

Case Report

Male Adolescent with Systemic Lupus Erythematosus

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ABSTRACT

Systemic lupus erythematosus (SLE) is uncommon in children, especially in male adolescents (10-20 years of age). We present the case of an 11 year old boy in whom the diagnosis was missed in the first instance because of vague initial presentation and lack of suspicion. Raised antinuclear antibody

(ANA) and anti-double stranded DNA (anti-ds DNA) antibody titers that were checked after appearance of malar erythematous rash provided the diagnosis. The patient responded well to corticosteroid therapy.

KEY WORDS: antinuclear antibodies, male adolescent, systemic lupus erythematosus

INTRODUCTION

Systemic lupus erythematosus (SLE) is uncommon in male children and young adolescents (10-20 years of age). Its reported incidence and prevalence is shown in Table 1. Although the American College of Rheumatology (ACR) include definitive clinical features amongst diagnostic criteria for SLE, yet it is known to present with protean, albeit initially, atypical manifestations^[1].

We came across a young male adolescent, 11 years old, in whom the diagnosis was delayed because of the vague initial manifestation and lack of suspicion. It merits presentation here because of the paucity of its occurrence in male adolescents and to promote awareness of this condition in patients of this age group.

CASE REPORT

NH, a previously healthy 11-year-old boy, presented with two weeks, history of fever, generalized fatigue and ill-health. On physical examination he looked sick and was febrile (temp 39° C). He was normotensive (BP 110/60 mmHg), and his pulse rate was 110/minute. He did not have any arthralgias, myalgias or skin rash. Systemic examination was normal. No obvious focus of infection was found. Investigations results (Table 2) were as follows: CBC showed pancytopenia with WBC count of $1.9 \times 10^9/l$, (neutrophils $0.8 \times 10^9/l$), hemoglobin 89 g/l and platelet count $125 \times 10^9/l$. The erythrocyte sedimentation rate (ESR) was markedly elevated (117 mm/1st hour, Westergren). CRP was normal. Biochemical profile showed

raised hepatic enzymes (ALT = 150 IU/l and AST = 350 IU/l).

In view of persistent fever, deteriorating clinical condition and no identifiable focus of infection, the patient was treated empirically with IV cefotaxime and gentamycin, but without any noticeable benefit. A week later, the patient developed extensive erythematous rash on the malar area of face, limbs, palms and soles (Fig. 1). He also developed oral ulcers and bleeding from the buccal mucosa. Though he complained of generalized body aches, there were no objective signs of any bone or joint involvement. At this stage the possibility of SLE was considered and this was confirmed with findings of conspicuously raised ANA (2647.6 IU/ml) and raised double stranded DNA antibody (2783.3 IU/ml) titers. Additionally, complement levels were also significantly reduced (C3 0.24g/l, C4 < 0.1 g/l). Serum protein electrophoresis showed polyclonal rise in gammaglobulins. The patient was given pulse therapy with IV methylprednisolone, 20 mg/kg/day for three days. He also received IV Immunoglobulin, 400 mg/kg for five days. The response was dramatic. He soon became afebrile, and the erythematous rash disappeared; he stopped complaining of generalized aches and looked well. Subsequently the patient was started on methotrexate and oral glucocorticoid therapy (prednisolone 20 mg/day). Until the writing of this report (one year since the diagnosis) the child continues to remain in remarkable remission on a daily maintenance dose of prednisolone 10 mg on alternate days.

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Table 1: Incidence and prevalence of SLE in different ethnic groups^[1]

Ethnic group	Incidence Rate (per 100,000)	Prevalence Rate (per 100,000)
US adults	2-5	120
US children < 15 years	0.5-0.6	
US white females < 20years	4.4	
Oriental females < 20 years	31	
Black females < 20 years	19.8	

DISCUSSION

Systemic lupus erythematosus is a well known auto-immune disease that presents with protean multi-organ manifestations. The incidence of SLE varies worldwide. In the United States, incidence of this disease among children younger than 15 years of age is 0.5-0.6 per 100,000 per year, with a prevalence rate of 14-50 per 100,000; the rates being higher in females than in males^[1-4].

SLE is uncommon in children and young adolescents (10-20years of age). Its occurrence is particularly rare in male subjects. This combined with the fact that its clinical presentation, especially initially, can often be varied and vague, the diagnosis of SLE in male children is frequently missed in the first instance.

SLE can present with a wide variety of clinical features, reflecting multi-organ involvement. This is matched serologically by the presence of a wide spectrum of auto-antibodies. The clinical markers include malar erythematous rash, arthralgias, myalgias or renal involvement^[5]. SLE should also be considered when generalized fatigue occurs in combination with mucocutaneous manifestations which evolve over some time.

However, elevated levels of ANA and anti dsDNA antibodies remain the sheet anchor for its diagnosis. The case presented here is an example. In this instance, no suspicion was raised initially because the patient who is a male young adolescent presented only with fever and was treated empirically for infection in absence of evidence of any specific cause of fever. It was when he developed erythematous rash, particularly on malar area of face that suspicion of SLE was raised and specific investigations like ANA, anti dsDNA and serum complement levels were undertaken. These led to the diagnosis of SLE. The ACR has laid down definitive clinical and laboratory criteria for the diagnosis of SLE. The patient should have at least four out of the eleven criteria that occur in the course of this disease^[5-7].

The most useful screening tests that are recommended for diagnosis of SLE include complete blood count, erythrocyte sedimentation rate and testing for antinuclear antibody^[8-10].

Table 2: Results of relevant laboratory investigations at admission and after four weeks of treatment

	At admission	After 4 weeks of treatment	Reference value
Hb (g/l)	86	11	115-155
WBCs (x10 ⁹ /l)	1.6	7.5	4.5-13.5
Neutrophils(x10 ⁹ /l)	0.7	5.4	3-5.8
Lymphocyte(x10 ⁹ /l)	0.7	1.6	1.5-3
Platelets (x10 ⁹ /l)	125	574	150-400
ESR(mm 1st hour)	115	8	0-10
CRP(mg/l)	6	<3	<3
BUN (mml/l)	5.3	6.2	2.5-6.6
S.Creatinine (mmol/l)	43	29	60-120
ALT (IU/l)	108	51	10-60
AST (IU/l)	293	38	10-42
ANA	1:160	1:160	1:80
Anti dsDNA(IU/ml)	2647.6	425.57	<50
ENA(Ribosomal RNP)*	+ve		-ve
C3 (g/l)	0.24	0.88	0.55-1.2
C4 (g/l)	<0.10	0.13	0.20-0.5

ANA = antinuclear antibodies, Anti dsDNA = anti double-stranded DNA, ENA(Ribosomal RNP) = Extractable nuclear antigen(Ribosomal ribonucleoprotein), C3 = complement 3, C4 = complement 4

However, children differ from adults *vis-à-vis* the criteria of diagnosis of SLE. In the early course of this disease, the child may fulfill only three criteria. The fourth criterion may evolve later^[11]

With this case report we intend to promote the awareness among pediatricians that SLE occurs not only in girls, but also in young males. Male adolescents should also be investigated for SLE, even when they present with generalized weakness and fever for which no focus of infection or other obvious cause of fever can be located. Awareness and active suspicion can work towards early diagnosis so that appropriate treatment can be initiated and several serious complications can be prevented.

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