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A RARE PRESENTATION OF CLASS V LUPUS NEPHRITIS IN A YOUNG MALE: A CASE REPORT

Nephrology

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ABSTRACT

Systemic lupus erythematosus (SLE) is a connective tissue disease that can rarely manifest as class V lupus nephritis, which frequently presents clinically as nephrotic syndrome and the pathological finding of subepithelial immune complex deposition on a renal biopsy. Although SLE disproportionately affects females, the lower incidence of the disease in males often leads to a delay in diagnosis. Therefore, it is crucial to maintain a high level of clinical suspicion for SLE in male patients who present with unusual renal symptoms.

KEYWORDS

Systemic lupus erythematosus (SLE), Lupus nephritis, Nephrotic syndrome

INTRODUCTION:

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune-mediated disorder. It is classically characterised by the circulation of autoantibodies and deposition of immune complexes across various organ systems and is known to present with a plethora of varied clinicopathological manifestations. While the aetiology of SLE is not completely clear, it is thought to have genetic and environmental involvement. (1) More so, various risk factors, including ethnicity, age and gender have been linked to the development and progression of SLE. This condition has a female preponderance, affecting women of childbearing age nine times more commonly than their male counterparts. (2) Males experience less typical mucocutaneous and musculoskeletal symptoms than women during diagnosis. Although it commonly emerges between the ages of 15 to 45, a broad spectrum of ages can be affected. Due to its wide variety of clinical presentations, lupus nephritis can present from asymptomatic urinary abnormalities to nephrotic syndrome and renal failure. Given the rarity of disease presentation in young men, diagnosing lupus nephritis is often challenging. (3)

Case Presentation:

Patient Information: A 22-year-old male patient presented to the general medicine OPD with complaints of shortness of breath over the past 8 - 10 days, along with generalised edema and progressively worsening fatigue over the past month. Upon eliciting a detailed history, the patient stated that the edema had gradually worsened and had initially affected the face and the upper limbs, which then progressed to the lower limbs.

Clinical Presentation: On clinical examination, he was found to be tachycardic (120 bpm), hypertensive with elevated systolic blood pressure (160/94mm Hg) and bilaterally decreased respiratory sounds in the lower lung fields. There were no other clinically significant findings.

Laboratory Findings

a) Serum and urine investigations:

Table 1: Investigations Findings Of The Patient Showing Mild Leucocytosis, Moderate Thrombocytopenia, Hypoproteinemia And Hypoalbuminemia, Low C3 And C4 And Positive Autoimmune Panel

Investigations	Reports (normal range)
Haemoglobin (g/dl)	13.60 (13-17)
Total leucocyte count/ mm3	10700 (4000 - 10000)
Platelet count /mm3	60000 (150000 - 400000)
CRP (mg/L)	< 2 (<6)
Total protein (g/dl)	4.8 (6.8 - 8.6)
Serum albumin (g/dl)	1.5 (3.7 - 5.2)
Serum creatinine (mg/dl)	1.1 (0.6 - 1.3)
Urea (mg/dl)	48 (11 - 40)
C3 complement (mg/dl)	28.84 (75 - 135)
C4 complement (mg/dl)	4.04 (10 - 40)
antinuclear antibodies, anti-Smith antibodies, anti-chromatin antibodies, anti-double-stranded DNA antibodies, and cytoplasmic perinuclear antibody (p-ANCA)	Positive
Urine protein creatinine ratio	5 (high)
Urine routine and microscopy	Trace proteinuria

b) Imaging

1) USG Abdomen + KUB: A bilateral renal ultrasound ruled out hydronephrosis and/or any obstructive causes. Mild ascites was noted on USG abdomen.

2) USG Chest: 100 cc of pleural effusion in the bilateral lower lobes.

c) Renal Biopsy:

The kidney biopsy revealed mild mesangiopathic glomerulonephritis with immune complex deposition, as indicated by positive immunofluorescence staining for IgG, IgA, IgM, C3, and C1q. These findings were highly supportive of making a formal diagnosis of class V lupus nephritis, which is classically characterised by podocytopathy and collapsing glomerulosclerosis.

Management And Outcome: The patient was started on a high-dose regimen of mycophenolate mofetil combined with prednisone, which showcased a positive clinical outcome. A low-dose mycophenolate mofetil regimen was continued as a part of maintenance therapy. The patient was advised to undergo monthly serum protein and renal function tests as a part of the follow-up plan. The patient's weight also dropped from 76kg to 66kg at the time of the one-month follow-up, implying a positive outcome of the treatment regimen.

DISCUSSION:

Systemic lupus erythematosus is a chronic multisystemic autoimmune condition wherein the individual's immune system generates antibodies targeting self-proteins, particularly those present within the cell nucleus. It is believed to be a type III hypersensitivity reaction, known to affect a variety of organ systems, most frequently the skin, musculoskeletal and hematological systems. While SLE affects both sexes, the symptoms associated with each sex are different. Females often experience a higher frequency of relapses, reduced white blood cell counts, increased prevalence of arthritis, Raynaud's syndrome, and psychiatric symptoms. In contrast, males tend to exhibit more seizures, kidney disease, serositis, skin conditions, and peripheral neuropathy. (4)

The histological hallmark of SLE is membranous glomerulonephritis with 'wire loop' abnormalities due to the immune complex deposition along the glomerular basement membrane, leading to a typical granular appearance in immunofluorescence testing. Renal

involvement in SLE patients is a significant cause of morbidity and leads to an overall 2.6% mortality rate. (5)

The various glomerular immune complex deposition patterns are classified based on the revised ISN/RPS classification system. The classification of lupus nephritis begins with class I as minimal mesangial lupus nephritis to class VI as advanced sclerosis lupus nephritis. These include predominantly mesangial deposits (classes I and II), subendothelial deposits with endocapillary hypercellularity or prominent glomerular basement membrane (GBM) duplication, and wire-loop lesions, which are frequently associated with necrotising and crescentic lesions (classes III and IV, focal and diffuse LN), or membranous forms. Membranous LN (class V) is seen in up to 15% of biopsied SLE patients, who usually present with nephrotic syndrome and can occur in combination with localised or diffuse LN (class III or IV). (6)

Treatment options for class V lupus nephritis are dependent on the range of proteinuria. Nephrotic range proteinuria includes blood pressure control, renin-angiotensin system blockade along with combined immunosuppressive therapy consisting of glucocorticoids and one other agent (mycophenolic acid analogues, cyclophosphamide, rituximab, azathioprine or calcineurin inhibitors) (3). The therapeutic goal is to produce a clinical remission with normalisation of renal functioning and proteinuria, along with a serological remission, which is the normalisation of anti-ds-DNA antibody, C3 and C4 levels. (3)

Other differentials to be kept in mind which could overlap with similar symptoms include primary membranous nephropathies, mixed connective tissue diseases, inherited conditions (Alport, Fabry's), etc.

CONCLUSION:

This case demonstrated a relatively rare presentation of lupus nephritis in a young male patient without typical SLE features, highlighting the importance of renal biopsy in confirming a diagnosis. Awareness amongst practitioners regarding the varied and multiple presentations of SLE can help initiate treatment sooner to reduce morbidity and mortality.

Declarations:

- **Ethical approval:** This is a case report. The GMERS Gotri Research Ethics Committee has confirmed that no ethical approval is required.
- **Informed consent:** Informed consent has been obtained from patient**.**
- **Consent to participate:** The participant has consented to the submission of the case report to the journal.
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Conflict Of Interest: On behalf of all authors, the corresponding author states that there is no conflict of interest.

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