ORIGINAL RESEARCH PAPER

Dermatology

A STUDY ON PATTERN OF CUTANEOUS MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS IN A TERTIARY CARE HOSPITAL OF NORTH INDIA

KEY WORDS: systemic lupus erythematosus, oral ulcer, malar rash, cutaneous manifesitations.

Dr Shweta Agarwal MD (Medicine), Associate Professor, Department of Medicine, Career Institute of Medical Sciences, Lucknow.

Rajeev Agarwal* MD (Dermatology), Associate Professor, Department of Dermatology, Career Institute of Medical Sciences, Lucknow *Corresponding Author

ABSTRACT

Background: Systemic Lupus Erythematosus (SLE) is an autoimmune connective tissue disease with multi-organ involvement with skin being the second most commonly affected organ. SLE with skin lesions can produce considerable morbidity resulting from painful skin
lesions, oral ulcers, disfigurement or toxic epidermal necrolysis like manifestations, etc. Skin lesions in patients with lupus may be specific (LE specific) or non-specific (LE non-specific) as per the Gilliam classification of skin lesion associated with Lupus Erythematosus (LE). Patients with acute cutaneous LE (Lupus specific) have high chances of having systemic disease.
Objective: This study was planned to evaluate the frequency and pattern of skin manifestations of SLE, in a tertiary care hospital of Lucknow in North India.
Material and Methods: This was a retrospective, observational study carried out in a tertiary care centre of North India over a period of 2 years (April 2017 - March 2019). 40 patients of SLE (5 male and 35 female), attending the Dermatology OPD of a tertiary care centre were enrolled in the study.
Results: The mean age of patients was 26 years; 87.5% patients were female and 12.5% were male. Out of 40 patients, oral ulcers were seen in 28 (70%), malar rash in 22 (55%), photosensitivity in 21 (52.5%) patients, discoid rash in 15 (37.5%), alopecia in 19 (47.5%), palpable purpura in 10 (25%), nail changes in 10 (25%), erythema multiforme in 7 (17.5%), Raynaud's phenomenon in 5 (12.5%), subacute cutaneous LE in 3 (07.5%), bullous eruptions in 3 (07.5%), telangiectasia in 2 (05%), facial oedema in 2 (05%), lupus panniculitis in 1 (02.5%), cheilitis in 1 (02.5%), lichen planus in 1 (02.5%).
Conclusion: SLE was predominantly seen among young female patients. Oral ulcer was the most common manifestation followed by malar rash and photosensitivity in our study.

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]

Lupus Erythematosus (LE) is the root designation for a diverse array of illnesses that are linked together by the development of autoimmunity directed predominantly at the molecular constituents of nucleosomes and ribonucleoproteins. These manifestations encompass a spectrum ranging from a few localized discoid LE lesions to the life-threatening systemic manifestations such as nephritis, central nervous system disease or vasculitis of systemic LE. Thus, it is important to Lupus Erythematosus (LE) is the root designation for a diverse array of illnesses that are linked together by the development of autoimmunity directed predominantly at the molecular constituents of nucleosomes and ribonucleoproteins. These manifestations encompass a spectrum ranging from a few localized discoid LE lesions to the life-threatening systemic manifestations such as nephritis, central nervous system disease or vasculitis of systemic LE. Thus, it is important to Lupus Erythematosus (LE) is the root designation for a diverse array of illnesses that are linked together by the development of autoimmunity directed predominantly at the molecular constituents of nucleosomes and ribonucleoproteins. These manifestations encompass a spectrum ranging from a few localized discoid LE lesions to the life-threatening systemic manifestations such as nephritis, central nervous system disease or vasculitis of systemic LE. Thus, it is important to recognise the varied clinical manifestations at early part of disease and prevent or decrease the subsequent morbidity and mortality.

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 Fc γ RIIIb or in the DNA exonuclease TREX1.

Skin is the second most common organ affected in Systemic Lupus Erythematosus (SLE). Cutaneous manifestations are often the first symptoms, which make the patient attend a dermatologist. The diagnosis of cutaneous manifestations of SLE is based on clinical, histopathological and immunohistology of skin lesions. In addition, serum autoantibodies are considered as immunologic markers for distinct clinical types of the illness.

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.

Aim and objectives:

This study was planned to evaluate the frequency and pattern of skin manifestations of SLE, in a tertiary care hospital of

Lucknow in North India.

MATERIAL AND METHODS:

This was a retrospective, observational study carried out in a tertiary care centre of North India over a period of 2 years (April 2017 - March 2019). 40 patients of SLE (5 male and 35 female), attending the Dermatology OPD of a tertiary care centre were enrolled in the study after Institutional Ethical Clearance. Informed consent was obtained from all patients. Detailed history about symptoms, duration and evolution of cutaneous lesions was obtained. The patients were examined clinically focussing the mucocutaneous manifestations.

Routine and specific laboratory investigations were carried out to confirm the diagnosis of SLE under Revised American College of Rheumatology (ACR) criteria 1997.

RESULTS:

In our study, male-to-female ratio was 1:7 and maximum number of patients were seen in age group of 21-30 years with a median age of 26 years. In our study oral ulcers were seen in 70%, malar rash in 55%, photosensitivity in 52.5%, alopecia in 47.5%, discoid rash in 37.5%, palpable purpura in 25%, nail changes in 25%, erythema multiforme in 17.5%, Raynaud's phenomenon in 12.5%, subacute cutaneous LE in 07.5%, bullous eruptions in 07.5%, telangiectasia in 05%, facial oedema in 05%, lupus panniculitis in 02.5%, cheilitis in 02.5%, lichen planus in 02.5%.

