

CUTANEOUS MANIFESTATIONS OF LUPUS ERYTHEMATOSUS FROM A TERTIARY CARE CENTRE IN COASTAL KERALA

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ABSTRACT

BACKGROUND

The aim of this study was to describe the Lupus Erythematosus (LE)-specific and LE-nonspecific cutaneous manifestations of lupus erythematosus.

MATERIALS AND METHODS

Cutaneous manifestations of lupus erythematosus in forty consecutive patients with LE who attended Dermatology department from January 2014 to June 2015 were studied.

RESULTS

Majority of the patients included in the study were female (65%). The most common age group affected was 20-40 years (47.5%). Photosensitivity was the most common symptom (45%). Forty five percent of patients in the study were diagnosed of systemic lupus erythematosus (SLE) according to the ACR criteria. LE-specific skin lesions were more common than LE-nonspecific lesions (95% vs. 40%). Chronic Cutaneous LE (CCLE) was the most common LE-specific skin lesion (85%). Only psoriasiform type of Subacute Cutaneous LE (SCLE) lesions were seen in the study group. Malar rash was the most common Acute Cutaneous LE (ACLE) lesion (87.5%). Features suggestive of systemic involvement were most commonly found in patients with ACLE and least in those with CCLE. Most common LE nonspecific lesion was non-scarring alopecia (32.5%) due to telogen effluvium. Cutaneous vascular disease in the form of palpable purpura of leucocytoclastic vasculitis (2.5%), periungual telangiectasia (7.5%), erythema multiforme (2.5%) and leg ulcers (5%) were the other LE- nonspecific skin lesions observed in the study. Patients with LE-nonspecific skin lesion had more systemic involvement compared to those without it.

CONCLUSION

LE-specific lesions are more common and can act as a diagnostic clue for lupus. LE-nonspecific lesions are more commonly associated with systemic disease.

KEYWORDS

Acute Cutaneous Lupus Erythematosus; Subacute Cutaneous Lupus Erythematosus; Chronic Cutaneous Lupus Erythematosus; LE-Specific and LE-Nonspecific Skin Lesions.

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BACKGROUND

Lupus erythematosus (LE) is a heterogeneous disease with a wide clinical spectrum and course that can vary considerably ranging from disease limited to skin to serious manifestations that can be found in systemic LE (SLE). Skin is the second most common organ affected; second only to musculoskeletal system.

Cutaneous lesions, even though rarely life threatening may persist for many years causing alopecia, scarring, pigmentary abnormalities and disfigurement. Thus, these are potentially disabling, limiting quality of life with several patients experiencing some form of vocational handicap.

Dermatological manifestations of LE are myriad and often possess diagnostic and therapeutic challenge. Skin lesions in LE can act as diagnostic clue as well as predictor of severity of systemic involvement. Since the subsets in cutaneous LE differ considerably in relation to their clinical course and systemic involvement, identification and classification of cutaneous lesions is important prognostically also. Though there are several studies on LE, most investigators have focused on patients with SLE alone. The present study attempts to describe the broad clinical spectrum of skin disease in LE including those patients without SLE.

MATERIALS AND METHODS

This descriptive study was conducted from January 2014 to June 2015 (18 months) including all clinically diagnosed patients with acute, subacute and chronic cutaneous LE or patients satisfying 1982 Revised Criteria for Classification of Systemic Lupus Erythematosus and its update in 1997 by American College of Rheumatology (ACR)^{1,2} among patients attending outpatient and inpatient departments of Dermatology, Venereology and Leprosy, at a tertiary care hospital in Alappuzha, India. Patients with LE-unrelated skin lesion alone, were excluded.

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Forty patients satisfied the study criteria. Informed written consent was taken prior to detailed history, systemic and dermatological examinations. Cutaneous manifestations of LE were charted out as LE specific and LE nonspecific according to Gilliam's classification.³ Punch biopsy of skin for histopathological and direct immunofluorescence study was done in clinically doubtful cases of cutaneous LE. Investigations including haemoglobin, total WBC count, differential count, ESR, platelet count, urine- albumin & microscopy, ANA, Anti-ds-DNA were done in all patients. Data was recorded in pre-structured proforma and analysed using SPSS 16.0 software. Qualitative data was analysed using percentages and quantitative data was analysed using mean. Permission to conduct the study was obtained from Human Ethical Committee and Institutional Research Committee.

RESULTS

Out of the 40 patients included in the study, 26 were female (Male: Female = 1:1.86). Ages of patients ranged from 13 years to 75 years (mean age = 39.8 years). LE specific lesions were seen in 38 patients (95%) and LE nonspecific lesions in 16 patients. Twenty-four patients had LE specific lesions alone (60%), 2 had LE nonspecific lesions alone (5%) and 14 patients (35%) had both LE specific and LE nonspecific lesions.

