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Case Series

Varied spectrum of lupus erythematosus: case series

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ABSTRACT

Autoimmune connective tissue disorders represent polygenic clinical disorders that have heterogenous and overlapping clinical features. Lupus erythematosus (LE) is a connective tissue disorder that affects multiple organ systems with increased incidence among female population compared to males (9:1 F:M ratio). Skin is 2nd most frequent organ to be affected in LE after joints. Cutaneous lesions are a source of disability and on many occasions, an indicator of internal disease and these lesions may be specific to lupus/seen in other conditions as well. Correlation between clinical, histopathology, immunofluorescence, and serologic profiles (Anti-nuclear antibody) remains crucial as no single clinical feature or lab abnormality can confirm the diagnosis of LE. Treatment consists of sun protection, topical sunscreens, systemic steroids, antimalarials, immunosuppressives and biologicals. Effective treatment should be initiated at the earliest to prevent complications. Herein we report 4 cases of LE with varied presentation.

Keywords: Autoimmune, LE, Cutaneous lesions, Joints

INTRODUCTION

Term " LE " refers to a collection of diverse illnesses that are characterized by production of antibodies against molecular components of nucleosomes and ribo-nucleoprotein. Only small percentage of patients develop life-threatening systemic signs of lupus, whereas others who share same basic underlying pathology only experience isolated skin lesions during the course of their illness. It is useful to consider of LE as a spectrum, with skin only disease at one end to those who are at risk of developing systemic manifestations of LE such nephritis, CNS illness, or vasculitis at the other end. As a result, LE can cause significant psychological stress, morbidity, financial burden and mortality.[1,2]

CASE SERIES

Case 1

A 28-year-old female presented with complaints of red raised lesions over nose with Scaling and redness over the scalp since 1 year. History of photosensitivity was present. On cutaneous examination solitary well defined erythematous to depigmented atrophic plaque surrounded by hyperpigmented border with mild scaling at the center was present over the nose (Figure 1). Crusting over the lower lip and mild scaling over scalp was noted. Histopathological examination revealed epidermis exhibiting hyperkeratosis, focal acanthosis, spongiosis, basal cell degeneration. Dermal edema and infiltration by lymphocytes at dermo epidermal junction and perivascular inflammatory infiltrate was noted. Final diagnosis of discoid LE was made.

Case 2

A 19 year old female presented with complaints of red raised lesions all over the body associated with loss of hair on scalp since 7-8 months and these lesions were associated with pain and itching over the lesions. H/o was photosensitivity present. On cutaneous examination well defined erythematous scaly indurated plaque with mild atrophy surrounded by hyperpigmented border present

over the bilateral malar region sparing nose. Multiple well defined atrophic scars of size 0.5 to 1 cm with central crusting present over the lower back (Figure 2). Solitary well defined ulcer of size 2×2 cm with central necrosis and crusting with surrounding erythema present over the lower back. Multiple depigmented atrophic scars with erythema seen over occipital area of scalp (Figure 3). Areas of Scaring alopecia present. Routine blood investigations revealed Pancytopenia, ANA profile was positive. Histopathological examination revealed epidermis exhibiting hyperkeratosis, focal parakeratosis, basal layer vacuolar degeneration. Dermal edema, perivascular peri adnexal lymphocytic infiltrate with interstitial mucin deposition was noted. Final diagnosis of disseminated discoid LE was made.

Case 3

19-year-old female presented with complaints of redness of cheeks with burning sensation on sun exposure, multiple joint pains and fever for 8 months. Also, complaints of breathlessness, difficulty in swallowing, blurring of vision and oral ulcers for 2 months. On cutaneous examination diffuse hyperpigmentation over the face sparing periorbital area, and irregular atrophic depigmented plaque with surrounding hyperpigmentation over B/L malar area was noted. Multiple well defined to ill-defined hyperpigmented patches over bilateral forearms, back (Figure 4) and atrophic scars over dorsum of hand was present. Erosions over the hard palate and lower lip were seen. Routine blood investigations revealed pancytopenia, ANA profile was positive for ds DNA and urinary protein 2+ was present. In view of the above clinical, laboratory findings and patient fulfilling ARA (7)/SLICC (9) criteria, Final diagnosis of systemic LE was made.

