

Clinical presentation of pediatric SLE at outpatient clinic

Abstract

Background: Systemic lupus erythematosus (SLE) is a rheumatic disease resulting in damage to any organ in the body. 10-20% of SLE patients diagnosed in childhood.

Objective: To define the pattern of disease expression in pediatric SLE.

Sitting: Outpatient clinic of pediatric SLE of Abu Alrich hospital-Cairo-Egypt.

Subject: 25 pediatric SLE patients who attend the clinic.

Design of study: observational descriptive case series report.

Data collection: Data collected by second author during her residency in Abu Alrich hospital to study rheumatology. The 25 patients randomly selected. She collects medical history information directly from the patients and their parents and reviews their files to collect the other information (e.g.: investigation results and clinical examination and finding during course of the disease).

Results: From 25 patients Female to male ratio 2.3:1. Age at diagnosis range from 4 to 15 years with mean of 9 years. Duration of symptoms before diagnosis established range from one month to 7 years with mean of 8 months. Frequency of SLE classification criteria as follows: +ve ANA 95%, renal disorder: 84%, immunological disorder 60%, photosensitivity 48%, oral ulcers 48%, Hematological disorder 44%, arthritis 36%, serositis 28%, malar rash 12%, discoid rash 4%, neurological disorder 0%. Other clinical manifestation of SLE include fever 80%, arthralgia and anorexia 84% each, hair fall 40%, abdominal pain 36%, other skin manifestation 24%, headache 20%, dyspnea and palpitation 16% each, Reynard's phenomena and lymphadenopathy 12% each. The most frequent hematological abnormality: elevated ESR: 100% anemia: 72%, leucopenia: 44%, thrombocytopenia: 20%, leukocytosis: 12%, thrombocytosis 8%, lymphopenia: 4%.

Conclusion and recommendation: The presentation and the clinical course of SLE have wide variability. Nephropathy, fever, anorexia and arthralgia were the most common presenting clinical manifestations in pediatric patients. There is marked delay between the starting of the disease manifestation and diagnosis, so alertness to the possibility of SLE as diagnosis in patients with variable manifestation should be there.

Keywords: Systemic lupus erythematosus, Rheumatic disease, Classification criteria

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Introduction

Systemic lupus erythematosus (SLE) is a rheumatic disease characterized by autoantibody directed against self-antigens, immune complex formation, and immune deregulation, resulting in damage to essentially any organ. Childhood SLE generally presents between the ages of 4 and 15 years, with girls outnumbering boys 4:1.¹ In 10-20% of SLE patients the diagnosis is made for the first time in childhood.²⁻⁶ The disease can affect any organ but the most frequent are the kidneys, skin, blood vessels and cells, joints, heart and SLE nervous system. The patients may present with many years of symptoms or with acute history of life-threatening disease.⁷⁻⁹ Common initial and chronic complaints include fever, malaise, fatigue, joint pain, and temporary loss of cognitive abilities. Because they are so often seen with other diseases, these signs and symptoms are not part of the diagnostic criteria for SLE. When occurring in conjunction with other signs and symptoms they are considered suggestive.¹⁰ The American College of Rheumatology established eleven criteria as a classificatory instrument in 1982¹¹ which were revised in 1997¹² in clinical trials.

They were not intended to be used to diagnose individuals and do not do well in that capacity. For the purpose of identifying patients

for clinical studies, a person has SLE if any 4 out of 11 symptoms are present simultaneously or serially on two separate occasions without other explanation. Classification criteria include:¹³ 1- malar rash. 2- discoid rash. 3- serositis. 4-oral ulcers. 5- photosensitivity. 6- arthritis. 7- haematological changes (hemolytic anaemia, leucopenia, lymphopenia, thrombocytopenia) 8- Renal disorder. 9- +ve ANA. 10- Immunological disorder (eg: +ve double strand DNA, anti phospholipids anti body, anti smith). 11- CNS changes (eg: seizures, psychosis). The treatment of SLE involves preventing flares and reducing their severity and duration when they occur. It includes corticosteroids and anti-malarial drugs (hydroxychloroquin). Certain types of lupus nephritis such as diffuse proliferative glomerulonephritis require bouts of cytotoxic drugs. These drugs include azathioprin, mycophenolate, cyclophosphamide. Intra venous immunoglobulin also can be used in patients with multi organ involvement.¹⁴⁻¹⁶ Regarding prognosis SLE is incurable, but treatable. In the 1950s, most people diagnosed with SLE lived fewer than five years. Today over 90% survive for more than ten years, and many live relatively asymptotically. 80-90% can expect to live a normal lifespan.¹⁷ We conduct this study to shed light on the clinical presentation of pediatric SLE.

