

Spectrum of Cutaneous Lupus Erythematosus – A Case Series

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ABSTRACT

Introduction: Lupus erythematosus (LE) is auto-immune, inflammatory disease characterized by multi-organ involvement with skin being second most commonly involved. Skin lesions in LE are classified into LE specific and LE non-specific lesions. LE Specific lesions are further divided into acute cutaneous LE (ACLE), Subacute cutaneous LE (SCLE) and Chronic cutaneous LE (CCLE).

Case series: We describe 4 cases of Lupus erythematosus presented to department of dermatology in one year to our institute. Mean age of onset of disease was 21 years and all four cases were females. Out of 4 cases, 2 cases were discoid LE and 2 cases were ACLE with systemic involvement. Non-specific skin lesions like Raynaud's phenomenon and diffuse non scarring alopecia with lupus hair were also seen in case 3 and case 4 respectively. All patients underwent skin biopsy with DIF and confirmed for LE. Various combination of treatment options was instituted depending on individual case basis.

Conclusion: Cutaneous manifestation of LE is very frequently observed with spectrum of disease varying from minor localized skin lesion to life threatening disease. Early diagnosis and prompt treatment will give good result.

Keywords: Cutaneous Lupus Erythematosus, Specific Skin Lesions, Non-Specific Skin Lesions, Discoid Lupus Erythematosus, Systemic Lupus Erythematosus

INTRODUCTION

Lupus erythematosus (LE) is auto-immune, inflammatory disease characterized by multi-organ involvement with skin being frequently involved i.e., second most common. Skin is involved in 70-85% of the patients with lupus erythematosus.¹ Skin lesions in LE are classified into LE specific and LE non-specific lesions by Gilliam and Sontheimer.² LE Specific lesions are further divided into acute cutaneous LE (ACLE), Subacute cutaneous LE (SCLE) and Chronic cutaneous LE (CCLE). ACLE has localized and generalized forms, common variants of SCLE are annular and papulosquamous forms whereas most common variant of CCLE is Discoid LE which is further divided into localized and disseminated form. Other variants of CCLE are LE profundus (LEP) and Chilblain LE (CLE). LE tumidus (LET) has also been described as separate entity.³ LE non-specific lesions are not specific for LE but also seen in other autoimmune diseases along with SLE. Common non-specific LE lesions are vascular in nature i.e., Leukocytoclastic vasculitis (Palpable purpura, urticarial vasculitis), livedo reticularis, periungual telangiectases, Raynaud's syndrome, thrombophlebitis, occlusive vasculopathy. Other non-specific LE lesions are non-scarring alopecia, calcinosis

cutis, papular mucinosis and erythema multiforme.³

There are various extracutaneous manifestation of Systemic Lupus Erythematosus (SLE) which involves multiple organ system i.e., Musculoskeletal, Haematological, Cardio-pulmonary, Renal, Neuropsychiatric, Gastrointestinal, Ocular, lymphatic system. American College of Rheumatology (ACR) SLE classification criteria and Systemic Lupus International Collaborating Clinics (SLICC) classification criteria are used to diagnose SLE. SLICC is the most recent one which includes clinical and immunological criteria. 4 out of 18 criteria are required to diagnose SLE as per SLICC SLE criteria.⁴ As per the treatment for SLE is concerned, there is no cure for it but it can be managed with various topical and systemic drugs. Prevention of the triggering events such as exposure to sunlight, smoking is important. Various treatment options for skin specific LE are topical steroids, topical calcineurin inhibitors, intralesional steroid injections, anti-malarial drugs such as hydroxychloroquine. Resistant cases can be treated by retinoids, methotrexate, thalidomide, mycophenolate mofetil, azathioprine and dapsone, cyclophosphamide, clofazamine. We here describe our case series of LE with varied spectrum.

CASE SERIES

We have assessed all patients presenting to dermatology department with skin lesions suggestive of LE from January 2019 to January 2020. All patients were examined by senior dermatologist and routine blood investigations, ANA profile, urine analysis, skin biopsy with direct immunofluorescence (DIF) were performed on suspected case of LE. Four cases of cutaneous LE were confirmed. Here is the description of the cases:

Case 1

A 28-year-old female presented with red raised lesions over the nose for 5 years and scaling with redness over the scalp for last one 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 over the nose were

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Figure-1: FIG 1A showing patient of localized DLE and FIG 1B showing histopathology of skin biopsy of the same patient.



