# THE SPECTRUM OF CUTANEOUS MANIFESTATIONS IN LUPUS ERYTHEMATOSUS: THE TERTIARY HOSPITAL EXPERIENCE

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**Summary**

*Systemic lupus erythematosus (SLE) is an autoimmne disease in which muco-cutaneous lesions occur in majority of patients. This study from Chittagong Medical College Hospital, Chittagong is conducted to find out the pattern and spectrum of cutaneous lesions in SLE patients.*

*Forty (40) patients with SLE fulfilling the clinical and laboratory criteria of the American Rheumatology Association 1997, are examined in the Department of Dermatology & Venereology and Medicine between July 2008 and June 2009 for the presence of cutaneous manifestations in SLE patients.*

*Common cutaneous manifestations observed are photosensitivity (95%), oral ulcer (87.5%), malar rash (82.5%), non-cicatricial diffuse alopecia (67.5%), cutaneous vasculitis (62.5%), DLE Lesions (57.5%), lesions in lip (30%) and urticaria (27.5%), Raynaud's phenomenon (12.5%). Purpura, bullous lesion, livedo reticularis, leg ulcer, dermal atrophy, lchthyosis, genital mucosal lesion, nail changes, maculopapular rash, erythema multiforme, lupus profundus, lichen planus, conjunctival ulcer, psoriasiform lesion, xeroderma, pitted scarring in both toes and digits, palmoplantar keratoderma are rare.*

*A different clinical pattern is noted in our patients than reported previously which will be helpful for the clinicians for early detection and intervention to prevent complications and to improve quality of life and thus life expectancy may be prolonged.*

**Key words :** SLE; cutaneous; chittagong

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## Introduction

In Latin "lupus" means wolf. The rash therefore was said to resemble the skin which had actually been bitten by a wolf. The name lupus erythematosus (red) is given due to the facial rash, called "Butterfly rash"which is common in this disease. When it was found that many organs (system) of the body were affected then it was renamed systemic lupus erythematosus [1].

Lupus erythematosus is a heterogeneous, multisystem, autoimmune disease characterized by the production of autoantibodies against several cell constituents. The skin is one of the target organs most variable affected by disease [2].

To the clinicians systemic lupus erthematosus is important because it is potentially fatal disease that is easily confused with many other disorders.To the immunologist lupus is intriguing because all the key components of the immune system are involved in the underlying mechanism of the disease. The diverse presentation of lupus range from rash and arthritis through anemia and thrombocytopenia to serositis, nephritis, seizures and psychosis. Lupus should be part of the differential diagnosis in virtually any patients presenting with one of these clinical problems, especially in female patients between 15 and 50 years of age [3].

The skin and mucus membrane are symptomatically involved at some points in over 80% of patients with SLE. There is a tremendous variability and diversity in the type of involvement ranging from classical "Butterfly rash" and atropic hyperkeratotic lesion of discoid lupus to bullae, alopecia and vasculitis of dermal vessels [4].

There is no sufficient published data in Bangladesh regarding cutaneous manifestation in SLE patients.

The main purpose of the study is to analyze the prevalence and clinical importance of cutaneous lesion in SLE patients. So far there is no cure for LE and since cause of LE is unknown, some preventive measures may be taken by early diagnosis after assessment of cutaneous lesions and prompt early intervention to prevent complications.

**Materials and methods**

This is a cross sectional study carried out in Chittagong Medical College Hospital, Chittagong, both in the department of Dermatology and Venereology, and Medicine between July 2008 to June 2009. Patients primarily diagnosed as SLE by ARA (American rheumatology association) criteria 1997 were enrolled in the study. The history, physical examination, findings was recorded after taking informed consent from the patient. All data was collected using a structured questionnaire containing all the variables of interest. Outdoor / indoor patients of both sexes were included accordingly & finally all reports were recorded into a case record form. True years are considered for age determination as no fraction was taken.