Case 4

A 25-year-old female presented with complaints of high-grade fever, facial and periorbital puffiness since 1week. Also, complaints of breathlessness on exertion, weakness over both lower limbs, multiple joint pains, redness of cheeks with burning sensation on sun exposure in the past 2 years. On cutaneous examination diffuse erythema, edema, crusting, hyperpigmentation, hypopigmentation present all over the face sparing periorbital area, nasolabial folds, upper lip (sun protected areas) (Figure 5). Crusting of upper and lower lips was also seen. Hyperpigmentation and Scaling was present over forearms and upper back. Thin, short, Fragile hair mainly over the frontal area noted (lupus hair**).** Routine blood investigations revealed pancytopenia, ANA profile was positive for ds DNA, SS-a/Ro, U1- Sn RNP, 24 hr urinary protein was 1240 mg/mol, Renal doppler revealed grade 1 renal disease. In view of the above clinical, laboratory findings and patient fulfilling ARA (5)/SLICC (7) criteria, Final diagnosis of the mixed connective tissue disorder (systemic LE with Sjogren syndrome) was made.

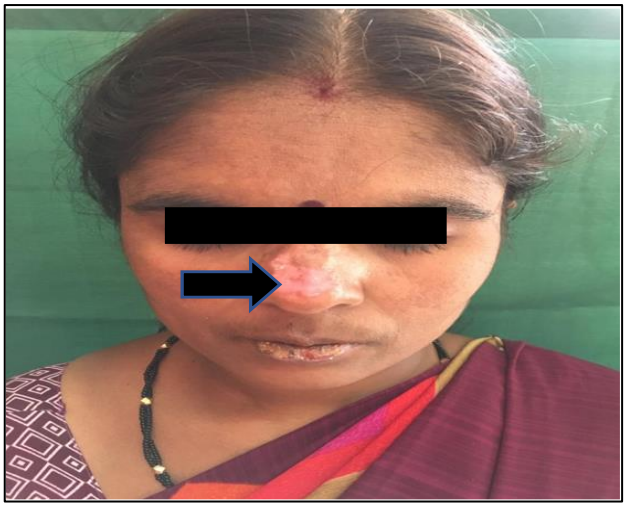

Figure 1: Depigmented atrophic plaque over the nose.

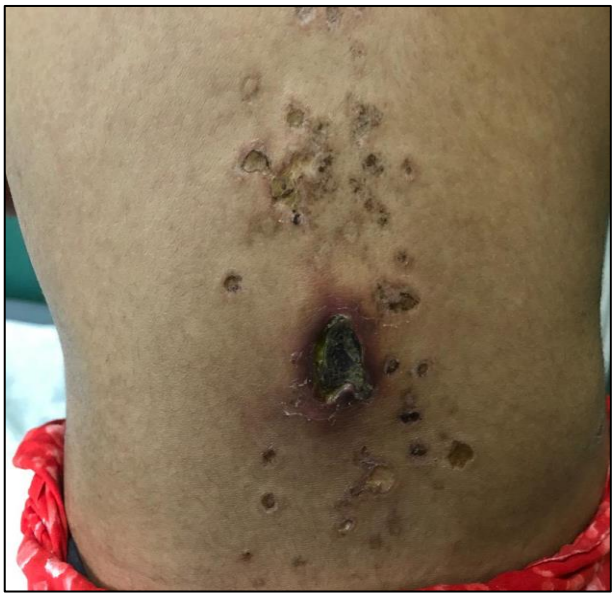

Figure 2: Atrophic scars of variable scars over lower back.

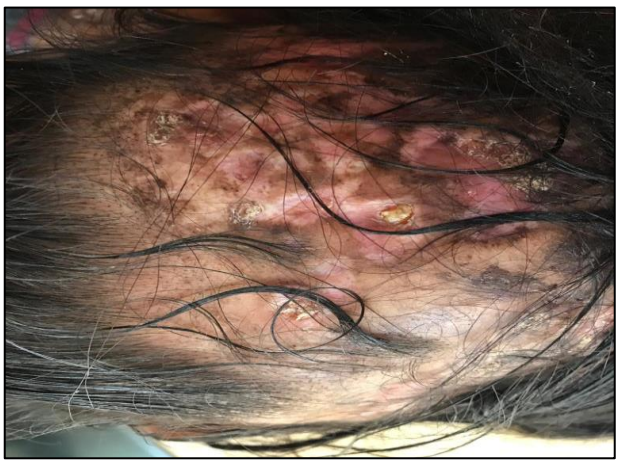

Figure 3: Multiple depigmented atrophic scars over occipital area of scalp.

