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A Case Series of Varied Clinical Presentations of Systemic Lupus Erythematosus

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Abstract

Systemic lupus erythematosus (SLE) is a chronic inflammatory disease of unknown cause that can affect virtually every organ. Immunologic abnormalities, particularly the production of a number of antinuclear antibodies, are another prominent feature of the disease. Women are affected more frequently than men.

SLE has protean clinical manifestations which can affect virtually every organ, and it can also vary dramatically from patient to patient. The clinical course of SLE is highly variable among patients and may be characterized by periods of remissions and of chronic or acute relapses. In this clinical case series, we present 5 patients who had diverse clinical presentations along with their management in our hospital.

Keywords: SLE, ANA, HEP.

Introduction

Systemic Lupus Erythematosus (SLE), is a multisystemic disease with an unknown etiology. It predominantly affects women of child bearing age. The prevalence of systemic lupus erythematosus (SLE) in the general population is 20-150 cases per 100,000 population[1]. The female to male ratio ranges from 7-15:1.

SLE has a vasculitic feature and may affect multiple organs. Clinical findings as well as laboratory tests are important for diagnosis. A variety of auto-antibodies are present in the course of systemic lupus erythematosus.[2]

The 209 EULAR CRITERIA is used for diagnosis of Systemic Lupus Erythematosus (SLE). At least 1 Clinical criterion is required to classify as (Systemic Lupus Erythematosus) SLE. Additional Additive (Clinical or Immunological) criteria are counted towards the total score.. The entry criterion is ANA at a titer of ≥1:80 on HEp-2 cells or an Equivalent Positive test. If ANA is absent, do not classify as SLE Additional criteria within the same domain will not be counted.3.

Clinical domains and criteria	Weight
Constitutional	
Fever	2
Hematologic	

Leukopenia	3
Thrombocytopenia	4
Autoimmune hemolysis	4
Neuropsychiatric	
Delirium	2
Psychosis	3
Seizure	5
Mucocutaneous	
Nonscarring alopecia	2
Oral ulcers	2
Subacute cutaneous or discoid lupus	4

Acute cutaneous lupus	6
Serosal	
Pleural or Pericardial effusion	5
Acute pericarditis	6
Musculoskeletal	
Joint involvement	6
Renal	
Proteinuria >0.5 g per 24 hours	4
Renal biopsy Class II or V lupus nephritis	8
Renal biopsy Class III or IV lupus nephritis	10
Immunology domains and criteria	Weight
Antiphospholipid antibodies	
Anti-cardiolipin antibodies or anti-beta-2GP1 antibodies or lupus anticoagulant	2
Complement proteins	
Low C3 or low C4	3
Low C3 and low C4	4
SLE-specific antibodies	
Anti-dsDNA antibody or anti-Smith antibody	6
A total score of ≥10 and ≥1 clinical criterion are required to classify SLE.	
Total score	

A total score of ≥10 and ≥1 Clinical criterion is required to classify SLE.

We report five cases of SLE with varied clinical presentations and their different multisystem associations, approach and management in our institution.[3]

Case Report 1

A 34 years old lady with no comorbidities presented with complaints of easy fatiguability, multiple small and large joint pains associated with redness and swelling, fever, exertional dyspnoea and bilateral lower limb swelling of 2 months duration. She was evaluated for the same at a local hospital and was diagnosed to have hypertension and hypothyroidism for which she was started on tablet amlodipine 5mg OD and tablet thyroxine 25 mcg OD. However, patient reported no improvement in clinical symptoms. On Day 3 of admission, patient developed epigastric discomfort, nausea and malena following which she noticed a reduction of urine output. The following day, patient had a single episode of Generalized Tonic Clonic Seizure (GTCS). Her Blood Pressure was recorded as 200/100 mm of hg, with tachycardia and a room air saturation of 88%. She also had pedal edema with bilateral basal crepitations. Blood gas analysis revealed metabolic acidosis. Neuroimaging done was suggestive of Posterior Reversible Encephalopathy Syndrome. Complete blood count showed normocytic

normochromic anemia with a Hemoglobin of 9.2g/dl, Platelets of 88,000 cells/mm^3, Total Leukocytes of 15900 cells /mm^3. Her creatinine was 1.5 mg/dl, Urine Routine showed 2+ albuminuria, 32 RBCs/hpf and granular casts. D-dimer was elevated at 6800 mg/L with Procalcitonin of >200ng/ml. ECG revealed a new onset right bundle branch block pattern. 2-Dimensional Echocardiography showed dilated right heart chambers with Pulmonary artery hypertension and an ejection fraction of 58%. CT Pulmonary Angiography did not reveal pulmonary embolism.

