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Original Article

Cutaneous manifestations of systemic lupus erythematosus – An experience from Bahawal-Victoria Hospital, Bahawalpur

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Abstract

Objective To document the frequency of cutaneous manifestation of systemic lupus erythematosus (SLE).

Methodos It was a case-series collected from Dermatology Department and all four Medical Units of Bahawal-Victoria Teaching Hospital of Quaid-e-Azam Medical, Bahawalpur. A total of 100 patients with diagnosis of SLE and fulfilling the inclusion criteria were included in the study. All the patients were evaluated for the cutaneous manifestations of SLE. All the information was collected in a specially designed proforma and analyzed with the help of SPSS version 10.

Results The mean age of the patients was 25.97±4.64 years. 92% patients were female and 8% were male. Out of 100 patients, 85% patient presented with cutaneous manifestations. Photosensitivity was seen in 40 (40%) patients, discoid rash in 35 (35%), malar rash in 25 (25%), and oral ulcers in 24 (24%).

Conclusion SLE is predominantly seen among female patients of young age. Photosensitivity is most common presentation, followed by discoid rash.

Key words
Systemic lupus erythematosus, cutaneous manifestations, malar rash, photosensitivity.

Introduction

Systemic lupus erythematosus (SLE) is referred to a chronic inflammatory connective tissue disorder that can involve multiple organs of the body including skin, mucous membranes, joints, kidneys and blood vessels. American Rheumatology Association (ARA) has developed criteria known as 'ARA criteria of SLE' based on the specific findings of patients including skin lesions: malar rash, discoid rash, photosensitivity, oral ulcers, non-erosive arthritis, serositis (pleurisy or pericarditis), renal disorder (persistent proteinuria or cellular casts), neurological disorder (seizures, psychosis), hematological disorder (hemolytic anemia, leucopenia or thrombocytopenia), lupus erythematosus cells (LE cells) or anti-ds DNA antibody or anti-Sm antibody or false-positive serology for syphilis and antinuclear antibody may be used as an aid to diagnosis. A patient is said to have SLE if 4 or more of 11 criteria are present, serially or simultaneously during any interval of observation.[1]

There is a great variation in prevalence of SLE according to geographical and racial background. Among the Caucasian population, SLE affects 1 in 2,000 individuals but is more common in other ethnic groups, such as Afro-

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Americans and Asians.[2] The disease is more commonly seen among female population. It is about 9 times as common in women as in men, with a peak age of onset between 20-40 years.[3] In England, the prevalence of SLE is 200 cases per 100,000 women (18 to 65 years of age). Overall U.S. prevalence of systemic lupus erythematosus is 40 cases per 100,000 persons.[4] The disease course is milder and survival rate higher among persons with isolated skin and musculoskeletal involvement than in those with renal and CNS disease.[5]

Etiology of SLE is still not fully known but there are several predisposing factors. Multiple genes confer susceptibility to disease development. Interaction of sex hormones, hypothalamic-pituitary-adrenal axis, and defective immune regulation, such as clearance of apoptotic cells and immune complexes, modify its susceptibility. The loss of immune tolerance, increased antigenic load, excess T cell help, defective B cell suppression, and shifting of Th1 to Th2 immune responses lead to cytokine imbalance, B cell hyperactivity, and the production of pathogenic autoantibodies. Finally, certain environmental factors are probably needed to precipitate the onset of the disease.[6]

Skin holds a vital position in the diagnosis of this systemic disorder and acts as important marker of SLE. Skin changes are easily picked up in clinical settings and hence are helpful in establishing the diagnosis. Cutaneous changes constitute 4 of 11 criteria of American Rheumatism Association (ARA) for diagnosis of SLE.[7]

Cutaneous changes of SLE are divided into two categories: LE-specific (acute, subacute and chronic) and LE-nonspecific, e.g. photosensitivity, Raynaud's phenomenon, vasculitis, hair change and others.[7] These patterns of cutaneous involvement are not only responsible for morbidity and disfigurement, which is caused by scarring, dyspigmentation, etc., but also may serve as parameter of underlying systemic activity. Acute cutaneous LE (lupus-specific) has a strong association with systemic disease and non-specific skin lesions always indicate disease activity for which patients present to rheumatologists and internists.[8]

There have been many studies conducted to look into the frequency of cutaneous lesions in SLE. One of the studies was conducted in Pakistan on 100 patients by Rabbani *et al*.[9] to evaluate the frequency of cutaneous manifestations in SLE. SLE specific lesions noted in this study were; malar rash (31%), discoid rash (15%), photosensitivity (33%) and maculopapular rash (20%). In another study, Parveen *et al*.[10] have reported the frequency of skin lesions as: malar rash (70%), discoid rash (10%) and maculopapular rash (19 %). They also concluded that pattern and skin manifestation of SLE may vary from place to place.

