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Case Report

Cutaneous Manifestations in Systemic Lupus Erythematosus: A Case Series

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ABSTRACT

Lupus erythematosus (LE) is an autoimmune disorder with diverse clinical manifestation ranging from mild cutaneous disorder to life-threatening systemic illness. In some patients, it remains to persist in the skin-limited form while in others it evolves into systemic lupus erythematosus (SLE). Here comes the role of identifying the markers of systemic involvement, which could determine the course and prognosis of the disease. Hence, we report a case series of two female patients who first presented only with cutaneous manifestations. Skin lesions in patients with SLE are important disease manifestations and proper understanding is essential for diagnosis and efficient management.

KEYWORDS: Lupus Erythematosus, Cutaneous Lesions

INTRODUCTION

Lupus erythematosus is not just a cosmetic deformity; causing psychological upset due to the disfigurement arising thereof, but at times can be catastrophic and can damage various vital organ systems leading to perpetuating organ dysfunction and/or failure and subsequent death.

It is the dermatologists who primarily manage the cutaneous LE (CLE); on the other hand systemic LE (SLE) remains the domain of rheumatologists or internists. It is important to realize that a person with CLE will die not of the cutaneous lesion but of the systemic involvement. There comes the importance of bridging the gap between dermatologists and internists. [1]

The prevalence rate of SLE vary within 17-48/10,000 population worldwide and CLE is thought to be 2-3 times more frequent than SLE itself. It is note-worthy that cutaneous illness often precedes the systemic involvement, giving the opportunity to the dermatologist to recognize the disease process much before the systemic complaints are expressed. This will allow institution of appropriate management strategies and timely intervention, which could prevent the subsequent morbidity and/or mortality and the social burden arising thereof. [2]

LE is further divided into three main subsets: Acute CLE (ACLE), sub acute LE (SCLE) and chronic CLE (CCLE), where classic discoid LE (DLE) is the most common form. According to Gilliam and Sontheimer, the cutaneous manifestations of LE can be divided into LE-specific and LE-non-specific skin manifestations based on histopathological findings. Acute cutaneous LE (Lupus specific) has a strong association with systemic disease and non-specific skin lesions always indicate disease activity. Therefore, a thorough understanding of the cutaneous manifestations of SLE is essential for most efficient management. [3]

CASE REPORT 1:

A 42 year old female patient first presented to medicine OPD with history of facial edema. She was treated and edema reduced. One month later she returned with c/o itching, redness and burning over face and arms. The onset was gradual, progressive with photo aggravation. There was past h/o fever, joint pain, or oral ulcers. Pt was hypertensive and on medication. On cutaneous examination, there was diffuse hyper pigmentation on the malar area and bridge of the nose with cheilitis. (Fig.1) There were multiple erythematous urticarial lesions on the extensor surface of both upper arms and forearms. (Fig. 2)

Patient's CBC showed hemolytic anaemia (Hb 10g/dl), leucopenia (< 4000/mm$^{3)}$, Urine R/E, RFT, LFT were within normal limits. ANA was positive (>1:32), VDRL negative. Skin biopsy for HPE showed thinning of epidermis with loss of rete ridges and at places, dense inflammatory infiltrate at DEJ with focal perivascular lymphocytic infiltrate and pigmentary incontinence was seen suggesting possibility of Cutaneous LE (Fig.3). Patient was diagnosed as Lupus Erythematosus according to SLICC criteria and was started on tab hydroxychloroquine 200 mg BD, vitamin C and topical mid-potent corticosteroid and strict photo protection was advised.

CASE REPORT 2:

A 15-year-old female presented to skin outpatient department with complaints of burning sensation and redness over face, trunk and extremities since two months followed by multiple raised lesions on arms, legs and other exposed areas since one month. The onset of lesions was gradual and progressive in nature which got aggravated on sun exposure. Patient also gave history of episodic fever, facial edema with oral ulcers present on buccal mucosa and pain in knee and elbow joints (Figure 4 & 5). History of loss of appetite and weight loss was present .Patient was diagnosed with SLE according to the SLICC criteria and started on oral prednisolone 0.5mg/kg body weight along with cyclophosphamide which was continued for 3months along with topical corticosteroids and photo protective measures were advised.

