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Brief Report

Cutaneous Manifestations of Systemic Lupus Erythematosus in Iranian Children

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Abstract
Systemic lupus erythematous (SLE) is an autoimmune process in which cutaneous lesions occur in majority of patients. This study was conducted to determine the pattern and prevalence of such lesions in SLE in Iranian children infected with SLE. Forty-eight patients, age between 3-16 yrs and male to female ratio of 7/1 were examined for the presence of cutaneous manifestations of SLE. The most common skin changes noted were malar rash, discoid eruptions, alopecia, and photosensitivity. Vascular lesions including peripheral gangrene, chronic ulcers, Raynauds phenomenon, urticaria, palmar erythema, and erythema multiform were rare. Anti nuclear antibodies and anti-ds-DNA were positive. The prominent part of SLE is skin change which may be used as helpful diagnostic means. The result of the present study indicated that cutaneous involvements were more common in Iranian children and these changes would provide helpful information in early diagnosis of juvenile systemic lupus erythematous.
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Keywords ● SLE ● cutaneous ● Raynauds phenomenon ● discoid eruptions ● autoimmune

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Introduction

Systemic lupus erythematous (SLE) is a chronic multisystem disease caused by tissue damages resulting from deposition of antibody and complement–fixing immune complexes.[1-3] This disorder usually is life-long and a potentially fatal autoimmune disease.[2] There are wide spectrum of clinical presentations which are characterized by remission and exacerbations with serious complications. Age, sex, and clinical features, with special attention to cutaneous manifestations, were noted.[2]

In SLE there is a preference for the clinical involvement of the joints, skin, kidney, brain, and serosa.[3] Skin manifestations are the most common symptoms and signs of SLE.[2] Cutaneous lupus erythematous has been classified into specific classic and non specific manifestations.[3-5] In this study we have tried to study in detail the signs of cutaneous lesions seen in Iranian children with systemic lupus erythematous.

Patients and Methods

Forty-eight children under 16-yrs of age were analyzed retrospectively during a 10-yr period diagnosed with SLE at the Department of Rheumatology of Children Medical Center of Tehran University of Medical Sciences, Tehran, Iran. Detailed information of the expression clinical signs and symptoms such

as, skin involvement, renal, musculoskeletal, cardiovascular, pulmonary, and hematological abnormalities were reviewed.

As a routine procedure we used their laboratory reports including complete blood count, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP). In addition we used other laboratory reports such as urinalysis, 24-hr urine protein, BUN and creatinine. In order to consolidate our diagnosis, we also used serology tests including antinuclear antibody (ANA), anti-double-stranded DNA (Anti-ds-DNA), and serum complements (C3, C4, CH50), chest X-ray, ECG and echocardiography.

Results

The study group comprised 42 girls (87%) and six boys (13%). The initial specific cutaneous manifestations and the involvement of their organs, at the onset of clinical manifestations, are presented in Tables 1. Butterfly skin rash was seen in 75% of the patients and 50% of them had photosensitivity. Alopecia was present in 29% and involvement of organs was encountered in the majority of patients. The kidney was the most common organ affected at onset (83%). In 30 cases, arthritis arthralgia was manifested. Myalgia and myositis were seen in 41%. Cardiovascular system (CVS) involvement was seen in 29%, respiratory system was involved in 12.5% and oral ulcer was present in 42% of the patients. During the period of the study three patients had septicemia, two were in the end stage of renal failure and five patients died.

Table 1: Clinical manifestations of SLE

Skin Manifestations	No (%)
Cutaneous	42 (87)
Malar rash	36 (76)
Photosensitivity	24 (50)
Cutaneous vasculitis	19 (39)
Alopecia	14 (29)
Discoid LE	7 (14)
Other organ manifestations	
Renal	40 (83)
Arthritis /Arthralgia	30 (63)
Myalgia/Myositis	20 (41)
Cardiovascular	14 (29)
Pulmonary	6 (12)
Oral ulcers	20 (41)

Laboratory findings are summarizes in Table 2. Anemia (41%) was the most common hematological abnormality at the onset of the disease; leukopenia was present in 20% and thrombocytopenia in 25% of the patients. The majority of the patients had hematuria (88%) and proteinuria (83%). BUN was high (more than 40 mg) in 20 patients (40%) and creatinine was elevated to 1.2 mg% in 12 patients (26%). ANA positivity was more than 1:160 in 45 (95%) and positive anti-ds-DNA in 33 patients (68%). The level of C3, C4 and Ch50 complements were lower than normal in 30 (62%) patients.

