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General Medicine

EVALUATION OF CLINICAL PROFILE OF SYSTEMIC LUPUS ERYTHEMATOSUS

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ABSTRACT

Objective: To evaluate the patterns and clinical characteristics of Systemic Lupus Erythematosus patients.

Methods: This was an observational cross-sectional study conducted at a tertiary care centre in Mumbai. Patients who were diagnosed with Systemic Lupus Erythematosus (SLE) were enrolled in the study from 01 Jan 16 to 31 July 16. After history & clinical examination these patients were subjected to various diagnostic procedures so as to confirm the diagnosis of SLE. The clinical and immunological profile of all these patients was mapped to have better understanding of the disease pattern.

Results: A total number of 11 SLE patients were enrolled with a mean age of 28.5 years. Gender distribution was found to be female preponderant (72.7%). Patients had involvement of various systems arthritis/arthralgia (64%), nephritis (45%), skin involvement/rashes (18%), adenitis (9%) and edema (9%). The patients were positive for antibodies like ANA, Anti Sm, Anti Ro, Anti La, U1RNP, Ro52 etc. As a standard of care, all the patients were previously taking dual or triple therapy comprising of a steroid with one or two of immunosuppressant for a mean duration of 2.9 years. The disease activity of these patients was evaluated using SLEDAI (SLE disease activity index); the mean value was 5.64. Consequently, all these patients were given steroids with steroid sparing agents to which they showed good to moderate response.

Conclusions: The observations of present study suggest that SLE patients may present with a wide variety of manifestations. Vigilant evaluation is required to diagnose the disease and its clinical pattern. Steroids and other immuno-suppressants can play an important role in management of these patients.

KEYWORDS : Systemic Lupus Erythematosus, Immune-suppressants, SLEDAI

INTRODUCTION:

Systemic lupus erythematosus (SLE), a heterogeneous autoimmune disease, may engage many different organs and show a variable clinical course (1). The tremendous heterogeneity of the disease has led some researchers to propose that SLE should be considered a syndrome rather than a single disease(2). It remains one of the most frequent systemic rheumatic diseases and concerns for the rheumatologists.

The incidence of lupus has nearly tripled in the last 40 years, mainly due to improved diagnosis of mild disease. Estimated incidence rates in North America, South America, and Europe range from 2 to 8 per 100 000 per year. Women are affected nine times more frequently than men and African American and Latin American mestizos are affected much more frequently than Caucasians, and have higher disease morbidity. The disease appears to be more common in urban than rural areas (2). There is anecdotal epidemiologic information regarding systemic lupus erythematosus among countries in Asia where prevalence rates generally fall within 30—50/100,000 population (3). The disease expression is largely influenced by the combined effect of genetic, environmental, demographic and geographical factors (4) Significant variation has been observed regarding various clinical manifestation of SLE among various ethnic groups as well as various geographical regions (4).

The diagnosis of SLE is based on characteristic clinical findings of the skin, joints, kidneys, and the central nervous system, as well as on serological parameters such as antinuclear antibodies (ANA), in particular antibodies to dsDNA (2). Currently both ACR (American College of Rheumatology) & SLICC (Systemic Lupus International Collaborating Clinics) sets of criteria are often applied simultaneously for the confirm diagnosis of SLE.

Prognosis of SLE in current time has definitely improved with the advances in diagnostics, optimized treatment (also of comorbidities), and regular monitoring of disease activity and damage. SLE is a chronic disease of variable severity with a waxing and waning course, with significant morbidity that can be fatal—if not treated early—in some patients (2). The target of treatment is remission or at least minimization of disease activity and avoidance of flares. (1).

Considering the high degree of variable presentations and scarcity of information in Indian context, the present work was undertaken to map the clinical profile of SLE patients focusing mainly on presenting symptoms, system involvement, diagnostic (including serological) approach and treatment usage pattern.