Data was processed and analyzed using computer software SPSS (Statistical Packages for Social Sciences)Version 12. Data is presented by table, bar diagram and pie chart accordingly. The patients are analysed according to their age, sex, clinical features with special attention to cutaneous manifestations. Laboratory investigations included complete blood count, serum creatinine, ESR, 24 hours urinary total proteins, anti nuclear antibody, anti dsDNA, rheumatoid factor, skin biopsy, serum complement level, chest X ray, ultrasound of the whole abdomen and echocardiogram.

**Results**

Among the forty SLE patients, females out number the males. Two (5%) are males and 38 (95%) were females. The age of the patients enrolled in the study group ranged from 18 to57 years with mean age 29 (±0.05) years.Precipitating factors included pregnancy (40%), oral contraceptive pill (35%), family history of rheumatic diseases (10%), sunlight (10%), anti-TB drugs (5%). At the time of presentation 50% patients had cutaneous lesions, 45% patients had cutaneus and systemic lesions and only 5% had systemic lesions. Spectrum of clinical manifestation in SLE are shown in table I.

**Table I**: Spectrum of cutaneous findings in SLE patients

| Cutaneous manifestations | (n = 40)[α] |
|---|---|
| 1.Photosensitivity | 38 (95)[β] |
| 2. Malar rash / Butterfly rash | 33 (82.5) |
| 3. Cutaneous vasculitis | 25 (62.5) |
| 4. DLE lesions | 23 (57.5) |
| 5. Urticaria | 11 (27.5) |
| 6. Raynaud's phenomenon | 5 (12.5) |
| 7. Livedo reticularis | 3 (7.5) |
| 8. Purpura | 3 (7.5) |
| 9. Bullous lesions | 3 (7.5) |
| 10. Leg ulcer | 2 (2.5) |
| 11. Dermal atrophy | 2 (5) |
| 12. Ichthyosis | 2 (5) |
| 13. Maculo papular rash | 1 (2.5) |
| 14. Erythema multiforme | 1 (2.5) |
| 15. Lichen planus | 1 (2.5) |
| 16. Psoriasiform lesion | 1 (2.5) |
| 17. Erythema nodosum | 1 (2.5) |
| 18. Palmo plantar keratoderma | 1 (2.5) |
| 19. Pitted scaring in both digits & toes | 1 (2.5) |
| 20. Xeroderma | 1 (2.5) |

α n will not correspond to 100%, because of multiple cutaneous manifestations.
β Figures in the parenthesis indicate corresponding %.

Among LE-specific cutaneous lesions noted (Fig 1) were photosensitivity (95%), malar rash (Fig 2) (82.5%), DLE lesions (Fig 3) (57.5%) and bullous lesions (7.5%), maculo papular rash (2.5%), Lupus propundus (2.5%), conjunctival DLE (2.5%) and psoriasiform lesions (2.5%).

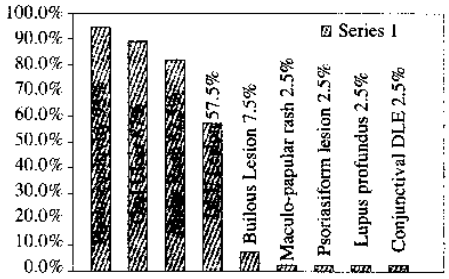


**Fig 1**: Shows LE-specific skin disease

Non specific lesions of SLE included (Fig 4) cutaneous vasculitis (62.5%), urticaria (27.5%), Raynaud's phenomenon (12.5%), livedo reticularis (7.5%), purpura (7.5%), leg ulcer (5%) (Fig 5), dermal atrophy (5%), ichthyosis (5%), erythema multiforme (2.5%) (Fig 6), lichen planus (2.5%), erythema nodosum (2.5%), palmo plantar

