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Original Research Article

A study of cutaneous manifestations of systemic lupus erythematosus in Malwa region of India

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ABSTRACT

Background: Systemic lupus erythematosus (SLE) is an autoimmune disease. SLE is the most common connective tissue disease. Its prevalence varies according to geographical and racial background from 3/10 000 in Caucasians to 20/10 000 in Afro-Caribbeans. Around 90% of affected individuals are women, and the peak age at onset is between 20 and 30 years. Limited studies are available in India 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 a tertiary care centre of Malwa region of India.
Methods: All demographic data including age, sex, weight was collected on a pre-designed proforma. All the patients were evaluated for cutaneous manifestations of SLE i.e. malar rash, discoid rash, oral ulcers, photosensitivity etc. All the information was recorded in designed proforma.
Results: Among 100 SLE subjects, 82 (82%) patients had skin manifestations, 18 (18%) patients had oral ulcer and 12 (12%) patients had raynaud's phenomenon at the time of presentation. Among dermatological manifestations, 42% patients had photosensivity, 35% patients had malar rash, 30% patients had discoid rash, 10% patients had alopecia and 2% patients had bullous lesion.
Conclusions: Skin lesions in patients with lupus may be specific or nonspecific. This study covers the SLE-specific cutaneous changes: malar rash, discoid rash, photosensitivity, and oral mucosal lesions as well as SLE nonspecific skin manifestations. A deeper thorough understanding of the cutaneous manifestations of SLE is essential for diagnosis, prognosis, and efficient management. Thus, dermatologists should be involved with other specialties to provide optimal care of SLE patient.

Keywords: Lupus, Skin, Photosensivity

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease. SLE is the most common connective tissue disease. Its prevalence varies according to geographical and racial background from 3/10 000 in Caucasians to 20/10 000 in Afro-Caribbeans. Around 90% of affected individuals are women, and the peak age at onset is between 20 and 30 years.[1] The cause of SLE is incompletely understood but genetic factors play an important role. There is a higher concordance in monozygotic twins and associations with multiple polymorphisms in the HLA locus on chromosome 6 have been identified. In a few instances, SLE is associated with inherited mutations in complement components C1q, C2, and C4; in the immunoglobulin receptor FcγRIIIb or in the DNA exonuclease TREX1. Recent studies have identified common polymorphisms that

predispose to SLE, including the ITGAM gene which encodes an integrin; the IRF5 and STAT4 genes which are involved in interferon signalling; and the BLK gene which is involved in B-cell signalling. From an immunological standpoint, the characteristic feature of SLE is the production of autoantibodies. These have specificity for a wide range of targets but many are directed against antigens present within the cell or within the nucleus. This has led to the suggestion that patients with SLE have defects in apoptosis or in the clearance of apoptotic cells, which causes inappropriate exposure of intracellular antigens on the cell surface, leading to polyclonal B- and T-cell activation and autoantibody production. This is supported by the fact that environmental factors that cause flares of lupus, such as ultraviolet (UV) light, pregnancy and infections, increase oxidative stress and/or stimulate apoptosis; by the association with complement deficiency; and by the defect in the TREX1 nuclease.

Patients often have non-specific symptoms. Some, such as fever, weight loss and mild lymphadenopathy, reflect active inflammatory disease, whereas others, such as fatigue, malaise and fibromyalgia-like symptoms, are not necessarily associated with flares in disease activity, at least as assessed by standard laboratory tests.

Rash is common in SLE and is classically precipitated by exposure to UV light. Three distinct types of rash can occur.[2,3] The classic butterfly facial rash (up to 20% of patients) is erythematous, raised and painful or itchy, and occurs over the cheeks with sparing of the nasolabial folds. Subacute cutaneous lupus erythematosus (SCLE) rashes are migratory, non-scarring and either annular or psoriaform. Discoid lupus lesions are characterised by hyperkeratosis and follicular plugging, and may cause scarring alopecia if present on the scalp. Diffuse, usually non-scarring alopecia may occur with active disease. Other skin manifestations include periungual erythema (reflecting dilated capillary loops), vasculitis and livedo reticularis, which is also a common feature of the antiphospholipid syndrome.[4] Other cutaneous manifestations related to, but not specific to, SLE include the following: Reynaud phenomenon, Livedo reticularis, Panniculitis (lupus profundus), Bullous lesions, Vasculitic purpura, Telangiectasias and Urticaria.

