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Original Research Article

A retrospective study of systemic manifestations in systemic lupus erythematosus in a tertiary care centre

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ABSTRACT

Background: Systemic lupus erythematosus (SLE) is an autoimmune disease involving multiple organ systems. Other than cutaneous manifestations, thorough examination and investigations for systemic involvement should also be done. The objective is to study the prevalence of various systemic manifestations in SLE patients in a tertiary care hospital.
Methods: A retrospective study was conducted from April 2009 to August 2018 on the patients who fulfilled the Systemic Lupus International Collaborating Clinics (SLICC) criteria for SLE. A total of 63 cases of SLE were extracted from the case record for the study. Each case was subjected to detailed history, examination, investigations including complete blood count, liver function test, renal function test, blood sugar, urine routine examination, ECG, X-ray of chest and involved joints, USG abdomen, pulmonary function test. Ours being a resource limited set-up, ANA, ENA profile and echocardiography were done whenever patient could afford.
Results: In present study of 63 cases of SLE 94% were females. Mean age was 30 years. Various systemic involvement was present, amongst them musculoskeletal system was the most common system involved with 60 (95%) patients, followed by hematological system 48 (76%), renal system 37 (59%), neuropsychiatric system 29 (46%), gastrointestinal system 28 (44%), reproductive system 27 (43%), cardiovascular system 25 (40%). Other systems with ophthalmic involvement 23 (21%), hepatobiliary system 22 (35%), respiratory system 17 (27%), endocrine involvement 13 (28%) and malignanacy was present in 2 (3%) cases.
Conclusions: SLE is protean in its manifestations. Thorough knowledge about the systemic involvement will help in tailoring the treatment in case of SLE.

Keywords: Systemic lupus erythematosus, Systemic manifestations

INTRODUCTION

The term "lupus erythemateaux" was first used by Cazenave in the mid-1800s. Due in part to observations of Hutchinson, Osler and Jadassohn, it was recognized that cutaneous lesions of LE maybe associated with significant internal abnormalities, including arthritis, nephritis, serositis, cytopenias and neurologic disease. In 1964 and during the following years, Dubois developed the concept of lupus as a spectrum of disease, ranging from isolated cutaneous lesionsto life-threatening, multiorgan disease.[1] Lupus erythematosus is potentially a multiorgan disease, although in few patients it is often only one or few organs that are significantly affected. The organ systems most commonly affected are joints, skin, hematologic, pulmonary, renal and the CNS.[1] Many of

the organ manifestations classically associated with lupus are part of the American College of Rheumatology (ACR) and Systemic Lupus International Collaborating Clinics (SLICC) criteria for SLE. The present study is an attempt to study the prevalence of various organ system involvement, their varied presentation and their abnormal investigatory parameters.

METHODS

A retrospective analysis of data collected from clinical records over a period of 10 years (April 2009 to August 2018) on the patients who fulfilled the American College of Rheumatology (ACR) and/or Systemic Lupus International Collaborating Clinics (SLICC) criteria for SLE. Those patients not willing to participate in the study were excluded. A total of 63 cases of SLE were extracted from the case record for the study. Patients presenting with history, symptoms and sign suggestive of SLE were subjected to available investigations. A detailed history regarding the onset, duration and progress of lesions and precipitating factors was noted. History regarding systemic complaints, e.g. joint pain, muscle weakness and tenderness, anorexia, nausea/ vomiting, chest pain, edema, palpitation, any eye complaints or neuropsychiatric manifestations was actively sought. In case of female patients, menstrual and obstretric history was noted. Thorough general, Cutaneous and systemic examinations were carried out.

All cases were subjected to routine investigations, comprising complete blood count, liver and renal function test, blood sugar, urine routine and microscopic examination, x-ray chest, USG abdomen and pelvis, ECG, RA factor. Antinuclear antibodies (ANA) and Extractable Nuclear Antigen (ENA) profile were done in as many cases as possible. Cases were also subjected to available special investigations as follows: x-ray of affected joints, pulmonary function test (PFT), echocardiography, MRI brain if required, fundus examination for eye.

RESULTS

63 patients of SLE were studied out of which 59 (94%) were females and 4 (6%) males. Male: Female ratio was 1:14.7. The maximum number of the patients at the time of presentation belonged to the age group of 21-30 years (38%). Mean age was 30 years, though the cases ranged from 16 to 55 years of age.

Amongst all the systemic manifestations, musculoskeletal system was the most common system involving 60 (95%) patients, of which 50 (79%) patients had joint pain (arthralgia) which was the commonest initial manifestation. Morning stiffness was present in 20 (31%) and X-ray changes of osteopenia was present in 10 (16%) patients. Generalized muscle weakness was seen in 9 (14%) and muscle tenderness in 1 patient.