We did not see any case of pyoderma gangrenosum, livedo reticularis, chilblain lupus, Degos-like lesion/atrophie blanche, thrombophlebitis, episcleritis, rheumatoid nodules, erythromelalgia, sclerodactaly. Mucosa other than oral was not affected. 26.31% patients presented with nail changes, and included paronychia (7.89%), nail fold telangiectasia (5.26%), onycholysis (5.26%), ragged cuticles (2.63%), leukonychia (2.63%), splinter haemorrhages (2.63%).

Table 1: Different cutaneous manifestations of SLE

Cutaneous lesion	Male	Female	Total	%
Oral ulcer	3	25	28	70.0
Malar rash	2	20	22	55.0
Photosensitivity	2	19	21	52.5
Alopecia	1	18	19	47.5
Discoid rash	1	14	15	37.5
Palpable purpura	0	10	10	25.0
Nail changes	0	10	10	25.0
Erythema multiforme	0	7	7	17.5
Raynaud's phenomenon	0	5	5	12.5
SCLE lesion	0	3	3	7.5
Bullous lesion	0	3	3	7.5
Telangiectasia	0	2	2	5.0
Facial oedema	0	2	2	5.0
Panniculitis	0	1	1	2.5
Cheilitis	0	1	1	2.5
Lichen planus	0	1	1	2.5

Table 2: Distribution of patients according to age.

Age group	Male	Female	Total
0-10	0	0	0
11-20	0	2	2
21-30	2	20	22
31-40	2	10	12
41-50	1	3	4
Total	5	35	40

DISCUSSION:

It was a retrospective, observational study where we enrolled patients based on modified American College of Rheumatology criteria.[4]

In current study, patients age ranged from 0-50 years with mean age of onset 26 years. Kole et al,[5] observed a mean age of disease onset as 25 years and male-to-female ratio as 1:14, Parveen et al[6] observed mean age as 30.04 years and male--female ratio as 1:5; Masi et al[7] observed a median age of disease onset as 31 years.

In our study oral ulcers were present in 70%, while in study by Kole et al oral ulcers were present in 56.67%[5].

Parveen et al 34%.[6] study by Patel et al 19%.[8] Rabbani et al 21%.[9] Kapadia et al 60%.[10] Yell JA et al 31.5%.[11] In our study, the frequency of malar rash was 55%, while in study by Kole et al 80%[5], Parveen et al 70%.[6] Patel et al 40%.[8], Rabbani et al 31%.[9], Kapadia et al 60%.[10] Yell JA et al 51%.[11].

In our study the frequency of photosensitivity was 52.5%, while in study by Kole et al 50%.[5], Parveen et al 75%.[6], Rabbani et al 33%[9], Kapadia et al 60%[10], Yell JA et al 63%.[11].

In our study the frequency of alopecia was 47.5%, while in study by Kole et al 86.67%.[5], Parveen et al 47%.[6], Patel et al 24%.[8], Kapadia et al 82.5%.[10] and Yell JA et al 40%.[11] In our study the frequency of discoid rash was 37.5%, while in study by Kole et al 20%.[5], Parveen et al 10%.[6], Rabbani et al 15%.[9], Kapadia et al 57.5%.[10] and Yell JA et al 18%.[11]

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.

SLE was predominantly seen among young female patients. Oral ulcer was the most common manifestation followed by malar rash and photosensitivity in our study.

Limitation of study:

As it was a single centre study the results cannot be generalized to entire population. Furthermore comprehensive and multi centric studies including meta-analysis of various earlier studies should be done, to have a more meaningful and high impact results.

REFERENCES:

1. Livingston B, Bonner A, Pope J. Differences in clinical manifestations between childhood-onset lupus and adult-onset lupus: a meta-analysis. Lupus. 2011;20(13):1345-55.
2. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum. 1997;40(9):1725.
3. American College of Rheumatology. 1997 Update of the 1982 American College of Rheumatology revised criteria for classification of systemic lupus erythematosus. Available at http://tinyurl.com/ 1997SLEcriteria. Accessed on 25 September 2017.
4. Tan EM, Cohen AS, Fries JF, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1982;25(11):1271 7.
5. Kole AK, Ghosh A. Cutaneous manifestations of systemic lupus erythematosus in a tertiary referral centre. Indian J Dermatol 2009;54(2):132 6.
6. Parveen N, Das DK, Haque M, et al. Cutaneous manifestations of systemic lupus erythematosus. J Medicine 2007;8:44-8.
7. Tan EM, Cohen AS, Fries JF, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1982;25(11):1271-7.
8. Kole AK, Ghosh A. Cutaneous manifestations of systemic lupus erythematosus in a tertiary referral centre. Indian J Dermatol 2009;54(2):132-6.
9. Parveen N, Das DK, Haque M, et al. Cutaneous manifestations of systemic lupus erythematosus. J Medicine 2007;8:44-8.
10. Masi AT, Kaslow RA. Sex effects in SLE: a clue to pathogenesis. Arthritis Rheum 1978;21(4):480-4.
11. Patel P, Werth V. Cutaneous lupus erythematosus: a review. Dermatol Clin 2002;20(3):373-85, v.
12. Rabbani MA, Ahmad A, Siddiqui BK, et al. Do clinical manifestations of systemic lupus erythematosus in Pakistan correlate with rest of Asia. J Pak Med Assoc 2006;56(5):222-7.

10. Kapadia N, Haroon TS. Cutaneous manifestations of systemic lupus erythematosus: study from Lahore, Pakistan. International Journal of Dermatology 1996;35(6):408 9.
11. Yell JA, Mbuagbaw J, Burge SM. Cutaneous manifestations of systemic lupus erythematosus. Br J Dermatol 1996;135(3):355-62.