Limited studies are available in India which has documented the frequency of cutaneous manifestations of SLE, so, this study was planned to evaluate the frequency of skin manifestations of SLE, in a tertiary care centre of Malwa region of India.

METHODS

Study design

This is a prospective study.

Study setup

This study is conducted at Department of Dermatology, Venerology and Leprosy of a tertiary care centre.

Study duration

The duration of study was one year; January-2017 to December-2017.

Sampling

Purposive sampling technique is used for selection of desired samples according to inclusion criterion.

Sample size

One thousand patients of SLE visiting Department of Dermatology, Venerology & Leprosy of a tertiary care centre were evaluated for possible inclusion in study.

Inclusion criteria

All adults of either sex, age ≥18 years who fulfilled the ACR criteria were enrolled in the study.

Exclusion criteria

Patients with drug-induced SLE (history of drug intake and appearance of skin lesions after receiving medication) were excluded from the study.

Methods

Written informed consent was taken from all subjects. All demographic data including age, sex, weight was collected on a pre-designed proforma. All the patients were evaluated for cutaneous manifestations of SLE i.e. malar rash, discoid rash, oral ulcers, photosensitivity etc. All the information was recorded in designed proforma.

Statistical technique

The demographic data of 100 subjects was analysed by statistical software, SPSS version 17.0. Microsoft Excel® and SPSS® 20 for Windows® were used for data storage and analysis. The qualitative data were expressed in percentages and quantitative data were expressed as mean ± standard deviation. Student's t test and Chi-Square test were used to determine statistical difference between variables.

Financial input and funding: The patient underwent procedures as per protocol laid down by our institution for management of such patients. Hence there was no financial burden on patient or institution. This project was not funded by any of pharmaceutical/diagnostic industry.

RESULTS

Total 100 SLE subjects were included in this study, among them 96 (96%) were female and 4 (4%) were male. Mean age of subjects was 28.8±12.8 years (Table 1) Mean (±SD) body mass index (BMI) was 23.8(±2.6) and 23.2 (±2.8) among males and females respectively.

Table 1: Demographic measures of all subjects.

Characteristics	SLE Patients
Number	100
Gender % (Female)	96%
Age (Mean±SD)	28.8±12.8 years
BMI (Mean±SD)	23.7 ±3.1 kg/m^2

Among 100 SLE subjects, 82 (82%) patients had skin manifestations, 18 (18%) patients had oral ulcer and 12 (12%) patients had raynaud's phenomenon at the time of presentation. Among dermatological manifestations, 42% patients had photosensivity, 35% patients had malar rash, 30% patients had discoid rash, 10% patients had alopecia and 2% patients had bullous lesion.(table 2)

Table 2: Skin manifestations of SLE patients

Type of Lesion	Percentage (%)
Photosensivity	42
Malar rash	35
Discoid rash	30
Alopecia	10
Bullous lesion	2

DISCUSSION

SLE is a disease of unknown cause that may produce variable combinations of fever, rash, hair loss, arthritis, pleuritis, pericarditis, nephritis, anemia, leukopenia, thrombocytopenia, and central nervous system disease. The clinical course is characterized by periods of remissions and acute or chronic relapses. Characteristic immune abnormalities, especially antibodies to a number of nuclear and other cellular antigens, develop in patients with SLE.SLE is an autoimmune disorder characterized by multisystem inflammation with the generation of autoantibodies. Although the specific cause of SLE is unknown, multiple factors are associated with the development of the disease, including genetic, epigenetic, ethnic, immunoregulatory, hormonal, and environmental factors.

SLE can occur at any age but has its onset primarily between ages 16 and 55. It occurs more frequently in women. In children, the female-male ratio is 1.4 to 5.8:1; in adults, it ranges from 8:1 to 13:1; and in older individuals, the ratio is 2:1. The prevalence of SLE is estimated to be between 4 and 250 cases per 100,000 populations. In the United States, the highest incidence is among Asians in Hawaii, blacks, and certain Native Americans (Sioux, Crow, Arapahoe). The risk of SLE developing in a black American female has been estimated to be 1:250. The prevalence is about the same worldwide; the disease appears to be common in China, in Southeast Asia, and among blacks in the Caribbean, but is seen infrequently in blacks in Africa. Limited observations suggest that the incidence of discoid lupus erythematosus is the same as that for SLE.