The second most common system involved was Hematological system 48 (76%). Amongst them anemia was present in 25 (39.6%) patients, neutropenia in 8 (12.6%), thrombocytopenia in 7 (11%) and pancytopenia in 8 (12.6%) patients.

Renal involvement was present in 37 (59%) patients. Proteinuria was seen in 30 (47.6%) patients, hematuria in 8 (12.6%) patients, pyuria in 18 (28%) patients, cast was present in 3 patients and altered renal function test in 23 (36.5%) patients. On ultrasound examination 2 patients had renal parenchymal disease and 5 patients had renal cyst.

Neuropsychiatric abnormalities were seen in 29 (46%) patients. Anxiety 10 (16%) and depression 6 (9%) were most common neuropsychiatric abnormalities observed followed by psychosis. Epilepsy and chronic headache was present in 2 patients each. 1 patient had peripheral neuropathy. One patient developed demyelinating neuropathy. Idiopathic intracranial hypertension and facial palsy was developed in 1 patient each.

Gastrointestinal system was involved in 28 (44%) patients. Amongst them 12 patients had anorexia, 8 had nausea/vomiting, 7 had abdominal pain and 1 patient had hematemesis. On ultrasound examination 7 patients had ascites.

Reproductive system was involved in 27 (43%) patients. Out of 59 female patients, 13 (22%) had menstrual irregularity, 10 (17%) patients had oligomenorrhea, and 4 (7%) had menorrhagia.

Involvement of cardiovascular system was present in 25 (40%) patients. Palpitation was most common symptom in cardiac involvement. Pedal edema was present in 10 patients. Chest pain was present in 5 patients, their chest X-ray showed cardiomegaly and echocardiograph was suggestive of pericardial effusion. Amongst them most common echocardiogram finding was mitral regurgitation, followed by tricuspid regurgitation, mitral valve prolapsed, left ventricular hypertrophy, pulmonary artery hypertension and atrial regurgitation.

Ophthalmic involvement was present in 29 (46%) patients. Lid edema 20% was the most common symptom followed by conjunctivitis 14%. Dryness of eyes was present in 7 (11%) patients, ptosis in 1 patient. Visual blurring was present in 8 (12.6%) patients, out of which on further investigation 6 patients had cataract, 1 patient had macular atrophy and 1 had papilledema.

Hepatobiliary system was involved in 22 (35%) patients. Amongst them, Liver function test was altered in 5 (8%) patients. On ultrasound examination 8 (13%) patients had fatty liver, 6 (9.5%) had hepatomegaly and 5 (8%) had asymptomatic hemangioma.

Respiratory system was involved in 17 (27%) patients. Dyspnea in 8 (12.6%) cases was the most common symptom in pulmonary involvement followed by pleuritic chest pain 5 (8%) and recurrent cough 4 (6.3%). Pulmonary function test (PFT) was impaired in 8 (12.6%) patients. On further investigation cause of dyspnea and altered PFT was pulmonary tuberculosis showed as consolidation on chest X-ray in 3 patients and pleural effusion in 5 patients.

Endocrine system was involved in 13 (28%) patients. Amongst them 8 (12.6%) suffered from hypothyroidism and 5 (8%) from diabetes mellitus.

Malignancy was noticed in 2 patients. One was diagnosed with post cricoid carcinoma and the other with bronchogenic carcinoma.

ANA was done in all the patients of SLE 100% positivity. Most common pattern was homogenous patterns (87.3%), followed by speckled pattern (11.11%) with nucleolar pattern in 1.5% of the cases.

DISCUSSION

In our study we included 63 patients of SLE who fullied the ARA/SLICC criteria. The mean age at presentation among those cases was 30 years, while that reported by Malaviya et al, Kole et al and Paul et al was 24.5, 25 and 21.6 years respectively.[2-4] The cases ranged from 16-55 years, with 38% of the cases presented between 21-30 years of age. Male: Female ratio was 1:14.7, while study by Malaviya et al showed a ratio of 1:8.[2]

Table 1: Systemic manifestations of SLE in present study.

Systemic manifestations	Present study (2018) in % (n=63)
Musculoskeletal system	95.2
Respiratory system	27
Cardiovascular system	40
Renal involvement	59
Hematological system	76
GIT system	44
Neuropsychiatric system	46
Eye involvement	21
ANA	100

Involvement of the joints occurs at some time in approximately 90% of the patients. Arthralgia is more common than arthritis.[5] In present study, musculoskeletal system was most common system involved of which 50 (79%) patients had joint pain (arthralgia) which was the commonest initial manifestation. Morning stiffness was present in 20 (31%), and X-ray changes of osteopenia was present in 10 (16%) patients. Muscle pain occurs in approximately 50% of the patients which is confused with the pain of arthritis. Muscle weakness is a less common feature. Generalized muscle tenderness was seen in 9 (14%) and muscle weakness in 1 patient[5]. In present study musculoskeletal system was most commonly involved which was followed by hematological system and renal system, which was consistent with the study by Salinas et al which showed 93.5%, 78.5% and 59% of case involvement respectively.[6] Whilst in Malaviya et al study, renal involvement was most common followed by musculoskeletal system and hematological system.[2] Paul et al reported, musculoskeletal system was most common followed by renal system and neuropsychiatric system.[4] While Kole et al study showed equal involvement of musculoskeletal system and hematological system (Table 1).[3]