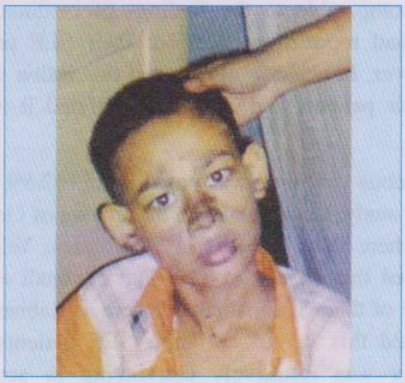

**Fig 2** : Malar rash

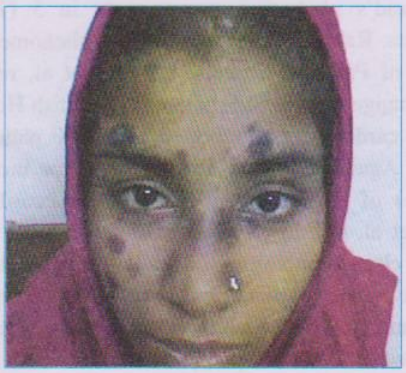

**Fig 3** : DLE Lesion

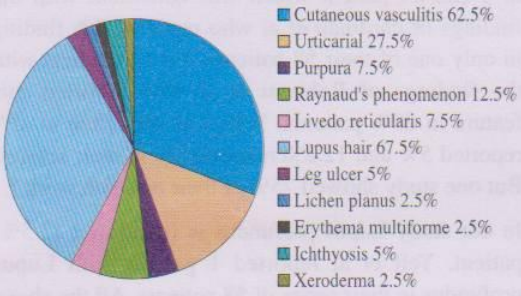


**Fig 4** : Shows pie diagram of LE non-specific skin disease

keratoderma (2.5%), pitted scarring in both digits and toes (2.5%) and xeroderma (2.5%) None of the patients had lesions of annular polycyclic lesions, hypertrophic DLE, lupus tumidus, thrombophlebitis, sclerodactyly, gangrene extremities, atrophae blanche, rheumatoid nodule, erythromelalgia, pyoderma gangrenosum.

Hyperpigmentation occurred in 2.5% patients. Hair changes occurred in 67.5% patients those included non-cicatricial diffuse alopecia (Fig 7) and lupus hair. Follicular plugging is not found. Five percent patients

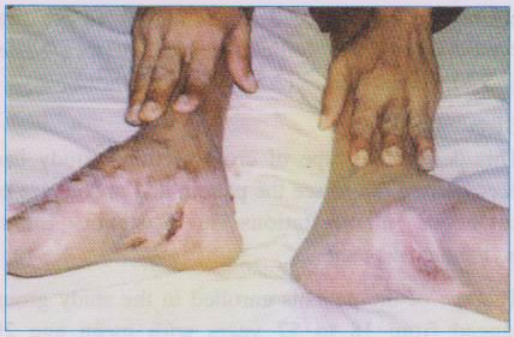

**Fig 5** : Leg Ulcer

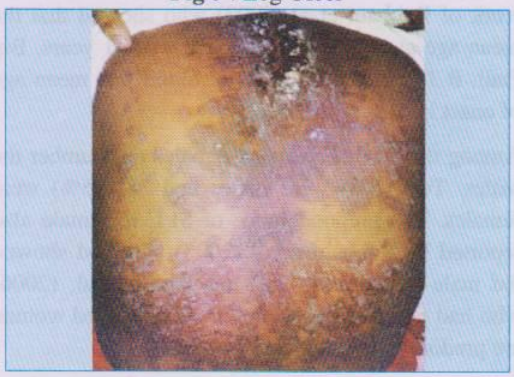

**Fig 6** : ErythemaMultiforme

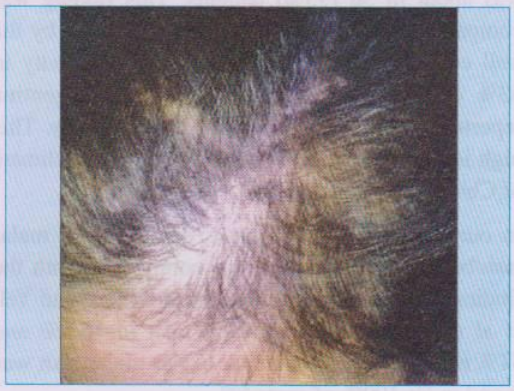

**Fig 7** : Alopecia

presented with nail changes and included panonychia (2.5%) and onycholysis (2.5%). Periungual telangiectasia, nail fold erythema, Red lunula, pitting, ridging, clubbing, leukonychia, splinter hemorrhage are not observed in our series of patients.

