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Medicine

Experience of Acute Systemic Lupus Erythematosus in Indoor and Outdoor Patients

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ABSTRACT

Objective: The aim of this study was to determine spectrum of clinical and Laboratory diagnosis of Systemic lupus erythematosus at tertiary care unit.

Study Design: Descriptive type, (cross sectional).

Place and Duration of Study: This study was conducted at the Department of General medicine and Paediatric Medicine in indoor and outpatients CMC, Shaheed Muhatarma Benazir Bhutto University Medical Larkana from February 2010 to February 2012.

Materials and Methods: We studied prospectively 44 patients with SLE who were seen consecutively either as inpatients or outpatients. All were met the American College of Rheumatology (formerly American Rheumatism Association, ARA) revised criteria for SLE and underwent medical interview as well as routine general physical examination by a researcher, and the laboratory investigations were carried out from single laboratory of Larkana, all these characteristics features of patients were collected in a protocol form. These investigations were includes the CBC, serology for ANA, anti dsDNA and urine for protienuria. X-ray chest and echocardiography was also done for pleural and pericardial effusion.

Results: Forty-four patients fulfilled the ACR criteria for SLE, most common presentation were cutaneous 30 (68.2%) cases, mucocutaneous ulceration 26 (59.1%), Fever 26 cases (59.1%), pallor 47 (67.14), cough 34(48.57%), swelling of body 12(17.14%), headache in 10 (22.7%) and Major physical signs were arthritis and arthralgia 30 (68.2%), Hepatosplenomegaly 20 (45.5%), generalized lymphadenopathy 12(27.3%), pleural effusion 08 (18.2%)and 02 (4.5%)patients had pericardial effusion, Raynaud's phenomenon 10 (22.7%), while direct comb test, ANA, anti dsDNA antibodies were found to be positive in 10 (22.7%), 42 (95.5%), 34(77.3%) cases respectively.

Conclusion: Most patients presented almost universally with fever and arthralgias or arthritis in combination with malar rash or oral ulcers and in some patients a combination of all of the above was observed. A combination of positive anti nuclear antibody test, increased ESR and proteinuria were found to be a sensitive and cost effective set of laboratory findings for the diagnosis of patients suffering from SLE. The set of these clinical and laboratory features would help in the correct and early diagnosis of patients suffering from SLE, a relatively rare disease, in the busy medical outpatient and inpatient departments in our set up.

Key Words: Systemic lupus erythematosus, Arthritis, Arthralgia, Raynaud's phenomenon

INTRODUCTION

Systemic lupus erythematosus (SLE) is a multisystem, autoimmune chronic inflammatory disorder characterized by the formation of antibodies to cellular components; the diagnosis can be established by four of 11 clinical and laboratory criteria which include such disparate manifestations as renal disease, arthritis, hematologic disorders, and skin manifestations – classified as having SLE.[1] As these disparate manifestations have great impact on the disease course, understanding their specific genetic etiology is of prime importance.[2] The disease can affect almost any part of the body and is characterized by remission and relapses. It is most common in women of reproductive age but can present at any age from 1 to 90 years and in men.[3] Antinuclear antibody titer is the primary laboratory test used to diagnose systemic lupus erythematosus. SLE is common connective tissue disorder, a study reported in England a prevalence of 200 cases per 100,000 women (18 to 65 years of age).[4] While in the U.S. prevalence of systemic lupus erythematosus is 40 to 50 cases per 100,000 persons.[5] Worldwide, 15 - 17% of patients present before 16 years of age, with a peak incidence at 10 - 14 years, but it is rare in children less than 4 years of age, and there is a female predominance in adolescence and adulthood.[6] Children usually present with skin rashes, arthritis, and nonspecific constitutional symptoms that may precede the finding of organ-specific disease. The incidence of renal pathology in children is high both on presentation and later in the disease process.[7] The reported data on SLE from our set up is very scanty this study comprises the clinical features of SLE in patients presenting at Larkana.

MATERIALS AND METHODS

Patient Selection: We studied prospectively 44 patients with SLE who were seen consecutively either as inpatients or outpatients between Feb. 2010 to Feb. 2012. All were met the American College of Rheumatology (formerly American Rheumatism Association, ARA)7 revised criteria for SLE.

