International Journal of Contemporary Pediatrics
Aier T et al. Int J Contemp Pediatr. 2025 Jan;12(1):100-104
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20243862

Original Research Article

Clinical and laboratory characteristics of paediatric systemic lupus erythematosus at secondary level hospital among local population in North East India

Tenukala Aier*, Chippy Hanna Joe, Sulanthung Kikon, Wonashi Tsanglao

Department of Pediatrics, Christian Institute of Health Science and Research (CIHSR), Dimapur, Nagaland, India

Received: 15 November 2024
Accepted: 11 December 2024

***Correspondence:**
Dr. Tenukala Aier,
E-mail: tenukala@yahoo.com

ABSTRACT

Background: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation and the presence of circulating auto antibodies directed against self-antigens with flare-ups and remissions. Childhood-onset SLE (cSLE) is a rare disease with an incidence of 0.3-0.9 per 100.000 children-years. The study among paediatric population of north east India were few and hence this study from north east India was undertaken. Objective of this study was to study the clinical and immunological profile of children with systemic lupus erythematosus (SLE). To study the distribution of Renal Lesions according to ISN/RPS Classification of Lupus Nephritis.

Methods: Retrospective observational single centre study at secondary care hospital in North East India. Medical records of children with SLE admitted in Paediatric department from the period of 2018-2024 through the hospital information management were analysed. Clinico pathological features and immunological profile were compared with other studies.

Results: Among 20 patients studied female to male ratio was 3:1. The mean patient's age at the time of presentation was 12.4 years. The mean duration of illness was 8.5 months. Most common systems involved were hematological (85%), followed by kidney (75%) and mucocutaneous (75%). All (100%) cases were ANA positive. 45% were anti smith antibody positive, 20% were anti- dsDNA positive. Focal and segmental proliferative glomerulonephritis (ISN/RPS class III) was the most commonly seen histological pattern, seen in 5 (83%) patients who underwent biopsy. Diffuse proliferative glomerulonephritis (ISN/RPS class IV) was seen in 1 (16%) patient.

Conclusions: Childhood onset SLE is still a challenge to diagnose and manage due to unpredictable clinical manifestations with variable disease activity at different age.

Keywords: Auto antibodies, Lupus nephritis, Systemic lupus erythematosus

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation and the presence of circulating auto antibodies directed against self-antigens with flare-ups and remissions.[1,2] Childhood-onset systemic lupus erythematosus (cSLE) is a term used to refer to individuals who developed SLE before the age of 18 years old.[3] The median age of onset for cSLE is between 11 and 12 years with more severe disease at presentation and it rarely affects children less than five years old.[4] Childhood-onset SLE (cSLE) is a rare disease with an incidence of 0.3-0.9 per 100.000 children-years and a prevalence of 3.3-8.8 per 100.000 children. A female-to-male ratio of approximately 4:1 occurs before puberty and a ratio of 8:1 occurs after puberty.[5,6] Due to the role of estrogen in etiopathogenesis of the disease, SLE is more common in female as compare with male. Genetic factor superimposed on certain environmental factors

plays a very pivotal role in manifesting abnormal immunological response.[6] Common presentations in children are fever, malar rash and cutaneous vasculitis.[7] Among paediatric studies lupus nephritis is common manifestation of SLE at presentation than adults and severity vary among different studies.[8] The study among local ethnic population of north east India were few, here by authors report here the profile of cases in this centre. This study was undertaken to study the clinical and immunological profile of children with systemic lupus erythematosus (SLE) and to study the distribution of renal lesions according to ISN/RPS Classification of Lupus Nephritis.

METHODS

This was a retrospective observational single centre study at secondary care hospital. Study protocol approval was obtained from CIHSR Ethics Committee. All data were retrieved from Hospital Information management (HIM).

Inclusion criteria

Those children who were diagnosed and treated as systemic lupus erythematosus were included in the study. The authors screened retrospectively all patients admitted in paediatric department from the period of July 2018 and March 2024 with discharge diagnosis of SLE and fulfilling the revised American College of Rheumatology (ACR) criteria (1997) for SLE were included in the study.[9] Authors collected information with respect to demographic characteristics, duration of disease and assessment of various organs involvement like cutaneous, musculoskeletal, renal, gastrointestinal tract, nervous and cardiopulmonary.