Oral mucosal ulcer is present in 87.5% cases. Ulcers in nasal septum and nasopharynx (15%), ulcer in glans penis (2.5%), ulcer in vulva (2.5%), and ulcer in conjunctiva (2.5%) are found. The rest of the mucosal surfaces of the body are not affected. Subcutaneous nodules, anetoderma, acanthosis nigricans, cutaneous mucinosis, calcinosis, facial

edema are not recorded. Infections noted are oral candidiasis (10%), folliculitis (5%), herpes zoster (2.5%), genital herpes (2.5%), genital wart(2.5%)

**Discussions**

This descriptive type of cross sectional study had been carried out to see the pattern and prevalence of cutaneous manifestations in Systemic Lupus erythematosus patients.

The age of the patients enrolled in the study group ranged from 18 to 57 years with mean age 29 (±0.05) years. This result was consistent with the work of Rabbani et al (2003) who showed that the mean age at presentation of SLE was 31 years. But Paul, B J et al (2003) of India found that mean age of onset is 21.6 years [5].

Among the 40 SLE patients females out number the males. Two (5%) were males and 38 (95%) were females. The preponderance of SLE in female also reported by Rabbani et al; (2003) who had showed the male-female ratio 1:8 and James et al; (2006) who had reported that young to middle aged woman are predominantly affected with SLE.

Most of the patients had got photosensitivity and it was about in 38 (95%) patients. This finding was comparable with that of the study conducted by the Yell et al (1996)[6] who reported photosensitivity in 63% cases. Saurit V et al (2003)[7] of Argentina reported 57.1% photosensitivity in their study. This high incidence was attributable to the warm climate of Chittagong, Bangladesh.

In our study 33 (82.5%) cases demonstrated malar rash/butterfly rash, which was comparable with the findings, reported by cardinali et al (2000)[8] and Yell et al (1996) who found malar rash in 46.5% and 51% respectively. In other studies as malar rash was found in 60-100% of cases [9-10].

The present study reveals that oral ulcer was present in 35 (87.5%) cases. It was higher than the finding of Yell et al who found oral ulcer in 31.5% cases. In other studies the incidence was 16-60% [9-12].

The present study shows that non-cicatritial alopecia with Lupus hair was present in 27 (67.5%) cases. Yell et al reported non-scarring alopecia in 40% cases, as well. This was consistent with the reports mentioned by other different authors [13, 14].

DLE lesion was found in 23 (57.5%) cases in the present study. It was similar to the findings of other studies where DLE was found in 57.5% patients [9]. It was comparable to the findings of cardinali et al who had reported in 32.6%of their SLE patients. However, Rabbani et al reported this lesion in 15% of their patients and Yell et al reported it in 25% cases.

Cutaneous vasculitis was noted in 25 (62.5%) cases in our study. It was nearer to the finding of Grigor et al[15] where they reported in 70% cases. Yell et al reported this change in 11% and cardinali et al in 13.7% of their SLE patients. Where as Rabbani et al reported this in 20% of their SLE patients. This finding was intuitively convincing in acute on chronic presentation of SLE patient.

Raynaud's phenomenon was seen in 5 (12.5%) patients. Rabbani et al reported this phenomenon in 2.5% of Pakistan patients, but Yell et al, reported this change in 60% SLE patient in a British Hospital, while cardinali et al reported in 39.6% patients in Itally. Again Paul et al noted a very low incidence (2.7%) of Raynaud's phenomenon. According to Paul et al, this low incidence is attributable to the warm climate in Northern Keralla, India. We believe in the same explanation for low incidence of Raynaud's phenomenon in our study, as Chittagong, Bangladesh was in tropical zone.

