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Cutaneous manifestations of Systemic Lupus Erythematosus

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Abstract Background: *SLE is a heterogeneous autoimmune connective tissue disease marked by diverse patterns of presentation, auto-antibody production with multi-organ involvement.* ***Aims & Objectives****: The study is aimed to evaluate the different patterns of skin manifestations in SLE.* ***Method****: 20 patients during a period of 8 months were enrolled for the study. A detailed history and examination findings were noted in a predesigned proforma. Haematological Investigations, urine analysis, antibody profile were done in all cases, HPE & DIF in few as per the requirement.* ***Result****: Out of 20 patients, 18 were females and 2 were males. The average age of presentation is 27, with youngest patient 9 yrs and oldest patient 50 yrs. Skin changes noted were as follows: LE specific lesions: malar rash in 12 patients, photosensitive dermatitis in 10 patients, generalized maculopapular rash in 2 patients, discoid rash in 4 patients. The LE non-specific lesions were non-scarring alopecia in 14 patients, scarring alopecia in 3 patients, mucosal ulcers including oral and genital in 11 patients, vasculitic lesions in 7 patients, bullous lesions in 1 patient, erythema multiforme lesions in 1 patient, and nail dystrophy in 3 patients, xerosis in 4 patients, cheilitis in 7 patients, peripheral gangrene in 2 patients.* ***Discussion & conclusion****: There is a great diversity in presentation of SLE both systemically and in cutaneous manifestations. Therefore, an open mind is required when diagnosing SLE cases and prompt treatment done to prevent the morbidities.*

Keywords: *cutaneous manifestations, systemic lupus erythematosus*

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I. Introduction

Lupus erythematosus is a group of heterogenous illnesses that have in common the development of immunity to self nucleic acids and their associated proteins, with skin only disease at one end of the spectrum and severe visceral involvement at the other[1]. Skin disease is the second most frequent manifestation of LE after joint inflammation. Cutaneous lesions are a sourse of disability and, on many occasions, an indicator of internal disease[2]. Lupus erythematosus (LE) is usually divided into two main types: discoid LE (DLE) and systemic LE (SLE), with a third group, subacute cutaneous LE (SCLE)[3]. The first classification criteria were developed by the ARA in 1971 and were modified in 1982. The Systemic Lupus International Collaborating Clinics (SLICC) in 2012 identified 17 criteria that resulted in greater sensitivity but lesser specificity for diagnosis of SLE than ACR criteria. According to SLICC, for the diagnosis of SLE, a patient must satisfy at least four criteria, including atleast one clinical criterion and one immunological criterion or the patient must have biopsy proven lupus nephritis in the presence of ANAs or anti-dsDNA antibodies.

Skin lesions may be specific, with interface dermatitis in histopathology or nonspecific and seen in other conditions as well. LE-specific lesions are malar rash, SCLE like lesions, DLE and its variants. LE non-specific lesions are Cutaneous vasculitis, photosensitivity, Raynauds phenomenon, alopecia, livedo reticularis, sclerodactyly, anetoderma, leg ulcers, papulonodular mucinosis, calcinosis cutis, LE non specific bullous lesions, rheumatoid nodules, mucosal erosions, nail dystrophy, cheilitis, erythema multiforme, periungual telangiectasia, thrombophlebitis, erythromelalgia, urticaria, acanthosis nigricans (type B insulin resistance).

II. Materials & Methods

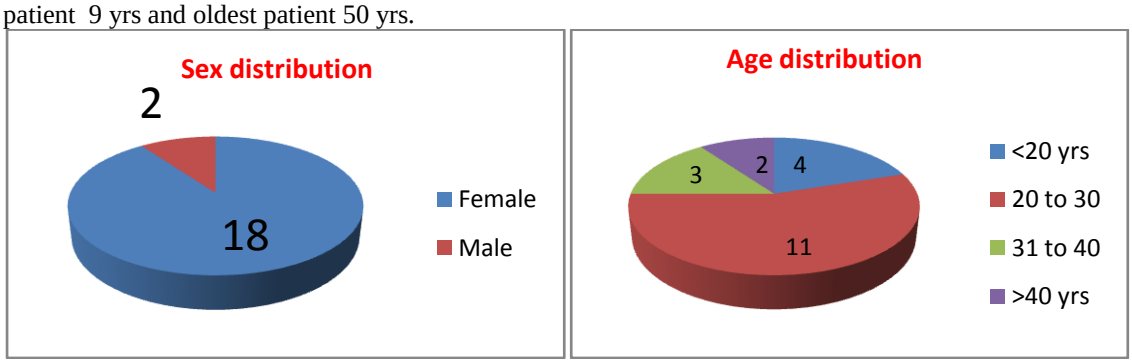
Study design**:** Prospective observational study. *Study period*: January 2017 - September 2017. *Study place*: Dept of DVL, GMC, Guntur. *Ethics committee approval*: Taken. 20 patients diagnosed on the basis of **SLICC** Criteria were enrolled for the study. Informed consent was taken. A detailed history and examination findings were noted in a predesigned proforma. Haematological Investigations, urine analysis (proteins), antibody profile were done in all cases, HPE & DIF in few as per the requirement.

