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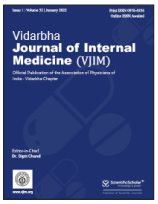

Case Report

Silent Lupus Nephritis in Adolescent Male: A Case Report

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ABSTRACT

Systemic lupus erythematosus (SLE) is a multisystemic disorder, one of several diseases known as 'the great imitators,' because it often mimics or is mistaken for other illnesses. SLE symptoms vary widely and appear and disappear unpredictably. Diagnosis can thus be elusive, with some patients having unexplained symptoms of untreated SLE for years. Autoimmunity plays a major role in the pathogenesis of lupus nephritis (LN). LN is one of the most serious complications of SLE since it is the major predictor of poor prognosis. The immunologic mechanisms include the production of autoantibodies directed against nuclear elements. Here is a case report of a young male patient who presented with fever, and alopecia and was diagnosed as an SLE with LN.

Keywords: SLE, Lupus Nephritis, Auto-Immunity

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disorder with a circulation of autoantibodies and deposition of immune complexes in various organs and systems. SLE is known for the heterogenicity of clinicopathological manifestations and life-threatening outcomes. According to the American Society of rheumatology SLE classification criteria, the diagnosis is based on the distinctive clinical features and presence of auto-antibodies. The disease aetiology remains unknown; however, numerous risk factors are associated with the onset and progression of SLE, such as ethnicity, age, and gender.[1] SLE tends to affect young women of child-bearing age and the incidence rate of SLE is 6–10 times more prevalent in women than in men.[2] Although SLE predominately affects females of childbearing age, it can occur in males and at any age. Women tend to peak between the ages of 20 and 30. For males, the peak is later, between the age of 45 and 60 years. One of the most important poor prognostic factors associated with an increased rate of mortality is the development of lupus nephritis (LN). Time from the disease onset and establishing a clinical diagnosis of LN was described to be more prolonged in males due to overlooking SLE symptoms.[3] More random presentation of SLE in males leads to less awareness and underestimating of SLE symptoms in male patients group. Moreover, morphological presentation and its progression to the end stage of renal disease among male patients were described as more severe rather than female counterparts.[2] However, opinions of the male gender as a risk factor for lupus nephritis varies. The onset of LN, progression, and also long-term prognosis in males is disputed in the literature.

CASE REPORT

A 15-year-old male presented with complaints of fever without chills and rigors, scarring lesion over face, back, and trunk, hair loss, and generalised weakness, since 10–12 months, and a decrease in appetite in the past 1 month. Patient had been receiving treatment outside for 3-4 months with no relief. He

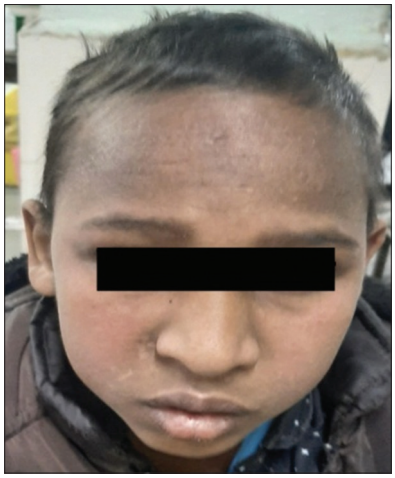

Figure 1: Scarring lesions over the face

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then came to our hospital and was hospitalised as a case of febrile illness for evaluation [Figure 1].

On general examination, the patient was afebrile, having Pallor, no icterus, no lymphadenopathy, heart rate of 112/min, BP was 130/90 mm of Hg in the right arm, and had oedema over feet.

Systemic examination was normal.

