ORIGINAL ARTICLE

VARIOUS MANIFESTATIONS OF SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) PRESENTING AT THE RHEUMATOLOGY CLINIC IN SOUTHERN PAKISTAN

Samar Abbas Jaffri [1], Tasnim Ahsan[2], Syeda Rida E Zehra[1], Sadia Sultan[3]

[1]Department of Medicine, Liaquat National Hospital & Medical College, Karachi - Pakistan

[2]Department of Medicine, Jinnah Postgraduate Medical Centre (JPMC), Karachi - Pakistan

[3]Laboratory services, National Medical Centre, Karachi - Pakistan

ABSTRACT

Objective: To know the various manifestations of Systemic Lupus Erythematosus in South of Pakistan.

Material and Methods: It was an observational, prospective study conducted at the Rheumatology clinic at medical unit II, Jinnah Postgraduate Medical Centre (JPMC), Karachi, Pakistan. The duration of the study was from February 2004 to February 2014.The inclusion criterion for this study was all patients with the SLE diagnosis according to American College Rheumatology (ACR) criteria.

Results: The total number of patients registered in a Rheumatology Clinic during the study period was 635, 42 (6.6%) were diagnosed with SLE. The mean age was 25.40 years. Nearly half of the patients presented in their third decade of life. The most common clinical manifestation was arthritis/arthralgia in 38 patients (90.5%) followed by malar rash in 34 (81%), oral ulcers in 15 (35.7%), lupus nephritis in 8 (19.5%), lupus cerebritis in 7 (16.7%) and serositis in 4 patients (9.6%).

Conclusions: The SLE patient's data in the clinic are similar to SLE data worldwide. The most common SLE clinical manifestation seen in the study's clinic is arthritis/arthralgia. A high index of suspicion is required for early diagnosis & prompt treatment to prevent life-threatening and disabling complications.

Keywords: Systemic lupus erythematosus, Rheumatological, Arthritis.

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INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that has protean manifestations and follows a relapsing and remitting course. More than 90% cases of SLE occur in women. The classic presentation of SLE is the triad of fever, joint pain, and rash. Women of childbearing age with the above-mentioned symptoms should require prompt investigation to reach the diagnosis[1]. Many of its clinical manifestations are secondary to the trapping of antigen-antibody complexes in capillaries of visceral structures or to autoantibody-mediated destruction of the host cell.

In the Asian population, a similar prevalence of SLE clinical manifestations exists; most commonly involving the skin & musculoskeletal system. Diagnosis of SLE requires clinical and serologic criteria. The female to male ratio peaks at 11:1 Error! Reference source not found. A correlation between age and incidence of SLE mirrors peak years of female sex hormone production. The onset of SLE is usually after puberty, typically in the third and fourth decade, with 20% of all cases diagnosed during the first 2 decades of life. There are some well-known studies about the mechanisms of SLE and its genetic associations[3,4,5]. At least 35 genes are known to increase the risk of SLE[4]. A genetic predisposition is supported by 40% concordance in monozygotic twins; if a mother has SLE, her daughter's risk of developing the disease has been estimated to be 1:40, and her son's risk, 1:250. Being the largest continent by area and population, Asia has different people of socio-economic culture. Studies have shown that Asian and

Dr. Sadia Sultan (Corresponding Author)
Consultant Hematologist
Laboratory Services, National Medical Centre, Karachi - Pakistan
E-mail: sadiasultan96@yahoo.com
Contact: +92 - 300-2071894
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black populations have the highest prevalence of SLE compared to the white populations. In this study, the clinical characteristics prevalence and SLE manifestations in JPMC, Karachi, Pakistan was discussed.

