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Case Report

Dual Origin of Hematuria in Concomitant Lupus Nephritis and Nutcracker Syndrome: A Case Report

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ABSTRACT

Background: Systemic lupus erythematosus is a multisystem autoimmune disease where lupus nephritis is a major cause of morbidity. Nutcracker syndrome, a rare vascular condition caused by left renal vein compression, presents with similar urinary findings. The coexistence of both is exceptional and creates a significant diagnostic challenge.
Case Presentation: A 17-year-old female diagnosed with active Systemic lupus erythematosus and renal involvement, including proteinuria and microscopic hematuria.
Investigations: Initial investigations revealed proteinuria (1.19 g/24h) and significant microscopic hematuria (84 RBCs/HPF). While renal biopsy confirmed class III-A focal proliferative lupus nephritis, persistent postprandial abdominal pain led to an abdominal computed tomography scan, which demonstrated narrowing of the left renal vein between the aorta and superior mesenteric artery, consistent with anterior Nutcracker syndrome.
Treatment: The patient received pulse methylprednisolone, oral corticosteroids, hydroxychloroquine, and mycophenolate mofetil.
Outcome and Follow-up: After six months, Systemic lupus erythematosus activity significantly decreased (SELENA-SLEDAI score from 15 to 4) and proteinuria resolved completely. However, while abdominal pain resolved, microscopic hematuria persisted (24 RBCs/HPF) despite immunosuppressive therapy.
Conclusion: This case illustrates a dual mechanism of hematuria, involving lupus nephritis associated with the mechanical effect of Nutcracker syndrome. Recognizing Nutcracker syndrome as an independent cause of persistent hematuria in treated lupus nephritis is essential to prevent unnecessary escalation of immunosuppressive therapy.

Introduction

Systemic lupus erythematosus (SLE) is a complex multisystem autoimmune disease characterized by autoantibody production and systemic tissue inflammation.[1] Renal involvement, or lupus nephritis (LN), is among the most frequent and severe manifestations, occurring in approximately 50% of patients during the disease course.[1] LN is a major contributor to morbidity and mortality and may progress to end-stage renal disease. Typical clinical features

include proteinuria, hematuria, and hypertension, reflecting glomerular inflammation and damage.

By contrast, Nutcracker syndrome (NCS) is a rare structural vascular disorder resulting from compression of the left renal vein (LRV), most commonly between the abdominal aorta and the superior mesenteric artery corresponding to anterior NCS.[2,3] This extrinsic compression leads to venous hypertension in the LRV, which may result in blood stasis, dilation of collateral veins particularly the gonadal vein, and rupture of the thin-walled veins of the renal fornix into the collecting system. Clinical manifestations of NCS include microscopic or macroscopic hematuria, left flank or abdominal pain, and occasionally orthostatic proteinuria.[3] It is important to distinguish the "Nutcracker phenomenon," a purely anatomical or radiological finding in asymptomatic individuals, from the "Nutcracker syndrome," which requires the presence of compatible clinical symptoms.[2] Diagnosis relies on Doppler ultrasonography and cross-sectional imaging, as there is currently no established gold standard. Phlebography, combined with pressure gradient measurement, may provide complementary information but should be interpreted alongside imaging and clinical findings.[4]

The coexistence of LN and NCS in a single patient is extremely rare and poorly documented. This overlap poses a diagnostic challenge, as both conditions share common urinary and clinical features, particularly hematuria and proteinuria. Differentiating the contribution of each condition is essential for appropriate patient management.

We report the case of a 17-year-old adolescent with biopsy-proven LN and coexisting anterior NCS, highlighting the diagnostic challenge of dual-origin hematuria and the importance of tailored therapeutic strategies.

Case Presentation

Initial Visit (July 2023)

A 17-year-old female high school student presented with inflammatory polyarthralgia of several weeks' duration, associated with intermittent low-grade fever, fatigue, and a characteristic malar erythema in a 'vespertilio' distribution. She also reported chronic intermittent abdominal pain, predominantly postprandial and localized to the left flank and pelvic regions. She had no personal or family history of autoimmune, vascular, or renal disease.