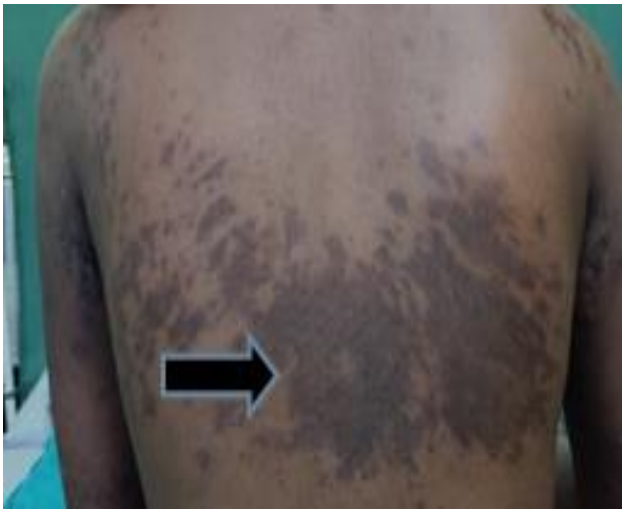

Figure 4: Well defined hyper pigmented patches over the back.

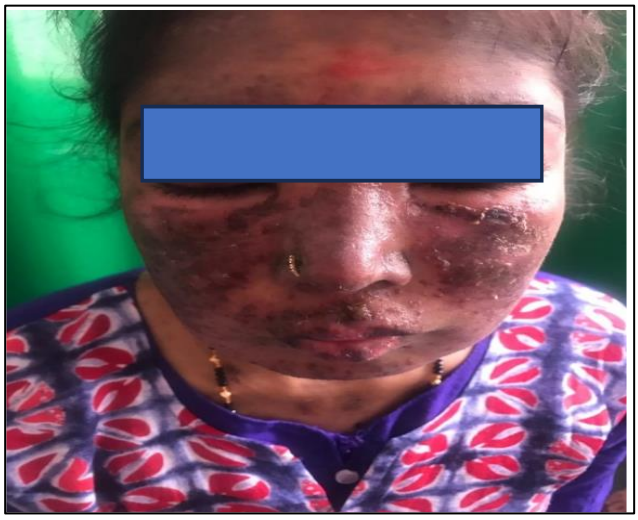

Figure 5: Erythematous to hyperpigmented plaque over the malar area.

DISCUSSION

LE is an autoimmune inflammatory disease that can be associated with severe systemic organ damage (systemic LE (SLE) or affect only skin cutaneous LE, (CLE). Four cutaneous LE subtypes can be defined based on clinical features, histological changes, and serological abnormalities, these are: (1) acute cutaneous LE (ACLE), (2) subacute cutaneous LE (SCLE) and (3) chronic cutaneous LE (CCLE), which includes discoid LE (DLE), LE panniculitis (LEP), and chilblain LE (CHLE).[3]

The pathogenesis of CLE is multifactorial and involves genetic predisposition, environmental factors and abnormalities of innate and adaptive immune responses.[4] The cutaneous signs associated with LE have diverse manifestations, leading to a broad classification into LE-specific and non-specific skin diseases, based on dermatologic features and analysis. histology. Skin signs specific to LE include acute CLE (ACLE), subacute CLE (SCLE), and chronic CLE (CCLE). Nonspecific cutaneous manifestations of LE include mucosal ulceration, non-scarring alopecia, cutaneous vasculitis, Raynaud's phenomenon, etc. It is important to examine for LE-nonspecific cutaneous manifestations in CLE patients, as they are indicators of internal lupus with high disease activity.[5-7] Currently, the diagnosis of CLE relies on the criteria for the classification of SLE established by the American college of rheumatology (ACR). The ACR guidelines require 4 out of 11 criteria to be met for a diagnosis of SLE.[8] the Systemic Lupus International Collaborating Clinics Classification Criteria (SLICC) was proposed as an updated method for diagnosing SLE, including the revised dermatologic criteria of ACLE, CCLE, oral ulcers, and nonscarring alopecia.[9] There are numerous treatment options available for CLE. Topical corticosteroids are the accepted first line therapy for all subtypes of the disease, but their use is limited by well-known side-effects, such as atrophy and telangiectasia. A safe and effective alternative topical treatment for CLE is the calcineurin inhibitors tacrolimus and pimecrolimus. Antimalarials, such as HCQ or CQ, are the first-line systemic treatment for disfiguring and widespread skin lesions and for the prevention of systemic disease. Systemic corticosteroids can be used in patients with acute and severe skin lesions, but should be time-limited due to the well-known side-effects. Further second line treatment options include MTX, retinoids and dapsone; MMF and MPA are third-line treatment options. Biologicals, such as belimumab or sirukumab, are promising new therapeutic options, but their efficacy and safety in the treatment of patients with CLE still needs to be evaluated in clinical trials.[10]

CONCLUSION

The above case series highlights the heterogeneity in presentation of LE and the role played by dermatologists in making a correct diagnosis and providing appropriate therapy.

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