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Original Research Article

Cutaneous manifestations and systemic correlation in patients with lupus erythematosus and its subsets: a study of 40 cases

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ABSTRACT

Background: The objective was to study various dermatological manifestations and clinical and laboratory features in patients of lupus erythematosus (LE) and its subsets.
Methods: This is a cross sectional observational study done in a tertiary care hospital in a rural setup in Piparia, Ta. Waghodiya, Dist. Vadodara, Gujarat. All the patients with clinical features of LE and its subsets were included in the study over a period of 16 months and were subjected to detailed history taking, complete cutaneous and general examination and laboratory investigations.
Results: Out of the 40 cases, 22 cases were systemic lupus erythematosus (SLE), 13 cases were discoid lupus erythematosus (DLE), four cases were of Rowell's syndrome and one case was of mixed connective tissue disorder (MCTD). 97.5% of cases had cutaneous involvement, i.e. photosensitivity (77.5%), oral ulcers, hair loss and malar rash. Systemic symptoms and abnormal laboratory parameters were present in the cases of SLE, Rowell's syndrome and MCTD, with hematological involvement being the most common in both SLE (77%) and Rowell's syndrome (100%) and positive anti-nuclear antibody (ANA) titer being the most common abnormal laboratory finding in both SLE (95.4%) and Rowell's syndrome (100%). The most commonly found antibody was anti-dsDNA (64.3%) in SLE and anti SS-A (100%) in Rowell's syndrome.
Conclusions: Cutaneous features though occasionally subtle, are pointers to a diagnosis of SLE. Hair loss, malar rash and photosensitivity alone or in association with altered hematologic/ANA profile are the key markers of the disease activity.

Keywords: Lupus erythematosus, Cutaneous manifestations, Anti-nuclear antibody

INTRODUCTION

Lupus erythematosus (LE) is an autoimmune connective tissue disorder which clinically presents with a variety of cutaneous and systemic manifestations.

The most common and first affected organ is skin. The skin lesions also vary in their morphology ranging from discoid lesions in DLE, malar rash in systemic lupus erythematosus (SLE) to toxic epidermal necrolysis (TEN) like lesions. Therefore, it is very important to identify the skin lesions and diagnose the disease at an early stage so that appropriate treatment can be started immediately.[1]

The laboratory parameters are altered in many cases and suspected cases should be subjected to investigations.

The present study focuses on various cutaneous and systemic manifestations that were present in the cases of LE who presented at dermatology OPD. The study also

shows various abnormal laboratory parameters that act as indicators of different system dysfunctions in LE.[1]

The present study was carried out with following aims and objectives:

- To study various demographic parameters related to LE and its subsets.
- To study dermatological manifestations in patients of LE and its subsets.
- To study various hematological and biochemical changes in patients of LE and its subsets.
- To study the ANA profile in patients of LE and its subsets.

The nosographic concept of LE includes 3 major subtypes: chronic cutaneous LE, subacute cutaneous LE, and systemic or acute cutaneous LE. Besides these 3 subtypes, other less frequent clinical varieties may occur.[2]

LE is a chronic, autoimmune disease that includes a broad spectrum of symptoms. Lupus is the Latin word for wolf and has been used to name various skin diseases at least since the 10thcentury. LE is included among the so called connective tissue diseases and is divided into one systemic form – SLE and one cutaneous form – CLE.[3]

METHODS

The present study of 40 cases of '(LE) and its subsets' was carried out over a period of 16 months (i.e. from May 2016 to August 2017) at Department of Dermatology, Venereology and Leprosy of Dhiraj Hospital, Piparia.

Study design

It is a prospective observational study.

Inclusion criteria

All patients who presented with clinical features of cutaneous LE and its subsets were classified according to the classification proposed by Gilliam et al and were subjected to appropriate investigations.[4]

The patients having clinical signs and symptoms of systemic LE were investigated and systemic lupus international collaborating clinics (SLICC) criteria were applied to them.[5]

All the cases thus confirmed of having cutaneous or systemic LE were included in the study.

