Clinical evaluation of Systemic Lupus Erythematosus patients admitted in a tertiary care hospital of Bangladesh.

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Abstract

Background: Systemic Lupus Erythematosus is a rare connective tissue disorder. It causes considerable morbidity and mortality among affected patients. **Objective:** To present the demographic profiles and clinical features of Systemic Lupus Erythematosus (SLE) patients admitted in Rajshahi Medical College Hospital . **Methods:** This was a cross sectional descriptive study conducted on 31 Systemic Lupus Erythematosus (SLE) patients admitted in Rajshahi Medical College Hospital from July2017 to December 2017 for a period of 6 months. Patients were diagnosed as having SLE on the basis of Revised American Rheumatism Association criteria. **Results:** A total 31 patients having SLE, 26 (83.9%) patients were female and 5 (16.1%) patients were male. Majority (77.4%) of the patients were young adults =30 years. All the patients had intermittent polyarthritis (100%). Other common presentations of them were skin lesions (83.9%), fever & constitutional symptoms (83.9%), hematological involvement (58.1%), ankle edema (32.3%), bed side proteinuria (32.3%), generalized swelling (29%). Antinuclear antibody (ANA) was positive among 29 patients (93.5%) and anti-double stranded DNA antibodies (anti ds DNA) was positive in 24 (77.4%) patients. **Conclusion:** Systemic Lupus Erythematous is a chronic disorder which affects the younger age group, mostly females. It not only causes increased morbidity but also reduce the quality of life. In clinical practice we should follow the ACR recommendation for testing ANA titer. And anti ds DNA antibodies have limited value in clinical correlation and in predicting disease flares and subset in SLE.

Key words: Systemic Lupus Erythematosus(SLE), ANA titer, anti-dsDNA .

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Introduction

Systemic Lupus Erythematosus (SLE) is a chronic, recurrent, potentially fatal multisystem inflammatory disorder that can be difficult to diagnose.[1,2] It is a connective tissue disease characterized by dysregulation of immune responses, autoantibody production often directed at components of the cell, nucleus, and widespread tissue damage. It is a rare disease with a prevalence that ranges from about 0.03% in Caucasians to 0.2% in Afro-Caribbeans. Some 90% of affected patients are female and the peak age at onset is between 20 and 30 years. Lupus is associated with considerable morbidity and a five-fold increase in mortality compared to age- and gender-matched controls, mainly because of an increased risk of premature cardiovascular disease.[3]

The diagnosis of Systemic Lupus Erythematosusis based on clinical and laboratory criteria. The criteria set developed by the American College of Rheumatology (ACR) is most widely used.[4,5] Elevation of the antinuclear antibody(ANA) titer to 1:40 or higher is the most sensitive of the ACR diagnostic criteria. More than 99 percent of patients with systemic lupus erythematosus have an elevated ANA titer at some point,[4,6] although a significant proportion of patients may have a negative ANA titer early in the disease.[2] The present study evaluated the clinical and laboratory features of a SLE cohort admitted in a tertiary hospital.

Methods

It was a cross sectional descriptive study conducted on 31 Systemic Lupus Erythematosus (SLE) patients admitted in Rajshahi Medical College Hospital from July2017 to December 2017 for a period of 6 months. Patients were diagnosed as having SLE on the basis of Revised American Rheumatism Association criteria.[3] Patients meeting 4 or more criteria including ANA or anti-double stranded DNA antibodies (Anti

Ds DNA) positive are included in this study. All patients were examined thoroughly and information were collected in a preformed data collection sheet. Data collection sheet was designed to record the information on demographic characteristics, clinical presentations and antibody titers. Data were entered in the computer and processed using SPSS 16. Descriptive statistics such as frequency distribution, computation of percentage etc. were applied.

Results

A total 31 patients having SLE, diagnosed on the basis of Revised American Rheumatism Association (ARA) criteria, 26 (83.9%) patients were female and 5 (16.1%) patients were male. Thus most of the affected patients were female. Majority (77.4%) of the patients were young adults =30 years and the rest 22.6% were older than 30 years. None of the patients were above 40 years.

Table 1: Clinical presentations of the SLE patients

Presentation	Present N (%)
Multiple joint pain & swelling	31 (100)
Skin manifestations	26 (83.9)
Fever and constitutional symptoms	26(83.9)
Haematological involvement	18 (58.1)
Ankle oedema	10 (32.3)
Bed side proteinuria	10(32.3)
Edema and generalised swelling	9 (29.0)
Abdominal pain and vomiting	8 (25.8)
Cardiopulmonary	7 (22.6)
Neurological menifestations	4 (12.9)
Eye involvement	4 (12.9)
Jaundice	1 (3.2)
Deep venous thrombosis	1 (3.2)
Lymphadenopathy	1 (3.2)
Flapping tremor	1(3.2)

All the patients presented with multiple joint pain (100%), followed by skin lesions (83.9%), fever & constitutional symptoms (83.9%), hematological involvement (58.1%), ankle edema(32.3%), bed side proteinuria (32.3%), generalized swelling (29%), abdominal pain & vomiting (25.8%), cardiopulmonary (22.6%), neurological involvement (12.9%) and others (Table 1).