One session of Saline Hemodialysis was done in view of oliguria and severe metabolic acidosis. Oesophago-duodenoscopy done in view of melena showed duodenitis with multiple ulcers.

Further Immunological tests revealed ANA of 4+, hypocomplementemia, dsDNA 2+, histone 1+, nucleosome 1+, Sm 1+ and SSA 1+. In view of features suggestive of SLE flare with multi-organ involvement, patient was started on pulse therapy of steroids followed by mycophenolate mofetil, hydroxychloroquine and tapering dose of oral steroids. The patient gradually improved and a follow up renal biopsy was done. It showed Proliferative Lupus Nephritis with crescents. Immunofluorescent staining showed coarse granular mesangial and capillary wall staining. Patient had no further episodes of seizures.

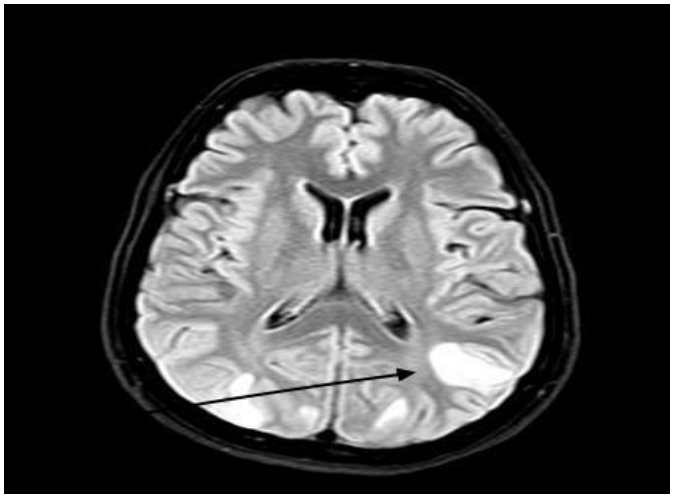

Figure 1

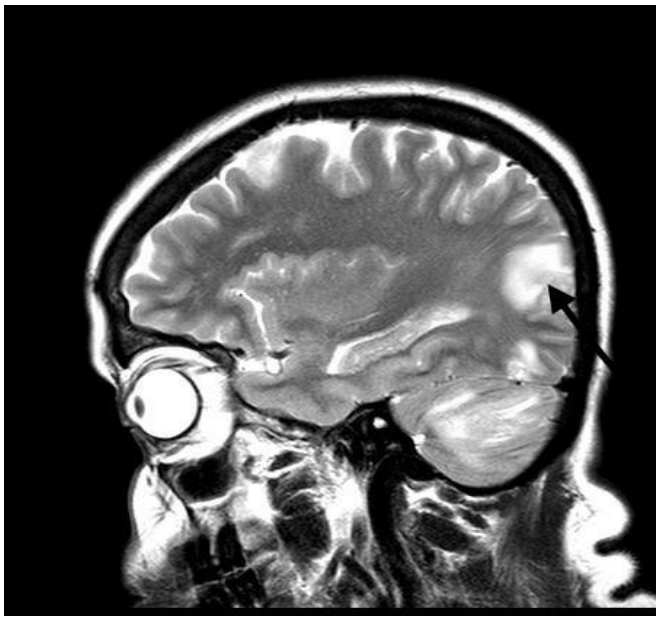

Figure 2

Figure 1 and 2 - MRI BRAIN showing T1 signal hyperintensity in posterior brain suggestive of posterior reversible encephalopathy syndrome. (Black Arrow)

Case Report 2

A 28 year old female presented with chief complaints of easy fatiguability of 1 month, pedal edema, abdominal distension, puffiness of face of 2 weeks and cough with expectoration of 5 days duration. Vitals at the time of examination were stable, except JVP being elevated.

