Original article

Cutaneous manifestations of systemic lupus erythematosus in Pakistani children

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Abstract ***Background*** Systemic lupus erythematosus (SLE) is a multisystem disease of autoimmune etiology. Cutaneous changes are seen in more than two-third of patients.

Objectives The present study was planned to evaluate cutaneous manifestations of SLE in children.

Patients and methods Fifteen cases of SLE were collected from the Department of Pediatric Dermatology, Children Hospital, Lahore. The diagnosis was based on American Rheumatism Association criteria. Cutaneous changes were recorded on a predevised pro forma.

Results Age of onset was 5-13 years in 14 (93.3%) children. One case of neonatal LE was seen. There were 8 (53.3%) females and 7 (46.7%) males. Malar rash was present in 10 (66.6%), photosensitivity in 8 (53.3%), diffuse hair loss in 6 (40%), hyperpigmentation in 5 (33.3%), vascular lesions in 5 (33.3%), mucosal lesions in 3 (20%), nail changes in 2 (13.3%), bullous lesions in 1 (6.7%), livedo reticularis in 1 (6.7%), and rheumatoid nodules in 1 (6.7%). The single case of NLE had generalized scaly lesions.

Conclusion Cutaneous changes in children are different from those seen in adults. Female preponderance was not seen in children. Photosensitivity and vascular lesions were less frequent while the discoid rash was rare. Peripheral gangrene, chilblain and Raynaud's phenomenon were not seen. Neonatal LE was a rare entity.

Key words SLE, neonatal LE, cutaneous manifestations.

Introduction

Systemic lupus erythematosus (SLE) is a multisystem disease of autoimmune etiology. Integument is affected in more than two-third of cases.[1-3] Cutaneous changes constitute four of 11 criteria of American Rheumatism Association (ARA) for the diagnosis of SLE. The disease usually affects adult females.

Childhood SLE is a rare entity with an incidence of 10-20 in 100000 white Caucasian children with a male to female ratio of 1:4.5 between the ages of 6 and 18 years.[4] It is reported that 15-17% of all patients with SLE, the disease starts before 16 years. Rarely the babies born to mothers with lupus may develop LE called neonatal lupus erythematosus (NLE), due to passive transfer of maternal autoantibodies to fetus. Cutaneous changes of all types occur in 60-90% of children at any time during the course of illness whereas these are seen in 60-78% of cases at the time of diagnosis.[4] Hence, the recognition of cutaneous changes of SLE is very important in the early diagnosis of this potentially dangerous

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disease which, otherwise may be missed by an unwary physician.

Many local studies [5-7] address the subject in adult patients with SLE; however, scanty data are available on the topic in children of Pakistani origin. The present study was conducted to evaluate the cutaneous manifestations of SLE in Pakistani children.

Patients and methods

This descriptive study was conducted at the Department of Pediatric Dermatology, Institute of Child Health/Children Hospital, Lahore, from 1st January, 2003 to 31st December, 2003. Fifteen children of SLE (diagnosed on the basis of ARA criteria) were enrolled in the study. A detailed history and physical examination were recorded. The relevant hematological, biochemical and immunological profile, radiological work-up and skin biopsy were carried out. Cutaneous changes were further subdivided as specific (histopathological changes characteristic of LE) and non specific.

Results

During the one-year period, 15 patients were diagnosed, 14 children had SLE and one had neonatal LE. There were 8 female (53.3%) and 7 (46.7%) male children, with male to female of 1:1.1. Age of onset ranged from 5-13 years in 14 children with SLE. The frequency of LE-specific lesions and LE-nonspecific lesions is shown in **Table 1** and **2,** respectively.

LE-specific included malar rash (**Figure 1**), mucosal ulcers (**Figure** 2) and photosensitivity. Oral mucosa was affected in 3 and genital in 2 patients (**Table 3**).

Table 1 Frequency of different LE-specific lesions in 15 case of SLE.

Lesions	*n (%)*
Malar rash	10 (66.7)
Photosensitivity	8 (53.3)
Psoriasiform lesions of subacute LE	1 (6.7)

Table 2 Distribution of different LE-nonspecific lesions

Lesions	*n (%)*
Vascular lesions	
Telangiectasia	5 (33.3)
Microinfarcts	2 (13.3)
Purpura	2 (13.3)
Chronic ulcers	2 (13.3)
Bullous Erythema multiforme	1 (6.7)
Livedo reticularis	1 (6.7)
Pigmentary changes	
Hyperpigmentation	5 (33.3)
Hypopigmentation	1 (6.7)
Hair changes	
Diffuse alopecia	6 (40)
Lupus hair	1 (6.7)
Others	
Ragged cuticle	2 (13.3)
Ichthyosis	2 (13.3)
Sclerodactyly	1 (6.7)
Erythematous papules	1 (6.7)
Rheumatoid nodules	1 (6.7)

Table 3 Mucosal sites affected (n=15)

Mucosae affected	*n (%)*
Lips	3 (20)
Palate	3 (20)
Buccal mucosa	3 (20)
Genital mucosa	2 (13.3)

Chronic discoid lesions were not seen in any case.

The single case of NLE had generalized psoriasiform eruption (**Figure 3**). She was the first baby of a 24-year-old mother who was found to be anti-Ro positive. She had fever, skin rash, hepatosplenomegaly and cardiac disease. Histopathology from the back showed changes compatible with LE. The baby was also anti-Ro positive.