Data collection

Authors collected data regarding investigation including complete blood count, erythrocyte sedimentation rate (ESR), serum creatinine, serum urea, Urine routine, 24-hour urinary protein, antinuclear antibodies (ANA), anti-double-stranded deoxyribonucleic acid (Ds DNA) antibodies, anti- smith antibodies, Coombs test, C3, C4, Renal Biopsy, chest radiograph and renal biopsy. Medical records of all SLE cases were collected in structured proforma. Clinico pathological features and immunological profile were compared with other studies.

Histological observation will be categorized according to the International Society of Nephrology/Renal Pathology Society (ISN/RPS) 2003 reclassification. There are 6 histological types of LN: 1) minimal mesangial LN, 2) mesangial LN, 3) focal LN, 4) diffuse proliferative LN, 5) membranous LN and 6) glomerulosclerosis.

Statistical analysis

Authors did a descriptive analysis of clinical features present in SLE patients and calculated the mean and percentage frequency of all clinical features, systems involved and histopathological type of nephritis in SLE patients. The data were analysed in Microsoft excel.

RESULTS

The study involved 20 patients (17 from the Naga ethnic tribes and 3 from Assam). The group included 15 girls and 5 boys (Male: Female–1:3). The patients' ages ranged from 9-16 years (Average age 12.4 years). The mean duration of illness was 8.5 months with a range of 0.25-60 months. The most common symptoms were fever (70%) and joint pain (45%). 45% of the cases were hypertensive. Arthritis was seen in 45% of cases. All of them had non erosive, non-deforming and symmetric arthritis.

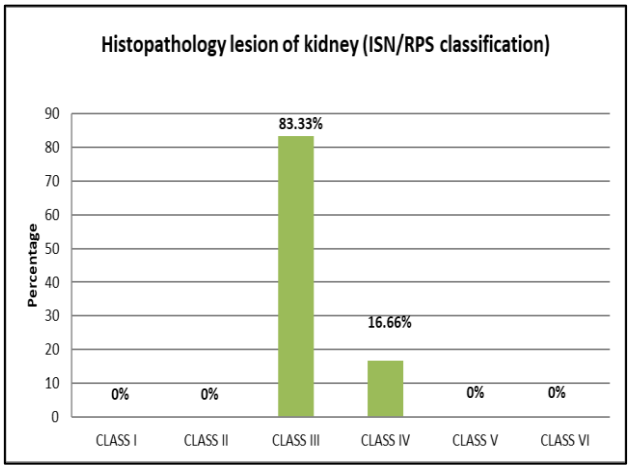

Figure 1: Frequency of histopathology lesions of kidney according to ISN/RPS classification.

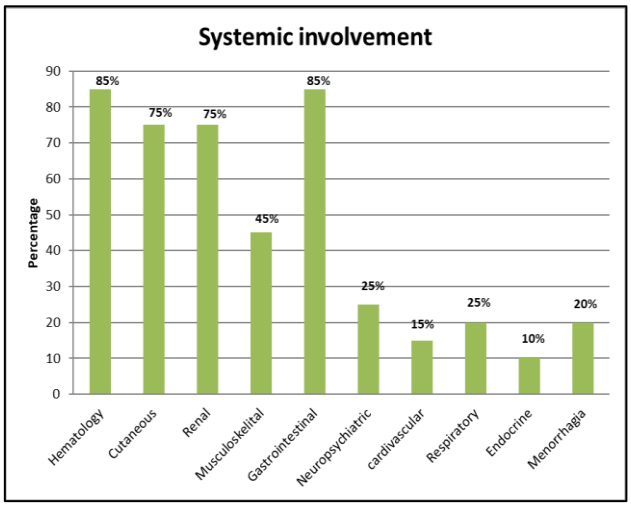

Figure 2: Frequency of various system involvement in SLE.

Haematological manifestations were observed in 17 (85%) of cases. 80% of the patients were anaemic. Autoimmune haemolytic anaemia was found in 7 (35%).

Mild anaemia was found in 3 (15%), moderate anaemia in 5 (25%), severe anaemia was found in 8 (40%).[10] Leucopenia was found in 1 (5%), thrombocytopenia in 9 (45%) cases.

