RESEARCH BRIEF

Clinical and Immunological Profile of Systemic Lupus Erythematosus

[$]V PRADHAN, M PATWARDHAN, *A RAJADHYAKSHA AND K GHOSH

*From the [$]Department of Autoimmune Disorders, National Institute of Immunohematology, Indian Council of Medical Research; and *Department of Medicine; King Edward Memorial Hospital, Mumbai, India.*

Correspondence to:
Dr Vandana Pradhan,
Department of Autoimmune Disorders,
National Institute of Immunohematology,
Indian Council of Medical Research,
13th floor, KEM Hospital, Parel,
Mumbai 400 012, India.
pradhanv69@rediffmail.com
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Pediatric onset systemic lupus erythematosus (SLE) is not uncommon and female to male ratio varies. Pediatric SLE patients have more severe disease at onset, higher rates of organ involvement and more aggressive clinical course than adults. We compared the clinical and immunological parameters among pediatric SLE and adult SLE from Western India. Twenty five children and 60 adult patients fulfilling American College of Rheumatology SLE criteria were included. Anti-nuclear antibodies, anti-dsDNA and complement (C3, C4) levels were tested. Of 25 pediatric SLE patients studied, 24% showed CNS involvement *vs*. 8.3% in adults SLE (*P*=0.0499). Lupus nephritis was seen in 75% adult patients *vs*. 52% among children. Hepatosplenomegaly was noted more among adult SLE 26.8% *vs* 12% among children. Alopecia was an exclusive features among adult SLE.

Key words: *Autoantibodies, Child, Lupus nephritis, India, Systemic lupus erythematosus*.

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Systemic lupus erythematosus (SLE) predominantly affects young women in their reproductive age [1]. Among 10-20% patients, the diagnosis is made for the first time in the childhood. A female-to-male ratio of approximately 4:1 occurs before puberty, and a ratio of 8:1 occurs after puberty [2-5]. The prevalence of pediatric SLE was reported more among non-Caucasian patients; these patients were significantly younger and more likely to have nephritis [6]. Among Caucasians, its incidence in children is between 6 and 18.9 cases per 100,000 individuals, whereas the prevalence among African–American was 30 cases per 100,000 [7,8].

Children usually have a more malar rash, severe renal disease at onset and higher rates of other organ involvement than adult SLE [9]. This study compares the clinical and serological manifestations of SLE in children and adults.

METHODS

We reviewed the case records of 25 children and 60 adult patients with SLE that were referred to this center. The mean duration of symptoms was less than six months before diagnosis. The diagnosis of SLE was based on the American College of Rheumatology criteria. Ethics committee approval and a written consent was obtained from these patients. Disease activity was assessed at the time of evaluation using the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) [12], and classified as mild, moderate and severe (mild <8, moderate 8-18 and severe >18). Criteria for renal involvement was persistent proteinuria >0.5 g/day. Renal biopsies were examined by light microscopy and classified according to revised WHO criteria [13]. Autoantibodies such as ANA and anti-ds DNA were detected by indirect immunofluorescence test.

Continuous variables were expressed as mean (SD). Pairs of groups were compared using '*t*' test for normally distributed continuous distribution; chi square test was used for the categorical variables. Statistical significance was set at $P<0.05$.

RESULTS

Out of 25 children with SLE, 21 (84%) were girls; female: male ratio was 5.2:1 *vs*. 9:1 in adults. The age was 9.2 years (range 5-14 years) as compared to 20-44 years in adults. Pediatric patients showed an increased central nervous system involvement (6 children 24%) as compared to 8% among adults. Seizures were present in 4 patients and headache was seen in 2/6 patients with CNS involvement. Renal manifestations in the form of lupus nephritis were noted among 13 (52%) children compared to 45 (75%) adult patients (*P*=0.037) (***Table* I**).

In pediatric SLE, 6 patients had severe disease (SLEDAI >18), 14 had moderate disease (SLEDAI 8-18) and 5 patients had mild activity (SLEDAI <8). Among the adult SLE patients, 13 had severe, 36 had moderate and 11 patients, had mild disease activity. Antinuclear

TABLE I CLINICAL MANIFESTATIONS IN CHILDREN AND ADULTS WITH SLE

Manifestations	*Children (n=25)(%)*	*Adult SLE(n=60)*	*P value*
Malar rash	11 (44)	20 (33.3%)	0.35
Cutaneous rash	9 (36)	16 (26.7%)	0.39
Arthritis	15 (60)	34 (56.7%)	0.06
Myositis	8 (32)	18 (30%)	0.86
Fever	4 (16)	18 (30%)	0.18
Neurological involvement	6 (24)	5 (8%)	0.05*
Low Complements levels	5 (20)	24 (40%)	0.07
Kidney involvement	13 (52)	45 (75%)	0.04 *
Hepatosplenomegaly	3 (12)	16 (26.8%)	0.14
Oral Ulcers	3 (12)	16 (26.8%)	0.14

antibodies were present in all pediatric patients; 12 had a homogenous pattern, 7 had speckled pattern, 2 each had homogenous nucleolar and mixed pattern. Anti-dsDNA antibodies were present in 21 children with titer varying from 1:80 to 1:320; children with anti-dsDNA titres above 1:160 showed more severe manifestations. Most 19/25 children with SLE (76%) had reduced complement levels as compared to 34 adults.

DISCUSSION

In different parts of India, clinical manifestations vary in adult and pediatric SLE cases. In Eastern and Northern India, pediatric SLE has higher frequency of malar rash, renal manifestations, hematological and neurological involvements than in adult SLE. On the other hand, photosensitivity and discoid skin lesions are prominent in adult SLE patients [12,13]. Our findings matches with these findings. Studies in different parts of the world have reports more arthritis in pediatric SLE, and arthralgias and myalgias in adult SLE.

Our finding of hepatospenomegaly which was significantly higher in pediatric SLE than in adult SLE had also been reported by White, *et al*. [14]. Studies worldwide have shown a higher incidence of LN in pediatric SLE than adult SLE, whereas our study showed significantly higher incidence of LN (P =0.037) among adults as compared to children. The reason for this could be that clinically the children remain under-diagnosed or late diagnosed. Further our study reported a diffuse proliferative glomerulonephritis (DPGN) in half of the children with LN with higher SLEDAI scores. Similar report had been earlier documented by Bruner, *et al*. [4] in Caucasian SLE children, where higher SLEDAI scores were reported among pediatric SLE.

ANA negative pediatric SLE had been reported in Indian children by Mondal, *et al*. [12] from Eastern India. However, none of our SLE patients were ANA negative. Reduced complement levels in pediatric SLE had been shown in various populations, our study also had similar findings.

India has a diverse population and varied environmental conditions leading to regional differences in clinical manifestations among pediatric SLE. LN cases were diagnosed comparatively late among pediatric SLE and more attention is needed for their early diagnosis for further treatment and management of the disease.

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