ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 06 | June - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

VARIED PRESENTATION OF SYSTEMIC LUPUS ERYHTHEMATOSUS - A CASE SERIES

Rheumatology

Dr. Nilima K. Shah* Professor & Head Of Unit, General Medicine, B.j Medical College, Ahmedabad B. J Medical College & Civil Hospital, Asarwa, Ahmedabad. 380016 *Corresponding Author

Dr. Nikunj V Dadhaniya Consultant Rheumatologist, B.j Medical College, Ahmedabad B.j Medical College & Civil Hospital, Asarwa, Ahmedabad. 380016

Dr. Parag V. Tarale 3rd Year Resident General Medicine, B.j Medical College, Ahmedabad B.j Medical College & Civil Hospital, Asarwa, Ahmedabad. 380016

Dr. Krupit H. Vinchhi 3rd Year Resident, General Medicine, B.j Medical College, Ahmedabad B.j Medical College & Civil Hospital, Asarwa, Ahmedabad. 380016

Dr. Milind R. Solanki 2nd Year Resident, General Medicine, B.j Medical College, Ahmedabad B.j Medical College & Civil Hospital, Asarwa, Ahmedabad. 380016

ABSTRACT

Background: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease of unknown etiology with female preponderance of reproductive age group. It is a multi system disease with highly variable presentation. We report four cases of Systemic lupus erythematosus with different organs affected presented to our hospital. We prospectively collected data of Systemic lupus erythematosus patients admitted to our department with unusual presentation. **Case presentation:** Our case series, there were different presentation of Systemic lupus erythematosus includes following. First one was twenty year old Indian female had constitutional symptoms with lymphadenopathy mimicking tuberculosis, second case was about thirty year old Indian female having autoimmune myelofibrosis, third case was of twenty year old Indian female with lupus myelitis and lastly a case of twenty one year old Indian female having polyneuropathy. All of them were treated successfully with steroids and other immunosuppressive agents. **Conclusion:** Systemic lupus erythematosus has variable clinical presentation and can affect any organ-system. Physicians need to be vigilant so as to make early diagnosis and timely treatment.

KEYWORDS

systemic lupus erythematosus, autoimmune myelofibrosis, myelitis, polyneuropathy, lymphadenopathy

INTRODUCTION

Systemic lupus erythematosus (SLE) is autoimmune disease characterized by multiorgan involvement and presence of auto antibodies. SLE has a broad range of presentations and manifestations, and as such, its course and organ involvement are unpredictable. The systems mainly involved are musculoskeletal, mucocutaneous, haematological, vascular, renal, pulmonary, and central and peripheral nervous system. The wide range of presentations and differential diagnosis often pose a diagnostic challenge for clinicians.

Case Presentation

Case 1

A twenty year old unmarried female presented with anorexia, fatigue, weight loss, joint pain, apthous oral ulcers and intermittent fever for last 8 months. On physical examination she had bilateral non-matted, nontender axillary, inguinal lymphadenopathy with normal other systemic examination. Routine investigations suggestive of anemia (Hb – 7.20 gm/l), and leucopenia (WBC- 3581 /cumm) Renal and liver function test were within normal range. Direct coombs test was grade 2 positive. Ultrasonography showed multiple sub-centimetric lymph nodes in mesenteric region, axillary and inguinal lymph nodes. Histopathology from axillary lymph node showed reactive hyperplasia without any evidence of tuberculosis or malignancy and CBNAAT was negative, raising suspicion of autoimmune disease. Her ANA was positive (Homogenous, 1:160 titer) and ANA profile was positive for dsDNA. She was diagnosed as having SLE with lymphadenopathy, constitutional and hematological involvement. She was started on prednisolone 20 mg and hydroxychloroquine 200 mg and haematinics. On follow up After 15 days Patient showed improvement in appetite and started gaining weight with reduced constitutional symptoms and improvement in the blood counts. Azathioprine 50 mg was added and steroid was tapered off over 4 months.

