Egyptian Dermatology Online Journal
Volume 6 Number 1

# Systemic Lupus Erythematosus in Male Masquerading as Pyrexia of Unknown Origin

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**Egyptian Dermatology Online Journal 6 (1): 15**

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**Sumitted:** April 8, 2010
**Accepted:** April 30, 2010

## Summary

Systemic lupus erythematosus (SLE) is known to present with diverse clinical features. The disease has a predilection for females. Here, we report a case of SLE in a male patient masquerading as pyrexia of unknown origin and culminating in multi-organ failure. The case is being reported for its unusual presentation and rarity in male gender. High index of suspicion is required for early diagnosis of the disease so as to avoid delay in initiation of treatment and minimise mortality associated with the illness.

## Introduction

SLE like most other autoimmune diseases has a predilection for female sex. The disease is characterised by the development of auto-antibodies directed against various body proteins. The disease can have a vague presentation at least initially which may result in delay in the diagnosis and an adverse impact on the prognosis. Here, a case of SLE in a male is being reported for its unusual presentation which probably led to the delay in diagnosis and poor outcome of the disease [1,2].

## Case Report

Forty years old male, farmer by profession, presented with complaint of fever for the last 3 months. Fever was low grade, intermittent not associated with rigors or chills.

Moreover, there was history of easy fatigability, malaise and myalgias.

On examination, patient had an oral temperature of 101o F with non-inflammatory and non- scarring patchy alopecia of scalp **(fig. 1)**. The patient also had significant cervical and axillary lymphadenopathy. Lymph nodes were soft, discrete, non-tender and not adherent to the underlying tissue or skin. The rest of the systemic examination was unremarkable.

Investigations of the patient revealed normocytic, normochromic anemia with normal total leucocyte and platelet count. However he had high ESR (37mm) and transaminitis.

Other routine investigations including kidney function test, fasting blood glucose, serum electrolytes, spot urine examination, chest skiagram and USG abdomen were normal. Retro viral serology was nonreactive. Axillary lymph node excision biopsy revealed non-specific lymphadenitis. CT chest did not reveal any adenopathy. A workup for chronic infections especially common in this region like tuberculosis and brucellosis was unrewarding.

During hospital stay, patient developed malar rash with oral ulceration **(fig. 2-3)**.

This was followed by pancytopenia. At this stage ANA & anti-ds DNA were positive in high titres by immuno-florescence. Subsequently patient developed epistaxis followed by bleeding from lymph node biopsy site and melena. Serum creatinine and blood urea of the patient started to rise. There was further decline in platelet count to 1000/mm3. It is pertinent to mention here that he maintained normal sensorium till end thereby negating the diagnosis of thrombotic thrombocytopenic purpura. With aforementioned clinical and laboratory sequence of events, a diagnosis of systemic lupus erythematosis masquerading as pyrexia of unknown origin and culminating in multi-organ failure was contemplated as he was fulfilling requisite criteria for the same [3]. Patient was put on methyl prednisolone 1gm/day pulse therapy. However patient's condition continued to worsen despite requisite therapy. Finally patient developed hemodynamic compromise, multi-organ failure and succumbed to the disease.

