Section: General Medicine

Case Series

CASE SERIES ON SLE

Dixitha A[1], Meenu C Nair[1], Lanord Stanley Jawahar M[2]

[1]Post Graduate, Department of General Medicine, SRM Medical College and Research Centre, Chennai, Tamil Nadu, India.
[2]Professor, Department of General Medicine, SRM Medical College and Research Centre, Chennai, Tamil Nadu, India.

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Corresponding Author:
Dr. Dixitha A,
Email: dixitha1@gmail.com
ORCID: 0000-0003-0640-0294

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Abstract

In this case series we discuss the various manifestations of systemic lupus erythematosus (SLE) in a tertiary care hospital. SLE is a chronic autoimmune disease of variable severity and development. The patients with SLE may present with numerous systemic manifestations. It may present as generalized seizures, cerebrovascular accidents, or neuropsychiatric manifestations. Early diagnosis will aid in treating the patient successfully. The general symptoms are fever, malaise, myalgias, arthralgias, headache, loss of weight and appetite. In new instances or recurring active SLE flare-ups, nonspecific fatigue, fever, arthralgia, and weight changes are the most prevalent symptoms. The most of patients can present with varied symptoms like fever and generalized weakness, and co-existing with other morbidity such as nephritis. This case series will assist in recognizing diverse manifestations of SLE and their precise treatment.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease of variable severity and course. Its history dates back to 916 AD when Herbernus of Tours applied the term “lupus” to a skin disease. In 1872 Kaposi subdivided this disease into two types discoid and systemic, from thereon SLE has been defined by its variety of cutaneous and systemic manifestations with potentially fatal outcomes in many cases.[1] There are four main types of lupus neonatal, discoid, drug-induced, and systemic SLE. Although the precise mechanism remains a mystery various genetic, hormonal, environmental factors and immune abnormalities have been identified.[2] This disease can affect the kidneys, lungs, skin, nervous system, and musculoskeletal system. CNS is clinically involved in approximately 40% of all SLE patients.[3] It may present as generalized seizures, cerebrovascular accidents, or neuropsychiatric manifestations. In this case series, we have taken 6 cases of SLE with different manifestations. The ACR: EULAR 2018 criteria have been used to diagnose SLE in all these cases.

CASE SERIES

Case 1: A 30-year-old female, came to the hospital with complaints of generalized anasarca, abdomen pain, right side hip pain, and rashes over her face for one-month duration. She was emaciated with severe pallor, malar rash, hyperpigmentation of hard palate, right iliac fossa tenderness, gross ascites, and bilateral pitting pedal edema. Peripheral smear showed severe normocytic normochromic anemia. The urine routine showed 3+ albuminuria with the granular cast. Ultrasound abdomen showed normal-sized kidneys, hepatosplenomegaly, and ascites. CT chest showed bilateral pleural effusion. USG guide pleural tapping was done and showed transudative effusion. ANA was found to be 3+ (Cytoplasmic fluorescence) and Anti dsDNA was low positive. The patient had low complement C3 and C4.

The patient's vitals were as follows: Hb was 7.9 on day 1, while 8.3 and 7.9 on day 7 and day 10, respectively. WBC count was 4080 on both day 1 and day 10, while it was 3430 on day 7. Platelet was 1,34,000, 91,500, and 2,03,000 on day 1, 7, and 10, respectively. ESR was 78, 43, and 27 mm/hr on day 1, 7, and 10, respectively. CRP was 130, 36, and 14 on day 1, 7, and 10, respectively. Urea was 64, 45, and 24 on day 1, 7, and 10, respectively, while Creatinine was 1.4, 1.3, and 0.8 on day 1, 7, and 10, respectively.

MRI of hip showed an intramuscular abscess with calcification in the right iliac and intramuscular chronic hematoma, with mild joint effusion in bilateral subcutaneous edema. 24-hour urine protein was done which detected moderate proteinuria subsequently renal biopsy was performed and it showed Grade IV lupus nephritis. The patient was treated with a pulse dose of steroids, immunomodulatory with cyclophosphamide, hydroxychloroquine, Ramipril, multivitamins, and sunscreen lotion.