Case 2

A thirty year old female presented with gradually progressive abdominal distension, dyspnoea on exertion and generalized weakness since 30 days. She had tachycardia, pallor and ascites. Other systemic examination was unremarkable. Routine investigation showed pancytopenia with Hb –5.3 gm/l, WBC - 600/cumm (neutrophils-400, lymphocytes 200, eosinophils-0, monocytes and basophils-0), platelets- 36000/cumm with normal renal and liver function tests except decreased albumin- 2.2 g/dL. Ultrasonography showed hepato-splenomegaly with gross ascites and bilateral moderate pleural effusion. Both ascitic and pleural fluid examination revealed exudative characteristic with normal cell count, normal ADA levels and negative CBNAAT. Her Vitamin B12 – 150 pg/mL , serum TSH – 7.5 mIU/L, Ferritin- 220 ng/mL , LDH -165 U/L, HIV and HCV were non reactive. Urinary examination was normal, 2D echo showed normal study. In view of low absolute neutrophil count, she was started on prophylactic antibiotics, antifungals. bone marrow biopsy examination showed hyper cellular marrow with Moderate fibrosis. Further JAK 2 and CALR gene mutation was done to rule out primary myelofibrosis which were negative. Considering young female with polyserositis and pancytopenia her ANA was sent, which was positive (1:80 + speckled pattern). Her dsDNA was positive. She was diagnosed as secondary autoimmune myelofibrosis. Patient started on tablet prednisolone 1mg/kg with injectable filgrastim 300 mg once a day. She had gradual recovery of hematological parameters. She was discharged on oral prednisolone 20 mg, hydroxychlororquine 200 mg and azathioprine 50 mg was added in the follow up.

Case 3

A twenty year old female presented with history of intermittent low grade fever, joint pain since 2 month, insidious onset bilateral lower limb weakness with difficulty in walking, bladder and bowel incontinence since last 15 days. CNS examination showed increased tone in bilateral lower limbs, reduced muscle power in all four extremities: the MRC grade of the upper limbs was 4/5, while in the lower limbs it was 0/5 in both proximal and distal muscles. Knee and ankle reflexes were exaggerated and normal in upper extremities. The laboratory examination showed Hb– 12 gm/l, WBC 4650/cu mm, platelet – 203 × 10^3/cu mm, ESR –70 mm/hr, RFT, LFT within normal range. MRI scan done which shows changes of myelitis involving cervical dorsal cord extending from C7- D12 levels. CSF examination reveals no significant findings. Aquaporin 4 receptor and myelin oligodendrocytes glycoprotein antibody testing was negative. Patients ANA titer levels were 1:160 with homogenous pattern. Her anti-dsDNA and anti-ribosomal P antibodies were positive. She was diagnosed as SLE longitudinal extensive transverse myelitis. Patient started on intravenous gamma-globulin for 5 days along with intravenous methylprednisolone 1000 mg for 5 days along with

cyclophosphamide 500 mg fortnightly for 6 pulses. At the 3-month follow-up, the patient's neurological deficit was completely resolved and was able to walk independently.

Case 4

A twenty one year old unmarried female presented with insidious onset and gradually progressive weakness and numbness over bilateral lower limb (left > right) and upper limb from last 3 months, burning sensation over bilateral soles from last 1 month and intermittent fever from last 4-5 days. On CNS examination patient conscious oriented with normal tone in all limbs with power 4/5 proximal and distally in all 4 limbs. Plantar reflex was bilaterally absent, deep tendon reflex was symmetrically absent in bilateral lower limb and normal in upper limb, touch, pain temperature, joint position, fine touch, vibration were absent below ankle joint bilaterally. The laboratory reports were as follows with Haemoglobin -7.10 gm/l, WBC– 13090/cu mm, platelets – 304000/cu mm with normal renal and liver function. EMG NCS revealed demyelinating plus axonal motor and sensory polyradiculopathy. MRI spine was normal and Brain revealed multiple tiny acute infarcts in bilateral subcortical white matter and in right cerebellar hemisphere. Her ANA was positive with 1:160 homogenous pattern and ds DNA by ELISA. Sural nerve biopsy showed predominantly axonal changes with perivascular inflammation and perinuclear thickening. Patient diagnosed with SLE with polyneuropathy.

Patient was given IVIg 0.4 gm/kg per day for 5 days and intravenous methylprednisolone 1000 mg for 3 days followed by oral prednisolone 50 mg for 4 weeks with further tapering and azathioprine was added. She has partial recovery in weakness and sensation.