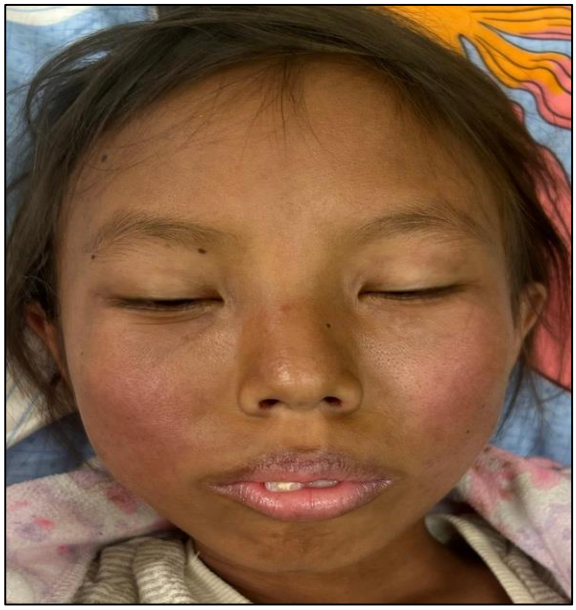

Figure 3: Malar rash, cutaneous manifestation of SLE.

Renal manifestations were observed in 75%. Proteinuria of nephrotic range was seen in 8 (40%) patients and hematuria was seen in 10 (50%). Elevated serum creatinine was seen in 7 (35%) cases. Renal biopsy was performed in 6 patients having indication for biopsy. Focal and segmental proliferative glomerulonephritis (class III) was the most commonly seen histological pattern, seen in 5 (83%) patients who underwent biopsy. Diffuse proliferative glomerulonephritis (class IV) was seen in 1 (16%) patient. The different classes of lupus nephritis according to ISN/RPS classification is graphically depicted (Figure 1). These patients were treated with Glucocorticoids, Cyclophosphamide, Mycophenolate mofetil depending on response. Three patients also received Rituximab.

75% of cases had dermatological manifestations, including malar rash (55%), painless oral ulcers (20%) and cutaneous lesions suggestive of SLE (45%). One patient was diagnosed with vasculitis, another presented with digital gangrene and a third exhibited symptoms of Raynaud's phenomenon. 30% of the patients presented with petechial rashes. Cardiovascular involvement was observed in 3 (15%) patients, of which one presented with severe cardiac tamponade, one had myocarditis (Raised NT-Pro BNP) and the other had pericarditis along with myocarditis. The skin manifestation is shown in (Figure 2) Malar rash. Neuropsychiatric manifestations were present in 5 (25%) patients. Two patients had headache on presentation; two of them had seizures and one patient developed psychosis during the course of treatment. One of our patients had dural venous sinus thrombosis with cerebral edema. Gastrointestinal manifestations were present in 17 (85%) of patients.

Immunological profile

All the patients were ANA positive. 45% were anti smith antibody positive. 20% were anti- dsDNA positive. Both Anti Smith and Anti dsDNA positivity was shown by 5%. Anti histone antibody was positive in a patient with drug induced lupus due to Anti-tubercular therapy.

Table 1: Various clinical manifestation in SLE.

S. no.	Component	N (%)
1	Fever	14 (70)
2	Hematological	17 (85)
	Anaemia	16 (80)
	Leukopenia	1 (5)
	Thrombocytopenia	9 (45)
3	Musculoskeletal	9 (45)
	Arthralgia	9 (45)
4	Neurological	5 (25)
	Headache	2 (10)
	Seizures	2 (10)
	Psychosis	1 (5)
	Sinus venous thrombosis and cerebral edema	1 (5)
5	Renal	15 (75)
	Hematuria	10 (50)
	Significant proteinuria	8 (53)
	Raised creatinine	7 (35)
6	Mucocutaneous	15 (75)
	Malar rash	11 (55)
	Petechia/purpure	6 (30)
	Cutaneous lesions of SLE	2 (45)
	Oral ulcer	4 (20)
	Vasculitis	1 (5)
	Digital gangrene	1 (5)
	Raynaud's phenomenon	1 (5)
7	Cardiovascular	3 (15)
	Cardiac tamponade	1 (5)
	Myocarditis	2 (10)
	Pericarditis	1 (5)
8	Pulmonary	4 (20)
	Pleural effusion	3 (15)
	Hemoptysis	1 (5)
9	Gastrointestinal	17 (85)
	Hepatomegaly	6 (30)
	Splenomegaly	6 (30)
	Ascites	17 (85)
	Gastrointestinal bleed	1 (5)
10	Endocrine	2 (10)
	Vit D deficiency	1 (5)
	Hypothyroidism	1 (5)
11	Menorrhagia	3 (20)