Table 2: Laboratory findings of SLE patients

Parameters	No (%)
Hb < 10mg/dl	20 (40)
Leukopenia	10 (20)
Lymphopenia	8 (16)
Thrombocytopenia	12 (25)
Hematuria	42 (87)
Proteinuria	40 (85)
BUN > 40	20 (40)
Creatinine > 1.2	12 (26)
ANA +>1/160	45 (95)
Anti-ds-DNA + > 5	33 (68)
Low C3, C4, Ch50 complements	30 (62)
ESR> 50	40 (85)
CRP > +++	34 (70)

Discussion

The clinical manifestation of juvenile systemic lupus erythematous (JSLE) is similar to those of adults. Most patients have multisystem involvement. Although, the ratio of female/male SLE infection is lower before puberty than during puberty,[5] in general females of all ages are more prone to infection.[1,2] The occurrence of JSLE infection is rare before the age of six yr, but it usually appears in older children over nine-yrs old. The ratio before and after puberty are different. In our patients the incidence of JSLE was low before puberty, with its peak of incidence occurring in one patient of 15-yrs-old. This observation was similar to previous reports presented from other countries.[3]

The general clinical features of childhood SLE included broad variations between the presence of rash, arthritis, constitutional symptoms, renal disease, and the involvement of cardiovascular and pulmonary system. Cutaneous manifestations were common in our patients (88%) which were similar to that of previous childhood SLE studies.[6,7] All of our patients had skin involvements at some stage of the disease. Within those features, butterfly skin rash, photosensitivity, cutaneous vasculitis, and alopecia were the most frequent findings. The frequency of butterfly skin rash in our patients was higher than what was reported previously, possible due to the race. The race of Iranians is classified as Caucasians and reports that exist indicate that butterfly skin rash is seen more frequently in this race.[8,9] Although, photosensitivity was less common in our patients, but it occur in 50% of all patients.

Systemic lupus erythematosus is associated with renal involvement in the majority of affected children. Renal involvement, with the stage IV of WHO renal disease, was seen in 83% of our patients, which was similar to its incidence in

patients living in western countries.[5] As stated in the result section, in children the renal involvements appear to be high and very severe, the estimated prevalence raging from 60% to 80% of all patients. As well the nephritic range proteinuria is more frequent in pediatric patients compared to adults.

Other manifestations included musculoskeletal involvement which was stable during follow-up period in 62% our patients, which was less frequent than what was reported by Schaller et al,[3] Arthritis occurred in 62% of our patients, which was of non-deforming and self-limiting polyarthritis type. Muscle weakness, myalgia, and myositis were noted in 41% of our patients. Cardiac abnormalities were the major morbidity and mortality risk factor in childhood SLE. The incidence of cardiovascular and pulmonary involvements, noted in 29% of our patients which was similar to others reports.[10]

Antinuclear antibodies are the hallmark of SLE. Titration of anti-ds-DNA antibodies are said to be the most useful test in diagnosing SLE.[12,13] Anti-ds-DNA antibodies were positive in 68% of our patients. Comparison of the auto-antibody profiles of adult and childhood-onset SLE revealed a higher frequency of elevated anti-DNA antibodies in children. ANA was positive in 95% of the patients and in about 62% of them the level of complements was bellow normal similar to what was reported previously.[11-13]

Some authors have shown a higher mortality for children during the first year of diagnosis,[14] while others have shown an even distribution of mortality over the first five years of infection.[2] In this study during 10-yr period (1990-2000) three patients had septicemia; two patients were in the end stage of renal failure and five patients died. The prognosis in our patients reflects the improved care for children of juvenile SLE and the longer life expectancy of these patients.

The result of the present observation, therefore, is indicating that cutaneous involvement of lupus erythematous is more common in Iranian children. Skin changes may provide helpful diagnostic information in early diagnosis of juvenile systemic lupus erythematous.

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