Figure-3: FIG 3A and FIG 3B showing patient of ACLE with SLE



Figure-2: FIG 2A and FIG 2B showing patient of disseminated DLE

noted and crusting present over the lower lip with mild scaling over scalp (FIG 1A). ANA was positive and skin biopsy showed findings of LE in epidermis (Hyperkeratosis, focal acanthosis, spongiosis, basal cell degeneration) and dermis (edema and lymphocytic infiltration at dermo-epidermal junction with perivascular infiltrate) (FIG 1B). She was treated with broad spectrum sunscreen, topical steroids and systemic treatment with hydroxychloroquine, clofazimine and oral steroids.

Case 2:

An 18-year-old female presented with red raised lesions all over the body and loss of hair over the scalp for last 8 months. History of photosensitivity was present and had fever for last one week. On cutaneous examination, well defined erythematous, scaly, indurated plaque with mild atrophy surrounded by hyperpigmented border were presented over both malar regions sparing the nose (FIG 2A). Multiple well defined atrophic scars with central crusting were presented over lower back (FIG 2B). Solitary well defined ulcer with central necrosis ad crusting with surrounding erythema presented over the lower back (FIG 2B). Multiple depigmented atrophic scars with erythema were seen over occipital area of scalp and areas of scarring alopecia were present. Routine investigation showed anaemia with leukopenia and ANA was positive. Skin biopsy showed findings of LE in epidermis (Follicular plugging, Hyperkeratosis, basal cell



Figure-4: FIG 4A showing patient of ACLE with SLE and FIG 4B shows lupus hair of same patient

degeneration) and dermis (perivascular and peri-appendageal lymphocytic infiltration). She was treated with oral steroid, topical steroids and hydroxychloroquine.

Case 3

A 19-year-old female presented with high grade fever, multiple joint pain and photosensitivity for last 8 months. Had history of breathlessness, difficulty in swallowing, blurring of vision and oral ulcers for last 2 months. Raynaud's phenomenon was positive and on cutaneous examination diffuse hyperpigmentation over the face, ear pinna and neck sparing periorbital region. Irregular atrophic depigmented plaque with surrounding hyperpigmentation over bilateral malar area (FIG 3A). Multiple well defined to ill - defined hyperpigmented patches present over bilateral forearms (FIG 3B). Atrophic scars present over dorsum of the hand and erosions present over hard and soft palate. She also had multiple hyperpigmented patches of varying sizes over back and lower limb. Blood investigation showed anaemia, leukopenia and thrombocytopenia, urine analysis showed albumin 2+, ANA positive for Anti ds DNA Anti RNP autoantibodies. Skin biopsy was consistent with lupus erythematosus. She was treated with topical steroids and systemic treatment with intravenous steroid pulse, oral steroids, cephalosporins and hydroxychloroquine.

Case 4

A 25-year-old female presented with photosensitivity for last 2 years and multiple joint pain for last 6 months. Had history of breathlessness and fever for last one week. Known case of hypothyroidism, restrictive lung disease

with severe pulmonary artery hypertension on treatment. On cutaneous examination diffuse erythema, edema crusting, scaling and hyperpigmented patch noted all over the face, sparing periorbital region, nasolabial fold and upper & lower lip (FIG 4A). Hyperpigmentation and scaling present over forearms and upper back. Hair was found to be thin, short, fragile predominantly over frontal area (FIG 4B). Blood investigation showed anaemia, leukopenia and thrombocytopenia, 24 urine protein was high, ANA positive for Anti ds DNA, SS-a/Ro, U1-Sn RNP autoantibodies. Skin biopsy again was suggestive of SLE. She was treated with methyl prednisolone pulse therapy, Cephalosporins, hydroxychloroquine, topical steroids and broad-spectrum sunscreens. Unfortunately, this patient succumbed due to cardiopulmonary complications.

