Original Article

Systemic lupus erythematosus clinical spectrum at King Abdul Aziz Specialists Hospital, Taif, Saudi Arabia

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Objective: To study the clinical spectrum of systemic lupus patients and outcome at King Abdul Aziz Specialists Hospital, Taif.

Methodology: This retrospective study was conducted at Department of Nephrology, King Abdul Aziz Specialists Hospital, Taif. It included 110 patients with age limit of 14 to 60 years of both genders. Patients diagnosed with SLE either initial presentation or during follow up were reviewed.

Results: Out of 110 patients, 101 were female and 9 males (F:M::9:1), with majority between 14 to 40 years of age. Only 25% of patients were diagnosed with SLE at initial presentation and another 25% within one year. In 50%, the time period from initial presentation to diagnosis varied between 1 to 20 years. Presenting clinical features were rheumatological (joint/skin) in 70 (63%), renal in 56(51%), hematological in 36(33%) mucocutaneous in 26 (24%), neuropsychiatric in 14 (13%) and cardiovascular in 18(16%) patients. On follow up, 75% patients had involvements of another organ not present initially. Common affection was renal in 20, pulmonary hemorrhage in 6, seizures in 8, and serositis in 8 patients. Renal presentation comprised of nephrotic syndrome in 18, rapidly progressive glomerulonephritis (RPGN) in 12, proteinuria in 8, end stage renal disease in 8, and acute nephritic syndrome in 6 patients. On follow up, another 20 patients had renal involvement i.e. nephrotic syndrome in 14, RPGN in 4 and proteinuria in two. Treatment comprised of steroids (oral±pulse) in 52, mycophenolate mofetil in 25, hydroxychloroquine in 15, cyclophosphamide pulses in 10, and other in 6 patients in different combinations. At the time of conclusion of study, 38 patients were in remission and 10 on dialysis. Causes of mortality in 10 patients were severe disease in 3, pulmonary hemorrhage in 5 and severe thrombocytopenia one and sepsis one each.

Conclusion: Diagnosis of SLE was delayed in significant number of patients. Clinical renal involvement during follow up approached 70%. Pulmonary hemorrhage and severe disease was cause of mortality. Management was still not uniform. (Rawal Med J 202;45:253-256).

Keywords: SLE, renal, steroids, pulmonary hemorrhage, ESRD.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a heterogeneous multisystem disorder categorized by autoantibody production and diverse range of clinical features. The mechanism by which it is caused is multifactorial, involving genetic, hormonal and environmental factors. Although the occurrence of SLE has almost tripled, improvement in the survival is statistically remarkable in past four decades.[1] Distribution of SLE is worldwide and is more common in black patients. It principally occurs amongst women of childbearing age with the ratio of female: male 9:1. Caribbean, Chinese. Asians and South American Indian women are mainly affected.[2] Although, it is present in many regions of world, its prevalence, range and nature of the disease vary worldwide.[3] ACR Criteria for Diagnosis of Lupus includes the presence of four or more of the following signs and symptoms to help attain 96% sensitivity for the diagnosis. Discoid rash, malar rash, photosensitivity, oral ulcers, pleuropericarditis, non-erosive arthritis, renal disease (proteinuria and/or cellular casts), Hematologic disorder, Central nervous system involvement (seizures or psychosis in the absence of precipitating circumstances), raised anti-DNA antibody, , false-positive antitreponemal test; Positive fluorescent antinuclear antibody test, Positive LE cell preparation, anti-Sm present. The

aim of this study was to look into different parameters of SLE at our institution.

METHODOLOGY

We included 110 cases in this study, with age from 14 to 60 years of both genders. Sample population was composed of those patients who were identified with SLE either at initial presentation or during follow up. They fulfilled the clinical and laboratory criteria of the American Rheumatism Association (ARA). King Abdul Aziz Specialists Hospital, Taif is one of the leading tertiary care hospital, which is accredited by Joint Commission International (JCI), and recognized by Saudi Commission for health specialties for Saudi board training. It is a teaching hospital of Taif University Medical College, Taif.

