International Journal of Contemporary Pediatrics
Richa et al. Int J Contemp Pediatr. 2020 Jan;7(1):220-223
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20195758

Case Report

Case report of a male child with lupus nephritis

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Received: 10 September 2019
Revised: 02 October 2019
Accepted: 04 November 2019

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ABSTRACT

Systemic lupus erythematosus is a chronic autoimmune inflammatory disease of unknown etiology that affects various organs, most frequently the skin, joints, kidneys, nervous, hematologic and cardiovascular systems. It affects females more often as compared to males. The kidneys are one of the most serious organs involved. Lupus nephritis may present as hypertension, proteinuria, and renal failure or it may also be asymptomatic. The recent reports suggest that childhood-onset lupus nephritis could be more severe than the late-onset disease. The occurrence of SLE in pediatric patients is very rare, especially in a male child. Here author report a case of an 8-year-old male child clinically misdiagnosed as a case of henoch schonlein purpura, who was thoroughly investigated and finally confirmed as a case of lupus nephritis.

Keywords: Antibody, Autoimmune, Biopsy, Lupus nephritis, Male, Systemic lupus erythematosus

INTRODUCTION

Systemic Lupus Erythematosus (SLE) is a multi-systemic autoimmune disorder which commonly observed in adults than children. In pediatric population, it frequently affects girls than boys. Childhood-onset SLE is more severe.[1] Lupus nephritis is one of the most serious organ involvements of the disease which is present in up to 80% of pediatric patients with SLE and is more severe than in adults.[1] The majority of children with SLE are adolescent females.[1] The wide spectrum of presentation of lupus nephritis includes nephrotic syndrome and renal failure to only asymptomatic urinary findings as well.[1] Because of varying presentations and the rarity of the disease in childhood, diagnosis can be difficult. Early diagnosis with appropriate treatment is essential to provide a favorable outcome for normal childhood and adolescent development.

CASE REPORT

An 8-year-old male child, 1st issue of a non-consanguineous marriage, previously diagnosed as a case of henoch schonlein purpura on the basis of rash and polyarthritis was referred to the pediatric OPD of hospital for further investigations and management. The patient was apparently asymptomatic 3 months back when he developed maculopapular rash all over the body which was denser bilaterally on the lower extremities. For the last 15 days patient had started complaining of joint pain which was moderate grade, insidious in onset, starting in the wrist and ankle joints bilaterally. The pain was non-migratory, with an early morning stiffness and was relieved with medications. There was no history of fever or joint deformities.

He was previously healthy without a remarkable past illness. There was no significant family history of such illness or history of contact with tuberculosis.

He had been diagnosed previously as a case of henoch schonlein purpura and started on non-steroid anti-inflammatory drugs for arthritis.

On Physical examination, the vitals were stable with a blood pressure of 90/60 mmHg (50th percentile).

Anthropometrically the child had a weight of 24 kg (50th percentile), height of 126 cm (50th percentile) and BMI of 15.11. The child had pallor, no edema but disseminated maculopapular non pruritic rash over the entire body which was denser on the lower limbs. On locomotor system examination (pGALS- Pediatric Gait, arms, limbs and spine) was normal. Other systemic examination was unremarkable.

The laboratory examination showed anemia with a normal WBC and platelet count and high erythrocyte sedimentation rate. The kidney and liver function tests were normal. However, the albumin was markedly low. Urine analysis was suggestive of albuminuria with a dipstick of 3+ while urine sediments showed microscopic hematuria.

The C3 level was markedly low. The ANA was strongly positive. In a strong suspicion of SLE, author got anti dsDNA antibody levels done which came out to be positive (Table 1).

Table 1: Laboratory findings of the patient showing anaemia with haematuria, low serum C3 levels, positive ANA and highly positive anti dsDNA.

Investigations	Reports
Hemoglobin (g/dl)	10
Total leucocyte count /mm^3	8500
Platelet count /mm^3	2.5
ESR (mm)	56
Serum albumin (mg/dl)	2.5
Urine routine and microscopy	20-30 RBCs/hpf, Albumin 3+
Blood urea (mg/dl)	18
Serum creatinine (mg/dl)	0.7
Serum sodium (mmol/l)	136
Serum potassium (mmol/l)	3.8
Serum C3 (g/l)	0.31 (0.90-2.10)
ANA	2.82 (>1.2 positive)
Anti dsDNA	12.50 (>1.10 highly positive)
24-hour urine protein creatinine ratio	1.8
USG abdomen	Normal

The renal biopsy revealed features of endo capillary proliferation and segmental necrosis as shown in (Figure 1) (class IV lupus nephritis) and immunofluorescence showed full house pattern.

Hence depending upon the clinical, laboratory findings and biopsy report, the child was diagnosed as a case of lupus nephritis-class IV. The patient was started on pulse methylprednisolone (five doses) and cyclophosphamide (six doses) for the induction therapy.

After the induction therapy, the urinary findings became normal. Then the patient was started on maintenance therapy consisting of low dose steroid and azathioprine for 12 months.

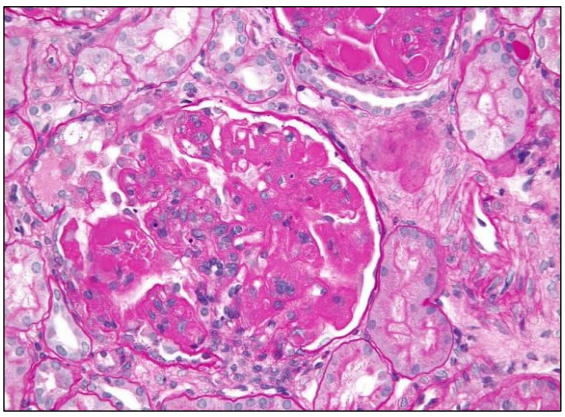

Figure 1: (Renal biopsy) This light microscopy pas stained, high power view of glomerulus showing features of endo capillary proliferation and segmental necrosis.