History

A detailed history was taken regarding the onset, duration and progression of the disease.

Other specific history pertaining to various clinical features of the disease was asked e.g. fever, joint pain/swelling, oral/nasal lesions, hair loss, weakness, Raynaud's phenomenon, photosensitivity, hematuria and other constitutional symptoms.

Any significant past, present, family or drug history was also asked.

Examination

Complete general examination was carried out, followed by a thorough cutaneous examination from head to toe, including all mucosa.

Systemic examination was done wherever indicated.

- Investigations
- Baseline investigations

All the cases were subjected to baseline investigations, which are as follows:

Complete blood count (CBC), erythrocyte sedimentation rate (ESR), urine examination (routine and microscopic), liver function tests (LFT), renal function tests (RFT), blood sugar levels (random, fasting and post prandial), anti-nuclear antibody (ANA) titres and profile in selected cases. Special investigations like skin biopsy, x-ray chest, electrocardiogram (ECG), ultrasonography (USG) - abdomen and pelvis, 24 hour urinary protein, rheumatoid arthritis (RA) factor, renal biopsy in certain cases.

RESULTS

The present study comprised of 40 cases, of which the most cases were of SLE 22 cases, followed by DLE 13 cases, Rowell's syndrome four cases and mixed connective tissue disorder one case.

Table 1: No. of cases.

Diagnosis	Number of cases (n=36)	Percentage (%)
SLE	21	58.33
DLE	13	36.11
MCTD	1	2.78
Rowell's syndrome	1	2.78

Skin lesions were the most common clinical manifestation, accounting for 39 out of 40 cases. The most common site involved was face and neck 37 cases, followed by upper limbs 18 cases, chest 16 cases, lower limbs 10 cases and scalp 7 cases, thus confirming the photosensitive nature of the disease.

Among the thirteen cases of DLE, eight patients had localized DLE, and five had disseminated DLE (Figure 1 A-C).

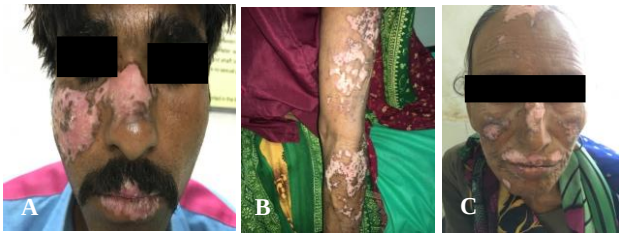

Figure 1: (A) Localised DLE; (B) disseminated DLE; (C) localised DLE.

Constitutional signs and symptoms were mainly seen in cases of SLE, Rowell's syndrome and MCTD. Out of the 27 cases of these, all the 27 cases had fever, while 25 cases had complaints of joint pain.

Table 2: Signs and symptoms of cases.

Cutaneous finding	No. of cases	Percentage (%)
Photosensitivity	27	75
Malar rash	16	44.44
Raynaud's phenomenon	8	22.22
Oral ulcer	21	58.33
Hair loss	19	52.78
Nail changes	9	25
Telegiecasia	5	13.89
Erythema multiforme	2	5.56
Edema	4	11.11

Nine cases had history of taking drugs for other diseases, among which four cases were known cases of pulmonary and extrapulmonary tuberculosis, and were taking treatment for the same. Out of the nine cases, six cases were of SLE and three cases were of Rowell's syndrome.

Abnormal laboratory parameters were observed in cases of SLE, Rowell's syndrome and MCTD. Among the 22 cases of SLE, the most common finding was a positive ANA titer in 21 cases, followed by anemia in 17 cases, elevated ESR in 15 cases and albuminuria in 13 cases. The most commonly involved system was renal in 13 cases, followed by cardiopulmonary in five cases and gastrointestinal in two cases. No cases of neuropsychiatric involvement were seen among the cases of SLE in present study.