Few unique pathologic features are associated with SLE. Biopsies of the malar erythema may reveal some minor basal layer abnormalities, as well as immune complex deposits at the dermal-epidermal junction. Photosensitivity, implying a rash after exposure to UVB light (e.g., sunlight, fluorescent light), occurs in more than 50% of patients. Some patients are also sensitive to UVA light--the clue, rash after exposure to sun filtered through glass. Fair-skinned individuals tend to be more susceptible. Photosensitivity may develop at any time or vary in intensity during the course of SLE. The classic butterfly rash, i.e., erythema over the cheeks and nose, develops after UV exposure in more than 50% of patients. The skin may feel warm and slightly edematous. Application of alcohol, found in many sunscreens, may cause vasodilation and thereby more erythema. The rash may last for hours or days and often recurs. A maculopapular eruption with fine scaling may ensue and last longer, although it generally heals without residue. Discoid lesions develop in 25% of patients with SLE but may also occur in the absence of any other feature of SLE. Discoid lesions are characterized by discrete round, annular, erythematous, slightly infiltrated plaques covered by a well-formed adherent scale that extends into dilated hair follicles. Follicular plugging is prominent. Lesions slowly expand with active inflammation at the periphery, and in their wake are left depressed scars, telangiectasia, and depigmentation; central scarring with atrophy is characteristic. Lesions tend to occur on the face, scalp, neck, and ears and around the shoulders. Some lesions may be hyperkeratotic and thus be confused with psoriasis. Patients with isolated discoid lupus have about a 10% chance of eventually developing SLE. Subacute cutaneous lupus erythematosus occurs in about 10% of patients with SLE. The lesions are small, erythematous, slightly scaly papules that evolve into psoriasiform or annular forms. Lesions appear typically on the forearms and upper part of the torso; atrophy or scarring rarely develops, although telangiectasia does. A strong association is seen with HLA-DR3 and anti-Ro antibodies. Lupus profundus/panniculitis is a rare manifestation of SLE. Typically, painful nodules develop under a skin lesion on the scalp, face, arms, chest, back, thighs, and buttocks and resolve as a depression. Ulcerations are uncommon. The presence of immune complex deposits at the dermal-epidermal junction helps distinguish these lesions from those of the Weber-Christian syndrome. Bullous lesions are rare and can be distinguished from other bullous diseases by the difference in serum antibodies and dermal immune deposits. Hair loss, on the scalp or elsewhere, occurs in

71% of SLE patients. The most common is premature hair loss (telogen effluvium) characterized by a diffuse thinning of the scalp. Such hair loss may follow a flare of SLE, stress, pregnancy, or the use of steroids; the hair generally grows back. Some patients have "lupus hair," hair that easily fractures and is thin and unruly. Discoid lesions of the scalp usually result in permanent hair loss.

Many studies have been conducted to find out the patterns and frequency of cutaneous manifestations of SLE. Almost every study has shown different results. In our study, dermatological manifestations were seen in 82% of SLE patients while in another study from south-east Asia it was 71%.[6]

In our study, malar rash was seen in 35% of SLE patients, while in a study by Rabbani et al the malar rash was present in 29% of SLE patients.[7] Other studies have found the frequency of malar rash as high as 60% by Edward et al.[8], 56% by Mok et al and 58.9% by Ward et al.[9,10]

In our study, discoid rash was seen in 30% of SLE patients while in other studies, it was observed 14% by Rabbani et al, 12% by Mok et al, 10% by Edward et al, 10% by Parveen et al and 7% by Ward et al.[6-10]

Photosensitivity was seen in 42% patients in our study. While it was seen only in 6% patients in a study by Rabbani et al.[7] The frequency of photosensitivity was 31% in a study by Edward et al, 35% in a study by Mok et al and 48% in a study by Ward et al.[8-10]

In our study, the oral ulcers were seen in 18% patients. When compared to other study, oral ulcer was seen in 20% of SLE patients in study by Rabbani et al and Edward et al.[7-8]

CONCLUSION

Skin lesions in patients with lupus may be specific or nonspecific. This study covers the SLE-specific cutaneous changes: malar rash, discoid rash, photosensitivity, and oral mucosal lesions as well as SLE nonspecific skin manifestations. A deeper thorough understanding of the cutaneous manifestations of SLE is essential for diagnosis, prognosis, and efficient management. Thus, dermatologists should be involved with other specialties to provide optimal care of SLE patient.

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