DISCUSSION

Systemic lupus erythematosus is a chronic autoimmune inflammatory disease of unknown cause that can affect any organ system, most frequently the skin, musculoskeletal system, hematological system. Clinically important renal involvement occurs in 40-50% of patients which is characterized by the production of multiple autoantibodies. The prevalence of SLE in children is more after the first decade of life where as it is rare before five years of age.[2] It affects females more often than males (8 to 9:1) ,even in the pre pubertal age group (4 to 5:1).[1-3] The worldwide prevalence rates range widely from 3.3 to 9.7 per 100,000 children and adolescents, depending upon population study and its ethnic distribution.

Though the cause of SLE remains unknown, genetic, immunologic, hormonal and environmental factors may play a role. The unique feature of childhood SLE is the identification of monogenic forms of SLE, mainly due to defects in the complement system: type I interferon pathway, aberrant nucleic acid repair, degradation and sensing, or abnormal B cell development.[4,5]

There were two classification criteria : one from the American College of Rheumatology (ACR), last revised in 1997, and another set of criteria titled SLICC (Systemic Lupus International Collaborating Clinics) group classification criteria published in 2012.[6] However in September 2019, ACR/ EULAR came up with a new classification of SLE (Table 2). Renal disease in SLE is usually asymptomatic, underscoring the need for careful monitoring of blood pressure and urinalyses where it often presents with proteinuria, hematuria and/or renal failure and the predominant symptoms being fatigue, edema, changes in urine color, and nausea/ vomiting.

Table 2: ACR/EULAR classification criteria for SLE-2019.

Entry criteria: ANA at a titer of ≥1:80 on hep-2 cells or an equivalent positive test (ever)					
⬇					
If absent, do not classify as SLE If present, apply additive criteria					
Additive criteria 1. Do not count a criterion if there is a more likely explanation than SLE. 2. Occurrence of a criterion on at least one occasion is sufficient. 3. SLE classification requires at least one clinical criterion and ≥10 points. 4. Criteria need not occur simultaneously. 5. Within each domain, only the highest weighted criterion is counted towards to the total scores.					
Clinical domain	% criteria	Weight	Immunological domain	% criteria	Weight
Constitutional	Fever	2	Antiphospholipid antibodies	Anticardiolipin antibodies or Anti ß2gp1 antibodies or Lupus anticoagulant	2
Hematologic	Leukopenia Thrombocytopenia Autoimmune hemolysis	3 4 4	Complement proteins	Low C3 or low C4 Low C3 and low C4	3 4
Neuropsychiatric	Delirium Psychosis Seizure	2 3 5	SLE specific antibodies	Anti-ds DNA antibody * or Anti-smith antibody	6
Mucocutaneous	Non scarring alopecia Oral ulcers Subacute cutaneous or discoid lupus Acute cutaneous lupus	2 2 4 6			
Serosal	Pleural or Pericardial effusion Acute pericarditis	5 6			
Musculoskeletal	Joint involvement	6			
Renal	Proteinuria >0.5 g/24 hrs Renal biopsy class ii or class v lupus Nephritis Renal biopsy class iii or class iv lupus nephritis	4 8 10			
Classify as systemic lupus erythematosus with a score of 10 or more if entry criterion fulfilled.					

The classification of lupus nephritis begins with class I as minimal mesangial lupus nephritis to class VI as advanced sclerosis lupus nephritis. Patient had diffused proliferative lupus nephritis which is a class IV lupus.

Treatment of lupus nephritis depends on the severity of the disease. In moderate to severe active nephritis (class III and IV), treatment with high-dose oral steroids and cytotoxic drugs is indicated. Prednisolone is initially given at a dose of 1-2 mg/kg daily for 4-6 weeks and then gradually tapered to a maintenance dose of 0.2-0.3 mg/kg daily, for 2-3 years or more.

Patients with class III and IV lupus nephritis are also given cyclophosphamide, preferably IV every 3-4 weeks for 6-7 pulses, or orally (2 mg/kg daily for 3-4 months). Subsequently cyclophosphamide is replaced either by azathioprine (1-2 mg/kg per day) or MMF. Patients with class V nephritis may be treated with a combination of oral prednisolone and either a calcineurin inhibitor (e.g. cyclosporine) or mycophenolate mofetil. The goal of the therapy is to produce a clinical remission with normalization of renal function and proteinuria, and a serological remission which is normalization of ant ds-DNA antibody, C3 and C4 levels.[7]

CONCLUSION

SLE with renal involvement in a male child is an unusual disease which is not only difficult to diagnose but manage also. The clinicians should be aware of the various manifestations and complications in children with SLE so that appropriate therapy can be initiated early to reduce morbidity and mortality.

ACKNOWLEDGEMENTS

Author would like to thank to all those who provided the possibility to complete this report. A special gratitude to Dr. Ramkesh Meena whose contribution in stimulating suggestions and encouragement, helped to coordinate this project especially in writing this report.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Richa, Sharma S, Abrol P. Case report of a male child with lupus nephritis. Int J Contemp Pediatr. 2020;7:220-3.