The laboratory data collected included erythrocyte sedimentation rate, serum creatinine, 24 h urinary protein and creatinine clearance, complete blood count, serum electrolytes. Antinuclear antibodies (ANA), antibodies to double-stranded DNA (dsDNA), rheumatoid factor (RF), anti-Smith (Sm) antibodies, anti-neutrophilic cytoplasmic antibodies (ANCA), antiphospholipid antibodies and serum complements levels were also considered. If there was indication, then radiographic studies were done. Biopsies were also recorded. The clinical course of the disease, their complications and its effect on morbidity and mortality were observed. These outcomes were then equated with other studies.

RESULTS

Out of 110 patients, there were 101 female and 09 males (F:M:9:1), with majority of patients between 14 to 40 years of age. Only 25% of patients were diagnosed with SLE at initial presentation and another 25% within one year. In 50%, the time period from initial presentation to diagnosis varied between 1 to 20 years. Presenting clinical features were Rheumatological (joint/skin) in 70(63%) patients, renal in 56(51%), hematological in 36(33%), mucocutaneous in 26(24%), neuropsychiatric in 14 (13%) and cardiovascular in 18(16%) patients. On follow up, 75% patients had involvements of another organ not present initially.

Common affection was renal in 20, pulmonary hemorrhage in 6, seizures in 8, and serositis in 8 patients. Clinical renal presenting features comprised of nephrotic syndrome in 18, rapidly progressive glomerulonephritis (RPGN) in 12, proteinuria in 8, end stage renal disease (ESRD) in 8, and acute nephritic syndrome in 6 patients. On follow up, another 20 patients had renal involvement i.e. nephrotic syndrome 14, RPGN 4 and proteinuria in two.

Treatment modalities comprised of steroids (Oral ±pulse) in 52, mycophenolate mofetil in 25, hydroxychloroquine in 15, cyclophosamide pulses 10 and different combinations in 6 patients. At the time of conclusion of study, 38 patients were in remission and 10 were on dialysis. Out of mortality in 10 patients, three had severe disease, five had pulmonary hemorrhage, and severe thrombocytopenia and sepsis was cause in one each.

DISCUSSION

There are 10.4 million expats in Saudi Arabia. This leads to its 3rd ranking for coping with foreign natives among other countries. Few studies are available which cover different aspects of SLE from different regions of the Kingdom.[4] All of the studies done in Arab countries are affirming that SLE is not uncommon in Arab world.[5] For instance, a retrospective study in Abu Dhabi reported that 71 patients were affected by SLE over a period of 12-year. Further deep into it, it showed that 56 patients had SLE while 15 were considered to have borderline SLE. Age and gender distribution were same in two subgroups.[6] Other studies on same topic had almost similar findings as in our study. Female gender was 91.8% of total population in our study, which is similar to other studies. Other clinical features of our study were also equivalent to international studies.[5,6]

SLE usually presents with different clinical features in different geographical groups and among different racial groups.[7] The fact that race is major predictor of clinical features, laboratory tests and disease related outcomes, is highlighted by The Hopkins Lupus Cohort, a prospective longitudinal study of SLE outcomes.[8] This finding was endorsed by several studies which showed that SLE had a higher prevalence in blacks of USA,[9,10] Jamaica,[11] native

Americans, especially Sioux Indians,[12] Polynesians,[13] Chinese,[14,15] and Asian Indians[16] than in whites. It was found that even the mortality was significant in African Americans,[17] Chinese[18] and Polynesians in New Zealand.[10] Variations between Caucasians and Chinese in morbidity, clinical features and laboratory findings were reported by Julian et al.[19]

The etiology of SLE is unknown, but it is known that genetics plays an important role particularly in Saudi Arabia, because consanguineous marriages constitute bulk of marriage's proportion. Qari et al identified that five out of seven SLE positive families had consanguineous marriages and they were transmitted through next generation by autosomal recessive mode.[20]

As this study was restricted to one tertiary care center, generalization cannot be done to the whole Saudi Arabia. However, SLE is much prevalent in young females of child-bearing age in Saudi Arabia. Though clinical spectrum was broad, still renal disease and nonspecific constitutional features occupied the spectrum in our and other International studies. Outcome and prognosis varies with infections and activity of disease. Obviously, prospective studies in collaboration with other regions might come up with further trends and details of disease.

CONCLUSION

Diagnosis of SLE is delayed in significant number of patients. Clinical renal involvement during follow up approaches 70%. Pulmonary hemorrhage and severe disease signify mortality. Management is still not uniform.

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