DISCUSSION

SLE is a chronic, occasionally life-threatening, multi-system disorder. Patients suffer from a wide array of symptoms and have variable prognosis that depends upon the severity and type of organ(s) involved. Females are more commonly affected in SLE. Women exposed to estrogen-containing oral contraceptives or hormone replacement has an increased risk of developing SLE. Peak incidence is typically seen mid-adulthood, but initial onset occurs later in affected men. [3] The disease is caused by a complex interplay of genetics, environmental factors, hormones, and ethnicity. There is also a strong association between CLE disease activity and ultraviolet (UV) radiation. The prevalence of SLE is higher among Asians, Afro-Americans, Afro-Caribbeans and Hispanic Americans compared with Americans of European descent in the United States and among Asian Indians, compared with Caucasians, in Great Britain. [4]

Cutaneous features of LE are classified as specific or nonspecific manifestations. Specific cutaneous lupus can be further classified into acute cutaneous LE (ACLE), SCLE, chronic cutaneous LE (CCLE), and bullous LE. Within CCLE are further subtypes, including discoid LE (DLE), lupus profoundus (LP), lupus tumidus (LET), and chilblain lupus. Acute cutaneous lupus erythematosus ACLE is typically confined to the face with malar erythema but can extend to a generalized distribution. The classical malar rash manifests as symmetrical erythematous plaques across the malar eminences and nasal bridge, with sparing of the nasolabial folds. Sub acute cutaneous lupus erythematosus SCLE is characterized by a nonscarring photosensitive eruption with psoriasiform or annular lesions on the face, V area of the neck, or extensor surfaces of the arms and upper back. Chronic cutaneous lupus erythematosus - DLE is the most common subtype of CCLE and presents with characteristic indurated discoid lesions with an overlying scale, predominantly on the face and scalp along with scarring alopecia. Nonspecific manifestations are secondary to the LE disease. These include vascular lesions, such as leucocytoclastic vasculitis, Raynaud's, and livedo reticularis. Nail changes, such as nailfold erythema, telangiectasia, clubbing, paronychia, pitting, leukonychia striata, and oncholysis, can be seen in all variants of CLE as well as SLE. [4,5]

DIAGNOSIS

The SLICC criteria for SLE classification requires: 1) Fulfillment of at least four criteria, with at least one clinical criterion AND one immunologic criterion OR 2) Lupus nephritis as the sole clinical criterion in the presence of ANA or anti-dsDNA antibodies.

Clinical Criteria

1. Acute cutaneous lupus
2. Chronic cutaneous lupus
3. Oral ulcers: palate
4. Non-scarring alopecia (diffuse thinning or hair fragility with visible broken hairs)
5. Synovitis (involving two or more joints, characterized by swelling or effusion OR tenderness in two or more joints and thirty minutes or more of morning stiffness)
6. Serositis (Pleural or Pericardial Effusion)
7. Renal (Proteinuria >0.5 gm / 24 hr)
8. Neurologic (Delirium, Psychosis, Seizure)
9. Hemolytic anemia (< 10 gm / dl)
10. Leukopenia (< 4000/mm3 at least once)
11. Thrombocytopenia (<100,000/mm3) at least once

Immunological Criteria

1. ANA above laboratory reference range
2. Anti-dsDNA above laboratory reference range, except ELISA: twice above laboratory
3. Anti-Sm
4. Antiphospholipid antibody

5. Low complement
6. Direct Coombs test in the absence of hemolytic anemia [4]

CLE can be managed but so far not cured. Avoidance of trigger factors is of utmost importance such as cessation of smoking and avoidance of sun exposure. The treatment is about the same for the different CLE subsets where first-line of treatment is sun-protection and local therapy with corticosteroids or calcineurin inhibitors. Antimalarials are the first choice of systemic treatment. With local therapy and antimalarial treatment about 75% of the CLE patients responds. For the refractory cases, a number of different treatments can be tried such as cyclophosphamide, metothrexate, thalidomide, mycophenolate, azathioprine and dapsone. [6]

CONCLUSION

Knowledge with regard to the pathogenesis of LE has progressed rapidly over the past decade. Given the chronic disease course of CLE, long-term treatment-related side effects must be minimized, and the introduction of new steroid-sparing agents are encouraging in this regard. It is also pivotal that each subtype of CLE be considered individually, given their variance in clinical presentation, prognosis for systemic disease, and response to treatment.

