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ORIGINAL ARTICLE

Clinical and Immunological Profile of Childhood Systemic Lupus Erythematosus: Observational Study

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ABSTRACT

Background: Childhood SLE is a severe autoimmune disease involving multiple systems that effect peoples globally.
Aim: To delineate the spectrum of clinical manifestations of childhood SLE at the Government Children's Hospital Lahore.
Study design: Descriptive observational study.
Methodology: Study was carried out at department of Pediatric medicine, unit 1, Children's hospital Lahore from January 2016 to December 2019. Applying a nonprobability convenience sampling technique, children between 1-16 years of age with childhood systemic lupus erythematosus attending rheumatology outpatient clinic or admitted through emergency were included in study. Written and verbal informed consent was obtained after explanation of study purpose.
Results: There were 59 (78.70%) female and 16 (28.3%) male patients enrolled in the study. Mean age at diagnosis was 10.89±3.4% years. Female predominance was noticeable. Amongst the clinical features fever (96%) was the commonest, followed by oral ulcers (74.70%), alopecia (72%), renal manifestations (69.30%), rash (61.30%), arthritis (56%), and hematological abnormalities (48%), photosensitivity (44%), neurological features (36%) in 24% of patients.
Conclusion: It was concluded that disease was prevalent among all races around the world but in South Asia there are few studies available on its prevalence. Timely diagnosis by a detailed history, physical examination and laboratory evaluation can improve the chances of prolonged disease-free periods and better survival of children.
Keywords: Childhood SLE, ACR and SLICC.

INTRODUCTION

Childhood SLE is a severe autoimmune disease involving multiple systems that effect people globally. According to one estimate, its annual incidence is 0.36-0.9 /100,000 children/ year[1]. Although the onset and pathology along-with its causes still remain a mystery but literature review revealed that Juvenile SLE development depends on environmental and genetic factors. Furthermore, different geographical regions of the world affecting various ethnic backgrounds and both sexes showed variable clinical features[2].

Only minor portion of SLE cases form Juvenile SLE (10-20%) that affect any organ system. Disease pattern is variable from mild to frequent flares requiring long term immunosuppressive therapy leading to considerable organ damage that depends on disease duration and time of onset[3,4].

Pakistan being a land of different socio-cultural backgrounds with people living in climatic extremes experience this disease commonly. Variable presentation is contributed by many reasons like unawareness, false beliefs, lack of medications and poverty. Although data on the clinical and laboratory characteristics is scarce but it is common in south east Asian region[5].

Recent advancement in medical sciences over past few years resulted in better survival of patients having juvenile SLE has improved upto 10 years survival rate. On the other hand patients have lower life expectancy than general population having fourfold greater risk of death[5]. However, juvenile SLE is more lethal in terms of mortality than adult SLE depending on disease duration[6,7]. Even with advancement, its correct diagnosis in children is a challenge for clinicians. Juvenile SLE tends to have a higher frequency of atypical, aggressive manifestations with high rates of organ involvement and subsequent morbidity and mortality[8]. American college of Rheumatology criteria (ACR) was used to diagnose the patients[9,10]. ACR classification was published in1982 and revised in 1997. It requires 4 out of eleven criteria for a patient to be diagnosed as childhood onset SLE.

The objective of the study was to delineate the spectrum of clinical manifestations of childhood SLE at the Government Children's Hospital Lahore.

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METHODOLOGY

Study was carried out in the Department of Pediatric Medicine, Children hospital & ICH Lahore after approval from IRB. It was a descriptive, observational study. Applying a nonprobability convenience sampling technique, children between 1-16 years of age of both sexes were included in the study. Those were either diagnosed as Juvenile SLE for the first time or were already registered in the rheumatology outpatient clinic. Their records were obtained after approval from ethical review board of Children's hospital Lahore. Children with isolated cutaneous lupus, Neonatal SLE, Drug induced lupus and other autoimmune disorders were excluded from the study population. Demographic data, clinical manifestations, laboratory and immunological profile (ANA, anti dsDNA, C3, C4 & direct Coombs test by ELISA technique) were recorded in a predesigned proforma. Written and verbal informed consent was obtained after explanation of study purpose. According to ACR criteria patients were diagnosed in current project[11].