Methods:

The study was conducted at a tertiary care centre in Mumbai. All patients satisfying the revised American College of Rheumatology criteria (1982) for SLE were included in the study over a period of 6 months starting from 01 Jan 16 to 31 July 16. We collected the detail information of all these patients with respect to demographic characteristics, duration of disease and assessment of various organs involvement like cutaneous, musculoskeletal, renal, gastrointestinal tract, nervous and cardiopulmonary. We collected data regarding various investigation including complete blood count, blood urea, chest radiograph, electrocardiogram, urine microscopy, 24 h urine protein excretion and serum creatinine. All the enrolled patients were also subjected to antinuclear antibodies (ANAs) and anti-double-stranded deoxyribonucleic acid (Ds DNA) antibody analysis.

A descriptive analysis of all demographic features of the patients was performed. We also analyzed the clinical features present in SLE patients and calculated the cumulative percentage frequency of all clinical features present in SLE patients. The cumulative percentage frequency of various systems involved in SLE patients was also calculated.

Results:

Age and sex distribution:

A total number of 11 SLE patients were enrolled with a mean age of 28.5 years. Most of the patients (approx 82%) were in the range of 24 to 32 years of age. Only one patient was of age more than 50 years. Gender distribution was found to be female preponderant with 72.7% contribution coming from females. The female to male patient ratio was calculated to be 2.7:1. The duration of disease in these patients ranged from 6 months to 4 years with an average duration of 2.9 years.

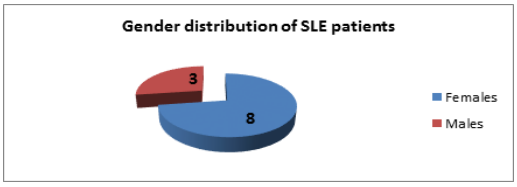

Chart 1: Gender distribution of SLE patients

Patient presentation and system involvement:

Arthritis or arthralgia was the most common presentation and was seen in 64% of patients. Most of the patients had involvement of multiple joints. Fever was the second most common symptom seen in 6 out of 11 patients (55.5%). Edema was complained by 5 patients (45%) out of

which 4 patients had pedal edema and one had whole body swelling. One patient had headache as well as hair loss and one patient each presented with weight loss, photosensitivity malar rash and lymphadenopathy.

As per the laboratory investigations, 5 patients (45%) had frank proteinuria. Two patients (18%) had low level of haemoglobin qualifying them to be diagnosed as anaemic. The mean value of haemoglobin in the patients was 13.2gm/dl. One patient reported leucopenia and one reported thrombocytopenia. None of the patient reported any abnormality in urine routine or microscopy examination.

Table: Clinical and immunological profile of SLE patients

	Number of patients	Proportion of Patients (%)
Age distribution		
<20 years	1	9
20-40 years	9	82
>40 years	1	9
Gender Distribution		
Male	3	27.3
Female	8	72.7
Clinical Features		
Mucocutaneous manifestation (Malar rash, photosensitivity)	1	9
Hematological (Anemia, Leucopenia, Thrombocytopenia)	3	27.3
Renal (Proteinuria)	5	45.45
Musculoskeletal (Arthralgia, Polyarthralgia, Arthritis, Joint Swelling)	7	63.33
Non-specific symptom (un-explained fever, pedal edema, Lymphadenopathy, body swelling, Headache, Hair loss)	10	90.90

Musculoskeletal was the most commonly involved system in 64% of patients. Kidney was the second most common organ involved with almost 45% of patients having nephritis. Cutaneous system involvement was seen in 18% of the patients. Adenitis and edema were seen in 9% of patient each. (Chart 2).

The disease activities of all patients were recorded as per SLEDAI (SLE disease activity index). The scores ranged from 4 to 8 in the subjects with the mean value of 5.64.

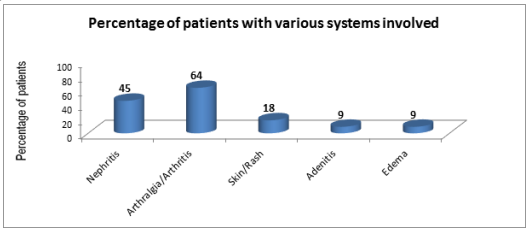

Chart 2: Involvement of various systems in SLE patients.