Local examination revealed a Malar rash over the face with thickened skin over bilateral feet. Auscultation

revealed Bilateral basal crepitations and an Ejection Systolic Murmur of grade 2 in the Neo aortic area. Complete hemogram revealed microcytic hypochromic anemia with hemoglobin of 3.8 g/dl. In view of severe anemia, 4 units of Packed Red Blood Cell transfusion was done. Chest X-ray showed Bilateral Reticulonodular Infiltrates and HRCT thorax showed features of Interstital Lung Disease. USG abdomen and pelvis revealed mild splenomegaly with mild ascites. Patient was started on Injection Ceftriaxone and Tablet Azithromycin suspecting a lower respiratory tract infection. 2 Dimensional Echocardiography done revealed Mitral valve prolapse with Moderate Mitral Regurgitation, Moderate Tricuspid Regurgitation, and Pulmonary Artery Hypertension with PASP of 54mm Hg. Urine routine showed 1+ albumin and RBCs 3/hpf. TSH was 5.476. A CT Pulmonary Angiogram done in view of Raised D-dimer showed an Embolus in the right main pulmonary artery SIZE). Patient was immediately started on Oral Dabigatran. Complete blood picture 1 week later, revealed a Hemoglobin of 10.4 g/dl. However, Thrombocytopenia was persisting with Platelet count of 1 lakh/mm^3. Coombs test was negative. ANA showed IF 2+, anti JO antibody 1+ with C3/C4 being normal. APLA workup showed Anti beta 2 microglobulin antibody positive. Patient was diagnosed as SLE-limited systemic sclerosis overlap.

Patient was started on tadalafil in view of pulmonary hypertension. In view of an SLE flare, patient was started on pulse therapy of methyl prednisolone. At the time of discharge the patient was initiated on mycophenolate mofetil and hydroxychloroquine. Patient had improved clinically.

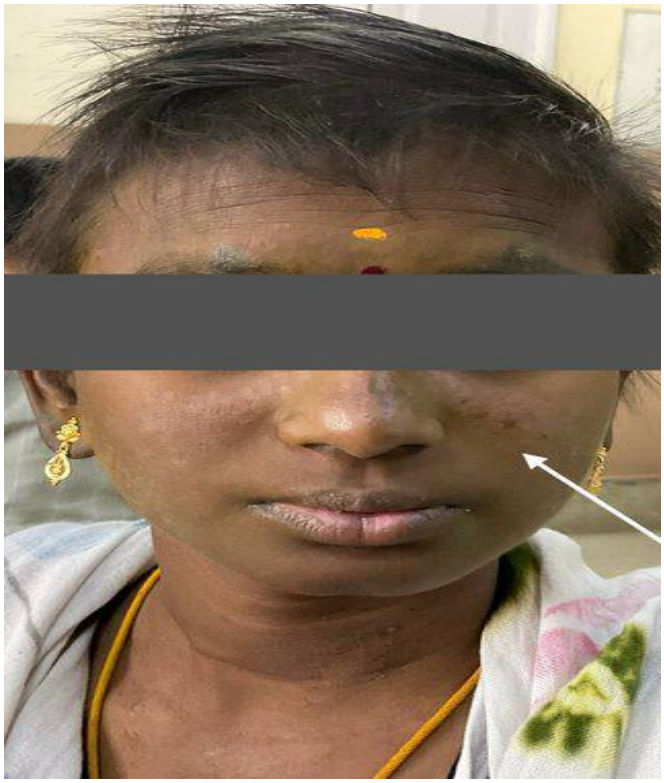

Figure 3: Image showing Malar Rash (white arrow)

