

Uncommon Presentation of SLE

Kumkum Sarkar¹, Madhuchhanda Mandal², Sudeshna Mallik³

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Abstract

Systemic lupus erythematosus (SLE) is a connective tissue disease whose etiology is unknown and it predominantly affects women of childbearing age. We report a case of male SLE with puffiness of face and mild pedal edema and mild yellowish discoloration of eyes. After exclusion of all causes of hepatitis all evaluations were used to establish a diagnosis of SLE. It is a great challenge to diagnose SLE among males as it has lower prevalence. Therefore, as primary care doctors, we need to have a high suspicion of SLE even in male. Thus, early treatment may help patients to prevent complications and improve their quality of life.

Keywords: Male, Lupus Erythematosus, Cutaneous, Antibodies, Antinuclear.

INTRODUCTION

SLE is an idiopathic autoimmune disease which involves multiple organs and has varied clinical presentations. Among all the organs involvement, the skin and kidney are more frequently affected. This disease is most common in women of reproductive age.¹ The female-to-male ratio as reported is 8-15:1, whereas the ratios prior to puberty and post menopause are significantly lower at 2-6:1 and 3-8:1, respectively.² Among different ethnic groups, the prevalence for Indians was 12 per 100,000 individuals, with an overall male-to-female ratio of 1:12.³ Though hormone plays an important role in the pathogenesis of the disease, there is

not yet an accurate explanation for the decreased occurrence in men.⁴ The common symptoms such as joint pain, extreme fatigue, and skin rashes are seen in both males and females, but males tend to experience a more severe clinical course.⁵ The approach to managing SLE frequently depends on the specific disease severity and the manifestations of individual patients. Hydroxychloroquine remains the primary long-term treatment component for all SLE patients.⁸ Corticosteroids (e.g. prednisone, methylprednisolone) are commonly prescribed for a short duration.⁹ Other medication options are non-biological disease-modifying antirheumatic drugs (DMARDs) such as mycophenolate, azathioprine cyclosporine, cyclophosphamide and methotrexate, and non-steroidal anti-inflammatory agents.¹⁰

Author's Affiliation: ^{1,2}Assistant Professor, ³Associate Professor, Department of Tropical Medicine, School of Tropical Medicine, Kolkata, West Bengal 700073, India.

Corresponding Author: Kumkum Sarkar, Assistant Professor, Department of Tropical Medicine, School of Tropical Medicine, Kolkata, West Bengal 700073, India.

E-mail: dr.kumkum.sarkar@gmail.com

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CASE

A 23-year-old male presented with puffiness of face and mild pedal edema and mild yellowish discoloration of eyes for 15 days. He gave history of low grade fever for 2 days before onset of these symptoms. He had associated anorexia and 2 kg weight loss during this 15 days. On examination he had mild pallor, icterus and edema. His blood pressure was high on repeated records though his previous blood pressure was normal. His systemic examination was normal. By keeping mind differentials as acute hepatitis and glomerulonephritis we sent all relevant investigations, which showed normocytic normochromic anemia, slightly high liver enzymes with normal bilirubin and proteinuria 1.6 gram per

day. His blood for HBsAg came positive but Anti HBc total antibody was nonreactive and HBV-DNA was not detected. USG whole abdomen and fibroscan of liver were normal. By the time ANA came positive and positive DsDNA but antismooth muscle antibody (ASMA), anti-liver-kidney microsome antibodies were negative. So we sent blood for HBsAg again for reconfirmation, which came negative this time. Now on thorough examination we found faint malar rash which we ignored earlier. We did kidney biopsy which revealed stage IV lupus nephritis. So, we diagnosed him as lupus nephritis with lupus hepatitis. We treated him with monthly cyclophosphamide with oral prednisolone. After 1 month his symptoms were improved, blood pressure was controlled, liver enzymes became normal and proteinuria decreased.



Fig. 1 & 2: Malar rash

DISCUSSION

SLE is a clinically heterogeneous autoimmune disease whose etiology is unknown. This disease is characterized serologically by autoantibodies that target self-proteins. Different organs and cells of affected individuals undergo damage mediated by tissue-binding autoantibodies and immune complexes.¹¹ Presentation of SLE are similar in men and women, particularly skin rash, extreme fatigue, and joint pain. But, the findings suggest that the disease course is more complex in men, and some studies have shown that renal impairment, central nervous system, and vascular disease are more common in men than in women.¹²

Hepatomegaly is quite common among patients with SLE. The most common cause of elevated liver enzymes in SLE patients is drug-induced (Non-steroidal anti-inflammatory drugs). Coincidental liver disease, thrombotic events with or without

lupus anticoagulant including Budd-Chiari syndrome and veno-occlusive disease are other common causes.¹³ Though it is still a controversy, there is compelling evidence in the literature that SLE itself is not associated with a specific, severe and progressive liver injury. But, several authors have pointed a role for SLE in triggering an often subclinical hepatopathy, referred to as "lupus hepatitis". They also had described that this disease as an asymptomatic hypertransaminasemia and commonly associated with exacerbations of the lupus disease, which returns to normal range after corticosteroid therapy.¹⁴

CONCLUSION

This case emphasizes the importance of considering SLE in a male patient specially those presented with uncommon symptoms of SLE. Primary recognition and quick diagnosis are vital

for the timely initiation of treatment to prevent the severity of future complications. The findings highlight different systemic complications of SLE, necessitating the individualization of treatment based on specific manifestations. We concluded that liver is often a target of SLE, and biochemical liver tests should be systematically carried out in these patients in regular interval.

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