Aim of study

To define the pattern of disease Expression (clinical and investigation) in patients with childhood onset systemic lupus erythematosus.

Patients and method

Settings

S L E Clinic (out patient clinic of pediatric SLE) of Abu Alrich hospital-Cairo Egypt.

Subject

25 pediatric SLE patients who follow the clinic.

Patients and method

Prospective- retrospective observational descriptive case series report.

Data collection

Data collected by second author during her residency in Abu Alrich hospital to study rheumatology. The 25 patients randomly selected. She collects medical history information directly from the patients and their parents and reviews their files to collect the other information (e.g.: investigation results and clinical examination finding during course of the disease).

Some important definitions and information for patient selections:

- All our patients were met the 1982 revised criteria for classification of SLE.
- Onset of disease, defined as the initial manifestation clearly attributable to SLE, occurred before the age of 14 years.
- Age at diagnosis, defined as the age when the patient fulfilled four or more of the 1982 revised criteria for the classification of SLE.
- We include any manifestation developed during time of evolution of the disease, (defined as the time from the onset until the time of data collection).

Results

- From 25 patients 17(68%) were females and 8 (32%) males. Male to female ratio 1: 2.3 (Figure1).

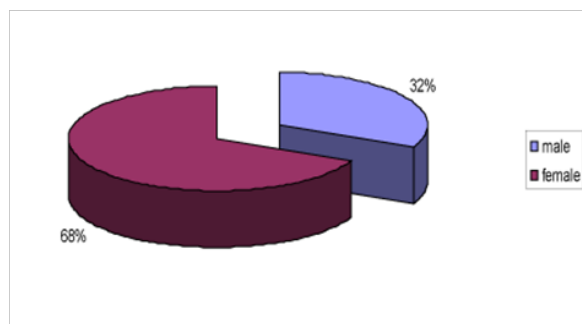


Figure 1 Male to female ratio.

- Age at diagnosis range from 4 to 15 years with mean of 9 years. (Only one patient aged 4 years).

- Duration of symptoms before diagnosis established range from one month to 7 years with mean of 8 months.
- Frequency of SLE classification criteria shown in Table 1.
- Other clinical manifestation of SLE Shown in Table 2.
- The most frequent hematological abnormality Shown in Figure 2.
- Regarding other investigations shown in Table 3.

Table 1 Frequency of 1982 revised criteria for classification of SLE

Criterion	Number of Pts	%
Malar rash	3	12
Discoid rash	1	4
photosensitivity	12	48
Oral ulcers	12	48
Arthritis	9	36
Serositis	7	28
Renal disorder	21	84
Neurological disorder	0	0
Hematological disorder	11	44
Immunological disorder	9/15	60
Antinuclear antibody	21/22	95

Table 2 other clinical manifestation of SLE

Clinical Presentation	Number of Pts	%
Fever	20	80
Arthralgia	12	48
Dyspnea	4	16
Palpitation	4	16
Abdominal Pain	9	36
Anorexia	12	48
Hair Fall	10	40
Raynaud's Phenomena	3	12
Other Skin Manifestation	6	24
Lymphadenopathy	3	12
Headache	5	20

Table 3 Other laboratory abnormality of SLE

Investigation	Number of Pts	Abnormal	Percentage
C3	17	14↓	82
C4	17	12↓	70
Anti double strand DNA	15	9 +ve	60
RFT	25	0	0
LFT	25	8	32

RFT, Renal function test; LFT, liver function test

- viii. 7(28%) patients had urinary complain: oliguria 3, dark colored urine 5, dysuria 1 patient.
- ix. 20 (80%) had urinary changes. The most frequent urinary changes shown in Figure 3.
- x. Renal biopsy done in 13 patients with urinary involvement: finding shown in Figure 4.

