

Prevalence of usual and unusual skin manifestations of systemic lupus erythematosus in a tertiary care hospital

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Abstract *Objective* To determine prevalence of different types of skin manifestations in SLE in our country.

Methods The study was carried out at the Rheumatology and Immunology Department, Shaikh Zayed Hospital, Lahore. 125 patients who were diagnosed with SLE on basis of American College of Rheumatology classification criteria for SLE and also having skin manifestations were included from June 2011 till June 2015 from outdoor and inpatient departments.

Results SLE specific skin manifestations were as follows: malar rash was seen in 83 (71.6%) patients, photosensitivity in 61 (49.12%) patients, oral ulcers in 39 (31.35%), maculopapular rashes in 29 (23.46%), discoid rash in 26 (20.7%), subacute cutaneous lupus erythematosus (SCLE) in 6 (5.18%) patients. The SLE non-specific skin manifestations included: vasculitic rash in 50 (40.0%), non-scarring alopecia in 39 (30.9%), palmar erythema in 26 (21.1%), vasculitic ulcers in 7 (4.9%), livedo reticularis in 5 (3.8%), digital gangrene in 3 (2.7%), and Raynaud's phenomenon in 3 (2.2%) patients. Patients having SLE specific skin manifestations like malar and discoid rash were more likely to have internal organ involvement, while with those having SLE non-specific skin manifestations were seen with acute flare of disease. ANA and anti-dsDNA antibodies were positive in 92% and 63% patients, respectively.

Conclusion Skin manifestations in SLE are important disease parameters which can give important diagnostic and disease activity information. Thorough focus on dermatological aspect is very important for diagnosis and treatment in this entity.

Key words

Skin manifestations, SLE-specific, SLE non-specific, prevalence.

Introduction

The discovery of lupus erythematosus cell by Hargraves in 1940s initiated many studies of SLE around the globe.¹ SLE is a chronic multisystem disease due to deposition of antigen-antibody complexes in various tissues causing a wide variety of organ specific symptomatology.²⁻⁴ This disorder usually is

chronic with relapsing remitting nature and a threat for fatal outcomes.³ Skin-related manifestations of SLE gives important information about diagnosis and disease activity. There is variation in prevalence, incidence, clinical diversity and disease severity between different regional, racial and ethnic groups. Environmental or genetic background may give clues to explaining and better understanding of these diversities.⁵ In SLE, other systemic involvement is of musculoskeletal system, renal system, central nervous system, cardiovascular system, pulmonary system and serosa.⁴ The skin and mucous membranes are

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symptomatically involved in over 75% of patients.⁶ Cutaneous lupus erythematosus has been divided into SLE specific and SLE non-specific manifestations.^{7,8} There is marked variability in the type ranging from malar rash and discoid lupus to bullae formation, alopecia and vasculitic rashes.⁹ Skin manifestations have a key role in classification of the disease as 4 out of 11 criteria in American College of Rheumatology (ACR) classification criteria are cutaneous.¹⁰ The main purpose of this study was to identify the prevalence and clinical importance of different skin manifestations in SLE patients in a tertiary care hospital.

Methods

The study was carried out at Rheumatology and Immunology department, Shaikh Zayed Hospital, Lahore. Patients were seen and selected in outdoor and inpatient departments for skin manifestations in SLE between June 2011 and June 2015. All these patients fulfilled the classification criteria of ACR for SLE.¹⁰ History and examination including symptoms and clinical signs comprising of skin, musculoskeletal, renal, cardiopulmonary and hematological abnormalities were documented. Laboratory tests were also carried out including ESR, complete blood counts, renal function tests, ANA, anti-dsDNA antibodies, 24 hours urinary protein and serum complement levels (C3 and C4).

Using SPSS software, the patients were analyzed according to their age, sex, and clinical features with focus on skin manifestations.

Results

Of the 125 patients of SLE, 113 (90.7%) were females and 12 (9.3%) were male. The male to female ratio was 1:9.7. Mean age at presentation was 31±11.2 years. Precipitating factors included endogenous (59%), poor drug compliance (22%), infections (15%) and pregnancy (4%). Anemia was present in 54 (43.2%) patients, thrombocytopenia in 34 (27.5%), leukopenia in 23 (18.4%) and proteinuria in 101 (80.5%). ANA was positive in 115 (92%) and anti ds-DNA antibodies in 90 (60%) patients. The level of C3 and C4 complements were low in 80 (64.3%) patients (**Table 1**).

Table 2 enlists the cutaneous manifestations in the study population. SLE-specific skin lesions were noticed in 101 (80.5%) patients. Malar rash was the most frequent finding seen in 82 (71.6%) patients, followed by photosensitivity in 61 (49.1%) patients. Oral ulcers occurred in 39 (31.4%) patients and it included superficial erosions, discoid lesions and erythema on the palate, buccal mucosa and gums. Less common changes included maculopapular rashes seen in 29 (23.5%) and discoid rash in 26 (22.7%) patients. SLE-nonspecific manifestations comprised vasculitic rash in 50 (40.0%) patients, scarring alopecia in 39 (30.9%), palmar erythema in 26 (21.0%), vasculitic ulcers in 7 (4.9%), livedo reticularis in 5 (3.8%), digital gangrene in 3 (2.4%), Raynaud's phenomenon in 3 (2.4%) patients. None of the patients had erythromelalgia, sclerodermatous changes, angioedema, bullous lesions, dermal mucinosis, nail changes, lichen planus, or pyoderma gangrenosum. Hyperpigmentation occurred in 40 (32%) of patients.