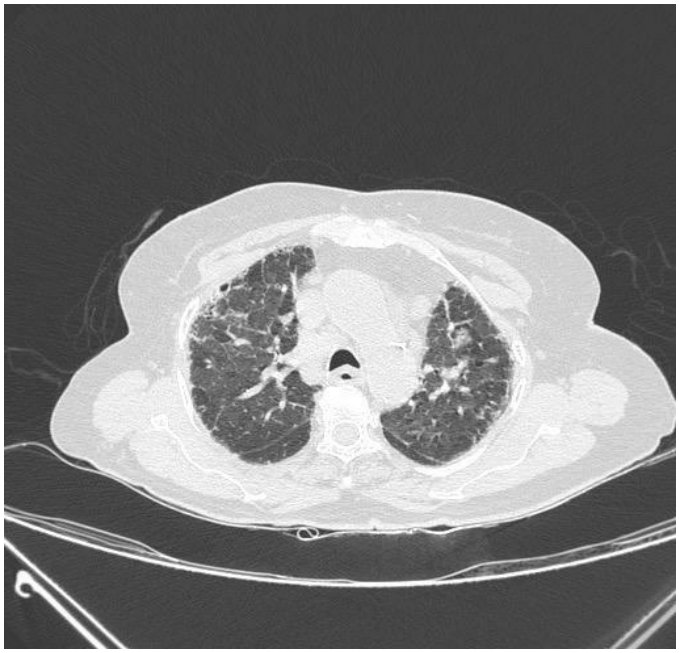

Figure 4: Contrast enhanced computed tomography of chest showing multiple bilateral ground glass opacities suggestive of Interstitial Lung Disease.

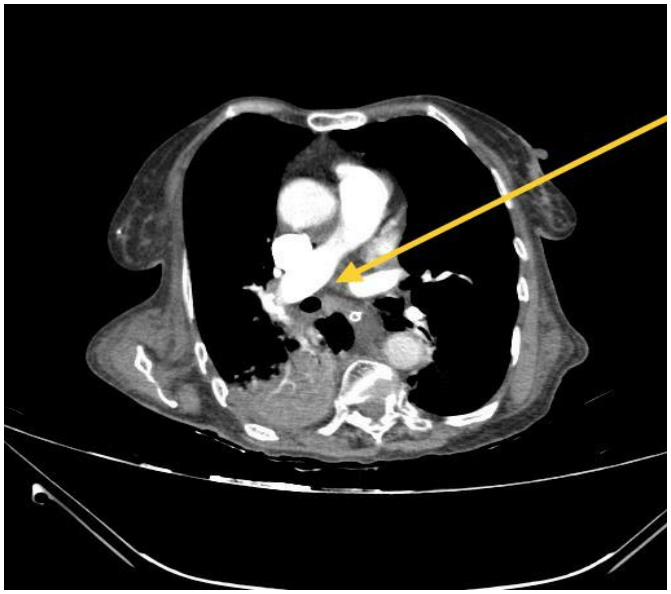

Figure 5: CT Pulmonary Angiography showing Thrombus in Right pulmonary artery (Yellow arrow)

Case Report 3

A 32-year-old lady presented with complaints of weakness of both lower limb followed by upper limbs of 3 months duration. Patient also gave a history of generalized body swelling and exertional breathlessness for 1 month prior to presentation, associated with difficulty in swallowing in the last 10 days. There was no history of fever, rashes or joint pains. Bilateral pitting edema was noted. Cranial nerve examination was normal. Motor examination showed hypotonia and proximal muscle weakness of all four limbs. Sensory examination was normal.

Investigations revealed anemia with hemoglobin of 10.2 g/dl, thrombocytopenia of 1.13 lakh/$mm^{3.}$ Thyroid profile revealed Subclinical Hypothyroidism with TSH 5.08.ng/ml. ECG done showed low voltage complexes. A 2-Dimensional Echocardiography done showed massive pericardial effusion with evidence of pulmonary hypertension. Pericardiocentesis was immediately done and patient was started on Tablet Thyroxine 25 mcg once daily. In view of motor weakness an MRI whole spine screening done showed no evidence of lesions/collections in the spinal cord or stenosis. MRI muscle in both upper and lower limb showed evidence of myositis. CPK was elevated at 6330 U/L. Considering an autoimmune etiology, workup done revealed ANA Positive, u1-RNP positive and Sm Antigen Strongly Positive 3+. ANCA, paraneoplastic panel, serum myositis profile was all negative. Nerve conduction study was normal. MRI of lower limbs done revealed myositis. A diagnosis of SLE With Overlap myositis - MCTD was inferred. Patient was treated with Intravenous pulse dose of methylprednisolone for 5 days followed by tapering doses of oral steroids and methotrexate. Patient showed gradual improvement of symptoms and is currently being followed up.