Table 1. SLICC criteria, 2012 (Rooks textbook of dermatology)

Clinical criteria	Definition
1. Acute cutaneous lupus	Including: lupus malar rash (do not include if malar discoid), bullous lupus, toxic epidermal necrolysis variant of SLE, maculopapular rash, photosensitive lupus rash in the absence of dermatomyositis; *or* subacutue cutaneous lupus
2. Chronic cutaneous lupus	Including: classic discoid rash, hypertrophic (verrucous) lupus, lupus panniculitis (profundus), mucosal lupus, lupus erythematosus tumidus, chilblain lupus, discoid lupus/lichen planus overlap
3. Oral ulcers	Palate, buccal, tongue or nasal ulcers in the absence of other causes
4. Non-scarring alopecia	Diffuse thinning or hair fragility with broken hairs in the absence of other causes
5. Synovitis	Involving two or more joints characterized by effusion or swelling *or* tenderness in two or more joints and at least 30 min of morning stiffness
6. Serositis: pleurisy or pericarditis	More that 1-day duration of pleural/pericaridal effusions or pleural/pericardial rub
7. Renal disorder: persistent proteinuria (>0.5 µg/day) or cellular casts	
8. Neurological disorder	Seizures, psychosis, mononeuritis multiplex, myelitis or acute confusional state in the absence of other causes
9. Haemolytic anaemia	
10. Leukopenia (<4000/ mm^3 at least once) *or* lymphopenia (<1000/mm^3)	
11. Thrombocytopenia (<100 000/mm^3 at least once)	
Immunological criteria	
1. ANA above reference laboratory range	
2. Anti-dsDNA antibody above reference laboratory range (or more than twofold the reference range if tested by ELISA)	
3. Anti-Sm: presence of antibody to Sm nuclear antigen	
4. Antiphospholipid antibody positivity	
5. Low complement (low C3, C4 or CH50)	
6. Direct Coombs' test in the absence of haemolytic anaemia	

III. Result

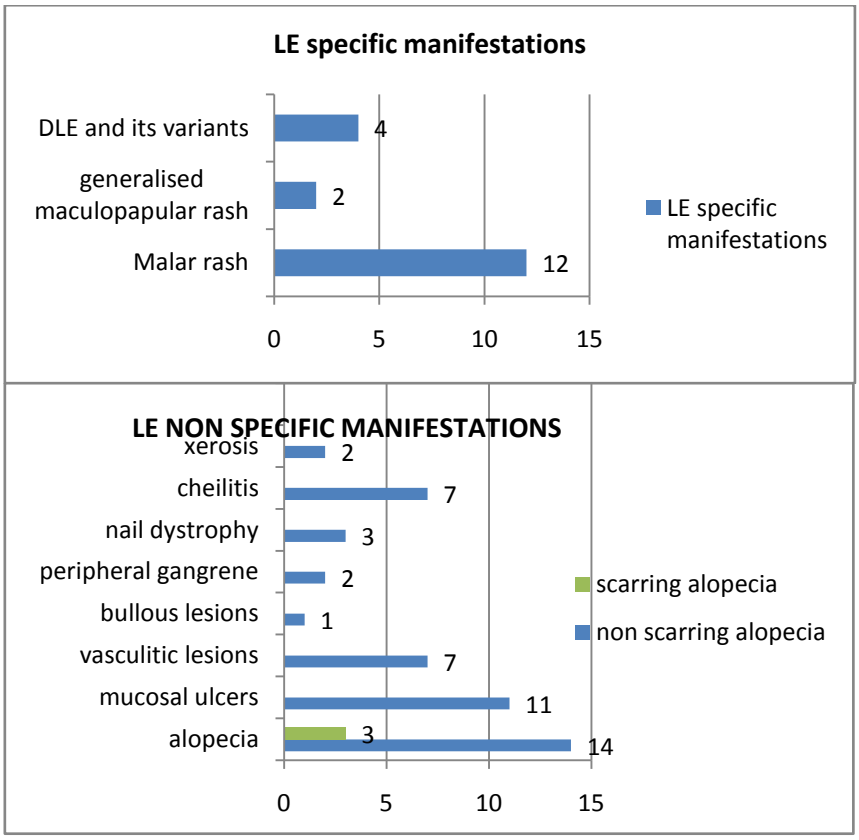
Out of 20 patients, 18 were females and 2 were males. The average age of presentation is 27, with youngest patient 9 yrs and oldest patient 50 yrs.