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Figure 1: Malar rash seen over cheeks and bridge of the nose with cheilitis

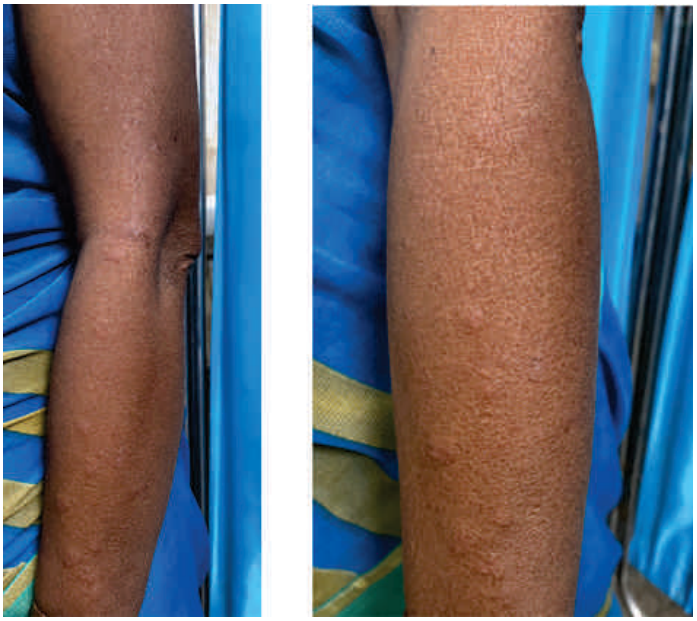


Figure 2: Urticarial plaques seen over extensor aspect of both forearms

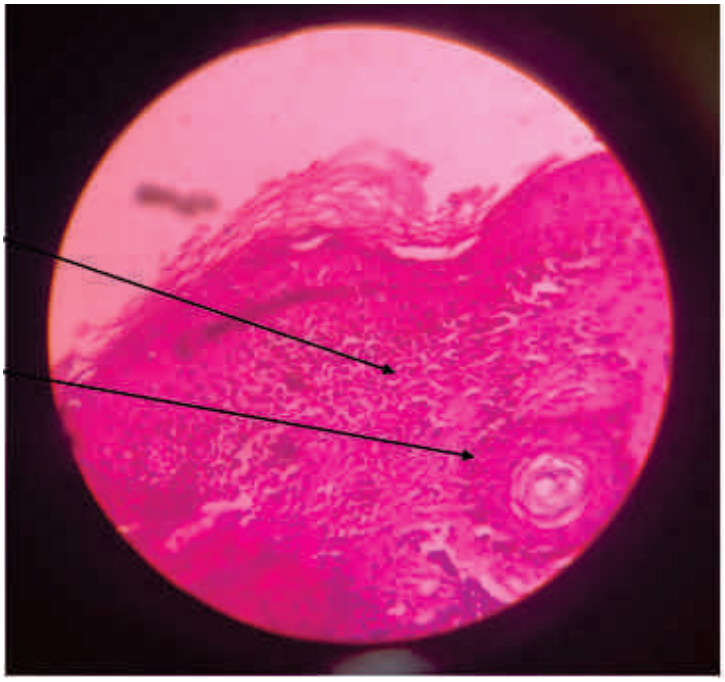


Figure 3: HPE shows thinning of epidermis with loss of rete ridges. At places, dense inflammatory infiltrate at DEJ with focal perivascular lymphocytic infiltrate and pigmentary incontinence

