

■ **Original Article**

Spectrum of cutaneous manifestations in lupus erythematosus -a dermatologist perspective

S Bhattarai¹, S Agrawal¹, A Rijal¹, S K Sharma², S S Dhakal²

¹Department of Dermatology and Venereology, ²Department of Internal Medicine
B. P. Koirala Institute of Health Sciences, Dharan, Nepal

Abstract

Background: The cutaneous manifestations of lupus erythematosus (LE) specific skin disease consists of acute cutaneous LE (ACLE), Subacute cutaneous (SCLE) and Chronic cutaneous (CCLE). **Objective:** To evaluate the spectrum of cutaneous manifestation in patients of LE. **Methods:** A case series of 41 clinically diagnosed cases of LE attending the outpatient department of Dermatology, BPKIHS were evaluated for the specific and non-specific skin changes. **Results:** All the patients enrolled in the study were female, with the age ranging from 14-64 years. ACLE was detected in 22/41(78.04%). Malar rash was the frequent skin lesion. CCLE was seen in 6/41 (14.63%) patients with classical discoid lesions (localized and generalized) in 4/6(66.66%) and 2/6(33.33%) respectively. Non specific skin lesions were found in 30/41(73.17%) of patients. Mucosal ulcers were seen in 23/41(56.09%), Facial telangiectasias 20/41(48.78%), Raynauds phenomena 22/41(53.65%), Chronic urticaria 9/41(21.95%), Nail changes 12/41(29.26%) and non scarring alopecia was seen in 6/41(14.63%) patients. Eye involvement was seen in 3/41(7.3%), cutaneous vasculitis in 5/41(12.19%) and scarring alopecia in 3/41(7.3%) patients. **Conclusion:** The cutaneous manifestations of patients with lupus erythematosus (LE) are very frequent, show a great variety and can occur at any stage of the disease.

Keywords: cutaneous manifestations, lupus erythematosus, specific and non-specific skin changes.

Introduction

Systemic Lupus erythematosus (SLE) is a chronic, autoimmune, inflammatory disease characterized by a spectrum of clinical forms with variable evolution with skin being the second most commonly affected organ.

Skin involvement occurs in 70-85% of all patients with LE.¹ According to Gilliam and Sontheimer², the cutaneous manifestations of LE can be classified as LE specific or LE nonspecific. The specific skin lesions can be further divided into acute cutaneous LE (ACLE), Subacute cutaneous (SCLE) and Chronic cutaneous (CCLE).

The most common form of CCLE is discoid LE (DLE)

which can be separated into two groups; localized and generalized.^{3,4} Psoriasiform and annular-polycyclic form are the most common variants of SCLE⁵ Similarly the most common manifestation of ACLE is malar erythema.⁶

LE-nonspecific skin lesions are those that in some way are related to the underlying LE process but are not specific for LE since the same lesions can be encountered in other autoimmune diseases.⁶

The identification of nonspecific but disease-related skin lesions is very important in LE since their presence implies systemic involvement.⁷ The most common non-specific skin lesions in LE are vascular lesions and in particular leukocytoclastic vasculitis, thrombophlebitis and livedo reticularis. Nonscarring alopecia, pigmentary abnormalities, sclerodactylia, calcinosis cutis, rheumatoid nodules, papulo-nodular mucinosis and cutis

Address for correspondence:
Dr Sabina Bhattarai, Associate Professor
Department of Dermatology and Venereology
B. P. Koirala Institute of Health Sciences, Dharan, Nepal
Email: sabeenab@gmail.com

laxa/aneoderma are other nonspecific skin diseases rarely observed in LE.⁶

There is great variation in incidence, clinical heterogeneity and severity of disease between different ethnic and racial groups in SLE due to environmental, cultural and genetic variability.⁸ SLE being a relatively uncommon disease has not been studied much from the dermatologist perspective in Nepal and this has prompted us to undertake this study to evaluate the pattern and prevalence of specific and non specific cutaneous manifestations among patients attending the Dermatology OPD at BPKIHS.

Methods

Forty-one clinically diagnosed cases of LE attending the out patient department of Dermatology, BPKIHS between July 2005 - July 2009 were retrospectively evaluated for the specific and non-specific skin changes.

The patients were analyzed according to their age, gender, clinical features and thorough cutaneous examinations. Data were recorded in a preset proforma. Laboratory investigations included complete blood count, erythrocyte sedimentation rate, urine analysis, anti nuclear antibody, anti ds-DNA, Chest X ray and ultrasonography of the kidney and abdomen.