As there are only few studies conducted in Pakistan which have documented the frequency of cutaneous manifestations of SLE, so, this study was planned to evaluate the frequency of skin manifestations of SLE, in southern areas of Province of Punjab, the catchment area of Bahawal-Victoria Hospital/ Quaid-e-Azam Medical College, Bahawalpur.

Methods

This case series was collected from Department of Dermatology and all four Medical Units of Bahawal-Victoria Hospital, Bahawalpur.

The calculated sample size was 100 cases with 7% margin of error, 95% confidence level taking expected percentage of discoid rash in patients of SLE i.e. 15%. Non probability purposive sampling technique was used. Patients of either sex, age ≥18 years who fulfilled the ARA criteria were enrolled in the study whereas patients with drug-induced SLE (history of drug intake and appearance of skin lesions after receiving medication) were excluded.

Patients admitted through out-patient and emergency departments, in the Skin Ward and four Medical Units of Bahawal-Victoria Hospital, who fulfilled the inclusion criteria, were included in the study. After taking informed consent from the patient, all concerned information was collected on a pre-designed proforma. General data including age and sex etc. were recorded. All the patients were examined regarding cutaneous manifestations of SLE i.e. malar rash, discoid rash, oral ulcers, photosensitivity etc. All the information was recorded in designed proforma.

Data analysis All the collected data were entered into SPSS version 10 and analyzed. The qualitative data like demographics and cutaneous manifestations of SLE i.e. malar rash (yes/no), discoid rash (yes/no), oral ulcers (yes/no) and photosensitivity (yes/no) were presented as frequency distribution. Quantitative data in the study, like age (in years) were presented as means and standard deviations. The main outcome variable was cutaneous manifestation of SLE i.e. malar rash, discoid rash, oral ulcers and photosensitivity. These were presented as frequency distribution.

Results

Regarding age distribution of these 100 patients, included in this prospective case series, mean age of 25.97±4.64, (range 18-36) years was observed (**Table 1**). The majority (83%) of patients were below 30 years of age. Male (n=10) to female (n=90) ratio was 1:9.

Table 1 Distribution of patients by age (n=100).

	N &%
Age (years)	
18-30	83
31-40	17
41-50	0
51-60	0
> 60	0
Sex	
Male	10 (10)
Female	90 (90)

Among the study population, there were 85 (85%) patients who presented with cutaneous manifestations of SLE while the remaining 15 (15%) patients did not show cutaneous manifestations at the time of diagnosis. Photosensitivity was seen in 40 (40%) patients, discoid rash in 35 (35%), malar rash in 25 (25%) oral ulcers in 24 (24%) patients, as shown in **Figure 1**.

Discussion

This prospective case series comprised of 100 patients who fulfilled the criteria of SLE according to ARA criteria. Cutaneous manifestations of SLE were seen in 85% in our patients. Of these, photosensitivity was seen in 40%, discoid rash in 35%, malar rash in 25% and oral ulcers in 24% of total patients.

Many studies have been conducted to find out the patterns and frequency of cutaneous manifestations of SLE. Almost every study has shown different results.

Parveen *et al*.[10] performed a study on 100 patients to find out the frequency and pattern of SLE. The mean age observed in this study was