Statistical analysis: Data was analyzed by SPSS v21. The results thus obtained were tabulated after measuring mean, percentage and standard deviation of the findings.

RESULTS

There were 59(78.70%) female and 16(28.3%) male patients enrolled in the study. Mean age at diagnosis was 10.89±3.4% years. The patients were divided in to 4 age groups.

Table-1: Distribution of patients parameters (n=75)

Variables	Groups	Frequency	%age
Age (years)	1-5	05	6.66
	5-10	23	30.6
	10-15	46	61.3
	>15	01	1.33
Mean age at diagnosis (years)	10.89±2		
Gender	Males	16	21.4
	Females	59	78.6

Five (6.66%) were between the age of 2-5 years of age, 23(30.66%) were between the ages of 2-5 years of age, 23(30.66%) were between the ages of 5.1-10 years, 46(61.33%) were between the ages of 10.1 to 15 years of age and only 1

patient (1.33%) was above 15 years of age (Table-1). The frequency of clinical features was estimated and fever (96%) was the commonest manifestation found (Table-2). However, frequency of clinical features was estimated according to ACR criteria.

Table 2: Clinical features of SLE patients

Clinical features	%age
Fever	96
Oral ulcer	74.7
Alopecia	72
Renal	69.3
Rash	61.3
Arthritis	56
Hematological abnormalities:	
Anemia	80.5
Thrombocytopenia	13.9
Leucopenia	5.6
Direct Coombs test	28
Immunological findings	
ANA	92
Anti-Ds DNA	89
Serum C3 &C4	57.3
Photosensitivity	44
Neurological features	36
Serositis	24

DISCUSSION

Systemic lupus erythematosus is an autoimmune disease with diverse clinical manifestations. Renal and neurological manifestations are the most severe ones, associated with an aggressive clinical course, morbidity and mortality[12,13].

On comparison of our study using the clinical components of ACR criteria of child hood SLE with 6 other multicenter studies, we found a female predominance with a female to male ratio of 3.6:1[14-16]. This is probably related to a stronger immune system in women than men and that is why they are prone to autoimmune disorders. Although our results underlie the rarity of lupus under five years of age but if it occurs in younger patients, a more severe disease phenotype and an extremely high mortality rate is expected[17-20].

The commonest constitutional clinical manifestation found was fever 96% that is comparable to other regional studies such as the ones done at Agha Khan hospital Karachi and Bangladesh[16,17]. Amongst the systemic manifestations, renal involvement was the most frequently observed 69.3%, followed by arthritis 56% and neurological manifestations i.e., 36%. A significantly higher frequency of patients with oral ulcers, alopecia and photosensitivity was identified in our study when compared with 5 other multicenter studies[17-20]. This difference is most probably due to the environmental, ethnic and racial differences[2].

Amongst the laboratory and immunological components of ACR criteria of child hood SLE, we found that the neurological, hematological and immunological features were also comparable to 6 other multicenter studies, except anemia, which was significantly higher than those. We can spiculate that this difference is attributable to underlying malnutrition which adds up to the hemolytic process[21]. The results were comparable to our six reference studies.

Limitations: Limitations included limited sample size, time frame, resources and financial constrains.

CONCLUSION

It was concluded that disease was prevalent among all races around the world but in South Asia there are few studies available on its prevalence. Timely diagnosis by a detailed history, physical examination and laboratory evaluation can improve the chances of prolonged disease-free periods and better survival of children.

Author's contribution: SQ&SN: Conceptualized the study, analyzed the data, and formulated the initial draft, **SB&RR:** Contributed to the proof reading, **AM&MYA:** Collected data, **TL:** Contributed to the proofreading the manuscript for intellectual content.

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