MATERIAL AND METHODS

It was an observational, prospective study conducted at the Rheumatology clinic at medical unit II, Jinnah Postgraduate Medical Centre (JPMC), Karachi, Pakistan. The duration of the study was from February 2004 to February 2014. The total numbers of patients registered in the Rheumatology clinic at JPMC was 602, out of which 42 were with the diagnosis of SLE. The inclusion criteria for this study was, all patients with the SLE diagnosis according to American College Rheumatology (ACR) criteria: malar rash, discoid rash, photosensitivity, oral ulcers, arthritis, serositis including pleuritis (history of pleuritic pain or rub, or pleural effusion) or pericarditis (documented by ECG or pericardial rub or evidence of pericardial effusion on echocardiography), renal disorder (elevated serum creatinine or persistent proteinuria >0.5 g/day or active cellular casts), neurological disorder (seizures or psychosis in the absence of other causes), haematological disorder (haemolytic anaemia with reticulocytosis or leucopenia defined as white cell count <4000/3mm on two or more occasions or lymphopenia defined as lymphocyte count <1500/mm3 on two or more occasions, or thrombocytopenia defined as platelet count <100000/mm3 not due to drugs or infection, immunologic disorder demonstrated by presence of anti-double stranded DNA (anti-ds-DNA), or anti smith nuclear antigen (Anti-Sm), antinuclear antibody (ANA) or antiphospholipid antibodies (APS). A patient was diagnosed with SLE if four of the above criteria were met. Clinical and laboratory data were collected according to an established protocol, which included a standard haematological and immunologic profile (complete blood count, erythrocyte sedimentation rate, serum electrolytes, serum creatinine, 24 hours urinary protein and creatinine clearance and various immunological tests including ANA, antibodies to double-stranded DNA (ds-DNA). This study was conducted in the Jinnah Postgraduate Medical Centre (JPMC), Karachi, Pakistan. Patients meeting inclusion criteria were enrolled in the study from the department of Medicine, JPMC. Informed consent was obtained from all the patients for assigning them to the study and using their data in research. Detailed clinical History and Physical Examination was done. Data was analyzed on SPSS Version 20.

RESULTS

The total number of patients registered in a Rheumatology Clinic during the study period was 635, 42 (6.6 %) were diagnosed with SLE. The female to male ratio was 13:1 and the mean age was 25.40 years (figure-1). Nearly half of the patients presented in their third decade of life. The most common clinical manifestation was arthritis/arthralgia in 38 patients (90.5%) followed by malar rash in 34 (81%), oral ulcers in 15 (35.7%), lupus nephritis in 8 (19.5%), lupus cerebritis in 7 (16.7%) and serositis in 4 patients (9.6%). On laboratory, the most common finding was a positive ANA in 38 patients (90.5%), followed by Anti-dsDNA in 34 (81%), lymphopenia in 15 patients (35.7%). Renal biopsy was performed in only 2 cases, which showed diffuse proliferative glomerulonephritis in both cases.