All underwent medical interview as well as routine general physical examination by a researcher. The laboratory investigations were carried out from single laboratory of Larkana. These investigations included the blood, serum and urine sample from each patient was collected for the CBC, immunological tests and for protienuria respectively. X-ray chest and echocardiography was also done for pleural and pericardial effusion Clinical and serological characteristics of all these patients were collected in a protocol form. Salient features included in this protocol were: (1) age at diagnosis, defined as the age when the patient fulfilled four or more of the 1982 revised ARA criteria for the classification of SLE, (2) cumulative clinical manifestations during the evolution of the disease, and (3) laboratory features at diagnosis. Information collected into the protocol forms was transferred to a computerized database program (SPSS-13).

RESULTS

Forty-four patients fulfilled the ACR criteria. There were 08(18.2%) boys and 36(81.8%) girls, reflecting a ratio of 1:4.5 overall, with a ratio of 1:1.2 under and 1:4 over 10 years. The mean age on presentation was 21.4 years, with a range of 8 - 36 years. (Table 1& 2)

Cutaneous signs were the commonest feature on our presentation 30 (68.2%) (Table 3) and 26 (59.1%) patients had mucocutaneous ulceration. Constitutional features were common in 26 cases (59.1%), but nonspecific Hepatosplenomegaly 20 (45.5%), generalized lymphadenopathy 12(27.3%), arthritis and arthralgia 30 (68.2%) were also common as were central nervous system (CNS) complaints, including persistent headache in 10 (22.7%) cases. Eight patients presented with the pleural effusion and 2 patients had pericardial effusion.

Table No.1: Sex of patients

Sex of Patients		Frequency	Valid Percent	Cumulative Percent
Valid	Male	8	13.2	13.2
	Female	36	81.8	100.0
	Total	44	100.0	

Ten (22.7%) patients suffered from Raynaud's phenomenon, 4 of 10 presented with gangrene. Lab studies show low Hb in twenty (50%) in cases, 14 (31.5%) cases were presented thrombocytopenia, 30 (68.2%) cases presented with low WBC count and albuminurea found in 20 (45.5%) cases while direct comb test, ANA, anti dsDNA antibodies were found to be positive in 10 (22.7%), 42 (95.5%), 34(77.3%) cases respectively. (Table 4)

The time duration from onset of symptoms to diagnosis was not always known. The time duration was available for 25 of the 44 patients (55%). The average time to diagnosis for these 20 patients was 16 months from onset of symptoms.

Table No.2: Age of patients

Sex of Patients	Frequency	Valid Percent	Cumulative Percent
8.00	2	4.5	4.5
9.00	2	4.5	9.1
10.00	2	4.5	13.6
11.00	2	4.5	18.2
12.00	2	4.5	22.7
17.00	2	4.5	27.3
18.00	2	4.5	31.8
19.00	4	9.1	40.9
20.00	2	4.5	45.5
22.00	2	4.5	50.5
25.00	6	13.6	63.6
26.00	8	18.2	81.8
26.50	2	4.5	86.4
30.00	2	4.5	90.9
35.00	2	4.5	95.9
36.00	2	4.5	100.0
Total	44	100.0	

Table No.3: Patients manifestation

Patients manifestation	No	Percentage
Cutaneous features	30	(68.2%)
Mucocutaneous ulceration	26	(59.1%)
Constitutional features (fever, myalgia, fatigue, weakness, weight loss)	26	(59.1%)
Renal disease (nephrotic syndrome)	06	(13.6%)
Arthralgia	30	(68.2%)
Arthritis	30	(68.2%)
Hepatosplenomegaly	20	(45.5%)
Lymphadenopathy	12	(27.3%)
Serositis (pleural effusion)	08	(18.2%)
Pericardial effusion	02	(4.5%)
CNS symptoms (seizures 1, encephalopathy 1, depression 2, hemiparesis 1, chorea 1, headache 4, psychosis 0)	10	(22.7%)
Raynaud's phenomenon (gangrene of feet 4)	10	(22.7%)

Table No.4: Patients manifestation

Patients manifestation	No	Percentage
Low Hb	20	(50%)
Thrombocytopenia	14	(31.5%)
Low WBC	30	(68.2%)
Urine for albuminurea	20	(45.5%)
Direct comb test,	10	(22.7%)
ANA	42	(95.5%)
Anti dsDNA Antibodies	34	(77.3%)

DISCUSSION

Systemic lupus erythematosus (SLE) is not uncommon problem in our population.

