Case Report

Systemic lupus erythematosus in a male patient in lower Assam

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ABSTRACT

Systemic lupus erythematosus (SLE) is one of the numerous diseases called "the great imitators" because it frequently resembles or is misdiagnosed as other illnesses. SLE is a chronic autoimmune inflammatory disorder that affects multiple systems and has a wide range of symptoms. The female to male ratio varies between 7 and 15 to 1. A number of observations point to an estrogen impact as the cause of this variance. Because sex hormonal impacts are likely to be modest in youngsters, the female to male ratio is 3:1. Renal involvement and seizures are more common in male SLE patients than photophobia and cutaneous symptoms. Males likewise appear to have a more devastating consequence. As a result, we feel that male lupus patients are uncommon, the illness's symptoms are life-threatening, and that early discovery of the disease will result in a better outcome for these patients. Here, we report the case of SLE with an unusual presentation in a 44-year-old man from lower Assam to emphasize the importance of a high index of suspicion in such patients.

Key words: Autoimmune, Mucocutaneous manifestation, Systemic lupus erythematosus, Systemic lupus international collaborating clinics criteria

Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory illness that can present with a variety of symptoms and can also mimic other medical disorders, making it difficult to identify without delay in the majority of instances. The condition is more common in women between the ages of 15 and 44 with a prevalence of 90% [1]. SLE affects 15–50 people/100,000 in the United States with African Americans having the highest prevalence among ethnic groups studied. Ninety percent of the patients are women who are planning to start a family [2]. Males have a far lower prevalence of SLE than females, especially after puberty [3]. Gender may also play a role in how SLE manifests [4].

Here, we report the case of SLE with an unusual presentation in a 44-year-old man from lower Assam to emphasize the importance of a high index of suspicion in such patients.

CASE REPORT

A 44-year-old male patient presented with multiple oral ulcers and atypical multiple mucocutaneous rashes over the face for 2 months for which he took treatment in a local nearby hospital. The rashes got a flare-up on the local application of cream (name not known). For this reason, he came to our hospital.

On presentation, the patient was afebrile (temperature of 99 F, pulse rate of 110/min regular, respiratory rate of 21/min). Multiple mucocutaneous rashes were present over the face (Fig. 1).

Laboratory investigation reveals pancytopenia with hemoglobin of 6.9 gm/dl. Total leucocyte count was 1770 (N-60.4%, L-35.6%, M-2.3, E-1.7%, B-0%), mean corpuscular volume was 84 fl, mean corpuscular hemoglobin was 30.7 pg, and mean corpuscular hemoglobin concentration was 36.5. Peripheral blood smear for cell morphology was predominantly microcytic with mild to moderate hypochromia and white blood cell series showing leukopenia. Serum iron was 50 Ug/dl, serum total iron-binding capacity was 207 Ug/dl, and serum ferritin was 280. Blood urea was 73.1, serum creatinine was 1.6, serum Na^+ was 138, and serum K^+ was 3.4. Total bilirubin was 0.8, conjugated bilirubin was 0.0, unconjugated bilirubin was 0.3, and delta bilirubin was 0.5. Total protein was 6.6, Albumin was 2.5, Globulin was 4.0, aspartate aminotransferase was 314, alanine transaminase was 69, alkaline phosphatase was 87, and gamma-glutamyl transferase was 26. Antinuclear antibody (ANA) reflex showed ANA Hep-2 positive nuclear homogenous. U1-nRNP/Sm was weekly positive, Ro52 was positive, ds-DNA showed strong positivity, nucleosomes and histones were strongly positive, Rib-P protein was positive, and AMA-M2 was positive.

Based on the systemic lupus international collaborating clinics (SLICC) criteria, he was scored more than 10 and was diagnosed as a case of male SLE. The patient was treated with

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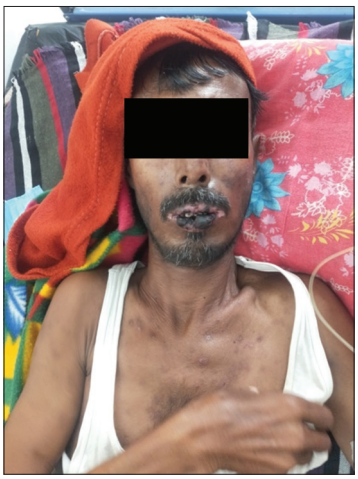

Figure 1: The patient with multiple mucocutaneous rashes

oral prednisolone, hydroxychloroquine, sulfasalazine, and cyclophosphamide pulse therapy. The patient was on regular follow-up and the symptoms improved after the completion of the course of drugs.

DISCUSSION

SLE is a chronic autoimmune inflammatory disorder with multifactorial etiology [5]. The presence of autoantibodies and specific clinical characteristics are used to make the diagnosis [2]. SLE affects more than 90% of women, with the majority of signs and symptoms being dermatological, musculoskeletal, and hematological in nature [6]. Males were more likely than females to have discoid lupus, thrombocytopenia, neuropsychiatric, and renal involvement at the time of diagnosis [7].

According to the SLICC criteria, the diagnosis of SLE requires fulfilment of at least four criteria, with at least one clinical criterion AND one immunologic criterion or lupus nephritis as the sole clinical criterion in the presence of ANA or anti-double-stranded deoxyribonucleic acid (dsDNA) antibodies. The clinical criteria of SLE include (a) acute cutaneous lupus, (b) chronic cutaneous lupus, (c) oral ulcers on the palate, (d) non-scarring alopecia (diffuse thinning or hair fragility with visible broken hairs), (e) synovitis involving two or more joints, characterized by swelling or effusion OR tenderness in two or more joints and thirty minutes or more of morning stiffness, (f) serositis, (g) renal, (h) neurologic, (i) hemolytic anemia, (j) leukopenia (<4000/mm^3 at least once), (h) thrombocytopenia (<100,000/mm^3) at least once. Immunologic criteria include (a) ANA above laboratory reference range, (b) anti-dsDNA above laboratory reference range, except ELISA: twice above laboratory, (c) anti-Sm, (d) antiphospholipid antibody: any of the following, (e) low complement, (f) and direct Coombs test in the absence of hemolytic anemia [8,9].

Our patient had extensive involvement of mucocutaneous rashes and based on the SLICC criteria, a diagnosis of SLE was made. In a study involving the population from upper Assam, mucocutaneous manifestation was 87.59% and other manifestations such as hematological (69.65%), renal (58.03%), and musculoskeletal (50.34%) manifestations were the common clinical complications, whereas, anti-dsDNA antibody was noted in 62.76% of the patients [8]. The patients with active SLE have fever, rash, fatigue, arthritis, serositis, central nervous system involvement, and renal involvement which is associated with high proteinuria. Symptoms related to active nephritis include peripheral edema associated with hypertension [9,10].

The patient was started on high-dose steroid and cyclophosphamide pulse therapy. Commonly used medications include corticosteroids, antimicrobials, NSAIDs, and immunosuppressants. Monoclonal antibodies are used widely [11,12]. This patient was treated with pulse steroid therapy-methyl prednisolone for 3 days and continued with oral prednisolone 40 mg/day which was tapered to 10 mg/day. Cyclophosphamide pulse therapy, hydroxychloroquine 200 mg twice daily, and sulfasalazine 1 g daily were also given and in due course, the patient improved.

CONCLUSION

The occurrence of SLE among males is rare and the initial presentation might vary with the presence of erythema multiforme which is even rarer. Hence, a high degree of suspicion is a clue for a physician to diagnose SLE in this situation. We like to report this case to make the physicians aware of the possibility of SLE in a case of erythema multiforme with systemic features.

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