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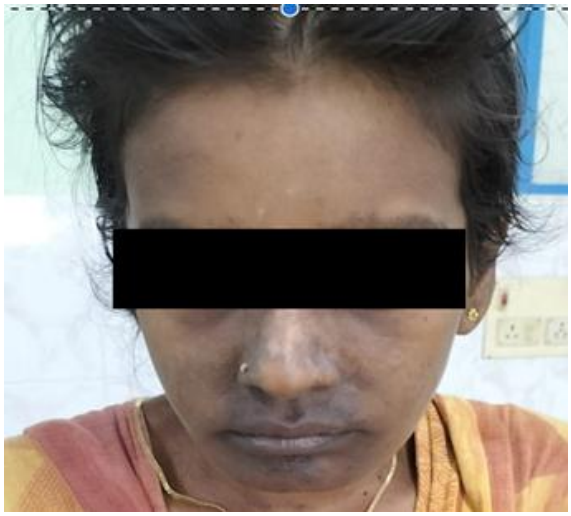

Figure 1: (Original) Malar Rash

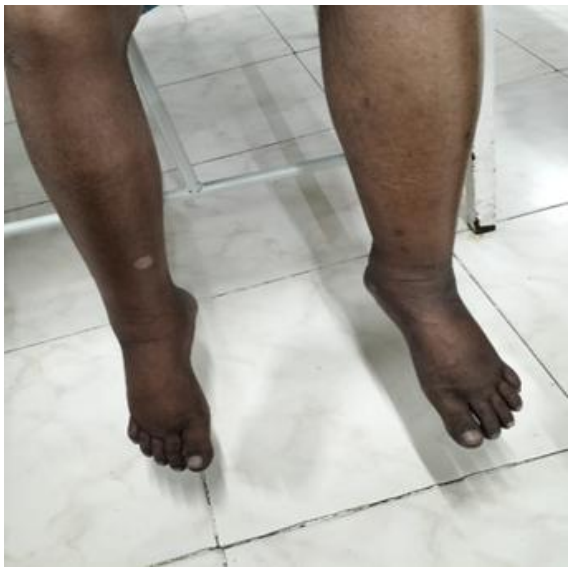

Figure 2: (Original) Bilateral pitting pedal edema

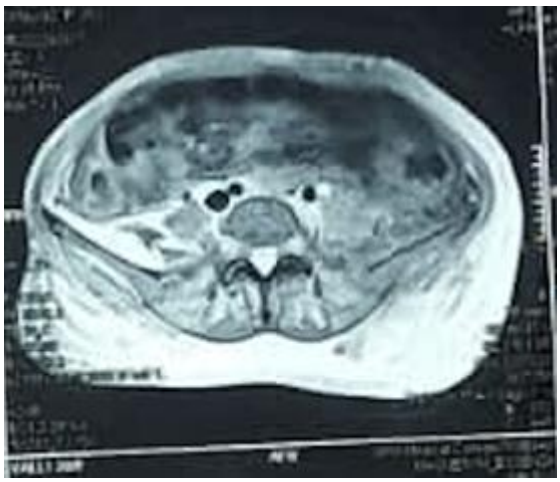

Figure 3: Right sided psoas abscess

Case 2: A 45-year-old female came to the hospital with fever, numbness of bilateral lower limbs, and generalized weakness of 2 weeks duration. She had pallor, and malar rash, sensory examination revealed vibration was reduced in bilateral lower limbs up to the knee. Upper limb sensory examination was normal.

The patient's vitals were as follows: Hb was 8.3 on day 1 and 8.9 on day 5. WBC count was 3040 on day 1 and 4350 on day 5. Platelet was 2,43,000 and 2,35,000 on day 1 and 5, respectively. Urea was 23 and 24 on day 1 and 5, respectively, while Creatinine was 0.7 and 0.8 on day 1 and 5, respectively. ESR was 58 and 29 mm/hr on day 1 and 5, respectively. CRP was 134 and 34 on day 1 and 5, respectively.

