



A RARE CASE OF SYSTEMIC LUPUS ERYTHEMATOSUS IN A 20-YEAR-OLD MALE PATIENT

Rheumatology

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ABSTRACT

Systemic lupus erythematosus (SLE) is a multi-system autoimmune disease characterized by a series of exacerbations and remissions. The disease involves antibody formation against DNA and nuclear proteins, which attack tissues of multiple-organ systems. SLE is 8 to 15 times more common in women of reproductive age compared with age-matched men. Being rarer in males, it can result in delayed diagnosis leading to poorer outcomes. When compared to women, men have a more aggressive clinical course and rapid development of organ damage, which leads to a worse prognosis. Studies have shown that male patients have an increased prevalence of cytopenia, hemolytic anaemia, nephritis, thrombotic events and seizures. This is why prompt diagnosis and treatment are important to prevent the progression of the disease. We report a case of SLE in a young male patient who presented with fever, joint pain, easy fatigability and oral ulcers. Prompt diagnosis and comprehensive laboratory workup can help an alert primary care physician be instrumental in ensuring better outcomes.

KEYWORDS

SLE, lymphadenopathy, male, fatigue

INTRODUCTION:

Systemic lupus erythematosus (SLE) is a chronic, systemic autoimmune disease that occurs due to the interplay of various factors such as genetic and epigenetic alterations, immunological factors, endocrine and environmental factors such as exposure to ultraviolet rays, viral infections, smoking etc. It is a multisystem disorder that can involve the renal system, nervous system, cardiovascular system, integumentary and musculoskeletal system. The disease involves the formation of auto-antibodies against the nuclear material (DNA and the nuclear proteins) of the cell.

SLE is 8 to 15 times more common in women of reproductive age compared with age-matched men [8]. SLE in males is uncommon, with only one male patient reported per nine females [11]. A possible explanation behind this sex predilection is the effect of androgens on T cells [6]. Eight out of every ten patients experience some kind of cutaneous or mucosal involvement. A common presentation is a facial malar rash. A "discoid rash" is seen in sub-acute to chronic forms of the disease [4]. Other dermatological features of SLE are oral and mucosal ulcers, alopecia, and cutaneous bullae. In comparison to females, males have a lesser chance of having prominent mucocutaneous features, psychosis and hypocomplementemia but a greater chance of infections like Tuberculosis.

Men with SLE are reported to have frequent serositis, cytopenia, hemolytic anemia, nephritis, antiphospholipid antibodies, thrombotic events and seizures [3, 5, 10, 12, 17]. Musculoskeletal involvement, Raynaud's phenomenon, photosensitivity, mucosal ulcers and lymphadenopathy are less common in men than in women [2]. The presence of lupus anticoagulant is more frequent in men, but other autoantibodies appear to occur at the same rate. In comparison to women with SLE, men often have a more aggressive clinical course and rapid development of organ damage, which leads to a worse prognosis. Below we present a unique case report of SLE in an Indian male which will help us study the diversity of Indian patients and also contribute to the Inception cohort for Research (INSPIRE) - the first prospective Indian SLE cohort [14].

Case Presentation:

A 20-year-old male patient presented with a history of intermittent low-grade evening-rise fever for the past month and multiple joint pain, easy fatigability and oral ulcerations for the past 7 days. History revealed an episode of Chikungunya fever one month back. Family

history was not significant for any medical illness. On general examination, the patient was conscious, cooperative and well-oriented to time, place and person. His body temperature was 98.6 degrees Fahrenheit, with a regular pulse rate of 96 beats per minute and blood pressure of 118/78 mmHg. Further general examination revealed generalized lymphadenopathy with palpable axillary, cervical and inguinal lymph nodes. Bilateral knee joint swelling and severe pallor was present. The remaining systemic examination was normal.

The laboratory investigations of the patient are as follows:

Table – 1
Laboratory Results

TEST	VALUE
Haemoglobin	6.4 g/dL
MCV	77.60 fL
RDW	23.10
Total Leukocyte count	8870/cu.mm
Platelet	70,000/cu.mm
B12	1278 mcg/dL
Iron	41 mcg/dL
E.S.R.	72 mm/hr.
Anti-ds DNA	100 U/mL
Anti-Nucleosome	100 U/mL
Anti-Sm Antibody	18 U/mL

The peripheral smear was suggestive of normocytic normochromic RBCs with the presence of anisopoikilocytosis and few target and teardrop cells. Liver enzymes were mildly elevated. Bilirubin levels were normal. Serology for HIV, HCV and Dengue was non-reactive. A negative screening for Chikungunya IgM antibodies was noted. ANA titres by immunofluorescence were sent which came significantly positive (titres mentioned in the table). Profile further showed anti-SSA and anti-SSB as positive. Anti-CCP was negative. DCT (Direct Coomb's Test) and ICT (Indirect Coomb's Test) both were negative. Urine for routine microscopic examination was normal with no proteinuria. A biopsy from the right Inguinal Lymph node showed reactive follicular hyperplasia with chronic inflammatory changes. Chest radiograph and ECG were normal with a normal 2D Echocardiography. USG abdomen revealed hepatosplenomegaly.

Based on the above clinical and laboratory as well as immunological studies, the patient was diagnosed as having SLE without any signs of

organ failure (as per SLICC and EULAR criteria). The patient was treated with oral Glucocorticoids (Tab. Prednisolone 60 mg) and HCQ and other nutritional and supportive measures. And he experienced resolution in symptoms of fever and myalgia. The patient was discharged on oral glucocorticoids on a tapering dose, Hydroxychloroquine and nutritional supplements. On the follow-up visit, the patient was symptom-free and showed a marked improvement in his blood investigations.

DISCUSSION:

SLE being an autoimmune disease with multisystem involvement shows a myriad of clinical presentations. Male SLE is associated with delayed diagnosis and poorer outcomes [8].

With improved detection of mild SLE, the incidence of SLE has gone up by three times in the last four decades [16]. Our patient presented with fatigue, a common symptom (seen in 4 of 5 individuals with SLE) that can be severe enough to interfere with day-to-day activity [7, 9, 13, 15]. Men with lupus tend to have a comparable rate of occurrence of skin disease, arthritis and central nervous system manifestations as women. However, they show a lesser prevalence of photosensitivity and higher prevalence of serositis, renal disease, later-age presentation and 1-year mortality. Active illness or infection caused by immunosuppression is the leading factor in early-life mortality. [1].

CONCLUSION:

SLE in males has a poor prognosis and more severe complications especially if there's a delay in diagnosis. Our case report illustrates how a high degree of suspicion at the primary point of care can help reduce diagnosis delays. A primary care physician must be vigilant when identifying SLE in male patients because in rural setups without a staff rheumatologist the primary care physician has an essential role in suspecting and either diagnosing or referring such cases as needed. Early diagnosis is crucial for improving prognosis in cases presenting with complications.

We also note that in a mild version of the disease without organ failure, treatment with anti-inflammatory medications such as Hydroxychloroquine, NSAIDs and low-dose oral glucocorticoids is sufficient enough to reduce symptoms and halt further disease progression. However, if organ failure occurs, treatment with high-dose steroids or immunomodulators is required.

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