Immunological profile:

Anti-neutrophil antibody (ANA) was positive in all 11 patients. 73% patients were positive for Anti RO antibodies. Anti Sm, Anti La, anti ds DNA and U1RNP were positive in 36%, 27%, 18% and 18% respectively. Ro52 and Anti centromere were found to be positive in one (9%) patient each. (Chart 3)

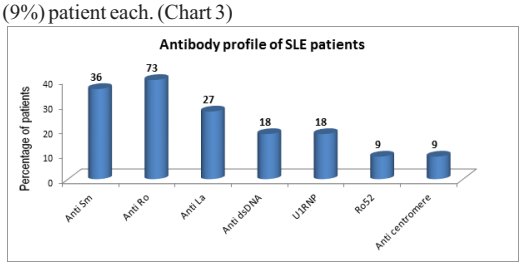

Chart 3: Antibody profiles of SLE patients

All these patients received one steroid (prednisolone or deflazacort) along with one or two of other immunosuppressants viz. hydroxychloroquine, mycophenolate mofetil, tacrolimus, leflunomide, methotrexate or azathioprine. Most of the patients (82%) gave good response to the conventional therapy while 18% patients responded moderately (chart 4).

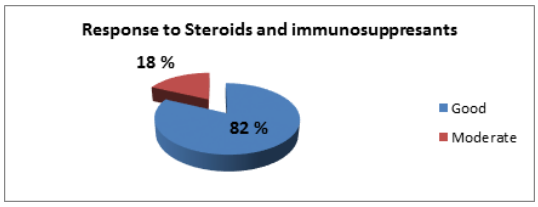

Chart 4: Response in SLE patients with Steroids and immunosuppressants

Discussion:

In our study of SLE patients, we found that the disease was more common in female patients especially during the child bearing age group. Our study showed higher frequency of immunological, musculoskeletal, renal & hematological involvement in SLE patients. Involvement of mucocutaneous, neurological, cardiovascular and respiratory system was found to be less common. Present study underscores the importance of detail clinical examination and focused investigation in patients suspected to have SLE, particularly in female patients of child bearing age(20-40 years age group) which is in complete agreement with the recently done research work by Agrawal et al.(4) in the central India.

A number of studies have been conducted in various part of our country regarding clinical and epidemiological profile of SLE which show wide range of geographical variation in clinical manifestations. A study done by Agrawal et al. analyzing 87 SLE patients from central part of India shows significant high proportion of patients presenting with mucocutaneous (83.9%) and renal (70%) manifestations. However, studies conducted by Binoy et al. and Kosaraju et al. from the south India have shown less prevalence of mucocutaneous manifestations(4). Our study results show that musculoskeletal, renal and nonspecific clinical manifestations are found in 63% and 45% and 90% of total patients respectively. In our study population we found diagnosis, based on clinical and immunological profile, of arthritis in 64% patients while lupus nephritis in around 36% patients.

The highly sensitive diagnostic test ANA (Sensitivity: 100%) can be used for screening the patients suspected to have SLE on clinical evaluation. However, because of low specificity it cannot be used for confirmation of disease. Above mentioned study by Agarwal et al. showed nearly 98% of patients were positive for ANA by indirect Immunofluorescence method, which was consistent with other studies done in different parts of India(4). Our study findings are in line with the published research work where we found ANA positivity in all of our study participants.

These days, antimalarials like hydroxychloroquine and glucocorticoids are basic treatment for patients with SLE, where in glucocorticoids are of even more importance in acute need. If reduction or tapering of glucocorticoids proves impossible, extended immunosuppression with azathioprine, methotrexate, or mycophenolate mofetil is recommended. Should the patient fail to respond, biologics (e.g. Belimumab) can be administered. In line with this standard of care we used various drugs like deflazacort, Prednisolone, Hydroxychloroquine, Tacrolimus, Mycophenolate Mofetil, Azathioprine, Leflunomide, Methotrexate etc. Almost all patients had glucocorticoids as the mainstay of treatment in our study. Most of the patients (82%) gave good response to the conventional therapy while 18% patients responded moderately.

Conclusions:

The observations of present study suggest that SLE patients may present with a wide variety of manifestations involving a number of systems viz. musculoskeletal, renal, coetaneous etc. Even the laboratory findings or immunological profile in these patients is highly variable hence; vigilant evaluation is required to diagnose the disease and its clinical pattern. Steroids and steroid sparing drugs can play an important role in management of SLE patients.

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