Table 2: Antibody titer among SLE patients

Autobnantibody	Present	Absent
ANA	29 (93.5%)	2 (6.5%)
Anti ds DNA	24 (77.4%)	7 (22.6%)

ANA was positive among 29 patients (93.5%) and anti ds DNA was positive in 24 (77.4%) patients (Table 2).

Discussion

We have assessed 31 patients with Systemic Lupus Erythematous during 6 months period in medicine department of Rajshahi medical college. Among them 83.9% patients were female and only 16.1% were male with male to female ratio of 1:5. All the female SLE patients were in reproductive age. This finding is inconsistence with other studies in Bangladesh and abroad.[7-9]

There are different hypotheses that might explain the development of SLE. 1st, patients with SLE have abnormally low total T cell DNA methylation (more activated genes). Because women have 2 X chromosomes, one of which has genes that are mostly inactivated, failure to inactivate affects women more than men. Demethylation of sites on an inactive X could contribute to female susceptibility to lupus. A child is conditioned by the inactivated X chromosomes in utero or early childhood event to be susceptible to SLE. 2nd, unmasking of susceptibility may require exposure to one or many environmental insults, such as a virus. Third. At female puberty (but not male puberty), high levels of estradiol may be permissive (or testosterone may be suppressive), allowing clinical disease to occur.[10] The present study findings i.e. female predominance of SLE and its occurrence in young adulthood goes infavour of third hypothesis.

All of the patients in the present study presented with intermittent polyarthritis, varying from mild to disabling, characterized by soft tissue swelling and tenderness in

joints, most commonly in hands, wrists, and knees. No joint deformities were present. This findings is consistent with other studies.[11] Skin involvement along with constitutional features also affecting majority of our patients. Schur PH and Gilboe IM[11], Husby G[12] also stated that Systemic lupus erythematosus most often manifests as a mixture of constitutional symptoms along with skin involvement. In our series skin involvement consist of butterfly rash, discoid lupus erythematosus (DLE), systemic rash, subacute cutaneous lupus erythematosus (SCLE), or "other." Most common haematological abnormalities in our series was anaemia, leukopenia, thrombocytopenia. Only one patient present with generalised lymphadenopathy. Renal involvement, in the form of bed side proteinuria found in 10 patients. Abdominal pain and vomiting was found in 8 of our patients, which can be manifestations of an SLE flare, as can diffuse abdominal pain caused by autoimmune peritonitis and/or intestinal vasculitis. Cardiopulmonary involvement in the form of pericarditis, pleural effusion were found in 7 patients. Cardiopulmonary involvement in SLE is usually due to accelerated atherosclerosis, which probably results from chronic inflammation and/or chronic oxidative damage to lipids and to organs. The most common pulmonary manifestation of SLE is pleural effusion. Most common neurological manifestations involving our patients were headache and one patient presented with acute confusional state. Nonspecific conjunctivitis occurs 4 of our patients. Jaundice, DVT and flapping tremor were found in one patient each.[13] However, it not only causes increased morbidity among the affected but also reduce the quality of life.

Elevation of the antinuclear antibody (ANA) titer to 1:40 or higher is the most sensitive of the ACR diagnostic criteria. Less than 1% of patients with SLE have not an elevated ANA titer at some point.[12,14] But more than 6% of the present study subjects had a negative ANA titer. It may be due to early stage of the disease process in some present study subjects. Because a significant proportion of the patients may have a negative ANA titer early in the disease.[2] However, in clinical practice we should follow the ACR recommendation for testing ANA titer i.e. ANA testing in patients with two or more unexplained signs or symptoms of systemic lupus erythematosus.

Anti-dsDNA antibodies have been a hallmark of lupus erythematosus for decades.[15] There are contradictory observations regarding the role of anti-dsDNA antibodies in SLE including predicting the disease flares and its subgroups. Most of the previous literatures identified anti-dsDNA as a pathogenetic role in the kidney injury and initiate the lupus nephritis.[16,17,18] But recent studies[19,20] revealed that these antibodies have limited value in clinical correlation and in predicting disease flares and subset in SLE. In addition, they are not likely to be the initiating auto-antibodies in lupus nephritis. The present study findings goes in favor of the recent observations. Because in this present study, though more than two third of the study subjects had anti-dsDNA positive but only less than one third of the study subjects had clinical features those correlate with lupus nephritis.

Systemic Lupus Erythematoss is a chronic disorder which affects the younger age group, mostly females. It not only causes increased morbidity among the affected but also reduce the quality of life. In clinical practice we should follow the ACR recommendation for testing ANA titer. And anti-dsDNA antibodies have limited value in clinical correlation and in predicting disease flares and subset in SLE.

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