ANA shows 3+ (Homogenous pattern) Anti dsDNA positive, C3 and C4 levels were low, Anti Ribosomal P positive, Anti Ro and La were low positive. A nerve conduction study showed bilateral Upper limb and lower limb polyradiculopathy, suggestive of sensory-motor polyradiculoneuropathy. The patient was treated with hydroxychloroquine, steroids, pregabalin and nortriptyline, sunscreen lotion, and multivitamins.

Case 3: A 29-year-old female presented with multiple joint pain, early morning stiffness, fever, chest pain, and generalized weakness. The patient had a history of generalized tonic-clonic seizures 6 months back for which she was started on antiepileptics with no further episode of seizures and a history of weight loss of 5 kgs in 1 month. Patient had bilateral distal interphalangeal joint and proximal interphalangeal joint and wrist joint tenderness and bilateral knee joint tenderness.

The patient's vitals were as follows: Hb was 5.4, WBC count was 6730, platelet was 2,34,000. ESR was 78, CRP was 96, Urea was 24, while Creatinine was 0.6.

Peripheral smear showed severe microcytic hypochromic anaemia. Chest X-ray showed bilateral mild pleural effusion. ECG showed low voltage complexes ECHO showed massive pericardial effusion and Pericardiocentesis was done and showed transudative pericardial effusion. ANA shows 2+ (speckled pattern), Anti dsDNA was positive and low C3, and C4 levels. The patient was treated with steroids, hydroxychloroquine, azathioprine, and multivitamin.

Case 4: A 37-year-old female presented to the hospital with joint pain, dyspnea on exertion, headache, and fever of 1-week duration. The patient had pallor. she had right middle finger swelling of the distal phalanx and was diagnosed with paronychia. The patient's vitals were as follows: Hb was 7.9, WBC count was 6500, platelet was 1,01,000. ESR was 45 and CRP was 76.

ANA was 3+ (homogeneous pattern), Anti dsDNA was positive and low C3 levels. 24-hour urine protein showedmoderate proteinuria. Direct Coomb's test was positive. She was diagnosed with autoimmune hemolytic anaemia. A renal biopsy showed grade 5 Lupus Nephritis. She was treated with steroids, MMF, hydroxychloroquine, amitriptyline, Ramipril, and multivitamins.

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Case 5: A 35-year-old female with a history of fever x 2 weeks, altered sensorium x 2 days. Fever, high grade associated with chills and rigor, myalgia, and headache. No diurnal variation. She also gave a history of severe headache and throbbing kind of pain. No history of joint pain or rashes. She is a known case of hypothyroidism on treatment, no other comorbidities. The patient developed an episode of generalised tonic-clonic seizures while in the causality for which antiepileptics were given. She was conscious, disoriented, and febrile, pallor was present and CNS Examination did not reveal any focal neurologic deficit or signs of meningeal irritation.

The patient's vitals were as follows: Hb was 9 on day 1, 7.8 on day 7, and 9.8 on day 14. WBC count was 5400, 2030, and 4030 on day 1, 7, and 14. Platelet was 1,54,000, 1,01,000, and 2,30,000 on day 1, 7, and 14 respectively. ESR was 78, 86, and 45 mm/hr on day 1,7, and 14, respectively. CRP was >130 on both day 1 and 7, while on day 14 it was 24. AST was 98, 97, and 45 on day 1,7, and 14, respectively. ALT was 77, 78, and 64 on day 1,7, and 14, respectively. Creatinine was 1.1, 1.2, and 1 on day 1, 7, and 14, respectively.

Peripheral smear showed normocytic normochromic anaemia initially and pancytopenia on day 7. Cultures were found to be negative. ANA was done and was found to be 3+ (speckled pattern). MRI Brain showed features suggestive of CNS lupus. The patient was started on high-dose pulse methylprednisolone therapy for 3 days with which the patient sensorium improved and there were no further seizure episodes. The patient sensorium came back to normal; fever spikes came down and steroids were tapered.