Clinical Findings

On physical examination, the patient was lean (BMI 22.7 kg/m²) and normotensive. Musculoskeletal assessment revealed tenderness of the metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints without swelling (Number of Tender Joints [NTJ] = 12; Number of Swollen Joints [NSJ] = 0), with a positive squeeze test. Cutaneous and mucosal examination confirmed a fixed malar erythema sparing the nasolabial folds, with no oral ulcers or peripheral edema. Abdominal palpation was unremarkable, showing a soft, non-tender abdomen without organomegaly or palpable masses. The remainder of the systemic examination was normal.

Timeline of clinical events

Date	Clinical Events and Interventions
July 2023	Initial presentation with polyarthralgia, malar rash, low-grade fever, and postprandial left flank pain.
August 2023	Contrast-enhanced computed tomography (CT) scan revealed compression of the left renal vein (NCS).
October 2023	Renal biopsy confirmed Class III (A) focal proliferative lupus nephritis.
October 2023	Initiation of immunosuppressive therapy (intravenous methylprednisolone pulses followed by oral corticosteroids, hydroxychloroquine, and mycophenolate mofetil).
April 2024 (6-month follow-up)	Resolution of joint, cutaneous, and abdominal symptoms. Normalization of proteinuria, with persistence of mild microscopic hematuria.

Diagnostic assessments

Initial laboratory evaluation demonstrated mild normocytic normochromic anemia (hemoglobin = 10.5 g/dL) and mild thrombocytopenia (platelet count = 118,000/mm³). Inflammatory markers showed an elevated erythrocyte sedimentation rate (ESR = 91 mm/h) with normal C-reactive protein (CRP <6 mg/L). Immunological tests revealed high-titre antinuclear antibodies (ANA = 1:1000), markedly elevated anti–double-stranded DNA (anti-dsDNA) antibodies (800 IU/mL; 26 times the upper limit of normal), negative anti-Smith (anti-Sm), anti-ribonucleoprotein (anti-RNP), anti-Sjögren's-syndrome-related antigen A antibody (anti-SSA), and anti-Sjögren's-syndrome-related antigen B antibody (anti-SSB), and reduced complement levels (C3 and C4). Antiphospholipid antibodies were negative.

Renal function was preserved, with a serum creatinine of 0.8 mg/dL. However, urinalysis and 24-hour collection revealed significant proteinuria (1.19 g/24h) and active microscopic hematuria (84 RBCs/HPF). Renal ultrasonography showed normal kidney size bilaterally and normal parenchymal echotexture.

Due to persistent abdominal pain, a contrast-enhanced abdominal and pelvic computed tomography (CT) scan was performed. It revealed narrowing of the left renal vein between the aorta and the superior mesenteric artery, associated with dilation of the left gonadal vein and pelvic venous congestion, consistent with anterior Nutcracker syndrome (Figure 1).

To rule out orthostatic proteinuria associated with left renal vein compression, split daytime and nighttime urine collections were performed, demonstrating no significant diurnal variation. Given the active urinary sediment and nephrotic-range/significant proteinuria, a percutaneous renal biopsy was performed. Light and immunofluorescence microscopy confirmed Class III (A) focal proliferative lupus nephritis, showing active endocapillary hypercellularity, mild mesangial expansion, and characteristic granular immune complex deposition.

Overall, the clinical presentation, serological profile, and histological findings fulfilled the 2019 ACR/EULAR classification criteria for SLE, with a total score of 38 points. At baseline, the modified

SELENA-SLEDAI score was 15, reflecting high disease activity driven by active mucocutaneous, articular, immunologic, and renal involvement.

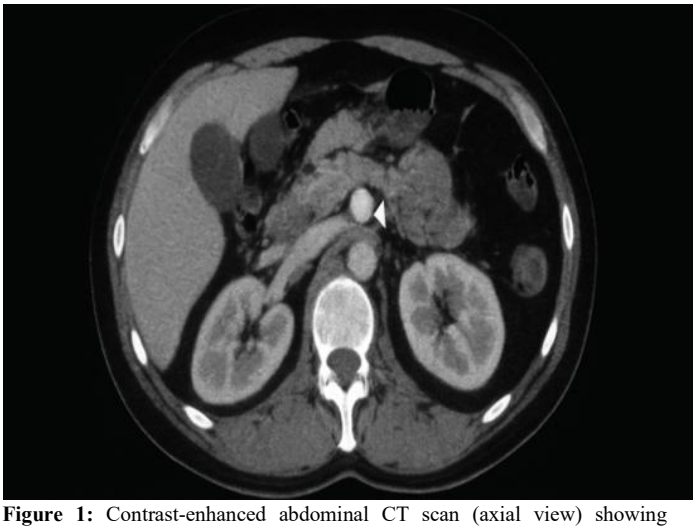

Figure 1: Contrast-enhanced abdominal CT scan (axial view) showing compression of the left renal vein between the aorta and the superior mesenteric artery, consistent with anterior Nutcracker syndrome. Both kidneys are well opacified without evidence of hydronephrosis.