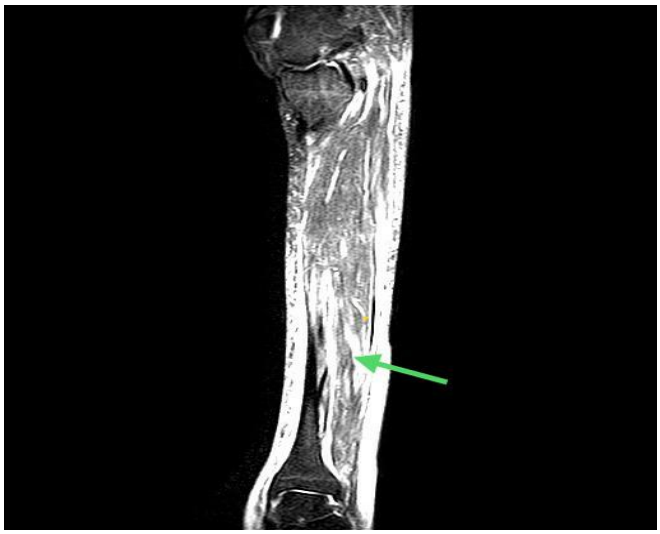

Figure 6: MRI Lower Limg Showing Myositis (Green Arrow)

Case Report 4

A 25 year old female patient presented to us with complaints of low grade fever on and off, loss of appetite and weight loss, multiple joint pains both small and large joints of 3 months duration. The patient also gave a history of oral ulcers and generalized weakness with rash over the face of 2 months duration. Patient also

gave history of intermittent pedal edema and cough with occasional blood streaked sputum.

On examination pallor was present with a malar rash over the face. Bilateral pitting pedal edema was noted. On auscultation, tubular bronchial breath sounds were heard in the left infra-axillary and infra-scapular areas.

Laboratory tests revealed pancytopenia with hemoglobin of 8.7gm/dl , total leukocyte count of 3560 cells/mm^3 and platelets of 1.44 lakhs/mm^3. Creatinine was mildly elevated at 1.35mg/dl. Urine routine showed albumin 2+ and RBCs 4+. Chest X-Ray was normal. An HRCT thorax done showed left lower zone consolidation with minimal left pleural effusion. A 2 Dimensional Echocardiography was done which revealed no vegetations, reported normal study.

Patient was treated with intravenous Meropenem 7 days for pneumonia.

Fever spikes responded to antibiotics with cough and hemoptysis subsiding gradually.

A 24 hour urine protein creatinine ratio done revealed a ratio of 1.79. Further work up for pancytopenia considering an autoimmune etiology revealed ANA 3+, dsDNA+, anti nucleosome+ and anti-histone 2+. P-ANCA was positive, C3 low, C4 was normal. In view of features suggestive of Lupus nephritis, a renal biopsy was done which showed proliferative Lupus Nephritis class III - IV. Patient was started on injection cyclophosphamide along with Mesna. Tapering doses of Prednisolone along with Hydroxychloroquine and Mycophenolate Mofetil were started. Patient improved and was kept on regular follow up.

