Case Report

Male systemic lupus erythematosus: Two distinct presentations

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Abstract

Systemic lupus erythematosus (SLE) multisystem autoimmune disease is characterised by a series of exacerbations and remissions. Tissues of multiple-organ systems are affected by autoantibodies. We report here two cases of SLE in male patients. Being rarer in males, diagnosis is often delayed and associated with poorer outcomes. An alert primary care physician can be instrumental in early diagnosis and ensuring better outcomes.

Keywords: End-stage renal failure, fatigue, male

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease of unknown aetiology. There is multi-system involvement. Systems that can be potentially involved include renal, nervous, integumentary and the musculoskeletal system. The autoimmunity is believed to be due to certain susceptibility genes in the body. SLE occurs worldwide, with wide variations in disease prevalence across racial groups. It is most common in African American women, the prevalence being 4 per 1000 females.[1] SLE predominantly affects young women, most presenting between 20 and 40 years of age.

Male SLE is rare, with only one male patient reported per nine females.[2] The effect of androgens on T cells has been proposed as an explanation.[3] There is plethora of presenting features, affecting different systems. Eight of 10 patients have some form of skin or mucosal involvement during the course of illness. The most widely known of these is the facial malar rash. Subacute forms of the disease present with the "discoid rash".[4-6] Oral and nasopharyngeal ulcers, alopecia, and cutaneous bullae can be other features of SLE. Many patients with SLE may have subclinical renal involvement that can be demonstrated with the help of renal biopsy. Renal involvement should be detected early in the disease and if left untreated can lead to end-stage renal disease.

Case 1

A 24-year-old male patient presented with a history of severe joint pain, fatigue and fever since 4 months and had been taking NSAIDs without any relief. He had visited multiple doctors but without being diagnosed. By the time he visited this hospital he had developed a facial 'Butterfly Rash' which was reddish-purple, present over both cheeks with sparing of nasolabial folds. He complained of sensitivity to sunlight as well. Upon further questioning he revealed a history of aphthous ulcers [Figure 1], hairfall and myalgias. Initial laboratory evaluation revealed the following numbers: Hemoglobin-11.7 gm/dL, ESR-76 mm in 1 h, total leukocyte count- 11800/cumm, Platelet count -310,000/cumm. Urinalysis - mild albuminuria, 24 urinary protein: 702 mg. CRP: 21.99 mg/dL. ANA-positive (speckled and cytoplasmic),

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Anti-dsDNA- 346IU/mL. C3 and C4 levels were decreased. Radiology (chest X-ray, ultrasound abdomen) and ECG were normal. He received Deflazacort, Hydroxychloroquine, Methotrexate and Folic acid. Fever subsided within 3 weeks and the rash in over a month.

Case 2

A 21-year-old male patient presented with the history of fever, generalised body pains and frothing in urine. He later developed rashes over the face (typical of a butterfly rash) and body [as shown in Figures 2 and 3 respectively] when he presented to the hospital. Initial laboratory evaluation revealed the following numbers: Hb-12 gm/dL, ESR-30 mm in 1 hr, TLC- 9900/cumm, Platelet count: 280,000/cumm. Urine routine showed mild albuminuria, absent casts. Urine proteins were high. CRP was 1.3 mg/dL. ANA-13.4 IU/mL (speckled and cytoplasmic), Anti-dsDNA- 58.5 IU/mL. C3 and C4 levels were normal. P-ANCA was positive. Renal biopsy showed Minimal Mesangial Lupus Nephritis. He was prescribed prednisolone, hydroxychloroquine and methotrexate and over a period of 1 month the rashes over the face and generalised body pains resolved.

Figure 1: Aphthous Ulcers over the lower lip

Figure 2: Butterfly rash: Barely visible as patient was already undergoing treatment

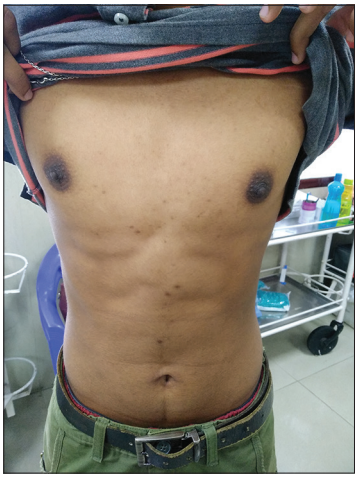

Figure 3: Scarring resulting from the discoid rash can be seen

Discussion

SLE is an autoimmune disease with multisystem involvement and varied clinical presentations.[7] Male SLE is associated with delayed diagnosis and poorer outcomes.[8] In our series, there were two patients, both young males.

The first patient presented with fatigue which is a frequent presenting complaint (seen in 4 of 5 patients presenting with SLE) and can be severe enough to impede day-to-day activities.[9-12] Because of improved detection of mild disease, the incidence of SLE has gone up by three times in the last four decades.[13] Men with lupus tend to have a similar frequency of occurrence of renal disease, skin disease, arthritis and central nervous system manifestations as women, lesser prevalence of photosensitivity and higher prevalence of serositis, older age at diagnosis and a higher 1-year mortality.[14,15] The largest cause of mortality in the early years of illness is active illness or infection due to immunosuppression.[15]

Both patients complained of an acute butterfly/malar rash. About half the patients mistake this rash with sunburn. The second patient had a discoid rash (red, scaly lesions with raised margins and atrophic centres) over chest, back and leg [as shown in Figure 3]. Discoid rash can cause scarring and disfiguration.[16,17] While a quarter of all patients with SLE have a discoid rash, only 5% of patients with DLE have SLE and tend to have low antibody titres.[18]

In the second case, the young boy presented with frothing of urine with other classical features of SLE. Urine routine showed proteinuria. The renal biopsy was suggestive of Minimal change Lupus Nephritis[19] Protienuria on urine routine is the most common and easily found sign of lupus nephritis.[20] The serum creatinine may be elevated, but this is not universal. Lupus Nephritis is usually the result of antigen-antibody complex-mediated glomerular injury.

Both cases are learning opportunities demonstrating that a high index of suspicion at the primary point of care will help prevent delay in diagnosis. In setups in remote areas of the country without a rheumatologist on staff, the primary care physician has an important role in suspecting and either diagnosing or referring such cases as needed. In cases presenting with complications, early diagnosis has a very important role in ensuring a better prognosis.

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Conflicts of interest

There are no conflicts of interest.

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