Among the four cases of Rowell's syndrome, all the cases had a positive ANA titer and anemia while three cases had a positive rheumatoid arthritis factor. Two patients had albuminuria. The most commonly involved system was hematological in four cases, followed by renal in two cases. There was one case with central nervous system involvement, which had features of cerebritis on CT scan.

ANA profile was done in 19 out of 40 patients, all of which were of SLE, Rowell's syndrome or MCTD. Out of the 19 cases, 14 cases were of SLE, in which anti-dsDNA antibodies were the most commonly found antibodies in nine cases, followed by anti-Sm antibodies in five cases.

All the four cases of Rowell's syndrome had antibodies against SS-A and three cases had antibodies against Sm, followed by antibodies against nucleosomes in two cases, histones in two cases, U1-RNP in two cases, SS-B in two cases and ribosome-P in one case.

There were five cases that had positive RA factor. Out of them, two cases were of SLE and three cases were of Rowell's syndrome. Thus, it can be said that there were two patients of SLE with concomitant rheumatoid arthritis.

The case of MCTD had fever, joint swelling, hair loss, vasculitic ulcer, proximal muscle weakness clinically. Along with that, anemia, increased ESR, positive RA factor and ANA titer were seen. In ANA profile, antibodies against nucleosomes, dsDNA, histones, U1-RNP, SS-A and SS-B were seen. Along with the features of LE, the case also had features of rheumatoid arthritis, myositis and was a known case of hypothyroidism.

DISCUSSION

SLE frequently presents with cutaneous signs which have been highlighted through this study. The importance of these signs cannot be underestimated and the identification of the same by a dermatologist should be given utmost priority, especially in resource poor settings in India. ANA titer was found to be the most sensitive laboratory parameter, and hence, it can be used as a screening test in suspected cases of LE. Of the ANA profile, presence of anti-dsDNA antibodies was proved to be the most important marker of SLE. Whenever erythema multiforme like lesions are observed in a patient, a thorough clinical and laboratory examination should be carried out along with a detailed history to rule out the possibility of underlying Rowell's syndrome.

Female to male ratio was 2.64:1 (29 females, 11 males). In SLE, it was 4.5:1 (18 females, four males). These findings were consistent with that of studies done by Parveen et al and Kole et al.[6,7] All the four cases of Rowell's syndrome were female.

Table 3: Comparison of female:male ration.

Study	Female:male ratio
Present study	4.2:1
Parveen et al[6]	5.25:1
Kole et al[7]	14:1

The average age of the cases was 36.7 years, ranging from four years to 60 years. The average age was slightly higher than that of other studies done by Parveen et al and Kole et al.[5,6] There were six cases of pediatric LE.

Table 4: Comparison of mean age.

Study	Mean age at presentation
Present study	37.11
Parveen et al[6]	30.4
Kole et al[7]	30

Among LE-nonspecific features, photosensitivity was the most common feature 31 cases, followed by oral ulcers 25 cases (Figure 2), hair loss 20 cases (Figure 3), malar rash 19 cases (Figure 4), nail changes 10 cases and Raynaud's phenomenon 9 cases. These findings were comparable with studies done by Parveen et al and Malaviya et al.[6,8] One case had cutaneous features suggestive of toxic epidrmal necrolysis (Figure 5 A, B).

The abnormal laboratory parameters and system involvements were comparable with studies done by Kole et al and Santhanam et al, except the CNS involvement, which was quite higher in other studies than the present one.[7,9]

Table 5: Comparison of laboratory findings.

