MEDICAL CHANNEL

Original Article

VARIOUS MODES OF PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS AT A TERTIARY CARE HOSPITAL

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ABSTRACT

Objective : This study was conducted to assess the various modes of presentation of systemic lupus erythematosus SLE in our setup at a tertiary care hospital in periphery of Sind.
Methadology **:** This retrospective, descriptive and analytical study was planned to study the various modes of presentation of SLE patients in our set up admitted in LUMHS hospital during the period of March 2006 to Jan 2011. Thirty three adult patients were included in this study. Special proforma were prepared to record the information from case sheets of patients including basic information, symptomatology and laboratory investigations. The criteria used in this study were from the American College of Rheumatology (ACR). Inclusion criteria in this study were, patients presenting with four of the eleven criteria.
Statistics:For statistical analysis of the results, statistical package SPSS version 10 was used. Data was presented in Mean ±SD and frequency (percentage in respective sex).
Results**:** In this study patient's age ranged from 20-65 years. The mean age was 35±7.5 years and median age was 30 years. Out of 33 patients 31 (94%) were females. Two (6%) were male patients. F/M ratio was 15.5:1. 23 (69.6%) patients presented with skin lesions particularly with malar rash and discoid lesions. 32 (97%) patients presented with arthralgia and arthritis. 17 (51.5%) patients had history of photosensitivity and 20 (60.6%) patients were found to have oral ulcers. 31 (94%) patients presented with high grade fever. 18 (54.5%) patients developed neuropsychiatric complications. 11 (33%) patients presented with renal manifestations. 18 (54.5%) patients had various infections. Anemia was found in 27 (81.8%) patients. 7 (21%) patients presented with mild pleural effusion and pleural rub. 3 (11%) patients presented with vasculitis. Splenomegaly was found in 25 (75.7%) patients. All 33 (100%) patients were ANA positive, and 22 (66.6%) patients had anti ds DNA positive, none was found to have false VDRL test positive. Demographic data and clinical features are shown in Table 1 and 2.
Conclusion: In this study it is concluded that by carefully observing the various modes of presentation of SLE in our setup in a tertiary care hospital, the diagnosis will be made early and complications may be prevented.

Key Words: Modes of presentation, Sysstemic lupus, Erythematous

INTRODUCTION

Systemic lupus erythematosus SLE is chronic multifaceted inflammatory disease. Worldwide a conservative estimate states that over 5 million people have SLE. The etiology of which is not well understood. When the disease develops, quality of life can be improved through flare prevention. The warning signs of an impending flare include increased fatigue, pain, rash, fever, abdominal discomfort, headache, and dizziness.

SLE can affect any part of the body, but most often harms the heart, joints, skin, lungs, blood vessels, liver, kidneys and nervous system. Cytokines play an important role in the pathogenesis of SLE and lupus nephritis[1]. SLE is one of the types of lupus which damages many organs of body[2]. Lupus can occur at any age, and is most common in women, particularly of non-European descent. Survival in patients with SLE in the United States, Canada, and Europe is approximately 95% at 5 years, 90% at 10 years, and 78% at 20 years[3]. There are several types of lupus; drug-induced lupus erythematosus, lupus nephritis, discoid lupus erythematosus,

Subacute cutaneous lupus erythematosus, neonatal lupus, Sjögren's syndrome. The environmental factors may not only exacerbate existing lupus conditions but also trigger the initial onset.

Antinuclear antibody (ANA) and anti-extractable nuclear antigen form the mainstay of serologic testing for lupus. Antiphospholipid antibodies occur more often in SLE and can predispose to thrombosis. More specific are the anti-Smith and anti-dsDNA (double stranded) antibodies. Both enzyme linked immunosorbent assay (ELISA) and immunofluoresencent (IF) techniques can be used for the measurement of Anti- double stranded DNA (anti-dsDNA) antibodies in clinical laboratories and these antibodies are not unique to SLE[4]. The American College of Rheumatology (ACR) established eleven criteria in 1982, which were revised in 1997 as a classificatory instrument to define the SLE in clinical trials. A useful mnemonic for these 11 criteria is SOAP BRAIN MD: Serositis$_{(8)}$, Oral ulcers$_{(4)}$, Arthritis$_{(5)}$, Photosensitivity$_{(3)}$, Blood Changes$_{(9)}$, Renal involvement (proteinuria or casts)$_{(6)}$, ANA$_{(10)}$, Immunological changes as anti-Smith, anti-ds DNA, antiphospholipid antibody, and/or false positive serological test for syphilis$_{(11)}$, Neurological signs (seizures, frank psychosis$_{)(7)}$, Malar Rash$_{(1)}$, Discoid Rash$_{(2)}$[5]. SLE is associated with manifestations other than those listed in the criteria[6, 7, 8]. SLE presents in different ways and because of not an uncommon disease it was planned to study the various modes of presentation of SLE patients presenting in a tertiary care hospital in our setup.