Skin changes noted were as follows:

LE specific lesions: malar rash in 12 patients, photosensitive dermatitis in 10 patients, generalized maculopapular rash in 2 patients, discoid rash in 4 patients.

The LE non-specific lesions were non-scarring alopecia in 14 patients, scarring alopecia in 3 patients, mucosal ulcers including oral and genital in 11 patients, vasculitic lesions in 7 patients including erythema multiforme lesions in 1 patient, bullous lesions in 1 patient, and nail dystrophy in 3 patients, nail fold necrosis in one patient, black pigmentation of nails in 5 patients, ridging of toe nail in 1 patient. xerosis in 4 patients, cheilitis in 7 patients, peripheral gangrene in 2 patients.



Systemic manifestations:
Various systemic manifestations observed in our patients were as shown in table 2

Table 2. Systemic manifestations

Systemic Manifestations	Number	Percentage
Polyarthritis	15	75%
Hematological abnormalities	10	50%
Nephritis	8	40%
Hepatomegaly	5	25%
H/o Spontaneous abortions	3	15%
Psychosis	2	10%
Serositis	2	10%
LV diastolic dysfunction	1	5%
Hemiplegia	1	5%
Retinopathy	1	5%

Antibody profile :
All patients were positive for ANA. Anti dsDNA was positive in 12 patients and Antiphospholipid antibodies were positive in 2 patients. Anti sm was positive in 5 patients.
Lupus nephritis was confirmed by Renal biopsy in 3 patients.

IV. Discussion

The ratio of female to male is 9:1 which is lower compared to study by Kole AK, Ghosh A[4], which was 14:1 and nearly equal to malaviya et al[5] which was 8:1. The average age of presentation is 27, similar to all previous studies which showed a peak in 3rd decade.

Comparison of incidence of cutaneous manifestations

Manifestation	**Our study n=20**	**Kole AK, Ghosh A n=150**	**Maheswari et al [6] n=110**
	LE specific		
Malar rash	12(60%)	120(80%)	73(66.36%)
Photosensitive dermatitis	10(50%)	75(50%)	NA
Generalized maculopapular rash	2(10%)	40(26.67%)	NA
SCLE like lesions	-	5(3.34%)	NA
DLE and its variants	4(20%)	30(20%)	22(20%)
	LE non specific		
Alopecia	17(85%)	130(86.67%)	95(86.36%)
Mucosal ulcers	11(55%)	85(56.67%)	77(70%)
Vasculitic lesions	7(35%)	50(33.34%)	29(26.36%)
Bullous lesions	1(5%)	15(10%)	NA
Erythema multiforme	1(5%)	10(6.67%)	7(6.36%)
Nail dystrophy	3(15%)	NA	18(16.36%)
Xerosis	4(20%)	NA	NA
Cheilitis	7(35%)	NA	NA
Peripheral gangrene	2(10%)	NA	4(3.36%)

1.Malar rash: Malar rash is characterised by an erythematous rash over the cheeks and nasal bridge typically sparing the nasolabial folds[7]. It was the most commonly encountered LE specific lesion, and was seen in 12 patients . It was associated with acute flare ups. It was observed to be the only skin manifestation in one patient. 8 patients had no present or past history of malar rash. It was observed in 60% patients in our study, while it was seen in 80% patients in a study conducted by Kole AK, Ghosh A.

2.DLE lesions: DLE lesions are disc shaped erythematous plaques with follicular hyperkeratosis. It may lead to permanent scarring, scarring alopecia and dyspigmentation later. They were seen in 4 patients. The female to male ratio was observed to be 3:1. It was localised to face and scalp in 2 patients and disseminated i.e., seen above and below neck in 2 patients. It was associated with scarring alopecia in 3 patients. Lesions of discoid lupus was 20% in this study is more or less same as the studies previously conducted by Kapadia [8]and Wysenbeek [9].