In this study, patients with SLE non-specific skin manifestations, especially maculopapular rashes, vasculitic rashes, and scarring alopecia were associated with more active disease in lieu

Table 1 Laboratory findings in 125 SLE patients.

Laboratory test	N (%)
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Hb <10mg/dL	54 (43.2)
Leucopenia	23 (18.4)
Thrombocytopenia	34 (27.2)
ANA	115 (92)
Anti-dsDNA antibodies	90 (72)
Low C3 and C4	80 (64)
Proteinuria	101 (80.8)

Table 2 Skin manifestations in 125 SLE patients.

Skin manifestations	N (%)
<i>SLE-specific skin manifestation</i>	
Malar rash	83 (66.4)
Photosensitivity	61 (48.8)
Maculopapular rashes	23 (18.4)
Oral ulcers	39 (31.2)
Discoid rash	26 (20.8)
Subacute cutaneous LE	6 (4.8)
<i>SLE-nonspecific manifestations</i>	
Vasculitic rash	50 (40)
Scarring alopecia	39 (31.2)
Palmar erythema	26 (20.8)
Vasculitic ulcers	7 (5.6)
Livedo reticularis	5 (4)
Digital gangrene	3 (2.4)
Raynaud's phenomenon	3 (2.4)

Table 3 Internal organ involvement in SLE patients (n=125).

Organ affected	No. (%)
Arthritis	105 (84)
Renal	79 (63.2)
Myalgia	34 (27.2)
Cardiovascular	36 (28.8)
Pulmonary	15 (12)
Lymph node enlargement	8 (6.4)

of clinical history and lab investigations and malar rash was associated with internal organ involvement.

Systemic involvement was also encountered in majority. Musculoskeletal system was the most common system affected in 105 (84.1%) patients. Renal problem was seen in 79 (63.3%), cardiovascular system in 36 (28.8%), myalgias in 34 (27.6%), pulmonary in 15 (12.3%), and lymphadenopathy in 8 (6.3%) patients, (**Table 3**).

Discussion

Skin manifestations occurring in this study matched with study by Hochberger *et al.*¹¹ and Koleet *al.*¹² Initial presentation of SLE as skin manifestations was comparable to that reported by Watson.¹³ The preponderance of female is comparable with other populations e.g. 28 out of 32 in India¹⁴ and 73 out of 78 in Australia.¹⁵ Mean age at onset was 31 years which was lower than observed in literature.^{16,17}

Among the SLE-specific skin manifestations, malar rash was most common which was 71.6%, higher than three studies done in Middle East countries.^{18,19,20} Discoid rash was noted in 25.9%, lower than a study in Pakistan (57%),²¹ but again higher in Middle East countries.^{18,19,20} Photosensitivity was reported in 49% which was comparable in prevalence with Middle East,²⁰ but lower than in Pakistan.²¹

Oral ulcers were reported in 31.4% which was lower than that reported in Pakistan²¹ and Middle East countries.^{18,19,20} Different percentage of prevalence of oral ulcers were also seen by Kole and Ghosh,¹² Dubois and Tuffanelli²² and Malaviya *et al.*²³ Maculopapular rashes and SCLE was noted in 23.46% and 5.18% of the cases respectively which were similar to Kole and Ghosh¹², but lower than Wysenbeek *et al.*²⁴ who reported these lesions in higher percentages. Raynaud's phenomenon was a less common skin manifestation in SLE i.e. 2.2%, while higher figures were seen by Kole and Ghosh,¹² Malaviya *et al.*²³ and Vaidya and Samant.²⁵ This variation may be attributable to diverse environmental conditions. Urticaria-like skin manifestations were not present in our study and it is rare in SLE patients.²⁶ Though Dubois²⁷ recommended that urticaria in SLE should be carefully evaluated as a clue for systemic disease flare. Erythema multiforme, sclerodermatous changes, and lichen planus were not seen in this

study. Hyperpigmentation was noted in 29 (23.2%) patients while it was lower in a study by Tuffanelli and Dubois(8.4%).¹⁷This difference may be due to excessive exposure to sunlight in our country. Scarring alopecia was 30.9% which was less frequent as compared to 86.7% by Kole and Ghosh,¹² 57% by Wysenbeeket *al.*²⁴ and 37% by Akhtar and Khan.¹⁶

The incidence of ANA-negative SLE was similar (8%) as compared to 4-13% reported previously.³¹Anti dsDNA antibodies were elevated in 63% of our patients similar to literature.³²ANA was positive in 92% similar to what was reported previously.^{33,34}

Conclusion

Skin manifestations in SLE are significant clues in diagnosis as observed by the fact that they account for 4 out of 11 criteria by American College of Rheumatology classification of SLE. Skin manifestations can give valuable information regarding diagnosis (SLE-specific), as well as, disease activity (SLE-nonspecific skin manifestations are associated with disease activity). Skin manifestations are responsible for increased morbidity too. Proper understanding of pathogenesis of skin manifestations will be further helpful for diagnosis and treatment of SLE in future.

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