Investigations	Value	Normal Range
CBC		
WBC count (×10³/μL)	3.2	4-10
Haemoglobin (g/dL)	8.3	12-16
Platelet count (×10³/μL)	176	150-450
Haematocrit (%)	23.2	40-52
Serum chemistry		
Sodium (mmol/L)	136	135-145
Potassium (mmol/L)	3.8	3.6-5.4
Calcium mg/dL)	7.9	8-11
Phosphorous (mg/dL)	5.5	2.5-4.5
Uric acid (mg/dL)	8.1	3.5-7.5
Urea (mg/dL)	56	6-24
Creatinine (mg/dL)	0.9	0.7-1.2
Total protein (g/dl)	5.7	5.5-7.8
Albumin (g/dl)	2.8	3.2-4.8
Total bilirubin (mg/dl)	3.3	0.1-1.0
ALP (U/L)	359	50-308
ALT (U/L)	38	<49
AST (U/L)	122	<46
LDH (U/L)	418	105-333
Ferritin (mcg/L)	2844	24-336
CRP (mg/L)	2.6	<10
CPKMB (U/L)	16.4	5-25
Triglycerides (mg/dl)	424	60-150
T. cholesterol (mg/dl)	256	130-200
HDL cholesterol (mg/dl)	84	30-70
LDL cholesterol (mg/dl)	87	80-180

Urine analysis	
Protein (mg/dl)	34.4 (high)
Creatinine (mg/dl)	7
UPCR	**4.49** (high)

*Interpretation – Nephrotic range Proteinuria.

PS: RBC – normocytic, normochromic, macrocytes, and microcytes present

TLC <4000/Cumm

Plt – Adequate

Parasite – Not seen

Sickling – Negative

Reticulocyte count – 1.3%

Fever profile (HRP2, DENGUE NS1 IGM IGG, SCRUB THYPHUS, and WIDAL) – NEG

HIV, HCV, HBSAG – NEG

URINE RM: **URINE PROTEIN (Albumin)- Present ++**

Amorphous Deposits – Present ++

CXR – WNL.

ECG had normal sinus rhythm no specific ST-T changes,

Echo-WNL.

After all routine workup, we suspected multisystem involvement and send ANA PROFILE.

ANA PROFILE S/O

Anti-ds-DNase was Positive,
Anti SmD1 positive,
Nucleosome antigen-positive,
Histone antigen positive
F/S/O SLE

As the case was young male to r/o kidney involvement I/v/o proteinuria, oedema feet, raise UPCR value, we did a renal biopsy.

Renal biopsy [Figure 2]

Microscopy

Renal glomeruli showed a mild-to-moderate increase in mesangial matrix and cellularity, and 15.3% showed segmental endocapillary cellularity. Subendothelial deposits, crescent formation, or active tuft necrosis were not observed in visualized glomeruli. None of the sampled glomeruli had global sclerosis. Tubular atrophy and interstitial fibrosis involved 15% of sampled cortex. Viable tubules showed vocally prominent tubular cytoplasmic vacuolar changes. Few hyaline and granular casts in tubular lumina, patchy interstitial oedema, and focal chronic interstitial inflammation were observed.

Arteries showed mild medial thickening and subintimal sclerosis. Arterioles revealed thickened wall focal hyalinosis lesion and vacuolisation in smooth muscle cells of media.

DIF-Parameter results

- IgA – 2+ Mesangial and segmental capillary wall granular
- IgG – 3+ Mesangial and segmental capillary wall granular
- IgM – 2+ Mesangial and segmental capillary wall granular
- C3 – + Mesangial and segmental capillary wall granular
- C1q – 2+ Mesangial and segmental capillary wall granular
- Kappa light chains – 3+ Mesangial and segmental capillary wall granular

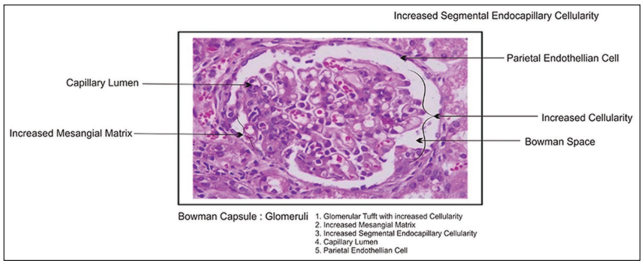

Figure 2: Renal biopsy

- Lambda light chains – 3+ Mesangial and segmental capillary wall granular.

Patchy granular staining for IGG, C1q, and kappa and lambda light chains is noted along tubular basement membranes.