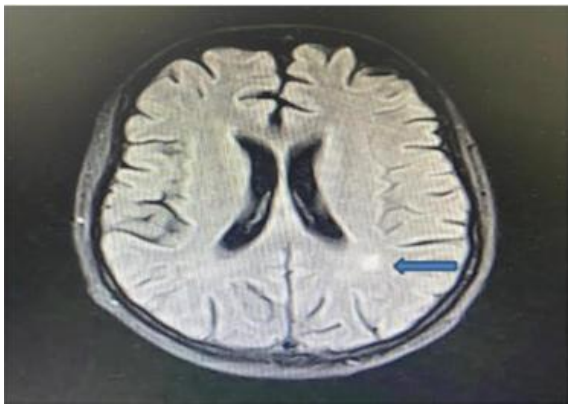

Figure 4: (Original) Shows chronic infarct in the deep white matter of periventricular region

Case 6: A 45-year-old female known case of SLE on treatment presented with complaints of generalized weakness, reduced sensorium, reduced urine output, rashes over the body no c/o fever or joint pains. At the presentation, she was conscious and oriented. Generalized malnourishment was present. Dark-pigmented maculopapular rashes were seen all over the body [Figure 5]. Painful oral ulcers were present.

The patient's vitals were as follows: Hb was 7.4, WBC count was 6030, platelet was 1,90,000, ESR was 45 mm/hr, CRP was 96, urea was 64, and Creatinine was 1.3. Peripheral smear showed microcytic hypochromic anemia. The urine routine showed 3+ albuminuria. ANA was positive 2+ (cytoplasmic pattern), positive anti-dsDNA and reduced C3, and C4 complement levels. She underwent a renal biopsy which showed Grade 3 lupus nephritis. She was started on steroids, MMF, and other supportive measures.

Figure 5: (Original) Maculopapular rash over the scalp and body

DISCUSSION

SLE is an autoimmune disease with multisystem involvement. It is predominantly seen in women in the reproductive age group.[4] The condition has several phenotypes, with varying clinical presentations from mild mucocutaneous manifestations to multiorgan and severe central nervous system involvement.[5] It is thought that single gene mutations causing deficiencies of the complement components C1q, C2, or C4 can promote SLE, possibly by impairing the clearance of immune complexes and apoptotic cell debris.[2] Several autoantibodies are identified for their diagnosis. The diagnosis of SLE is based on signs and symptoms, laboratory testing, and diagnostic testing tailored to each patient.

In our case series of cases, we were able to see the different presentations of SLE. Case A presents with cutaneous, arthritis, serositis, and nephritis as a manifestation of lupus erythematous. Case B is a maternal relative of Case A and presents as neuropathy rather than with serositis. Case C presents as massive polyserositis. Case D presents as a

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hematological, cutaneous, and nephropathy presentation of the disease. Case E presents as pyrexia of unknown origin and later made our diagnosis of SLE. Case F presents as a CNS manifestation leading us to make a diagnosis of lupus.

Serositis, one of the criteria of SLE may be present in patients with pleuritis, pericarditis, and pleural and pericardial effusion. The main reason for serositis is immune-mediated inflammation of serous membranes caused by autoantibodies.[6] Lupus nephritis can lead to nephrotic syndrome and renal failure in developed patients by autoantibodies. Nephritis is divided into 5 pathologic classification groups by World Health Organization.[7] An increase in dsDNA antibody titer is associated with an increased risk of renal flare and with hypocomplementemia (with an inverse correlation with C3). Antibody titers drop in response to treatment, with a 50% decrease in dsDNA antibody titer reducing renal flare rate by 52-53% in two treatment cohorts.[8] The diagnostic criteria of the ACR are widely used for the diagnosis of SLE. The application of these criteria for the diagnosis of Japanese patients has been reported to have a sensitivity of 97% and specificity of 89%.[9,10]

CONCLUSION

This case series highlights the different clinical presentations of the SLE patients. According to this investigation, the most frequent clinical symptoms were hematological, renal, and musculoskeletal in nature. Patients with pleuritis, pericarditis, and pleural and pericardial effusions may exhibit serositis, one of the SLE criteria. Serous membrane inflammation mediated by the immune system and brought on by autoantibodies is the primary cause of serositis. The most prevalent autoantibody among SLE patients was anti-dsDNA antibody, which has been linked to hypocomplementemia and a higher risk of renal flare. Additionally, our research implies that SLE patients with high anti-dsDNA titers may be more likely to experience a variety of clinical symptoms, which would add to the complexity of their disease.

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