Renal involvement in present study was 59%. The comparative data in the series with Malaviya et al, Paul et al, Kole et al and Salinas et al was 73%, 33.3%, 46.6% and 59% respectively.[2-4,6]

Hematological involvement was highest in Kole et al[3] study (90%), present study showed 76% involvement which was consistent with Salinas et al study, and was less common in Malaviya et al and Paul et al study.[2,4,6] Anemia (39.6%) was the commonest hematological abnormality detected in our study. Leukopenia was seen in11% cases and thrombocytopenia in 12% cases which was consistent with Paul et al study (Table 2).[4]

Table 2: Hematological manifestation in SLE of present study.

	Anemia	Leucopenia	Thrombocytopenia
Present study (%)	39.6	12.6	11

In present study cardiovascular system was involved in 40% while Malaviya et al and Paul et al study showed 5% involvement.[2,4] Respiratory involvement was present in 27% of cases in our study which was consistent with Salinas et al, while Malaviya et al, Paul et al and Kole et al reported 17%, 8%, 13.4% involvement respectively.[3,4,6,7]

Kole et al reported 73.34% of neuropsychiatric involvement, while our study showed 46% involvement.[3] Malaviya et al and Paul et al reported 15% and 13.3% respectively.[2,4]

In present study, 44% cases showed GIT involvement whilst Kole et al showed 73.34% involvement.[3] Our study showed ophthalmic involvement in 21% of cases which was consistent with Salinas et al.[6] Endocrine system was the least common system involved in our study. Malignancy was present in 2 cases, one with post cricoid carcinoma and other with bronchogenic carcinoma. ANA was positive in 100% in our study as well as in Kole et al and Maheshwari et al, whilst Malaviya et al and Salinas et al showed 98% positivity and in Paul et al study ANA was positive in 93.35%.[2-4,6,7]

Table 3: Systemic involvement in SLE in present study.

Systems involved					SLE (63)
Musculo-skeletal system	Joint pain without swelling: 42 Joint pain with swelling: 8	Morning stiffness: 20	X-ray changes: Osteopenia:4	50 (79%)	60 (95%)
	Muscle weakness: 9	Muscle tenderness:1		10 (16%)	
Pulmonary system	Recurrent cough: 4 Dyspnea: 8 Pleuritic chestpain: 5	Impaired PFT: 2	X-ray: PLE- 5 Consolidation 3		17 (27%)
Cardio-vascular system	Palpitation: 15 Pedal edema: 10 Chestpain: 5	ECG changes: 25 X-ray: Cardiomegaly: 5	ECHO: Valvular involvement: 31		25 (40%)
Renal system	Proteinuria: 30 Hematuria: 8 Pyuria: 18 Cast: 3	Altered RFT: 23	USG: RPD-2 Renal cyst-5		37 (59%)
Hematologic system	Anaemia: 25 Leucopenia: 8	Thrombocytopenia: 7 Pancytopenia: 8			48 (76%)
Git system	Anorexia: 12, Nausea/vomiting: 8 Abdominal pain: 8, Hemetesis: 1				28 (44%)
Hepato-biliary system	Raised LFT: 5	USG: Fatty changes: 8, Hepatomegaly: 6 Asymptomatic Hemangioma:3, LPD:5, Ascites:7			22 (35%)
Neuro psychiatric system	Epilepsy: 2 Chronic headache: 2	Facial palsy: 1 PN: 1 DN: 1	IIH: 1 TB meningitis:1		9 (14%)
	Psychosis: 4, Depression: 6, Anxiety: 10				20 (32%)
Endocrine system	Hypothyroidism: 8	Diabetes Mellitus: 5			13 (28%)
Eye	Lid edema: 13 Conjuctivitis: 9 Visual blurring: 8	Dry eyes (schirmer's test): 7 Ptosis:1	Cataract: 6 FUNDUS: MA-1, PE-1		29 (46%)
Reproductive system	Menstrual regularity: 13, Oligomenorrhea:10, Menorrhagia: 4				27 (43%)
Malignancy	Post cricoid CA: 1	Bronchogenic CA: 1			2 (3%)

PFT- Pulmonary function test, PLE: pleural effusion, RPD: renal parenchymal disease, PN: peripheral neuropathy, DM: demyelinating neuropathy, IIH: idiopathic intracranial hypertension, LPD: Liver parenchymal disease, MA: Macular Atrophy, PE: papilloedema.

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