DISCUSSION

SLE is a multisystem autoimmune disease with manifestation ranging from subtle rash to life threatening disease. Hereby we have described 4 different cases with various organ system involvements.

1st case had initial presentation of lymphadenopathy with low grade fever. Initial impression was that of the tuberculosis. On further evaluation tuberculosis was ruled out and based on constitutional symptoms and investigation, SLE was diagnosed. In 2nd case patient presented with pancytopenia and splenomegaly raising suspicion of primary hematologic disorder. Workup for primary myelofibrosis was negative so workup for autoimmune disease was done and was diagnosed as SLE with secondary myelofibrosis. 3rd case had findings of longitudinal extensive transverse myelitis for which work up for Neuromyelitis optica was negative. The Autoimmune work up turned out to be positive for SLE. Our 4th case presented with chronic symmetric progressive neuropathy which was confirmed by nerve biopsy, and on further investigation for cause revealed dsDNA positive and diagnosed with SLE with polyneuropathy. Systemic lupus erythematosus is autoimmune disorder with variety of clinical presentation from mild to severe in course. It needs strong suspicious for early diagnosis and treatment to prevents further complications.

Conflicts of interest disclosures-

The authors declare that they have no conflict of interest

Funding-

Not applicable

Ethics approval -

Not applicable

Written informed consents -

written informed consent was obtained from the patient for publication of this case series and any accompanying images. A copy of the written consent is available for review by the editor in chief of this journal.

REFERENCES

1. Isenberg D. Myositis in other connective tissue disorders. Clin Rheum Dis. 1984 Apr;10(1):151-74. https://pubmed.ncbi.nlm.nih.gov/6610527/
2. Budman DR, Steinberg AD. Hematologic aspects of systemic lupus erythematosus. Current concepts. Ann Intern Med. 1977 Feb;86(2):220-9. doi: 10.7326/0003-4819-86-2-220.
3. Appel, G. B., Silva, F. G., Pirani, C. L., Meltzer, J. I., & Estes, D. (1978). Renal involvement in systemic lupud erythematosus (SLE): a study of 56 patients emphasizing histologic classification. Medicine, 57(5), 371–410. https://doi.org/10.1097/00005792-197809000-00001
4. Khare S, Rajadhyaksha A. Profile of neurological manifestations in systemic lupus erythematosus, Indian Journal of Rheumatology, Volume 5, Issue 2, 2010, Pages 59-65, https://doi.org/10.1016/S0973-3698(10)60542-0
5. Zoma A. Musculoskeletal involvement in systemic lupus erythematosus. Lupus. 2004;13(11):851-3. doi: 10.1191/0961203303lu2021oa.
6. Stahl NI, Klippel JH, Decker JL. Fever in systemic lupus erythematosus. Am J Med. 1979 Dec;67(6):935-40. doi: 10.1016/0002-9343(79)90633-8.
7. Melikoglu MA, Melikoglu M. The clinical importance of lymphadenopathy in systemic lupus erythematosus.Acta Reumatol Port. 2008 Oct-Dec;33(4):402-6 https://pubmed.ncbi.nlm
8. Petri M. Monitoring systemic lupus erythematosus in standard clinical care. Best Pract Res Clin Rheumatol. 2007 Aug;21(4):687-97. doi: 10.1016/j.berh.2007.01.003. .nih.gov/19107085/
9. Konstantopoulos K, Terpos E, Prinolakis H, Kanta A, Variami E, Kanellopoulou G, Vaiopoulos G, Floros A, Androulaki A, Meletis J. Systemic lupus erythematosus presenting as myelofibrosis. Haematologia (Budap). 1998;29(2):153-6. https://pubmed.ncbi.nlm.nih.gov/9728808/
10. Martínez-Cordero E, Riebeling-Navarro CG, López-Zepeda J, Martínez-Miranda E, Negrete-García MC. Systemic lupus erythematous in children with renal and hematological manifestations. Relation to anti-nDNA antibodies. 1989 Jan. 46(1); 11 – 21. https://pubmed.ncbi.nlm.nih.gov/2785391/