Amongst nonspecific lesions vascular manifestations were the most frequent

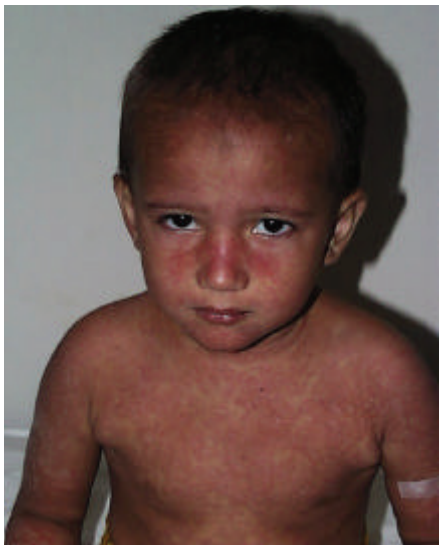

Figure 1 Malar rash, photosensitive generalized maculopapular eruption and non-cicatricial alopecia in a child with SLE.

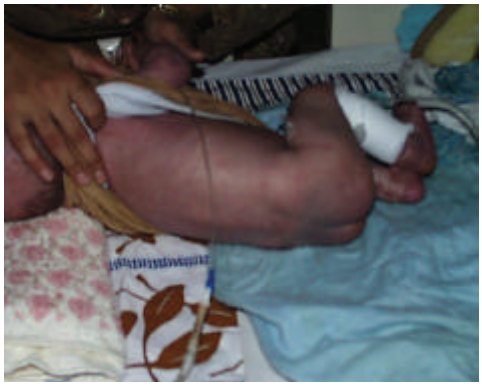

Figure 3 Generalized psoriasiform eruption in the female infant with neonatal LE.

(**Figure 4**). The frequency of other types of lesions is shown in **Table 2**. Peripheral gangrene, chronic ulcer, chilblain, and Raynaud's phenomenon were not seen.

Discussion

The present study affirms the high prevalence of cutaneous changes in SLE patients. However, the frequency was 100% in our series than previously reported[4] since all of our cases were collected from the dermatology out-patient clinic. Secondly, a diverse spectrum of cutaneous changes can be seen. We noticed an equal sex ratio whereas

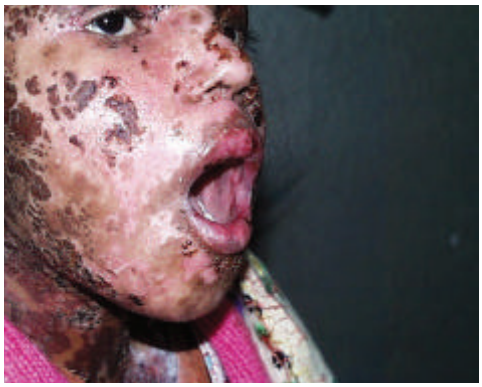

Figure 2 Ulcers over lips and buccal mucosa. Face and neck is als o affected.

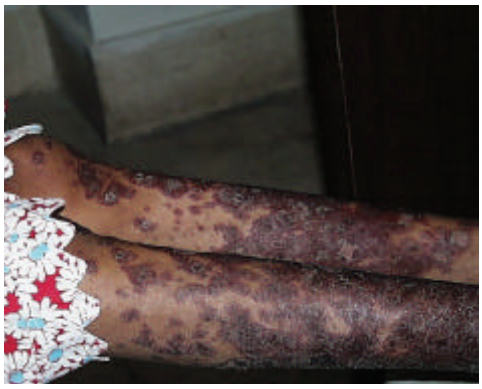

Figure 4 Bilateral purpuric eruption on the legs of a girl. Mild scaling is also evident.

previous reports give a female preponderance.[4]

Malar rash was the most frequent finding. This is in accordance with the previous reports.[4,8,9] Malar rash is seen in 30-80% of patients and it is present in 22-60% of cases at the time of diagnosis.[4] The incidence of malar rash increases with disease progression.[9] Photosensitivity was rare in our series. Similarly, none of our patients had discoid lesions. **Table 4** depicts the comparison of cutaneous changes in our patients with that by Font *et al*.[8] The difference between two can be due to different racial background.

Psoriasiform eruption seen in the single patient of NLE was in consonance with literature.[4] This signifies that a psoriasiform eruption in an infant in the

Table 4 Comparison with study in children.

Lesions	*Present study (2003)* (n=15)	Font *et al.* [4] Spain, 2004 (n=34)
Malar rash	66.7	44%
Photosensitivity	53.3%	23%
Oral ulcers	20%	9%
Psoriasiform lesions of SCLE	6.7%	-
Livedo reticularis	6.7%	3%

Table 5 Comparison with study in adults.

Lesions	*Present study (2003)* (n=15)	Rabbani *et al.* [5] Pakistan, 2003 (n=34)
Malar rash	66.7	31%
Photosensitivity	6.7%	33%
Vascular lesions	33.3%	20%
Mucosal lesions	20%	20%
Pigmentary changes	33.3%	22%
Discoid rash	-	15%

presence of systemic features should not be taken lightly.

Table 5 compares the cutaneous changes in children of our series with those in Pakistani adults. In comparison to adults, malar rash, photosensitivity and vascular lesions were more frequent whereas discoid lesions are seen more frequently.

Our results conclude that cutaneous changes in children are different from those seen in adults. Female preponderance is not seen in children. Photosensitivity is less frequent. Discoid rash is rare. Peripheral gangrene, chronic ulcer, chilblain, and Raynaud's phenomenon were not seen. Neonatal LE is a rare entity.

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