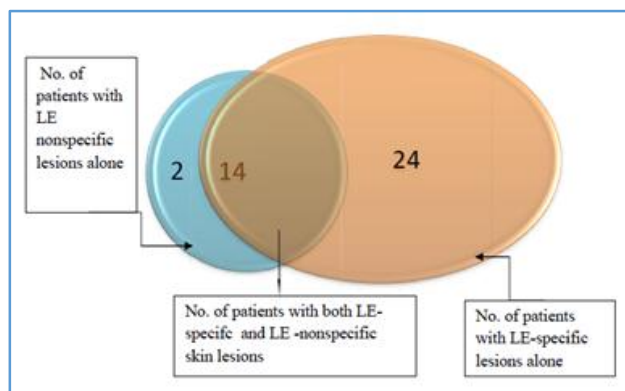


Figure 1. Distribution of LE Specific and LE Nonspecific Skin Lesions

Of the 40 patients, features suggestive of systemic involvement were as follows: photosensitivity was seen in 18 patients, oral ulcers in 12 patients, arthralgia in 14 patients, neurological disorder in 2 patients, renal disorder in 4 patients, haematological disorder 16 patients, immunological abnormalities in 18 patients. Serositis in the form of cardiopulmonary disease was not observed in the study. Eighteen patients (45%) in the study satisfied the ACR criteria for diagnosis of SLE. Ratio of patients with isolated cutaneous LE to SLE was 1.22:1.

	ACLE	SCLE	CCLE
Number of patients	8	2	34
Percentage of patients	20.00	5.00	85.00

Table 1. Distribution of LE Specific Skin Lesions

	Non-scarring Alopecia	Cutaneous Vascular Disease	Leg Ulcers	Erythema Multiforme
Number of patients	13	4	2	1
Percentage of patients	32.5	10	5	2.5

Table 2. Distribution of LE Nonspecific Skin Lesions

Among LE specific lesions, the most common type was Chronic Cutaneous LE (CCLE) seen in 34 patients followed by Acute Cutaneous LE (ACLE) in 8 patients and Subacute Cutaneous LE (SCLE) in 2 patients. Out of 8 patients with ACLE, 5 had malar rash alone, 1 patient had maculopapular lupus rash alone and 2 had both malar rash and maculopapular lupus rash. Only psoriasiform type of lesions were observed in SCLE.



Figure 2. Subacute Cutaneous LE- Psoriasiform Lesions on Forearms.

Out of the 34 patients who had chronic cutaneous LE lesions, the most common type was classic Discoid LE (DLE) seen in 30 patients followed by mucosal DLE in 5 patients and hypertrophic LE in 3 patients. Other variants like lupus profundus, lupus tumidus, chilblain LE and lichenoid DLE were not observed in the study.

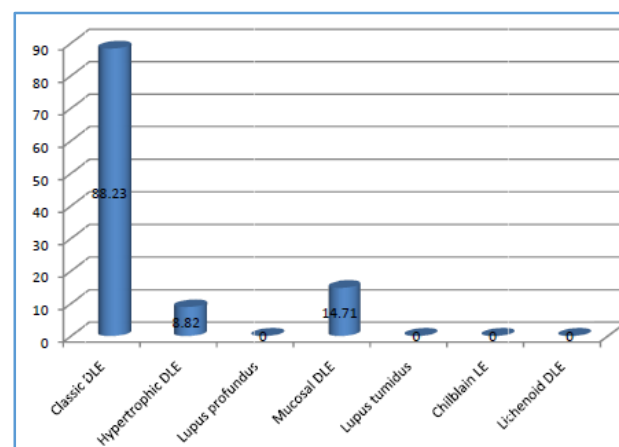


Figure 3. Distribution of Variants of CCLE in Percentage



Figure 4. Mucosal DLE Lesion on Hard Palate (arrow).

Photosensitivity was most commonly observed in ACLE patients (100%) in the study and least in CCLE patients (32.35%). Oral ulcers were also most commonly found in ACLE patients followed by SCLE (50%) and CCLE (23.52%). Arthralgia was present in all patients (100%) with SCLE and 62.5% patients with ACLE. But only 23.52% of CCLE patients had arthralgia. None of the patients had cardiopulmonary involvement. Neuropsychiatric disorder in the form of psychosis was seen in 12.5% of ACLE patients and 2.94 % of CCLE patients. Haematological disorders were present in all patients (100%) with ACLE and SCLE lesion and 29.4% of CCLE patients. Renal disorder was present mostly in ACLE patients (37.5%) followed by CCLE (8.8%). Immunological abnormalities were found in all patients (100%) with ACLE and SCLE, and 35.3% of CCLE patients.