Impression

1. **Focal** LN; ISN/RPS (2018 modified) **Class III**
2. Indices (modified NIH) of **disease activity 3/24 & chronicity 2/12.**

	Activity indices	Score
a.	Endocapillary hypercellularity	1
b.	Neutrophils/Karyorrhexis	1
c.	Fibrinoid necrosis	0
d.	Hyaline/wire loop deposits	0
e.	Cellular/fibrocellular crescent	0
f.	Interstitial inflammation	1

	Chronicity score	
a.	Total glomerular sclerosis score	0
b.	Fibrous crescent	0
c.	Tubular atrophy	1
d.	Interstitial fibrosis	1

The patient was treated with pulse therapy of steroids (Injection Methylprednisolone 1 gm OD for 3 days) and hydroxychloroquine 200 mg od. He was discharged on hydroxychloroquine and steroids with tapering doses and advice to follow-up.

DISCUSSION

The patient was diagnosed as SLE and presented with multiorgan involvement mainly kidney. He had laboratory abnormalities such as low albumin levels, and urinary protein with sediment suggesting active LN. Symptoms related to active nephritis were peripheral oedema secondary to hypertension or hypoalbuminemia. Extreme peripheral oedema is more common in diffuse or membranous LN, as these renal lesions are commonly associated with heavy proteinuria.[4] Patients with active LN often have other symptoms of active SLE, including fatigue, fever, rash, arthritis, serositis, or central nervous system disease. These are more common with focal proliferative (as in this case) and diffuse proliferative LN.[5]

Evaluating renal function in patients with SLE to detect any renal involvement early is important because early detection and treatment can significantly improve renal outcomes.[6] Renal biopsy should be considered in any patient with SLE who has clinical or laboratory evidence of active nephritis, especially on the first episode of nephritis.[6] Hence, the patient's renal biopsy was done and the patient was diagnosed as a case of LN Stage III according to the classification revised by the International Society of Nephrology (ISN) and the Renal Pathology Society (RPS) in 2004. This classification is based on light microscopy, immunofluorescence, and electron microscopy findings from renal biopsy specimens.

The principal goal of therapy in LN is to normalize renal function or, at least, to prevent the progressive loss of renal function. Therapy differs depending on the pathologic lesion.[4,6] Corticosteroid therapy should be instituted if the patient has clinically significant renal disease. The use of immunosuppressive agents, particularly cyclophosphamide, azathioprine, or mycophenolate mofetil, if the patient has aggressive proliferative renal lesions, as they improve the renal outcome. They can also be used if the patient has an inadequate response or excessive sensitivity to corticosteroids.[6]

The first guidelines for managing LN have been issued by the American College of Rheumatology.[7] Patients with clinical evidence of active, previously untreated LN, should have a renal biopsy to classify the disease according to ISN/RPS criteria. All patients with LN should receive background therapy with hydroxychloroquine unless contraindicated. This recommendation was based on a prospective controlled trial showing lower flare rates in those who continued hydroxychloroquine, compared with those who switched to placebo. However, opinions of the male gender as a risk factor for lupus nephritis varies. Glucocorticoids plus either cyclophosphamide intravenously (IV) or mycophenolate mofetil orally for induction in patients with ISN class III/IV disease. Patients with ISN/RPS classes I and II nephritis do not require immunosuppressive therapy. Administer ACE inhibitors or angiotensin-receptor blockers if proteinuria is 0.5 g/24 h or more. Maintain blood pressure at or below 130/80 mm Hg. Patients with end-stage renal disease, sclerosis, and a high chronicity index based on renal biopsy findings are unlikely to respond to aggressive therapy. In these cases, focus therapy on extrarenal manifestations of systemic lupus erythematosus (SLE) and on possible kidney transplantation.

CONCLUSION

All the patients of SLE with proteinuria should be promptly investigated and suspected of LN and treatment should be started at the earliest after diagnosis is established. Although it is expected that LN usually manifests around 4–5 years after diagnosis, the early presentation can occur in patients with a relatively shorter duration of illness.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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