METHODOLOGY

This retrospective and descriptive study was planned to study the clinical features of SLE patients in our set up admitted in LUMHS hospital during the period of March 2006 to Jan 2011. 33 adult patients were included in this study. Special proforma were prepared to record the information from case sheets of patients including basic information, symptomatology and laboratory investigations. The criteria used in this study were from the American College of Rheumatology (ACR). The eleven criteria for diagnosis of SLE were; Malar Rash$_{(1)}$, Discoid Rash$_{(2)}$, Photosensitivity$_{(3)}$, Oral ulcers$_{(4)}$, Arthritis$_{(5)}$, Renal involvement (proteinuria or casts)$_{(6)}$, Neurological signs (seizures, frank psychosis$_{)(7)}$, Serositis$_{(8)}$, Blood Changes$_{(9)}$, ANA$_{(10)}$, Immunological changes as anti-Smith, anti-ds DNA, antiphospholipid antibody, and/or false positive serological test for syphilis$_{(11)}$. Inclusion criteria in this study were, patients presenting with four of the above eleven criteria.

STATISTICS

For statistical analysis of the results, statistical package SPSS version 10 was used. Data was presented in Mean ±SD and frequency (percentage in respective sex).

RESULTS

In this study patient's age ranged from 20-65 years at time of presentation. The mean age was 35±7.5 years and median age was 30 years. Out of 33 patients 31 (94%) were females. Two (6%) were male patients. F/M ratio was 15.5:1. 23 (69.6%) patients presented with skin lesions particularly with malar rash and discoid lesions. 32 (97%) patients presented with arthralgia and arthritis. 17 (51.5%) patients had history of photosensitivity and 20 (60.6%) patients were found to have oral ulcers. 31 (94%) patients presented with high grade fever. 18 (54.5%) patients developed neuropsychiatric complications. 11 (33%) patients presented with renal manifestations. 18 (54.5%) patients had various infections, 3 (11%) had associated pulmonary tuberculosis. Mild to moderate normocytic and normochromic anemia was found in 27 (81.8%) patients. Coomb's positive hemolytic anemia was found in 6 (18%) patients. 7 (21%) patients presented with mild pleural effusion and pleural rub. 6 (18%) patients presented with chest pain. 3 (11%) presented with vasculitis. Splenomegaly was found in 25 (75.7%) patients. All 33 (100%) patients were ANA positive, and 22 (66.6%) patients had anti ds DNA positive, none was found to have false VDRL test positive. Demographic data and clinical features are shown in Table 1 and 2.

TABLE 1
VARIOUS MODES OF PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS AT A TERTIARY CARE HOSPITAL (N33)

DEMOGRAPHIC DATA

Parameters	No of Patients n33	Percentage
Age	20 Years to 65 Years	
Mean age	35± 7.5 years	
Median age	30 years	
Females	31	(94%)
Male	2	(6%)
F/M Ratio	15.5/1.0	

TABLE 2
VARIOUS MODES OF PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS AT A TERTIARY CARE HOSPITAL (N33)
CLINICAL FEATURES

Parameters	No of Patients n33	Percentage
High grade fever	31	94%
Oral ulcers	20	60.6%
Chest pain	6	18%
Malar and Discoid Rash	23	69.6%
Photosensitivity	17	51.5%
Infections	18	54.5%
Arthritis	32	97%
Renal involvement	11	33%
Neurological signs	18	54.5%
Serositis	7	21%
Blood Changes	27	81.8%
ANA	33	100%
anti-ds DNA	22	66.6%