Out of the 40 patients included in the study, 16 (40%) had LE nonspecific lesions, all of whom had features of SLE as well. Eighty nine percent of the SLE patients (n=14) had LE nonspecific lesions. The most common LE nonspecific lesion observed was non-scarring alopecia. Lupus hair (3 patients), telogen effluvium (13 patients) and alopecia areata (2 patients) were the types of non-scarring alopecia. Other LE nonspecific lesions were cutaneous vascular disease (palpable purpura-1 patient, periungual telangiectasia-3 patients), leg ulcers (2 patients) and erythema multiforme (1 patient). Patient with erythema multiforme also had SCLE, positive ANA, Anti-Sm antibody, Anti-Ro antibody and satisfied the ACR criteria for diagnosis of SLE.



Figure 5. Erythema Multiforme

Features suggestive of systemic involvement were more commonly found in patients with LE nonspecific skin lesions.

DISCUSSION

Lupus erythematosus comprises of a wide spectrum of clinical manifestations. Cutaneous manifestations of LE have been evaluated by multiple authors highlighting various clinical presentations of the disease. Malaviya⁴ et al, Das⁵ et al, Kole and Ghosh⁶ have extensively described the cutaneous manifestations of LE in Indian patients. This study evaluated the dermatologic manifestations of LE in patients from central Kerala. The study provides a preliminary data for comparing cutaneous manifestations of LE to other populations in India and the world.

Female gender was more frequently associated with LE and clinical findings in this study, similar to the observations reported by Biazar⁷ from Europe (3.3:1), Moghadam-Kia⁸ et al from US (3.2:1) and Das⁵ et al (4:1) from Kolkata. The mean age of patients in this study was 39.8 (range 13-75 years). Similar findings were reported by Tebbe and Orfanos⁹ in their study on 97 LE patients, with commonest age of presentation between 21 and 50 years.

A significant number of patients with cutaneous disease also have systemic involvement. Forty five percent of the patients in this study had features satisfying ACR criteria for SLE. Similar findings were reported by Biazar⁷ et al (40.7%) and Meuth¹⁰ et al (44%). Among the clinical features suggestive of systemic involvement, the most common signs/symptoms in the study population were photosensitivity (45%) and immunological abnormalities (45%). The prevalence of photosensitivity varies among different populations. Foering¹¹ et al reported 68% prevalence of photosensitivity in US patients and Sanders¹² et al found it to be 92%.

The most common LE-specific lesion in this study was CCLE (85%). Cardinali¹³ et al also had a similar observation with 72.5% patients of CCLE, 15% patients of ACLE and 8% patients of SCLE. Majority (87.5%) of ACLE patients had their lesions limited to the malar area. Cardinali¹³ et al (96%) and Meuth et al¹⁰ (100%) reported similar distribution of ACLE. Most studies^{8,13,14,15} report psoriasiform lesions as the commonest type of SCLE lesion. Both the SCLE patients in this study had psoriasiform lesions. The most common type of CCLE lesion was classic DLE (88.2%) similar to Moghadam-Kia et al⁸. Systemic involvements were common in ACLE and rare in CCLE. Photosensitivity, immuno-haematologic and renal disorders were the commonest findings in ACLE patients. Compared to other reports, incidence of systemic involvement in SCLE patients was higher in the study group.^{11,14,15} Similar observations in ACLE and CCLE were made by others.^{7,10,16}

LE-nonspecific skin lesions were seen exclusively in patients with SLE. The most common LE nonspecific lesion was non-scarring alopecia with predominant telogen effluvium. Similar findings have been reported from India by Cole⁶ and Ghosh⁶ and Biazar et al⁷ from Germany. The incidence of cutaneous vascular disease was lower compared to other reports.^{13,17} Association of LE with EM and positive Anti Ro in one patient with SCLE could suggest a diagnosis of Rowell's syndrome in this patient. Rowell's syndrome is reported in less than 100 patients worldwide.¹⁸ Photosensitivity, oral ulcers, arthralgia, renal, haematological

and immunological disorders, and ANA positivity were significantly associated with LE nonspecific skin disease. This is consistent with the observation by Cardinali¹³ et al that presence of nonspecific skin disease implies systemic disease.

CONCLUSION

Cutaneous manifestations of Lupus erythematosus are most commonly seen in adult females. CCLE is the commonest cutaneous manifestation of LE. Presence of LE nonspecific lesions can be a predictor of systemic involvement. A proper understanding of dermatological manifestations of lupus can act as a valuable aid in making diagnosis (LE specific lesions) as well as predicting prognosis (Presence of LE nonspecific lesions indicating systemic involvement).

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