Table 1: Laboratory findings at the time of admission and 6 months later

Parameter	At admission	At 6 months	Reference range
Hemoglobin (g/dL)	10.5	12.1	12–16
White blood cell count (×10⁹/L)	6.4	5.0	4–10
Platelet count (×10⁹/L)	118	283	150–400
Erythrocyte Sedimentation Rate(mm/h)	91	20	<13
C-reactive protein (mg/L)	<6	<6	<6
Serum creatinine (mg/dL)	0.8	0.64	0.6-1.2
Estimated GFR (mL/min/1.73 m²)	98	102	>90
Urine routine	Protein +++, 84 RBCs, No casts, no bacteria	Protein -, 24 RBCs, No casts, no bacteria	
Proteinuria (g/24 h)	1.19	0.049	<0.15

Therapeutic Intervention

Induction therapy was initiated with oral hydroxychloroquine (400 mg/day) and high-dose intravenous methylprednisolone pulse therapy (500 mg/day for three consecutive days). This was followed by oral prednisone (1 mg/kg/day, tapered per protocol) and mycophenolate mofetil (MMF) at an initial dose of 3 g/day.

Follow-up and outcomes

The patient was monitored over a 9-month period from July 2023 to April 2024. Within six weeks of starting induction therapy, articular and cutaneous manifestations resolved, abdominal discomfort improved markedly, and proteinuria dropped significantly (<150 mg/24h).

Corticosteroids were progressively tapered to a maintenance dose of 10 mg/day, while MMF was maintained at 2 g/day. At the 6-month evaluation, the patient achieved renal remission with low systemic disease activity (modified SELENA-SLEDAI score decreased from 15 to 4). Abdominal pain had fully subsided and 24-hour urinary protein excretion resolved to physiological levels (48.7 mg/24h, negative for proteinuria). However, isolated microscopic hematuria persisted on urinalysis (24 RBCs/HPF), consistent with the persistent mechanical effect of left renal vein compression (Table 1).

Discussion

The coexistence of LN and NCS presents a complex diagnostic dilemma. Because both glomerular injury and mechanical venous hypertension can manifest with hematuria and proteinuria, distinguishing between these two mechanisms is clinically vital. Overlooking an anatomical cause of persistent hematuria risks exposing the patient to an inappropriate and potentially harmful escalation of immunosuppressive therapy.

Rarity and novelty of the case

The overlap of active LN and NCS is exceptionally rare. While literature extensively covers both LN and isolated NCS individually, reports describing their coexistence remain extremely limited.[5] A systematic literature search was performed in PubMed, Scopus and Google Scholar using the keywords “systemic lupus erythematosus”, “lupus nephritis”, and “Nutcracker syndrome”. Only two previously published cases fulfilling these criteria were identified. [6,7] Our case represents the third reported case and further contributes to the limited evidence on this uncommon association. It illustrates that persistent hematuria in patients with SLE may result from two distinct mechanisms: glomerular inflammation due to LN and venous hypertension caused by NCS.

Hematuria: dual origin

In our patient, urinary red blood cell morphology was not assessed because phase-contrast microscopy was not routinely available. Therefore, the distinction between glomerular and non-glomerular hematuria relied on renal biopsy findings, treatment response, and imaging studies. The initial marked hematuria (84 RBCs/HPF), in the context of active disease (SELENA-SLEDAI score 15), was consistent with glomerular involvement and was confirmed by renal biopsy.