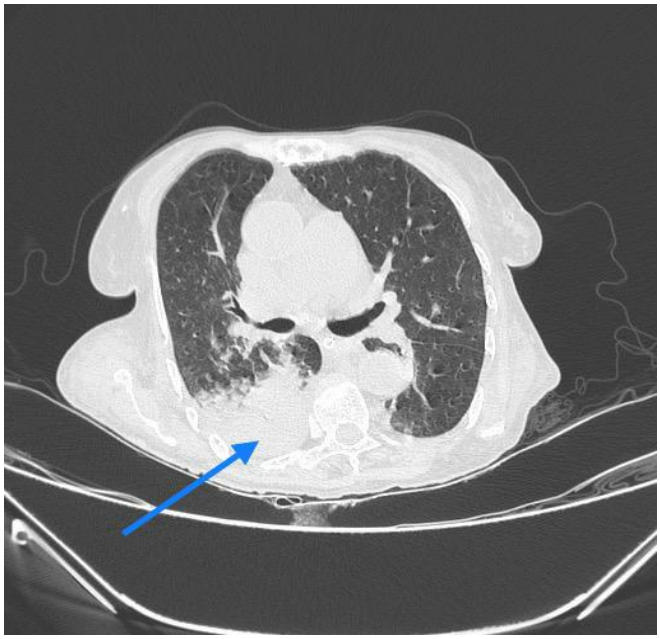

Figure 7: CECT THORAX showing Left Lower Zone Consolidation (Blue Arrow)

Case Report 5

A 33-year-old female with no known comorbidities was admitted with complaints of heaviness of both feet. The following day, patient complained of inability to move her feet. It was also associated with tingling and burning paresthesias which was sudden in onset, non-progressive. Patient also gave a history of hematuria. There was no history of bowel involvement. There was no history of weakness in the upper limbs, fever, rashes, arthralgia or oliguria.

On examination, the patient was conscious, oriented, afebrile. Skin, scalp, oral cavity were normal. Vitals at the time of examination revealed a blood pressure of 150/90 mm Hg. Motor system examination revealed nil power in bilateral ankles with power of ⅘ in bilateral knees with normal power in hip and upper limbs.

Sensory system examination revealed graded sensory loss over bilateral foot and legs till below the knee. Other systemic examination was within normal limits.

Initial workup revealed normocytic normochromic anemia Hb of 7.6 g/dl , Direct Coombs test was positive. LFT revealed transaminitis with AST of 120 IU/ml, ALT of 73 IU/ml and hypoalbuminemia. Urine examination revealed red blood cell casts in urine analysis. 24 hrs urine protein was 550mg/day.

MRI whole spine screening showed no evidence of nerve root compression, spinal canal stenosis or enhancing lesion/collections involving the spinal cord or vertebrae. Bilateral lower limb doppler showed no features of deep vein thrombosis or cellulitis. In view of multi system involvement a diagnosis of connective tissue disorder/vasculitis was considered. ANA 2+ , anti-Smith/RNP 2+.

Immune work up ANCA was negative. An ENMG done was suggestive of severe motor and sensory axonal neuropathy in the lower limbs. 2 Dimensional Echocardiography was normal. In view of progressive symptoms and multisystem involvement (AIHA/ peripheral neuropathy-mononeuritis multiplex/ autoimmune hepatitis/ nephritic range proteinuria) after ruling out other causes, a diagnosis of lupus was made. Patient was initially started on intravenous pulse dose of methylprednisolone for 3 days and patient clinically improved. Patient was later started on pulse cyclophosphamide therapy along with a tapering dose of steroids. Patient was discharged with immunosuppressant therapy and had no further deterioration on follow up.

Discussion

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease characterized by a high expression of autoantibodies and the systemic involvement of various systems, with different clinical manifestations and a complex pathogenesis.[4] SLE is predominantly a disease which presents in women of child bearing age group.[5] The disease can present with slowly or rapidly progressive active disease.

The disease is prone to relapses and remissions resulting in considerable morbidity due to disease flares activity and accumulated damage.[6] SLE diagnosis still represents a challenge, remaining largely based on clinical judgment.[7] It is important to ensure that the diagnosis of lupus is appropriate before considering treatment, given the wide variety of clinical manifestations/organ involvement that can occur and in the differential diagnosis of many acute and sub-acute presentations. A strong index of suspicion and early diagnosis is needed to prevent rapid progression of the disease.

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