In the present study maculo papular rash was present in 1 (2.5%) patient which was consistent with the findings of cardinali et al who reported this finding in only one of their 58 patients, but contradicts with the findings of Rabbani et al who reported this feature in 20% patients. Yell et al and Vitali et al[16], reported 5% and 12.2% respectively in their studies. But one study showed 75% in their research work [9].

In our study Lupus profundus is found in 1 (2.5%) patient. Yell et al reported 1 patient with Lupus profundus in their series of 58 patients. All the above findings were identical.

Livedo reticularis was found in 3 (7.5%) patients. But it was similar to finding of Cardinali et al who has found Livedo reticularis in 8.6% patients & Yell et al had reported in 4% cases while other studies found 16.7% and 16.4% respectively[13-14]. Saurit V et al had found Livedo reticularies in 11.7% patients. Rabbani et al found Livedo reticularis in 3% patients.

Urticaria had been seen in 11 (27.5%) patients. Rabbani et al (2003) also reported 10% patients with urticaria, while Yell et al had found urticaria in 44% cases. Other studies reported urticaria in 7.5% and

13% cases respectively[14-15]. Bullous lesion also noted in 3 (7.5%) patients. Cardinali et al had found 3.5% patients with Bullous LE, but it was consistent with the findings of Yell et al where they had found in 8% patients. Others found Bullous lesions in 1.7% to 2% cases [14-15].

Purpura was seen in 3 (7.5%) patients. It was similar to the findings of Cardinali et al. They had found Purpura in 6.8% cases. In one study this sign noted in 24% of their patients [15].

Leg ulcer was seen in 2 (5%) patients. It was consistent with the findings of other studies where leg ulcer was found in leg ulcer in 5.3% & 8% respectively in their patients[15]. Cardinali et al hae found one patient with chronic leg ulcer.

Dermal atrophy was found in 2 (5%) patients. In one study one patient with dermal atrophy following healed DLE lesion was noted [17].

Acquired Ichlthyosis as noted in 2 (5%) patients. It was similar to the findings of Rabbani et al, they had found Acquired Ichlthyosis in 1% cases.

Hyper pigmentation was seen in 1 (2.5%) patient which was similar to the result obtained by Cardinal et al, who reported Hyper pigmentation in one patient out of a series of 58 patients. In one study it was found in 8.4% to 37.5% of patients [18].

Psoriasiform lesion, lichen planus, Erythema multiforme, Xeroderma and pitted scarring in both toes and digits are noted in 1 (2.5%) patient in each category. Cardinali et al reported 3.4% patients with papulo-squamous lesions in their study. Rabbani et al noted erythema multiforme in 1% case.

In our study 2 (5%) patients are noted with nail changes. 1 (2.5%) patient with paronychia and 1 (2.5%) patient with onycholysis. It was comparable with the findings of Rabbani et al in which they had found paronychia in 10% patients and onycholysis in 7% patients. No nail fold infarcts are found in our study.

Two (5%) patients had got genital lesions, 1 (2.5%) in penis and 1 (2.5%) in vulva. These were rare mucosal findings of SLE patients, where all other causes of genital ulcer had been excluded clinically. Conjunctival ulcer was noted in 1 (2.5%) patient. It was nothing but Conjunctival DLE.

Lip lesions were found in 12 (30%) patients, which included DLE and Blistering lesions. Lesions of annular polycyclic lesion, hypertrophic (verrucous) DLE, Lupus tumidus, Thrombophlebitis, Sclerodactyly, Gangrene extremities, Periungual telangiectasia, nail fold erythema, Red Lunula were not found in SLE patients under this study.

