www.jmscr.igmpublication.org
Impact Factor (SJIF): 6.379
Index Copernicus Value: 79.54
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossrefDOI: https://dx.doi.org/10.18535/jmscr/v7i1.12

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Case Report

A Case of Male SLE with unusual presentation

Authors
Dr Balasundaram Padmakumar, Dr Baraneedaran S, Dr Meenu M Tergestina
Dr Vrinda Vijayan
Corresponding Author
Baraneedaran S
Email: *selva.baraneemmc@gmail.com*, Mobile: 9487782324

Introduction

Systemic lupus erythematosus (SLE) is a complex autoimmune disease with significant heterogeneity and periods of relapse and remissions. It is more prevalent in women particularly in their reproductive years. The prevalence of SLE was found to be 3.2 per 100,000 population in a study conducted near Delhi in India[1].The etiology of SLE remains unknown and is clearly multifactorial. The diagnosis is based on characteristic clinical features and presence of autoantibodies. Of all the characteristic clinical features of lupus, it is the extreme sex skewing that remains least understood. Francis Fatoye et al[2] did a population based study in Alberta Canada, which estimated the prevalence to be 27.3 cases and 3.2 cases per10,000 for females and males, respectively.

Female predominates the disease with 9:1 ratioin a study done by weckerle C et al[3] in 2011. The female to male ratio as studied by pande et al[4] in 1993 in India is also 9:1.We present a case of male SLE which is a rare clinical presentation admitted in our tertiary care center with short febrile illness.

Case Report

A 47 year old male, with history of dyslipidemia on regular treatment and a chronic smoker presented with complaints of high grade fever for 7 days, myalgia and calf muscle pain for past 5 days, anuria for past 2 days. On examination patient was in hypotension, icteric, mild pedal edema was present and was started on inotrope support. Blood routines showed Hb-11.2g/dL,TC-8900/µL with 89% neutrophils, platelets-11000/µL, ESR-115mm/hr, serum creatinine-5.1mg/dL, blood urea-180mg/dL, total bilirubin-8.6mg/dL, SGOT/SGPT-381/138, urine pus cells 18 to 20. Patient was taken for emergency hemodialysis, 3 cycles of dialysis were given. A clinical diagnosis of probable Leptospirosis was made and patient was put on Injection Ceftriaxone 1gm IV BD for 5 days. Chest X-ray, ECG and USG abdomen were normal. Viral markers were negative. Peripheral smear was showing moderate thrombocytopenia. Blood sample was sent for IgM ELISA for Leptospirosis on the seventh day of illness and it was positive. Patients general condition improved, blood pressure was stable off inotrope support, urine output was adequate.

Platelets count, RFT and LFT were also improving.

When we planned to discharge the patient, he was complaining easy fatigability and dizziness. He was pale and Hemoglobin was found to be 6.5g/dL. Hemolytic work up was done. Peripheral smear showed no evidence of hemolysis, direct and indirect Coombs test were negative. Serum LDH was 289(120-220U/L) with mild elevation. Serum iron was 55(60-170µg/dL) and TSAT was 16%. Reticulocyte count was 2%, ESR - 150mm/hr. Though there was no evidence of hemolysis in peripheral smear we considered the possibility of hemolysis due to the sudden fall of hemoglobin from 11.2 to 6.5g/dl.In view of persistent elevation of Acute phase reactant ESR, retrospectively when we asked the history patient was revealing multiple joint pains associated with swelling on and offwe considered the possibility of underlying rheumatological condition. On examination there was no joint deformities and no erosions on x-ray.Autoantibodies like ANA and Anti dsDNA were also positive. The diagnosis of SLE was made due to the presence of following four criteria as per the revised ACR classification criteria for SLE.

1) Non erosive Arthritis
2) Hematological abnormalities: anemia and thrombocytopenia
3) Positive ANA by Immunofluroscence
4) Positive Anti dsDNA.

Patient was started on steroids and hydroxy chloroquine 400 mg/day. Patient significantly improved and there was no further fall in hemoglobin.

Discussion

The classification criteria for systemic lupus erythematosus were updated in 1997. At least four of these eleven criteria (in Table 1) are required to classify patients as having systemic lupus erythematosus.

Table 1 Criteria for the Classification of Systemic Lupus Erythematosus[5]

At least 4 of the following are required to classify patients as having SLE in reports of clinical research:

Malar rash
Discoid rash
Photosensitivity
Oral ulcers
Arthritis
Serositis
Renal disorder
Leukopenia (< 4000/µL), lymphopenia (< 1500/µL), haemolytic anaemia, or thrombocytopenia (<100,000/µL)
Neurologic disorder
Positive test for anti-DNA, anti-Smith, or antiphospholipid antibodies
Antinuclear antibodies in high titres

This patient's presentation fulfilled four of these eleven criteria for a definite diagnosis of systemic lupus erythematosus. Certain aspects of this patient's presenting features are quite atypical for classic SLE at initial presentation and could have delayed the diagnosis. One of the interesting thing to consider in studying gender disparities in SLE is the nature of its initial presentation in male and female patients. The greater awareness of SLE as a potential diagnosis in females, may lead to a greater delay in diagnosis in men with similar symptoms. The mean age range at diagnosis in males was 26–55 years[6,7], with females being diagnosed at a mean age of 27.9–42.6 years[7,8]. To address the clinical phenotype at presentation, a number of groups have specifically assessed organ involvement at disease onset. Although significant heterogeneity exists, the most consistent findings are a lower incidence of musculoskeletal symptoms, Raynuad's phenomenon, alopecia and photosensitivity in men at diagnosis, with the suggestion of more prevalent serositis and discoid lupus[9].