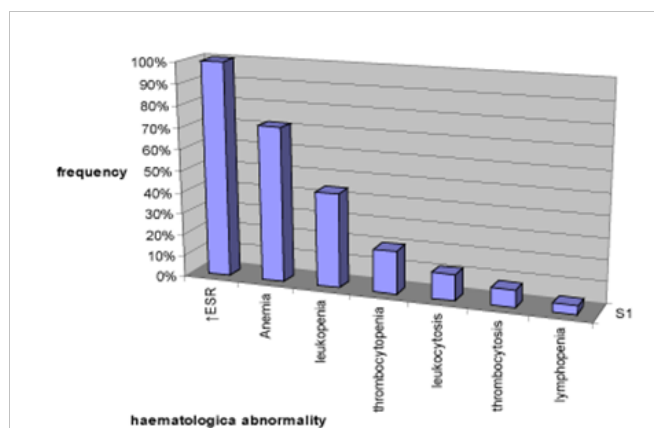


Figure 2 Hematological abnormality of SLE patients.

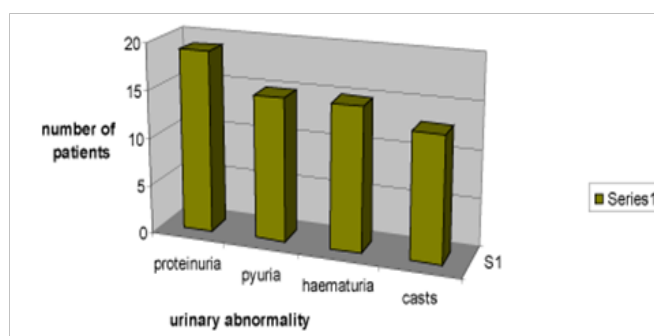


Figure 3 Urinary abnormality of SLE patients.

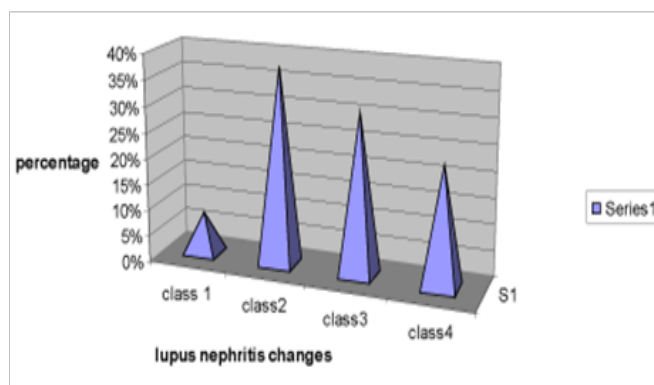


Figure 4 Renal biopsy changes of 15 patients with SLE nephritis.

Discussion

There have been several studies dealing with childhood onset SLE and their results suggested that age at onset modifies the expression of the disease in terms of clinical presentation, pattern of organ

involvement and serological findings.¹⁸ We used 14 years as cut off age as the most often used cut off ages are 14 or 16 years at onset of disease.¹⁹⁻²¹ The onset of SLE is rare before the age of 5 years.²² Mean age 11 years (range 5-14).²³ Regarding age our study show similar results. Female predominant male but our ratio less than others²⁴ may be because we didn't take all of the patients but we choose random sample of them. Clinical presentation of our patients comparable to others²⁵⁻²⁷ but malar rash less and fever more. Our study show high percentage of renal involvement (84%) but lupus nephritis is probably present to some degree in all children with SLE and is a major determinant of a long term outcome.²⁸⁻³⁰ In our study no patient with neuropsychiatric disorder Except for headache which made the most common neuropsychiatric disorder in SLE patients.³¹ Although the existence of a specific lupus headache and the optimal approach to headache in SLE cases remains controversial.³² And that's why we didn't include headache in neuropsychiatric manifestation of SLE. Regarding investigations our results similar to others.^{19,33,34} However, the small number of patients that have been analyzed, the disparity in selection criteria for patient inclusion, or the definition of the variables, and our study didn't include all the SLE patients following the clinic make comparison of findings with previous reports difficult.

Conclusion and recommendation

The presentation and the clinical course of SLE have wide variability. Nephropathy, fever, and arthralgia and anorexia were the most common in pediatric patients as presenting clinical manifestations. There is marked delay between the starting of the disease manifestation and diagnosis, so alertness to the possibility of SLE as diagnosis in patients with variable manifestation should be there.

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