Hepatomegaly and splenomegaly were present in 6 (30%) and 6 (30%) of the cases respectively. 85% patients had ascites which was secondary to nephritis associated with disease progression and flare. One of the patients presented with gastrointestinal bleed. Pleural effusion was present in 3 (15%) of our patients and one had hemoptysis. Hypothyroidism was diagnosed in one patient and was started on thyroxine. Another patient had severe Vitamin D deficiency.[3] patients presented with severe menorrhagia. The frequency of occurrence of various organ involvement described above is graphically depicted (Figure 3) and the same is depicted in detail according to number and percentage of involvement in tabular form (Table 1).

DISCUSSION

We encountered 20 cases whose clinical manifestations ranged from 0.25–60 months before they were diagnosed. The mean duration of illness prior to diagnosis was 8.5 months in our study which was earlier as compared to studies by Singh S et al and Sashidhara VS et al where the duration of illness prior to diagnosis was one year.[11,12] The patients' ages varied from 9-16 years with average age of symptoms onset being 12.4 years which was similar to another study conducted in South India where the average age of symptoms onset was 12.1 years.[8] The female to male ratio was 3:1, which was lower as compared to ratio of approximately 4:1 that occurs before puberty.[13,14]

The most common presenting symptom was fever (70%) which was similar to a study conducted in south India.[12] The most common clinical manifestation was haematological with anaemia in 85% cases, haemolytic anaemia was found in 35% of cases similar to other studies.[11,16] The second most common clinical manifestation were both renal (75%) and mucocutaneous lesions (75%). Studies by Singh et al, 56%, Shashidhara VS et al, 88% and Indira Agarwal et al, 77.1% also reported renal as the second most common clinical manifestation.[11,12,15]

The most common histopathological lesion observed was class III lupus nephritis (83%) followed by class IV lupus nephritis (16%). This finding was different as compared to studies from south India where class IV lupus nephritis was more common.[12,16] Class IV lupus nephritis is the predominant histopathology of lupus nephritis in Asian children (39.4% to 54.0%).[20]

Cardiovascular (CVS) manifestations involving pericardium, myocardium and endocardium are known to occur in up to 30% of children.[1,2] 15% cases had CVS involvement in our study which was lower as compared to studies by Shashidhara et al, (34%) and Dubois et al.[12,17] The incidence of neuropsychiatric manifestations (25%) was similar to other study by Shashidhara et al, (24%) but lower than the Singh S et al, study.[11,12] ANA positivity was seen in 100% of our patients, similar to study by Pradhan S V et al.[18] However, studies by Shashidhara et al and Mondal R et al, had reported 9% of cases being ANA negative.

Anti dsDNA antibody titres were positive in 20% of the cases which was very low as compared to 85% in study by Shashidhara et al.[12] Anti smith was positive in 50% of the cases which was similar to another study by Madhavan R et al.[19] Out of 8 cases whose complement levels were checked 5 cases were low (62%). These finding was low as compared to 92% Hypocomplementemia in studies by Shashidhara et al.[12] Out of 20 patients, 5 (25 %) expired within 2 years of diagnosis. All expired patients had lupus nephritis.