**Conclusion**

A different clinical pattern of SLE had been noted in this study than reported previously. There are many cutaneous manifestations in systemic lupus erythematosus, photosensitivity, oral ulcer, malar rash, DLE lesion, maculopapular rash, psoriasiform lesion, lupus profundus, conjunctival DLE are specific for SLE. Alopecia, cutaneous vasculitis, urticaria, Raynaud's phenomenon, purpura, Bullous lesions, Livedo reticularis, leg ulcers, lichen planus, Erythema multiform are non-specific for SLE. Recognition of both types of lesions is important in the early diagnosis of SLE for better management. LE non-specific skin lesions are those that in some way are related to the uderlying LE process, but are not specific for LE since the same lesions can be encountered in other autoimmune diseases. The ability to identify slight skin modifications at the onset or even during the remission of the disease and to differentiate from them LE like cutaneous lesions is probably a peculiarity of the dermatologist. In conclusion, we believe the fuifillment of strict interdisciplinary collaboration might be important not only for a careful clinical evaluation of the patients with SLE, but also for the optimisation of the therapeutic protocols.

**Disclosure**

All the authors declared no competing interestes.

**References**

1. Haq S A SLE: A disease with many faces-Medinews. www.generalpharma.com 2005; l: 3-10
2. Alceu LCB. Cutaneous lupus erythematosus-Clinical and Laboratory aspects. An Bras Dermatol 2005; l.80: 119-131
3. Rahman A. Systemic lupus erythematosus-The new England Journal of Medicine 2008; 358: 929-939
4. Rabbani MA. Cutaneous manifestations of systemic lupus erythematosus in Pakistan patients-Journal of Pakistan Medical Association. 2003; 53: 539-541
5. Paul B J. clinical profile of systemic lupus erythematosus in Northern kerala, Journal of Indian Rheumatology Association. 2003; 11: 94-97

6. Yell J A. Cutaneous Manifestations in systemic lupus erythematosus. Br. J. Dermatol.1996; 135: 355-362

7. Saurit V. Muco Cutaneous Manifestations in systemic lupus erythematosus patients. Medicina (B.Alres). 2003; 63: 283-287

8. Cardinale C. The Spectrum of Cutaneous manifestations in systemic lupus erythematosus the Italian exprience. Lupus. 2000; 9: 417-423

9. Kuhn A, Sticherling M, Bunsmann G. Clinical manafestations of cutaneous lupus erythematosus. JDDG. 2007; 5: 1124-1140

10. Kuhn A, Hintz Dr, Oslislo C, Ruzicka T, Megahed M, Lehmann P. lupus erythematosus Tumidus-A neglected subset of cutaneous lupus erythematosus: report of 40 case. Arch Dematol. 2007; 136-1033-1041

11. Orteu C H. systemic lupus erythematosus presenting with oral mucosal lesions easily missed. British Journal of Dermatology. 2001; 140: 1219-1223

12. Silvia V L. lupus erythematosus clinical and histopathological study of oral Manifestations and immuno histochemical profile of the inflamatory infiltrate. Journal of cutaneous. Pathology.2007; 34: 558-564

13. Siriwan W, Dhevy W, Prapai P. Cutaneous manifestation of childhood systemic lupus erythematosus. Pediatric dermatology. 1998; 15: 342-346

14. Albrecht J. Dermatology position paper on the revision of the 1982 ACR criteria for systemic lupus erythematosus.lupus.2004;13: 839-849

15. Gilboe LM. Sicca symptoms and secondary Sjogrens symptoms in systemic lupus erythematosus: comparison with Rheumatoid arthritis and correlation with disease variables. Ann Rheum Dis.2001; 60: 1103-1109

16. Rothfield N. lupus erythematosus : systemic and cutaneous manifestations. clinics in Dermatology.2006; 24: 348-362

17. Nyberg F, Hasan T, Puska P, Stephansson E, Hakkinen M, Ranki A et al. Occurrence of polymorphous light eruption in lupus erythematosus. British Journal of Dermatology. 1997; 136: 217-221

18. Doria A.Biasinutto C, Ghiradello A, Sartori E, Rondinone R, Piccoli A et al. photo sensitivity in systemic lupus erythematosus "laboratory testing of ARA/ACR definition". lupus.1996; 5: 2