DISCUSSION

SLE is a disease which is autoimmune in nature producing various auto-antibodies, which affects own body leading to multi-organ involvement. The spectrum of this disease varies from minor cutaneous lesions to life threatening multiorgan involvement.⁵ Cutaneous manifestation commonly occurs in SLE and it can occur at any stage of disease, sometimes it's the first sign of disease.³ Skin lesions in LE produce significant problems due to scarring, disfigurement, alopecia etc which leads to major disability.⁶ Mean age of onset in our case series is 21 years and mean age of onset in our two cases SLE is 21 years i.e., 3rd decade, which is similar to Indian study by Malaviya et al.⁷ All our patients were females and females are affected more than males with different studies quoting different female to male ratio. Indian study on SLE by Malaviya et al quoted female to male ratio of 8:1.⁷ DLE is the most common form of CCLE and localized DLE (60-80%) occurs much more frequently than generalized DLE (20-40%).⁸ In our case series we have two cases of DLE, out of which one is localized form (Case 1) and another is generalized form (Case 2). ACLE is almost always associated with systemic disease and localized form of ACLE is commoner than generalized ACLE. Generalized ACLE is characterized by widespread erythematous macular and papular lesions all over body, mainly over sun exposed areas and post-inflammatory hyperpigmentation without scarring is seen in chronic cases.^{9, 10} Two cases of our series had generalized ACLE with systemic involvement (Case 3 & 4) and one patient (Case 3) also had Raynaud's phenomenon. Diagnosis is by Clinical and immunological methods, ACR and SLICC criteria are used to diagnose SLE. But, for cases with only skin lesions without other signs of SLE, Skin biopsy with DIF is very valuable.⁹ All our cases underwent skin biopsy with DIF and diagnosis was confirmed with it. Various treatment options are available for CLE as described earlier and combination of treatment options are to be used appropriately depending on the type of lesion and individual patient needs. Complications are to be expected, mainly in systemic involvement which should be detected and treated promptly.

CONCLUSION

Lupus erythematosus is a chronic autoimmune disease affecting multiple organ systems. Skin is the second most organ to be affected in LE after joints. The spectrum varies from only mild localized cutaneous manifestations to life threatening SLE. Early diagnosis can be obtained by thorough clinical examination, blood investigations and immunological methods like ANA profile. Final confirmation with skin biopsy aided with direct immunofluorescence is of great help. Effective treatment should be initiated at the earliest to prevent complications.

REFERENCES

1. McCauliffe DP. Cutaneous lupus erythematosus. *Semin Cutan Med Surg* 2001; 20:14-26.
2. Gilliam JN, Sontheimer RD. Skin manifestations of SLE. *Clin Rheum Dis* 1982; 8:207-18.
3. A. Kuhn, M. Sticherling, and G. Bonsmann, "Clinical manifestations of cutaneous lupus erythematosus," *JDDG* 2007;5:1124–1137.
4. Petri M, Orbai A-M, Alarcón GS, Gordon C, Merrill JT, Fortin PR et al. Derivation and validation of the systemic lupus international collaborating clinics classification criteria for systemic lupus erythematosus. *Arthritis Rheum* 2012;64:2677–2686.
5. Kole AK, Ghosh A. Cutaneous manifestations of systemic lupus erythematosus in a tertiary referral center. *Indian J Dermatol* 2009; 54:132-136
6. O'Quinn SE, Cole J, Many H. Problems of disability in patients with chronic skin disease. *ArchDermatol* 1972; 105:35-40.
7. Malaviya AN, Singh RR, Kumar De A, Kumar A, Aradhya S. SLE in Northern India 1988;36:476-80.
8. Cardinali C, Caproni M, Bernacchi E, Amato L, Fabbri P. The spectrum of cutaneous manifestations in lupus erythematosus-the Italian experience. *Lupus* 2000; 9:417-23.
9. Ribero S, Sciascia S, Borradori L, Lipsker D. The Cutaneous Spectrum of Lupus Erythematosus. *Clin Rev Allergy Immunol*. 2017; 53:291-305.
10. Gronhagen CM, Nyberg F. Cutaneous lupus erythematosus: An update. *Indian Dermatol Online J* 2014; 5:7-13.

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