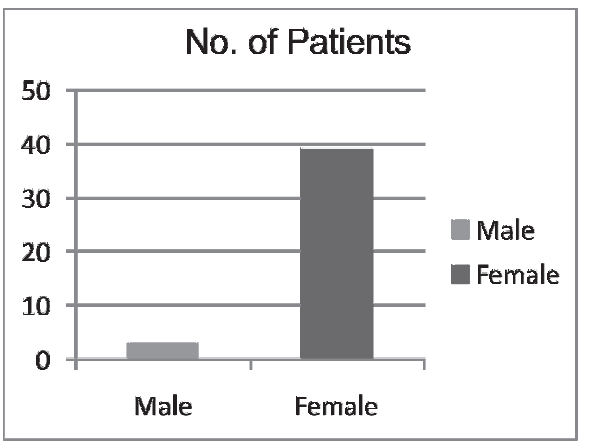


Fig 1

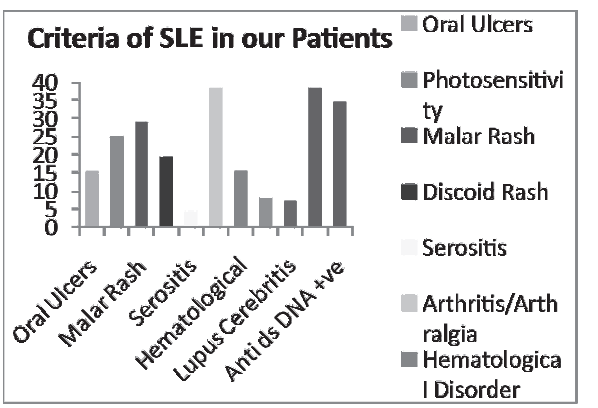


Fig 2

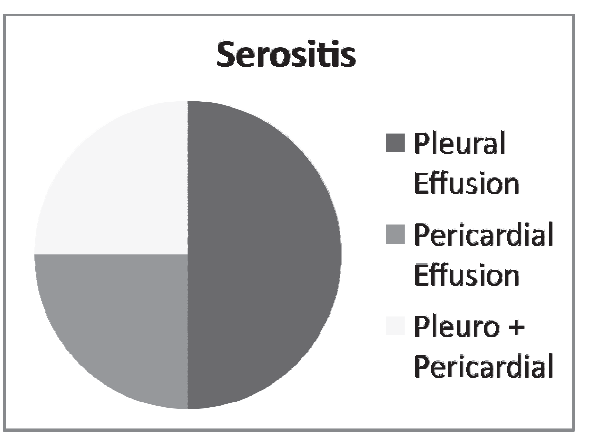
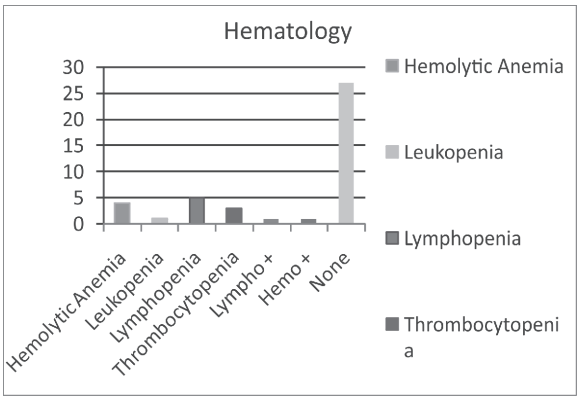


Fig 3



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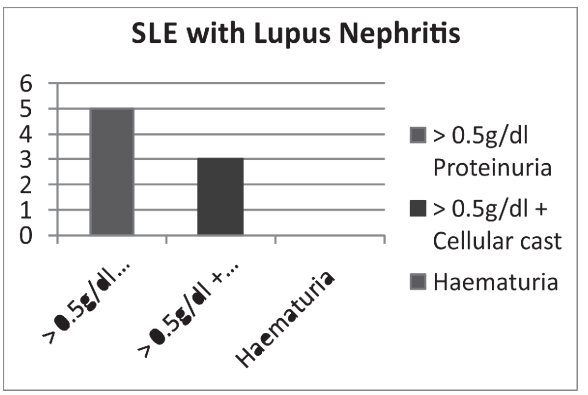

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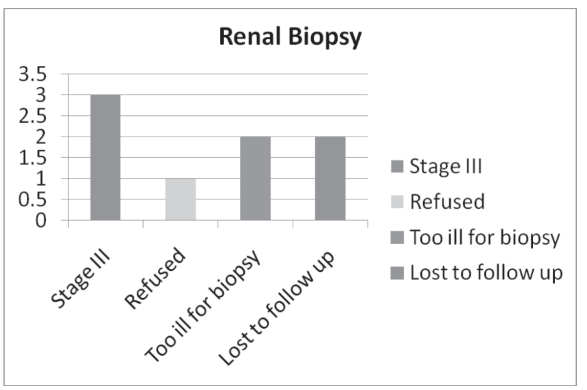

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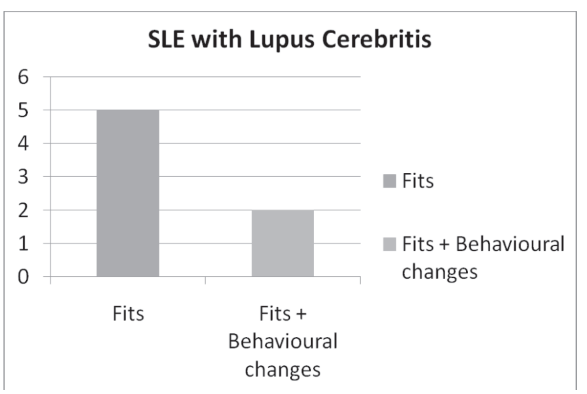

Fig 7

DISCUSSION

SLE is a chronic, multifaceted autoimmune inflammatory disease that can affect any part of the body. SLE is a disease of unknown etiology with a variety of presenting features and manifestations. The pathology majority in SLE relates to deposits of immune complexes in various organs. Diagnosis can be difficult because SLE mimics many other diseases; it requires clinical and serologic criteria. Systemic lupus erythematosus (SLE) is a disease that can affect people of all ages, ethnic groups, and both sexes, but more than 90% of new patients presenting with SLE are women in the childbearing age[6].

SLE is a disease that affects multiple systems. Joint pain is one of the most common reasons for the initial clinical presentation in patients with SLE. Arthritis and arthralgia have been noted in up to 95 percent of patients with SLE [7].