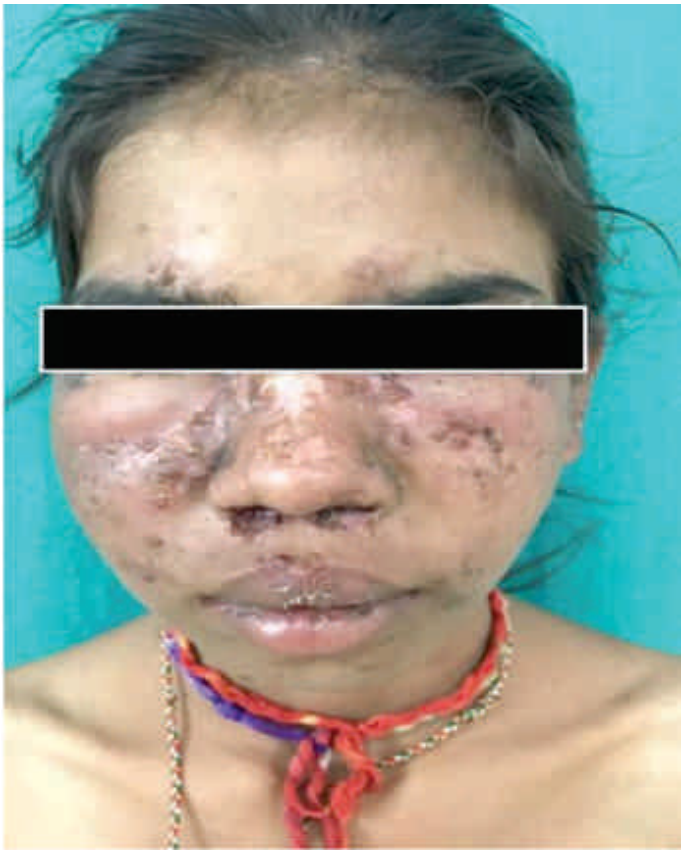
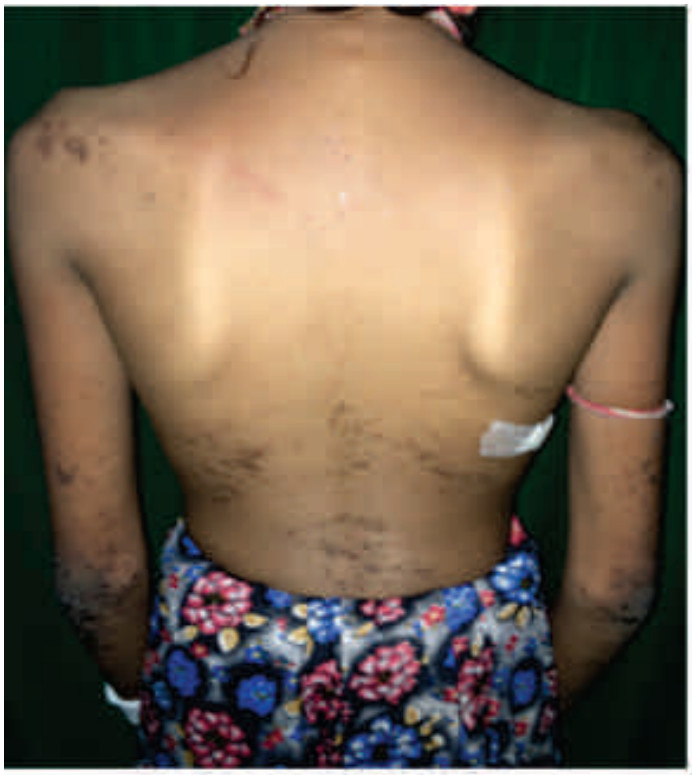


Figure 4: Edematous and erythematous crusted plaques present over cheeks, bridge of nose, eyebrows and lips



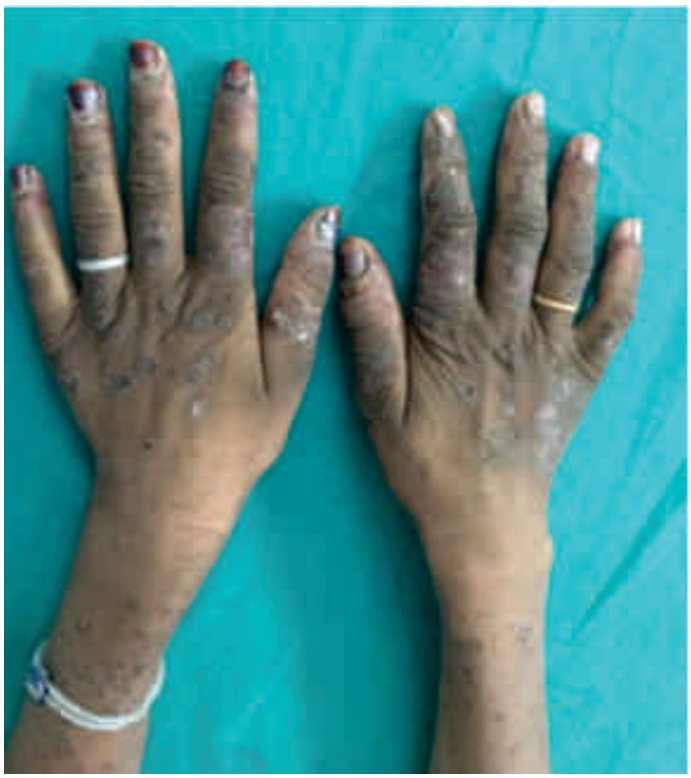


Figure 5: Hyperpigmented papules and plaques present over sun-exposed areas