Previous studies have reported that Asian patients have higher rates of renal involvement, more active renal disease and higher rates of nephritis-associated autoantibodies in comparison with predominant white populations. Mok *et al.*[10]found

that, after a median disease duration of 103.6 months in males (101.6 months in females), no significant differences in major organ involvement were identifiable, despite a trend towards less Raynuad's phenomenon, alopecia and arthritis in men.

In Europe Stefanidou et al[11] and Voulgari et al[12] examined the clinical phenotype of SLE in Greek patients. The latter found that men had significantly more serositis ($P < 0.01$), less photosensitivity ($P < 0.05$), oral ulcers ($P < 0.01$), RP ($P < 0.05$), thrombocytopenia ($P < 0.05$) or increased ESR ($P < 0.01$) in comparison with women[12]

There is also difference in the frequency of occurrence of some common features of SLE in Indian population and western population as shown in Table 2[13].

Frequency of clinical features of SLE:

Features(%)	Indian data(%)	Western data(%)
Arthritis	72-92	86-94
Alopecia	52-80	50
Skin rash	74-90	60
Photosensitivity	10-62	33-62
Malar rash	37-76	72-90
Oral ulcers	41-61	30
Fever	74-91	80
Lymphadenopathy	26-47	50
Neuro-psychiatric	19-63	20-45
Renal	35-73	29-73
Cardiac	10-29	20-30
Pleuropulmonary	9-54	36-57

This patient was admitted in our tertiary care center for evaluation of short febrile illness and found to have Leptospirosis. Incidentally due to the sudden fall in hemoglobin in spite of clinical improvement of his infection we considered the possibility of SLE.As SLE patients may presents with highly variable clinical features like constitutional symptoms, cutaneous manifestations, musculoskeletal features, glomerulonephritis, and neuropsychiatric disease we have to consider this auto immune disease in the differential diagnosis of patients presenting with varying clinical manifestations including febrile illness. Even though SLE is clearly a female predominant disease it can rarely occur in male also as shown in our case.

Conclusion

Systemic lupus erythematosus is uncommon in males and atypical presentation may make the diagnosis extremely difficult. This case report seeks to draw clinicians attention in India to the probability of systemic lupus erythematosus in males as well as the possibility of presentation with atypical features.

References

1. Malaviya AN. Systemic Lupus Erythematosus. In Association of physicians of India Text book of Medicine Sainani GS(ed)New Delhi(5th ed)1992;1125-32
2. Fatoye F, Gebrye T, Svenson LW. Real-world incidence and prevalence of systemic lupus erythematosus in Alberta, Canada[published online ahead of print, 2018 Jul 9]. Rheumatol Int. 2018; 38(9):1721–1726.
3. Weckerle, C. and Niewold, T. (2010). The Unexplained Female Predominance of Systemic Lupus Erythematosus: Clues from Genetic and Cytokine Studies. Clinical Reviews in Allergy & Immunology, 40(1), pp.42-49.
4. Pande I,Sakharan NG,Kailashet al. Analysis of clinical and laboratory profile in childhood SLE and its comparison with SLE in adults LUPUS 1993; 2:83-87
5. American College of Rheumatology. 1997 Update of the 1982 American College of Rheumatology revised criteria for classification of systemic lupus erythematosus.
6. Keskin G, Tokgöz G, Düzgün N, et al. Systemic lupus erythematosus in Turkish men, Clin Exp Rheumatol, 2000, vol. 18(pg. 114-5)
7. Prete P, Majlessi A, Hamideh F. Systemic lupus erythematosus in men: a retrospective analysis in a Veterans Administration healthcare system

population J Clin Rheum , 2011, vol. 7 (pg. 142-50)

8. Molina JF, Drenkard C, MolinaJ, et al. Systemic lupus erythematosus in males. A study of 107 Latin American patients, Medicine , 1996, vol. 75(pg. 124-30)
9. Font J, Cervera R, Navarro M, et al. Systemic lupus erythematosus in men: clinical and immunological characteristics. Ann Rheum Dis. 1992;51(9):1050-2.
10. Mok C, Lau C, Chan TS, et al. Clinical characteristics and outcome of southern Chinese males with systemic lupus erythematosus, Lupus , 1999, vol. 8 (pg. 188-96)
11. Stefanidou S, Benos A, Galanopoulou V. Clinical expression and morbidity of systemic lupus erythematosus during a post-diagnostic 5-year follow-up: a male:female comparison, Lupus , 2011, vol. 20 (pg. 1090-4)
12. Voulgari P, Katsimbri P, Alamanos Y, et al. Gender and age differences in systemic lupus erythematosus. A study of 489 Greek patients with a review of the literature, Lupus , 2002, vol. 11 (pg. 722-9)
13. Joshi. V.R, Balakrishnan C.Systemic Lupus Erythematosus- Manual of Rheumatology-IRA Publication1999 227-248.