Study	Renal (%)	Hematology (%)	Cardiology (%)	Gastrointestinal (%)	CNS (%)	ANA titre (%)
Present study	57.14	76.14	23.81	9.52	0	95.23
Kole et al[7]	46.67	83.34	13.34	46.67	73.34	100
Santhanam et al[9]	45	32	30	NA	46	100

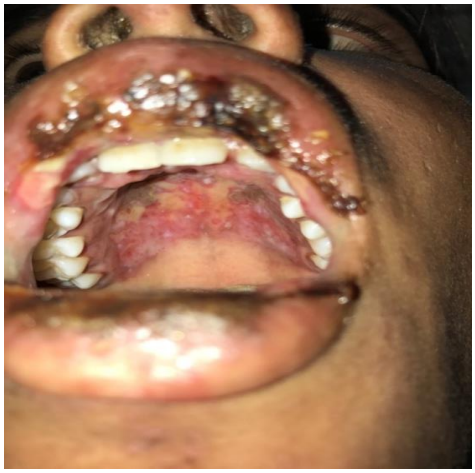

Figure 2: Oral ulcers in SLE.

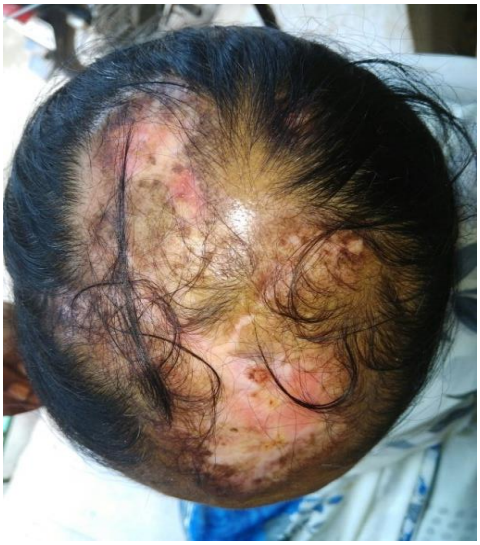

Figure 3: Scarring alopecia in DLE.

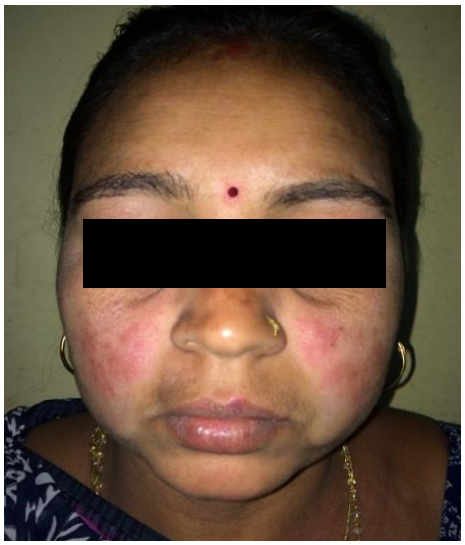

Figure 4: Malar rash.

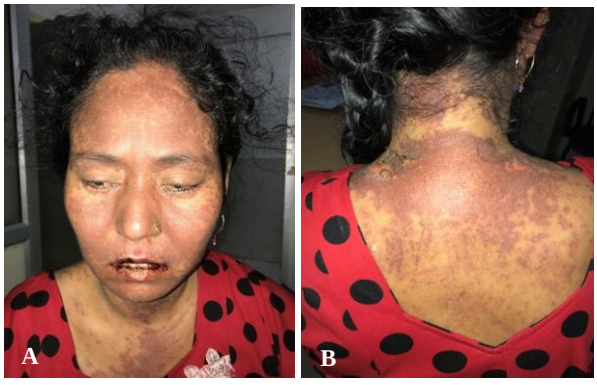

Figure 5 (A, B): SLE with ten like lesions.

Other antibodies that were found were against U1-RNP in three cases, SS-A in three cases, nucleosomes, SS-B, ribosome-P and histones two of each. These findings

were consistent with the studies done by Malaviya et al and Saigal et al.[6,10]

It is important to note that though four cases had history of taking anti-tubercular drugs, it couldn't be commented upon if the disease activity in these patients were triggered by the drugs.

With all this said and done, we conclude that a detailed history and complete clinical examination along with appropriate investigations are necessary to diagnose the cases of LE early in its course, which may help to alter its progression with prompt treatment.

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Conflict of interest: None declared
Ethical approval: The study was approved by the institutional ethics committee

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