Data were tabulated and interpreted in terms of percentage, mean and standard deviation in the computer using SPSS version 10.0.

Results

Demographics

All the patients enrolled in the study were female and 24/41(58.53%) were of the Mongolian origin. The age of the patients ranged from 14-64 years (mean 31.85± 12.5years) and the duration of the disease from 1-144 months (mean 18.68± 27.41 months)

LE specific skin lesions

ACLE was detected in 22/41(78.04%) patients and was the most common cutaneous manifestation. Malar rash was the frequent skin lesion seen in 100% of the patients with ACLE. Wide spread maculo-papular lesion with malar erythema was seen in 15/41(36.58%) patients.

CLE was seen in 6/41 (14.63%) patients with classical discoid lesions (localized and generalized) in 4/6(66.66%) and 2/6(33.33%) respectively.

LE non specific skin lesions

Non specific skin lesions were found in 30/41(73.17%) patients. Photosensitivity was seen in 31/41(75.6%), facial telangiectasias in 20/41(48.78%), Raynauds phenomenon in 22/41(53.65%), chronic urticaria in 9/41(21.95%) and cutaneous vasculitis in 5/41(12.19%) patients respectively. Purpura in 6/41 (14.63%) patients and chronic ulcers were seen in 5/41(12.19%) patients.

Scarring alopecia was seen in 3/41(7.3%) patients and non scarring alopecia in 6/41(14.63%) patients each. Similarly Lupus hair was seen in 19/41 (46.34%) patients. Nail changes were seen in 12/41(29.26%), the most common change being hyperkeratosis and ragged cuticle in 11/41(26.82%) patients. Other changes included ridging in 5/41(12.19%), pitting in 4/41(9.7%) and splinter haemorrhages in 2/41(4.8%) patients respectively.

Mucosal ulcers on the lips, buccal mucosa and gums were seen in 23/41(56.09%) patients and eye involvement was seen in 3/41(7.3%) patients.

Systemic involvement

Other systems involved noted were constitutional symptoms in the form of fever, anorexia and malaise in 29/41 (70.73%) patients, polyarthritis in 33/41 (80.48%) patients, nephritis in 15/41 (36.58%) patients with active sediments in urine in 7/41(17.03%) patients, proteinuria in 4/51 (27.45%) patients, cardiopulmonary involvement in 7/41 17.03%) patients with 2/41 (4.87%) having pleural effusion, haematological disturbances in 21/41 (51.21%) patients and neuropsychiatric involvement in 9/41 (21.95%) patients.

Thirty nine (95.12%) patients were ANA-positive and anti ds-DNA was detected in 30/41 (73.17%) patients.

Discussion

LE is a chronic, autoimmune, inflammatory disease characterized by a multifactorial etiology and by a spectrum of cutaneous manifestations. The specific cutaneous lesions are represented by DLE, SLE and ACLE.

Women are affected twice as often as men according to data from the literature.³ Only females with SLE presented to the OPD in our study and 24/41(58.53%) were of the Mongolian origin. This finding supports that genetic factors other than HLA and complement component deficiencies may also be involved.⁹

The butterfly rash is the most common form of ACLE and coincides with the exacerbation of the systemic disease. It was seen in 100% of the patients in our studies as compared to 80% of patients as reported by Kole et al¹⁰, 46.5% by Cardinali¹¹, 60% by Kapadia et al¹² and 51% by Yell et al⁷ This could be explained by the acute presentation of the patient and their health seeking behaviour.

Typical DLE lesions may be present at the onset of SLE in about 6-10% of the patients while, during the course of SLE, about 25-33% of patients may develop DLE lesions, usually of the generalized type.¹¹ In our series DLE was present in 14.63% where the localized variant was present at the onset of SLE in 4/6 (66.66%) and the generalized type in 2/6 (33.33%) patients. The results vary from 12% -57.5% in different studies.⁷⁻¹²

SCLE is usually characterized by the appearance of papulo-squamous and/or annular-polycyclic lesions. In our study we observed no patients who presented with similar lesions.

The identification of nonspecific but disease-related skin lesion is important because their presence implies systemic disease and they are often useful indicators of systemic disease activity.⁷

Vascular lesions are very frequent in SLE and can involve both small dermal vessels and larger subcutaneous vessels with an incidence ranging from 11-33% (Table 1).