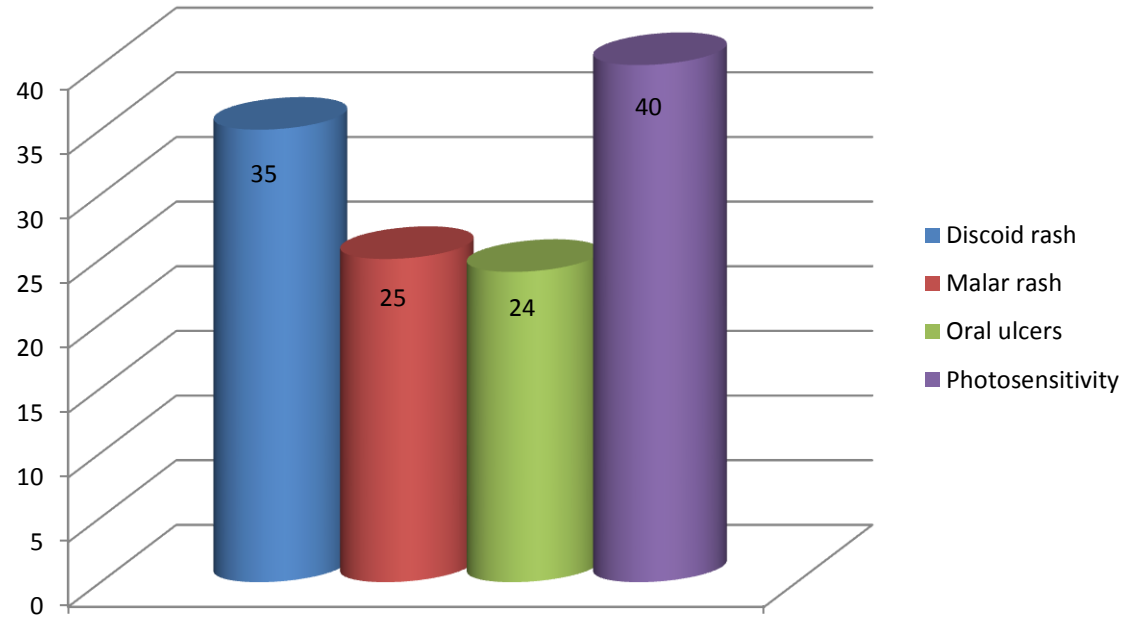

Figure 3 Distribution of patients by cutaneous features of SLE (n=100).

30.04 years and male to female ratio 1:5. In our study, the mean age of the patients was 25.97 years, and male to female ratio 1:9. These figures reflect the female preponderance and the prevalence of this disease in younger age population.

The female preponderance is also confirmed by other studies across the globe. George *et al.*[11] also described SLE in 28 females out of 32 patients in an Indian study. Weinsten *et al.*[12] showed male to female ration as 1:14.5%. In a study by Rabbani *et al.*[13] SLE was seen in 86.9% females while male population consisted of only 13.1%.

The mean age of the patients in our study was 25.97 years. Although the literature varies a lot in respect to age of the patients at presentation, most of the patients with SLE are younger. This has also been confirmed by Akhtar and Khan[14]; the mean age of patients in their study was 30 years. The average age of the patients described by Weinstein *et al.*[12] was 38 years. The mean age at presentation described by Rabbani *et al.* was 31 years. In our study, the mean age of the patients is lesser than the other studies however it is consistent with other studies in keeping the view that SLE is frequent in younger age group.

In our study, the cutaneous lesions were observed among 85% of the patient population. While it was approximately 71% in study by Parveen *et al.*[10] which further explained that 15 (15.0%) patients had only cutaneous lesions, 56 (56.0%) patients had both cutaneous and systemic lesions and 29 (29.0%) had only systemic lesions.

In our study, the frequency of malar rash was 25%, while in study by Rabbani *et al.*[13] the malar rash was present in 29% of patients. In another study by Rabbani *et al.*[9] the frequency of malar rash was 30%. Other studies have described the frequency of malar rash as 60% by Edward,[16] 56% by Mok and Lau[17] and 58.9% by Ward and

Studenski.[15] The highest frequency of malar rash was seen in a study by Parveen *et al*.[13] i.e. 70%. The results of our study are in consistent with other local studies like by Rabbani *et al*.[9,13] However, it has been shown in a higher frequency in other international studies.

The frequency of discoid rash in our study was 35%, while in other studies, it was observed 14% by Rabbani *et al.*,[13] 12% by Mok and Lau,[17] 10% by Edward,[16] 10% by Parveen *et al.*[10] and 7% by Ward and Studenski.[15] The above discussion shows that the frequency of discoid rash in SLE patients was from 7% to 35%, the highest being in our study.

Photosensitivity was seen in 40% patients in our study. While it was seen only in 6% patients in a study by Rabbani *et al*.[9] The frequency of photosensitivity documented by Edward[16] was 31%, by Mok and Lau[17] was 35% and by Ward and Studenski[15] was 48%. These results were also very close to the other studies across the world. However, the other local study[14] has documented a very low frequency i.e. 6% as compared to 40% in our study.

In our study, the oral ulcers were seen in 24% patients. When compared to other local study by Rabbani *et al.* which confirmed its presence in 20% patients population.[13] Oral ulcers were seen among 20% patients by Edward[16] and 11% and 55% by Mock[17] and Ward and Studenski,[15] respectively.

The above discussion suggests that cutaneous manifestations have shown great diversity across different parts of globe. In our study the frequency of cutaneous manifestation in SLE was higher (85%) than other studies. This can be attributed towards more sun exposure in the desert area, cosmetic disfigurement and patients' trend to report more frequently to the dermatology out-patient department as compared to medicine department for cutaneous lesion. The frequency of cutaneous manifestations may be higher than mentioned in our study as most of the patients in our set up do not present to tertiary care units due to negligence, poverty and lack of facilities. This may also be due to delayed referral by quacks.

Conclusion

SLE is more frequently seen among younger age group of female patients. More than two third of the patients with diagnosis of SLE present with cutaneous manifestations, the most common of which is photosensitivity followed by discoid rash. However, there is a need for large, multicentre, randomized trials to know the more precise figures in our population.

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