CONCLUSION

To conclude, SLE can present with diverse, unpredictable clinical manifestations with variable disease activity at different age. Childhood onset SLE is still a challenge to diagnose and manage in remote parts of the country due to many unforeseen challenges. The outcome of treatment is also greatly affected due to delay in referral and aggressive nature of the disease when renal system is involved.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

REFERENCES

1. Petty RE, Laxer RM. Systemic lupus erythematosus: In: Cassidy JT, Petty RE, Laxer ML, Lindsley CB, Eds. Textbook of Pediatric Rheumatology, 5th edn. Philadelphia: Elsevier Saunders; 2005:342-91.
2. Hiraki LT, Benseler SM, Tyrrell PN, Hebert D, Harvey E, Silverman ED. Clinical and laboratory characteristics and long-term outcome of pediatric systemic lupus erythematous: a longitudinal study. J Pediatr. 152:2008:550-6.
3. Taxonomy for systemic lupus erythematosus with onset before adulthood. Silva CA, Avcin T, Brunner HI. Arthritis Care Res (Hoboken). 2012;64:1787-93.
4. Clinical features, disease activity and outcomes of Malaysian children with paediatric systemic lupus erythematosus: a cohort from a tertiary centre. Lim SC, Chan EW, Tang SP. Lupus. 2020;29:1106–14.
5. Schur PH. Clinical features of systemic lupus erythematosus. In: Textbook of Rheumatology, 4th edn. Eds. Kelly WN, Harris ED, Ruddy S, Sledge CB. Philadelphia, W.B. Saunders Company. 1993:1017-42.
6. Rebecca E. Sadun, stacy p.ardoin and laura E. Schanberg. Systemic lupus erythematosus: Robert M Kleigmann. Nelson text book of pediatrics. 1 ed south Asia; 2016: 1176-80.

7. Childhood-onset systemic lupus erythematosus: Southeast Asian perspectives. Tang SP, Lim SC, Arkachaisri T. J Clin Med. 2021;10:559.
8. Shashidhara VS. Study of paediatric systemic lupus erythematosus in tertiary care center in South India. Int J Contemp Pediatr. 2020;7(5):1008-12.
9. Hochberg MC. Updating the American college of rheumatology revised criteria for the classification of systemic lupus erythematosus, Arthritis Rheum. 1997;40:1725.
10. WHO. Haemoglobin concentrations for the diagnosis of Anaemia and assessment of severity. Vitamin and Mineral Nutrition Information System. Geneva, World Health Organization, 2011. Available at: https://www.who.int/vmnis/indicator. Accessed on 18 August 2024.
11. Singh S, Kumar L, Khetarpal R, Aggarwal P, Marwaha RK, Minz RW et al. Clinical and immunological profile of SLE: Some unusual features: Indian Pediatr. 1997;2:34.
12. Shashidhara VS, Kumar AH, Kariyappa M. Study of paediatric systemic lupus erythematosus in tertiary care center in South India. Int J Contemp Pediatr 2020;7:1008-12.
13. Schur PH. Clinical features of systemic lupus erythematosus. In: Textbook of Rheumatology, 4th edn. Eds. Kelly WN, Harris ED, Ruddy S, Sledge CB. Philadelphia, W.B. Saunders Company;1993: 1017-42.
14. Rebecca E. Sadun, stacy p.ardoin and laura E. Schanberg. Systemic lupus erythematosus: Robert M Kleigmann. Nelson text book of pediatrics: first south asia ed; 2016: 1176-1180.
15. Yadav V, Bhardwaj P. Clinical and immunological profile of systemic lupus Erythematosus in a pediatric population. North India Egyptian Rheumatol Rehabilit. 2014;41:148-51.
16. Agarwal I, Kumar TS, Ranjini K, Kirubakaran C, Danda D. Clinical features and outcome of systemic lupus erythematosus. Indian Pediatr. 2009;46(8):365.
17. Hematological involvement in pediatric systemic lupus erythematosus: a multi-center study. Akca ÜK, Batu ED, Kısaarslan AP, et al. Lupus. 2021; 30:1983–90.
18. Dubois EL, Tuffanelli DL. Clinical manifestations of SLE. Computer analysis of 520 cases. J Am Med Assoc. 1964;190:104-11.
19. Pradhan SV, Patwardhan M, Rajadhyaksha A, Ghosh K. Clinical and immunological profile of systemic lupus erythematosus. Indian Pediatr. 2013:50:405-7.
20. Madhavan R. The Madras Experience. J Assoc Phys India. 1988;36:481-4.

Cite this article as: Aier T, Joe CH, Kikon S, Tsanglao W Clinical and laboratory characteristics of paediatric systemic lupus erythematosus at secondary level hospital among local population in North East India. Int J Contemp Pediatr 2025;12:100-4.