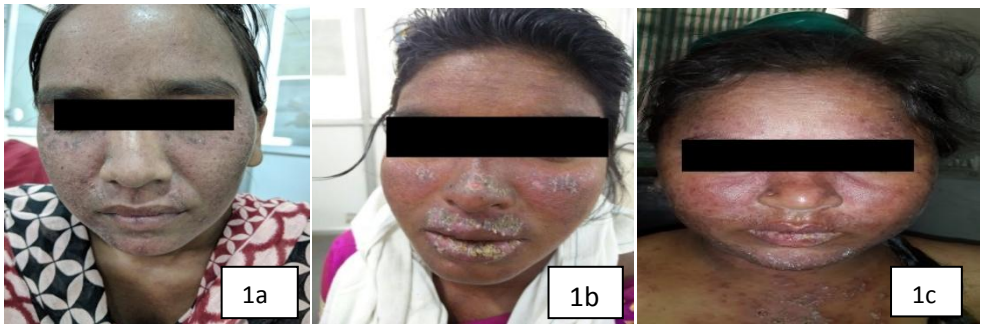


Figure 1a. Malar rash as the only skin manifestation; 1b. Malar rash and cheilitis; 1c. Typical malar rash

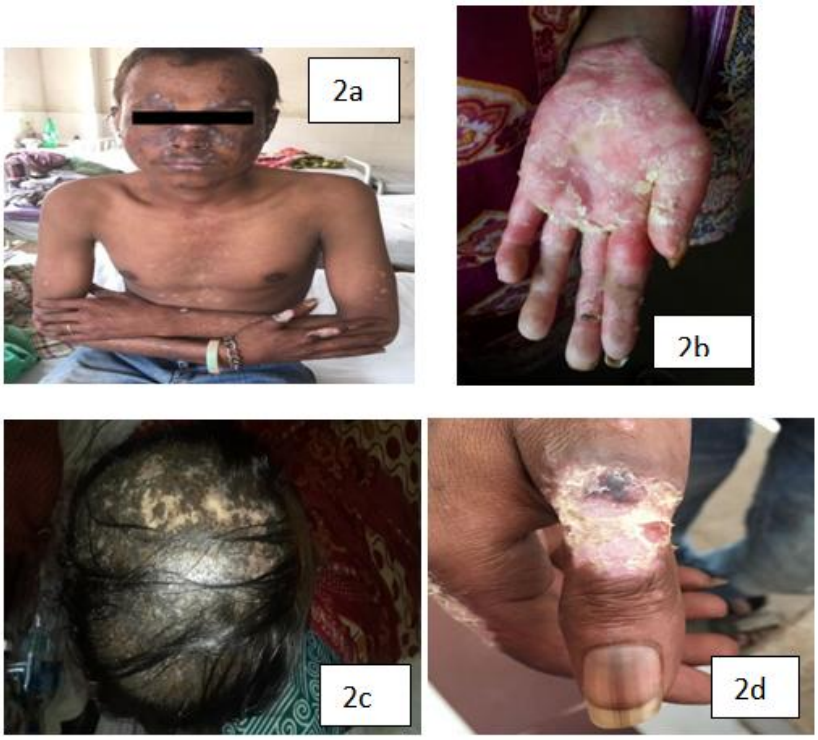


Figure 2a. DLE lesions on face, ears, chest, arms, forearms and finger; 4b. DLE lesions on palms; 4c. DLE lesion on scalp with scarring alopecia; 4d. DLE lesion on dorsum of finger

3.Generalised maculopapular rash: Generalised maculopapular rash was seen in 2 patients and It was associated with severe disease activity. Diffuse maculopapular rash was noted in 10% of the cases compared to 26.67% in Kole AK and 59% of the cases as reported by Wysenbeek, *et al.*

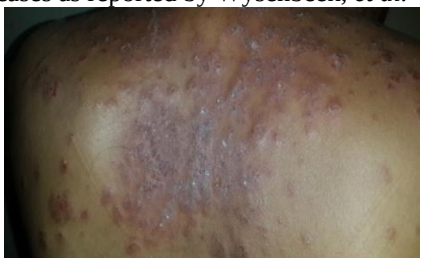


Figure 3. Maculopapular rash seen on back of trunk

4.Bullous lesions: Bullous lesions are rare blistering conditions occurring in less than 5% of patients with SLE in isolation or in combination with other skin lesions. Generalised bullous eruption was seen in 1 patient. Associated with oral and genital mucosal erosions and tender erythematous macules on palms and soles.