In contrast, hematuria in NCS results from elevated venous pressure in the LRV, leading to rupture of fragile venous collaterals into the collecting system.[3,7] This bleeding is typically intermittent, may be orthostatic, and is independent of inflammatory activity. In our patient, proteinuria resolved following immunosuppressive therapy, whereas hematuria persisted at lower levels (24 RBCs/HPF). The persistence of hematuria despite remission of proteinuria and improvement of lupus activity suggested an additional mechanical source. Contrast-enhanced CT, performed to investigate persistent abdominal pain, demonstrated characteristic findings of anterior NCS,

including compression of the LRV between the aorta and the superior mesenteric artery, gonadal vein dilatation, and pelvic venous congestion. Although Doppler ultrasonography is generally considered the first-line imaging modality for NCS, it was not performed in our patient. Quantitative CT parameters, such as the aortomesenteric angle and the hilar-to-aortomesenteric diameter ratio, were not available in the radiology report, representing a limitation of this case. In addition, cystoscopy was not performed because an alternative explanation for the persistent hematuria had been identified and the patient had no lower urinary tract symptoms. Likewise, venography and renocaval pressure-gradient measurement were not performed because the diagnosis was considered sufficiently supported by the clinical presentation and CT findings;[4] however, the absence of haemodynamic confirmation should be acknowledged as a limitation. Ultimately, this case illustrates that persistent hematuria in treated LN should not automatically be interpreted as refractory disease, thereby preventing unnecessary and potentially hazardous escalation of immunosuppressive therapy.

Proteinuria: predominantly lupus-related

Proteinuria in SLE reflects glomerular immune complex deposition and inflammation.[1] Our patient presented with proteinuria of 1.19 g/24h associated with biopsy-proven class IIIA nephritis. Immunosuppressive therapy led to a rapid decline to <150 mg/24 h within six weeks and complete remission (<50 mg/24 h), supporting LN as the primary driver.

Although NCS may be associated with mild, often orthostatic proteinuria,[3,7] split daytime and nighttime urine collections showed no circadian variation in our patient, arguing against a significant vascular contribution. Thus, unlike hematuria, proteinuria was almost exclusively related to LN.

Abdominal pain and Nutcracker syndrome

Abdominal and pelvic pain in NCS results from venous hypertension and the development of collateral circulation, particularly involving the gonadal vein.[2,8] The chronic postprandial left-sided pain reported by our patient is consistent with this mechanism. Interestingly, symptoms resolved after initiation of immunosuppressive therapy. Several mechanisms may explain this improvement:

- Reduction of perivascular inflammation and oedema, decreasing LRV compression.
- Corticosteroid-induced weight gain, increasing retroperitoneal fat and widening the aortomesenteric angle.[2,3]
- A possible minor contribution of low-grade mesenteric vasculitis ("lupus enteritis"), although CT findings were not suggestive.[9]

These factors may have converted a symptomatic Nutcracker *syndrome* into an asymptomatic Nutcracker *phenomenon*.

Management of NCS

Management depends on symptom severity. Conservative strategies—including observation, hydration, analgesia, and weight gain—are recommended in adolescents and young adults with preserved renal function and mild symptoms.[3,4] Interventional procedures, such as endovascular stenting or surgical LRV transposition, are reserved for severe or refractory cases, including those with significant hematuria, transfusion-requiring anaemia, debilitating pain, or impaired renal function.[4]

In our patient, conservative management was appropriate, as abdominal pain resolved, and hematuria remained mild without functional impact.

Conclusion

This case underscores that persistent hematuria in patients with treated lupus nephritis should not automatically be interpreted as ongoing renal disease activity. Alternative causes, including Nutcracker syndrome, should be considered to avoid unnecessary escalation of immunosuppressive therapy.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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Ethics Statement

Formal approval from an institutional ethics committee was not required for this single case report in accordance with local institutional regulations. The study was conducted in accordance with the principles of the Declaration of Helsinki. Written informed consent for publication of the clinical data was obtained from the patient's legal guardian.

Data availability

All data generated or analyzed during this study are included in this published article

Author Contributions

AH contributed to conceptualization of the case report, supervision of patient management, drafting, and critical revision of the manuscript; HA contributed to clinical assessment, literature review, drafting, and editing of the manuscript; HB contributed to interpretation of laboratory and diagnostic findings, nephrology expertise, and manuscript revision; FH contributed to critical revision of the manuscript for important intellectual content and final approval. All authors read and approved the final version, meet the ICMJE criteria for authorship, and agree to be accountable for all aspects of the work.

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