It is a multisystem autoimmune connective tissue disorder with a variable clinical presentations. There is a peak age of onset among young women between the late teens and early 40s, and a female to male ratio of 9:1.[1,2,8]

Diagnoses of SLE have increasing and it seems attribute to improved diagnostic tests and the recognition of mild disease manifestation. Although constitutional symptoms, arthritis, arthralgia and rash remain the most frequent signs of SLE, there is also an increase in the frequency of serositis and haematocytopenias in older patients. The most frequent presentation SLE in our population is with constitutional symptoms arthritis and arthralgias followed by renal symptoms, malar rash, serositis, and neurological symptoms. These clinical features were even more frequent in overall Asian populations.[3,4,9]

Forty-four patient were included in our study, from which mostly were >18years (68.2%) and <18years (31.8%), male comprises (18.2%) and female were (81.8%), reflecting a ratio of 1:4.5 overall, with a ratio of 1:1.2 under and 1:4 over 10 years. The mean age on presentation was 21.4 years, with a range of 8 - 36 years consistent with other studies in Pakisktan (Rabbani MA. et al).[10]

Skin and mucous membranes 140/198 (70%) were commonest (46%) compairable with the result reported by (Rabbani MA et al)10, followed by arthritis (38%), nephritis (36%), pericarditis (12%), lung involvement (17%) and CNS involvement (30%). 83% Patients were detected to have hematological disturbances with anemia (71%), leukopenia (20%) and thrombocytopenia (26%). ESR was raised in nearly 100% patients. Other laboratory findings were included positive ANA (93%), anti dsDNA (83%), protienuria (24%), and RBC and casts in the urine (32%).

In our study, cutaneous manifestation were the commonest feature 30 (68.2%) (Table 3) and twenty six (59.1%) patients found to have mucocutaneous ulceration. Constitutional features were common in 26 cases (59.1%), but nonspecific Hepatosplenomegaly in 20 (45.5%), generalized lymphadenopathy 12(27.3%), arthritis and arthralgia 30 (68.2%) were also common as were central nervous system (CNS) complaints, including persistent headache in 10 (22.7%) cases. Eight patients (18.2%) presented with the pleural effusion and 2(4.5%) patients had pericardial effusion. Ten (22.7%) patients suffered from Raynaud's phenomenon, 4 of this 10 presented with gangrene.

Lab studies show low Hb in twenty (50%) in cases, 14 (31.5%) cases were presented thrombocytopenia, 30 (68.2%) cases presented with low WBC count and albuminuria found in 20 (45.5%) cases while direct comb test, ANA, anti dsDNA antibodies were found to be positive in 10 (22.7%), 42 (95.5%), 34(77.3%) cases respectively and similar figures have been shown by (Faller G et al).[7,10]

In study by (Meunier BB, et al) the commonest presentation was hematologic (72%), cutaneous (70%), musculoskeletal (64%), renal (50%), and fever (58%) the erythrocyte sedimentation rate was raised in 100%. While Antinuclear antibodies in 97% an, anti-double stranded DNA antibodies in 93% were found.[11]

A study in Aga Khan University Hospital by (Rabbani MA et al) 86.9% was females and only 12.1% were male patients. While Malar rash was present in 29%, alopecia in 22% patients, 53% patients were febrile, and serosal involvement was noted in 29% patients. Pleural effusion was found in 19% patients and pericardial effusion in 9%.While arthralgias were noted in all patients, and arthritis in 38% patients and CNS involvement in 29% patients. Lab wise Complete blood count revealed leucopenia in 22% and thrombocytopenia in 26%. In renal involvement proteinuria was found in 74%. ANA in 86% patients, Anti-dsDNA test in 74% while 50% tested positive for anti-Sm antibodies.[9]

CONCLUSION

Number of limitations were found during the course of the study mostly which were related to nature of the study. This is hospital based study confined to one tertiary care unit; this spectrum could not be generalized to the entire country. Therefore it can be concluded that the classification set forth by the American rheumatology association in I 984 is applicable to SLE patients in our setting. The patients data used in our study provided us with information regarding the clinical presentation of SLE, most of which fitted into ACR criteria. Thus it is recommended, that doctors continue to use those criteria to help in diagnosis of SLE.

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