Approximately 20% of them have skin lesions as an initial presentation. Pathognomonic lupus or butterfly rash across the nose occurs in only 30% of patients with SLE. The kidney is the most commonly involved visceral organs in SLE. Although only approximately 50% of patients with SLE develop clinically evident renal disease, biopsy studies demonstrate some degree of renal involvement in almost all patients. The neurological manifestations of lupus are reported in 25 to 75% of patients and can involve all parts of the nervous system. Pleurisy with pleuritic chest pain with or without pleural effusions is the most common feature of acute pulmonary involvement in SLE. Thyroid dysfunction is more frequent in SLE patients than the general population and may have a genetic basis; 3%-24 % of patients with lupus also have autoimmune thyroid disease. Leukopaenia and, more specifically, lymphopaenia are common in SLE[8, 9].

A study of 928 patients with diagnosed SLE, the mean age is 31.4 years in young age with 5 & 10 years survival rate of 99.5% & 99.9% respectively. In the late onset group, the mean age is 51.7 years with 5 & 10 years survival rate of 98.1% & 91 % respectively[10]. In this study in JPMC, Karachi, Pakistan, SLE was more frequent in the 3rd decade of age group with the mean age group of 25.40 years.

In the study of 42 SLE patients, the most common clinical manifestation was arthritis in 85% of patients.

Skin manifestation (malar rash as more common) was found in 71%. Patients with haematological manifestation were around 35% of which lymphopaenia was seen in 14.2%. Lupus nephritis & cerebritis was found in 19% each respectively. ANA was positive in 90.47% & anti-dsDNA in 80.9% of patients.

The overall prevalence of major neuropsychiatric SLE varies among series. It ranges widely between 22% and 95% in children to reflect variable diagnostic criteria and the difficulty in defining psychiatric abnormalities as well as differences in the selection of patients for study[9].

One of the studies of medical records of 239 male and 2116 female patients with SLE in Iran showed initial manifestations in men were musculoskeletal (44%) and mucocutaneous involvements (43.5%). During the course of the disease, arthritis (61.1%) and malar rash (59%) were the most common manifestations in men[12].

A Study conducted on the diagnosis of SLE in 16 years (1996 – 2012) in Turkey, the majority of the 428 patients diagnosed with SLE within the study period were females. The most common clinical manifestations were arthritis, photosensitivity and Raynaud's phenomenon. Thrombocytopenia was found in 18% of patients diagnosed with SLE, 96.7% of SLE patients had a positive ANA. Lupus nephritis was found in 30% of patients[13].

Eighty-nine patients (82%) had haematological manifestations at presentation out of which thirty-eight only had a hematological abnormality as the first manifestation in one study conducted in India. The next common presentation was arthritis in 44 patients (40.7%) followed by lupus nephritis in 25 patients (23%) [14].

In a retrospective study of Filipino SLE patients, the most common presenting manifestation was arthritis (68%), followed by malar rash (49%), renal involvement (47%), photosensitivity (33%), and oral ulcers (33%). The following clinical syndromes were recorded prior to or during SLE diagnosis; nephrotic syndrome in 30%, renal manifestations in 47%[15].

A total of 624 cases of SLE were identified during the 27-year period in one of the study conducted in Saudi Arabia. The study cohort comprised of 566 females (90.7%) and 58 males (9.3%). The most frequent were hematological abnormalities occurring in 516 (82.7%), followed by arthritis in 502 (80.4%) and mucocutaneous symptoms in 401 (64.3%) patients. Malar rash occurred in 299 (47.9%), discoid rash in 110 (17.6%), lymphopenia (40.3%), and leucopenia (30.1%). Regarding the prevalence of different antibodies, it was discovered that ANA was positive in 622 (99.7%) patients[16].

CONCLUSION

The most common SLE clinical manifestation seen is arthritis/arthralgia. Younger patients having evidence of inflammatory arthritis should be evaluated for the possibility of SLE. A high index of suspicion is required for early diagnosis & prompt treatment to prevent life-threatening and disabling complications.

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AUTHOR'S CONTRIBUTION

Following authors have made substantial contributions to the manuscript as under:

Jaffri SA: Contributed substantially to the conception and design of the study, the acquisition of data,analysis and interpretation of data.

Ahsan T: Drafted and critical revised the article.

Zehra SRE: Manuscript writing and final approval of the version to publish.

Sultan S: Manuscript writing and final approval of the version to publish.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.