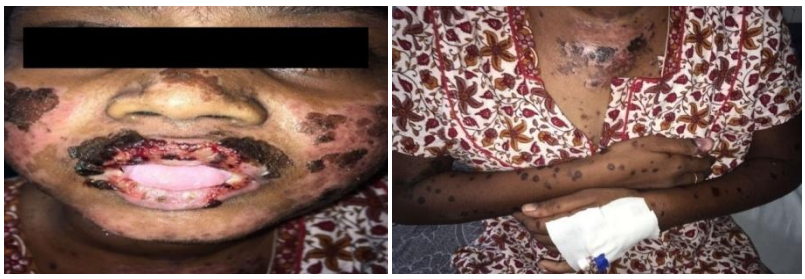


Figure 4. Crusted erosions following bullous lesions

5.Vasculitis and vasculopathy: Raynauds phenomenon is a commonly seen vasculopathy seen in SLE with systemic involvement or with U1RNP antibody positivity. It was seen in 3 patients, followed by peripheral gangrene in 2 of them. It was seen in 15% patients which is similar to 15.5% patients seen in Vaidya et al[10] and lesser compared to 32% in Malaviya et al .

Small vessel vasculitis is commonly seen in SLE as leucocytoclastic vasculitis or palpable purpura in lower limbs. Generalised vasculitic lesions were seen in 2 patients while vasculitic lesions localised to palms are seen in 4 patients. Erythema multiforme lesions were seen in one patient localised to palms and soles as the only cutaneous feature along with biopsy proven lupus nephritis. Other vasculitic features like livedo reticularis, urticarial vasculitis were not seen.

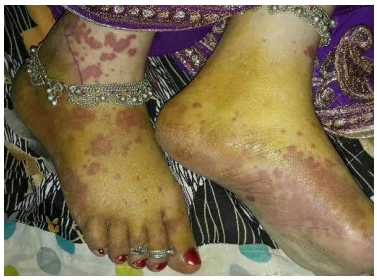
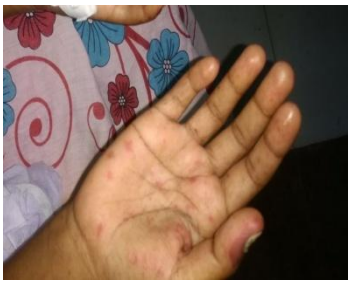



Figure5. Leucocytoclastic vasculitis Figure6. Erythematous macules and vasculitic lesions on hand

6.Peripheral gangrene: Peripheral gangrene is a rare manifestation of SLE. It was seen in 2 patients, was associated with raynauds phenomenon. It had led to amputation of all four extremities in one patient leading to severe morbidity. The other patient who was admitted in view of hemiplegia was promptly started on immunosuppressive therapy and good response was observed[11].

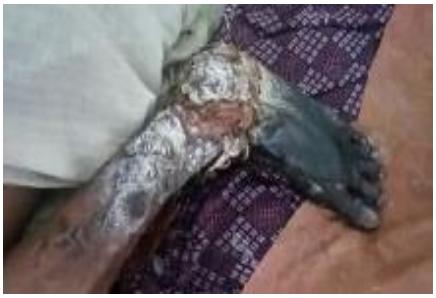
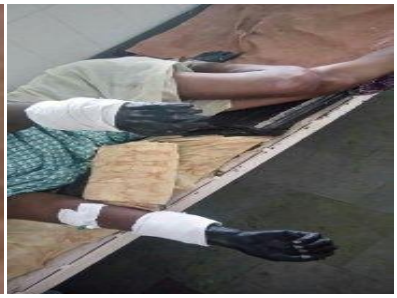



Figure 5. Severe peripheral gangrene of all extremities

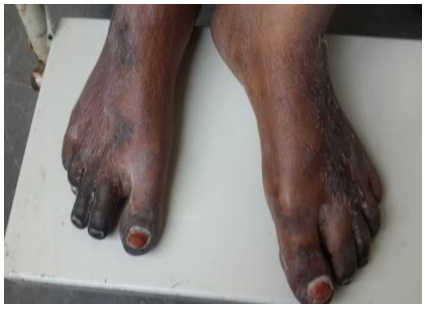



Figure 6. Peripheral gangrene of tip of finger and feet

7.Alopecia: Alopecia was the most common LE non specific manifestation. Diffuse non scarring alopecia was present in 14 patients (70%) compared to 86.6% patients in study by Kole AK, Ghosh A. Scarring alopecia as a result of DLE lesions was seen in 3 patients (15%) while it was only seen in 6.67% cases in Kole AK, Ghosh A.

