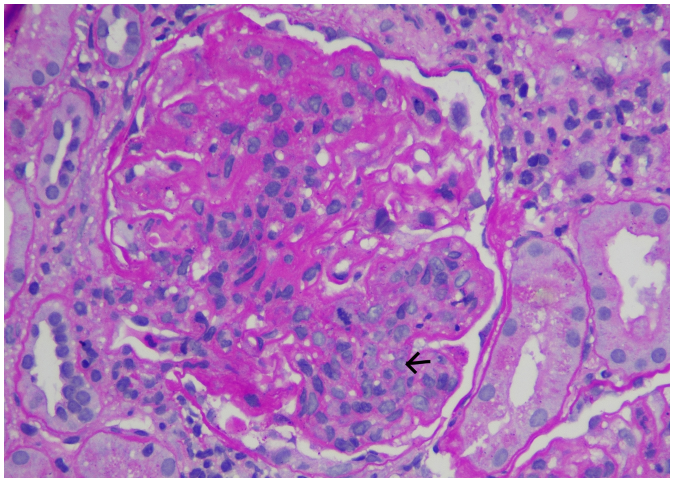

**Figure 2.** PAS stain showing mesangial and endocapillary hypercellularity(Arrow) with double contours of capillary walls (magnification: 400×). PAS, periodic acid–Schiff.



Urine culture was negative for bacteria and fungi and for mycobacteria tuberculosis by GeneXpert (a nucleic acid amplification test). Cystoscopy revealed grade 2 bladder trabeculations and colonoscopy revealed nonspecific inflammation and mucosal edema. Bladder biopsy was not performed and he did not consent for surgical intervention for hydroureteronephrosis. Urodynamic study revealed subnormal bladder capacity of 250 mL. Rectal and colonic biopsy revealed nonspecific enteritis. Kidney biopsy was done due to nephrotic proteinuria and hematuria, which revealed enlarged glomeruli with endocapillary and mesangial hypercellularity with double contours of the capillary walls (Figure 2).

Immunofluorescence revealed C1q (2+) (Figure 3), immunoglobulin M (2+), immunoglobulin A (2+), immunoglobulin G (3+), and C3 (3+), which ultimately led to a diagnosis of class 4 lupus nephritis with NIH activity index 12/24 and chronicity index 0/12.

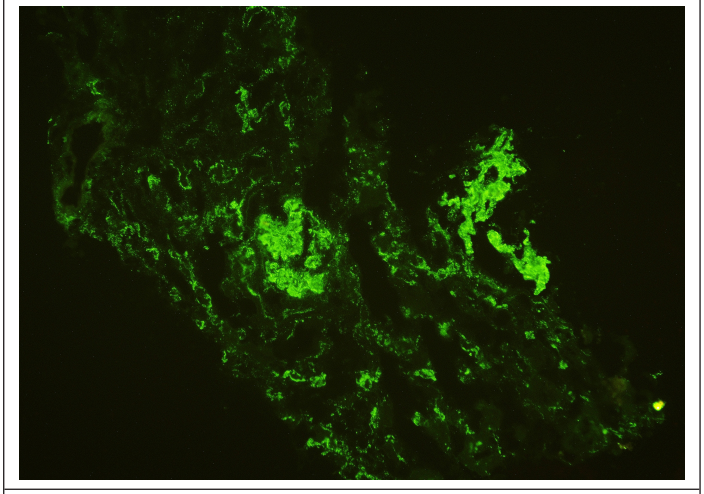

**Figure 3.** Immunofluorescence shows coarse C1q staining along capillary walls and mesangium of the glomerulus.

The patient's creatinine was between 1.6 and 1.8 mg/dL during hospital stay. He was started on intravenous cyclophosphamide pulse with steroids as per Euro-Lupus regimen with telmisartan. After 4 months on review, serum creatinine was 1 mg/dL, and proteinuria had reduced to 300 mg/day, with resolution of gastrointestinal symptoms. A repeat magnetic resonance urogram revealed reduction in the bilateral HUN. He is currently on a maintenance regimen of mycophenolate sodium and low-dose steroids.

## DISCUSSION

Lupus nephritis with enteritis and cystitis presenting with bilateral HUN is a challenging clinical entity. Vasculitis associated with immune complexes[3] leads to bladder wall fibrosis and a small-capacity bladder, which may result in irreversible dysfunction in lupus cystitis. Bladder biopsy is diagnostic in lupus cystitis, which shows mononuclear infiltration in smooth muscles of bladder with a full house pattern of immunoglobulins on immunofluorescence.[3] The differential diagnosis of lupus cystitis includes tuberculosis, vasculitis, inflammatory bowel disease, and mixed connective tissue disorder.[5] The presence of subnormal bladder capacity with bilateral HUN, positive ANA and dsDNA levels with depressed complement levels, negative urine GeneXpert for tuberculosis, nonspecific enteritis on colonic biopsy, and kidney histopathology of class 4 lupus nephritis corroborated a diagnosis of lupus cystitis with enteritis in our case, without bladder biopsy. Our patient had clinical and radiological resolution with immunosuppressants and did not require surgical intervention. Early diagnosis of lupus nephritis with cystitis and enteritis with time-appropriate treatment will lead to fruitful clinical resolution, prevent bladder fibrosis, and retain bladder capacity.

## CONCLUSION

Lupus nephritis with cystitis and nonspecific enteritis is an uncommon clinical presentation of SLE. Prompt recognition, appropriate investigations, and timely interventions can salvage kidney function and prevent irreversible bladder dysfunction.

**Ethics Committee Approval:** N/A.

**Informed Consent:** Written informed consent was obtained from patient who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept – G.G.M.; Design – G.G.M., D.B.; Supervision – G.G.M.; Data Collection and/or Processing – M.S.; Analysis and/or Interpretation – G.G.M., D.B.; Literature Search – G.G.M., D.B.; Writing Manuscript – G.G.M., D.B.; Critical Review – G.G.M., D.B.

**Declaration of Interests:** The authors declare that they have no competing interest.

**Funding:** The authors declared that this study has received no financial support.

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