The involvement of the postcapillary venules of the surface dermis is expressed with two kinds of skin disorders: urticarial vasculitis and palpable purpura. We detected cutaneous vasculitis in 12.19% and purpura in 14.63% of our SLE patients.

Large subcutaneous vessel involvement may appear with subcutaneous nodules, chronic and recurrent ulcers, livedo reticularis and peripheral gangrene.¹³

Chronic ulcers were reported by Wysenbeek and coworkers¹⁴ in 8% of their SLE patients.

We noted the leg ulcers in 12.19% of our patients.

The occurrence of Raynaud's phenomenon has been reported in 2.5%¹² to 60%⁷ of patients with SLE. Our study showed the phenomena in 53.65%. This variation could be due to the climatic condition of the place as most patients had come from the hilly cold areas.

Urticaria and angioedema are common cutaneous vascular reaction patterns. Idiopathic urticaria may well be autoimmune¹⁶ and as such; it is not surprising that the rate in SLE is high. Our study showed 21.95% as compared to studies in which the prevalence varied between 6.67%¹⁰ - 44%⁷.

Different prevalence rates have been reported for oral ulcers in patients with SLE; in fact they have been reported in 18.4%¹⁵ to 60%.¹² Oral ulcers were seen in 56.09% in our study. This again could be attributed to the acute presentation of the patients.

Table 1: Cumulative incidence of muco-cutaneous manifestation with SLE

	Present study	Kole et al ¹⁰	Cardinali et al ¹¹	Kapadia et al ¹²	Yell et al ⁷
No. of Patient	41	150	58	40	73
Year of study	2009	2009	2000	1996	1996
Malar Rash	41(100%)	120 (80%)	27 (46.5%)	60%	37 (51%)
Discoid Lupus:					
Localized	4/6(66%)	30 (20%)	7 (12%)	57.5%	18 (25%)
Generalized	2/6(33.33%)	NA	12 (20.6%)	NA	NA
Photosensitivity	31 (75.6%)	75(50%)	NA	60%	46 (63%)
Oral Ulcer	23 (56.09) %	85 (56.67%)	3 (5.1%)	60%	23 (31.5%)
Non Scarring Alopecia	6 (14.63%)	130 (86.67%)	18 (31%)	82.5%	29 (40%)
Raynand's Phenomena	22 (53.65%)	10 (6.67%)	23 (39.6%)	2.5%	44 (60%)
Urticaria	9 (21.95%)	10 (6.67%)	NA	7.5%	32 (44%)
Cutaneous Vasculitis	5 (12.19%)	50 (33.34%)	8 (13.7%)	NA	8 (11%)
Nail changes	12 (29.26%)	2 (1.34%)	12 (20.6%)	22 (55%)	NA

NA: not available

Hair loss is a common and characteristic finding in SLE. It may be scarring, if preceded by DLE, or non-scarring. "Lupus hair" describes short irregular frontal hair, producing a dishevelled appearance, a result of

growth retardation, rather than hair breakage.¹⁷ Lupus hair was seen in 46.34%, scarring alopecia in 7.3% and non scarring alopecia in 14.63% respectively. The prevalence of Non scarring alopecia ranged from

31%¹¹ -86.6%¹⁰ in the other studies.

Nail changes were seen in 29.26%, the most common change being hyperkeratosis and ragged cuticle in 26.82% patients. Other changes included ridging in 12.19%, pitting in 9.7% and splinter haemorrhages in 4.8% patients respectively. It is interesting to see that periungual telangiectases was reported to be 20.6%¹¹ by Cardinali et al while Callen and coworkers demonstrated telangiectases in 76.4%¹⁸ while they were absent in our cases.

Other systemic involvement included constitutional symptoms in 70.73%, polyarthritis in 80.48%, nephritis in 36.58%, cardiopulmonary involvement in 17.03% with 4.87% having pleural effusion, haematological disturbances in 51.21% and neuropsychiatric involvement in 21.95% patients. We also report 4.8% of patients who presented with hypothyroidism accounting to the auto immune nature of the disease.

Conclusion

The cutaneous manifestations of patients with lupus erythematosus (LE) are very frequent, show a great variety and can occur at any stage of the disease and yield valuable diagnostic as well as prognostic information regarding the disease.

The ability to identify slight skin modifications at the onset or even during the remission of the disease and to differentiate from them LE-like cutaneous manifestations is probably a peculiarity of the dermatologist. So more interaction between rheumatologist and dermatologist will lead to more specific diagnosis of cutaneous lesions in patients with LE.

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