DISCUSSION

SLE is a deadly chronic autoimmune problem affecting many organ systems. In this disease immune system attacks the body's cells and tissues, resulting in inflammation and tissue damage. The SLE has various modes of presentation. In this study 31 (94%) patients presented with high grade fever. In a study at Karachi fever was the most common symptom at the time of presentation (80.0%)[9]. Fever was in more patients in our study. The frequency of anemia in our study is 27 (81.8%) while another study was comparable to our study that the frequency of anemia was documented to be 79.37%[10]. In another study twenty eight (93.33%) patients presented with anaemia, 14 (46.66%) patients were of mild anaemia, 8 (26.66%) patients were of moderate grade anaemia and 6 (20%) patients had severe anaemia and 5 (16.66%) patients had Coomb's positive haemolytic anaemia[11]. The Lupus Foundation of America estimates that anemia as; iron deficiency may develop in as many as half of patients. Low platelet and white blood cell counts may be due to the disease or a side effect of treatment.

In our study 23 (69.6%) patients presented with skin lesions particularly with malar rash and discoid lesions. It has been discussed in literature that cutaneous changes in children are different from those seen in adults. Photosensitivity and vascular lesions are less frequent while the discoid rash is rare[12]. The source of IFN-á in SLE has been looked in plasmacytoid dendritic cells of SLE patients, and increased IFN-á expression in the skin has been demonstrated[13]. This study shows that 32 (97%) patients presented with arthralgia and arthritis. The Lupus Foundation of America estimates that more than 90 percent will experience joint and/or muscle pain at some time during the course of their illness. In this study 18 (54.5%) patients developed neuropsychiatric complications. Literature shows that about 10% of patients may present with seizures or psychosis. One-third may test positive for abnormalities in the cerebrospinal fluid[14].

In this study 18 (54.5%) patients had various infections, while in another study bacterial infections accounted for more than 50% of all infectious episodes in patients admitted with SLE, comparable to our study. Staphylococcus aureus was the commonest organism isolated, followed by Escherichia coli and Klebsiella spp[15]. One study has shown that the clinical characteristics of SLE patients in our country may be different to those of other Asian races[16]. But this study revealed that characteristics of SLE in our setup are similar to other parts of world. The most common cause of death in SLE is infection due to immunosuppression as a result of medications used to manage the disease. ANA is the most sensitive screening test, while anti-Sm (anti-Smith) is the most specific. The ds-DNA antibody is also fairly specific and often fluctuates with disease activity; therefore sometimes useful to diagnose or monitor acute flares or response to treatment. In our setup ANA was positive in all the cases and anti dsDNA in 66.6% cases.

Chest pain was present in 6 (18%) patients presented in our setup. Cardiovascular disease is a major cause of morbidity and mortality in SLE[17]. Atherosclerosis also tends to occur more often and advances more rapidly in SLE patients than in the general population[18]. In our study 11 (33%) patients presented with renal manifestations. It has been discussed in various studies that renal involvement leads to painless hematuria or proteinuria which may often be the only presenting renal symptom. The prevalence of lupus nephritis in our population is an intermediate between Caucasians and other Asians. It has been observed that the level of cytokines will increase, along with SLE disease activity, in line with the finding that serum levels of tumor necrosis factor (TNF) and interferon-á (IFN) correlate[19]. TNF is also over expressed in active lupus nephritis, where again it correlates with disease activity, and has been found in lupus skin lesions[20]. The patients with lupus nephritis should be treated with improved ancillary medical therapies and more effective immunosuppressive regimens[21]. Statins have been shown to limit progression of lupus nephropathy and decrease the TNF concentration and disease activity in pilot studies[22,23]. Treatment with statins in lupus may be a unique opportunity to correct many risk factors with a single drug[24].

In this study only 2 male patients had SLE, and F/M ratio was 15.5:1. Prognosis is normally worse for men and children than for women; fortunately, if symptoms are present after age 60, the disease tends to run a more benign course. A high index of suspicion for SLE in males may permit early diagnosis and dictate the need for more aggressive therapy[25].

CONCLUSION

In this study it is concluded that by carefully observing the various modes of presentation of SLE in our setup in a tertiary care hospital, the diagnosis will be made early and complications may be prevented.

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