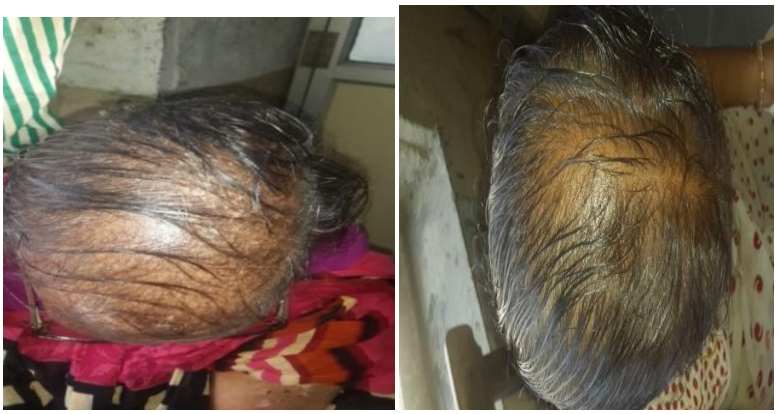


Figure 7. Non scarring alopecia- lupus hair

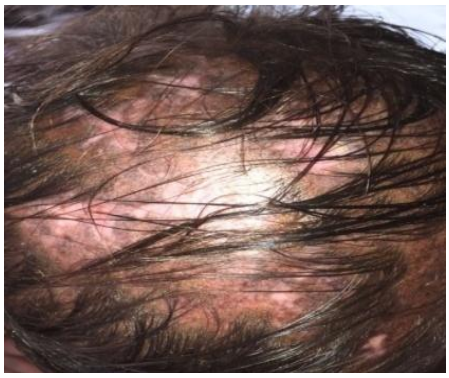


Figure 8 Scarring alopecia

8.Mucosal erosions: Palatal ulcers are most specific for SLE. Oral erosions were seen in 9 patients involving the buccal mucosa and hard palate. Genital erosions were seen in 2. Oral erosions were painful in 4 patients and were painless in 5 patients. Associated with disease activity. Mucous membrane lesions were seen in 55% patients similar to 56.67% in Kole AK,GhoshA and less than 70% as reported by maheswari et al.

9.**Nail changes:** Brown black pigmentation was seen in five patients it may be due to deposition of melanin or secondary to anti malarial drug therapy. Nail dystrophy was seen in 3 patients (15%) similar to 16.6% in a study by maheswari et al. Loss of nail following bullous lesion was seen in 1 patient. Nail fold necrosis was seen in 1 patient. Ridging of toe nail seen in 1 patient.

One patient presented with cheilitis, erythematous macules on palms and soles. Other non specific lesions like sclerodactyly, anetoderma, papulonodular mucinosis, calcinosis cutis, rheumatoid nodules, thrombophlebitis, erythromelalgia, were not encountered.

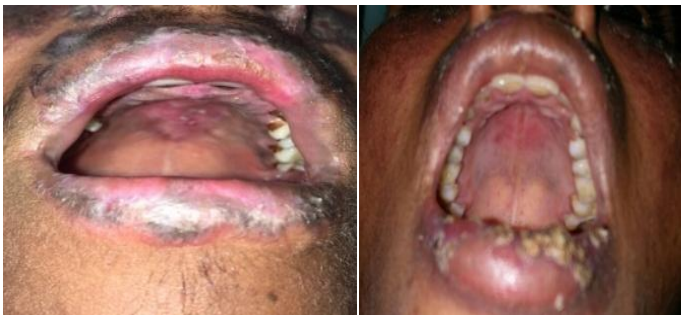


Figure 9. Palatal ulcers and crusted lips

Figure12a. loss of nail following bullous lesion; 12b. Disruption of cuticle; 12c. Black pigmentation of nails; 12d. Nail fold necrosis; 12e. Ridging of toe nail

V. Conclusion

There is a great diversity in presentation of SLE both systemically and in cutaneous manifestations making the diagnosis difficult. Therefore, an open mind is required when diagnosing and prompt treatment done to prevent the morbidities.

Conflict of interest: nil
Support: nil

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