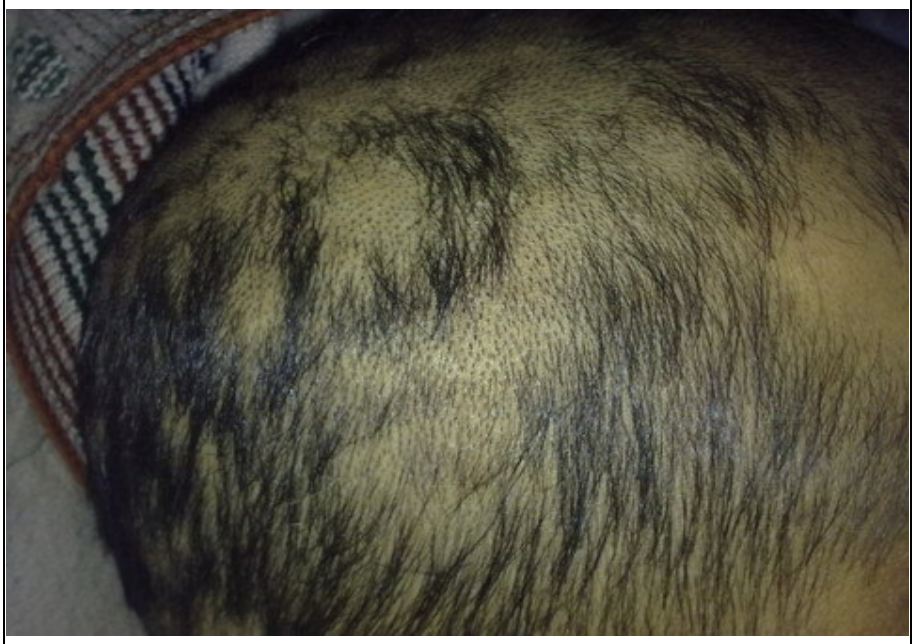

**Fig 1:** Non scarring alopecia of scalp.

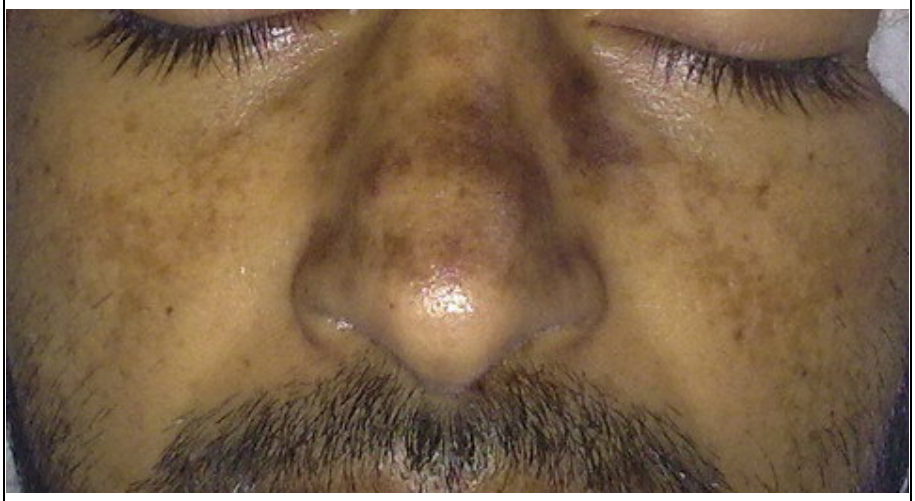

**Fig 2:** Malar rash.

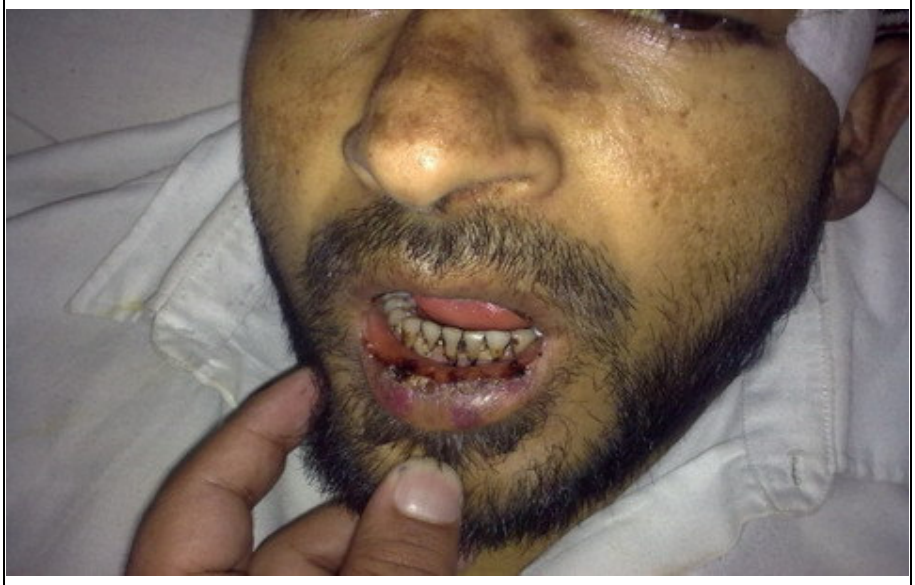

**Fig 3:** Oral ulceration.

## Discussion

Systemic lupus erythematosus (SLE) is a multi-organ system autoimmune disease with numerous immunological and clinical manifestations. It is characterized by an auto-antibody response to nuclear and cytoplasmic antigens. The disease mainly involves the skin, joints, kidneys, blood cells, and nervous system Systemic lupus erythematosus (SLE) has traditionally been considered a disease of women, and is uncommon in men. For all ages, the female to male ratio is 7:1 and 11:1 during the childbearing years [4]. As a rare auto-immune disease of males, lupus is commonly misdiagnosed or under-diagnosed leading to life impacting complications. The initial clinical course of systemic lupus erythematosus (SLE) is variable, ranging from relatively minor manifestations progressing over years to rapid onset of fulminate.

Several studies have tried to characterize lupus in men, in particular searching for any clinical differences between men and women. SLE in males tends to occur at an earlier age than female counterparts [5]. Male patients usually have a more severe disease, with higher morbidity, specially related to renal disease [6]. Renal disease occurs with an increased frequency and is usually more severe than females [6,7,8]. Moreover there is higher prevalence of serositis and thrombocytopenia [7,8,9]. However the prevalence of alopecia, arthralgia, Raynaud's phenomenon and psychosis are lower than female patients [8]. Among the neurological manifestations, males have higher frequency of seizures and peripheral neuropathy whereas females tend to have more psychiatric symptoms and headache [5].

Male patients of African- American race are more likely to have nephritis as their first clinical symptom and they usually progress from

initial clinical manifestations to SLE diagnosis more rapidly than other ethnic or gender groups [10].

It has been seen that male SLE patients from India have a higher incidence of mucocutaneous and renal involvement and a lower incidence of neuropsychiatric, gastrointestinal and hematological disease in comparison to those published from the developed countries. Moreover higher frequency of infection, particularly tuberculosis, was seen in male patients, which was the cause of death in some [5].

Potential causes of the female predilection for SLE included the effects of estrogen and its hydroxylation, decreased androgen levels, hyper-prolactinemia, and differences in

gonadotropin- releasing hormone (GnRH) [7]. In men and women with SLE and in their first-degree relatives estrone is preferentially hydroxylated at the C-16 position, resulting in the accumulation of 16-hydroxylated metabolites, which have sustained high estrogenic activity.

Men and women with SLE might have too much estrogenic and too little androgenic hormone, shifting their immune system toward increased responses. Prolactin levels are elevated in some individuals with SLE and may increase disease activity [11].

## Conclusion

SLE can have bizarre clinical presentation. High index of suspicion is needed to arrive at a proper diagnosis at the earliest, especially in male sex, in order to minimise mortality associated with the disease.

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