ISSN: 0975-8585

Research Journal of Pharmaceutical, Biological and Chemical Sciences

Clinicopathological Study Of Discoid Lupus Erythematosus.

T Aarthi Priya[1], S Sangeetha[2], and R Prabhakar Singh[3]*.

[1]Assistant Professor, Department of Rheumatology, Government Kilpauk Medical College, and Hospital, Chennai, Tamil Nadu, India.
[2]Senior Assistant Professor, Department of Anesthesia, Government Kilpauk Medical College and Hospital, Chennai, Tamil Nadu, India.
[3]Associate Professor in Orthopedics, Department of Arthroscopy and Sports Injury in Orthopedics, Tamil Nādu Multi-Super Specialty Government Hospital (TNGMSSH) Omandurar Estate, Chennai, Tamil Nadu, India.

ABSTRACT

Discoid lupus erythematosus is a benign disorder of the skin characterized by well-defined, erythematous atrophic plaques covered by a prominent, adherent scale that extends into the orifices of dilated hair follicles. The disease affects twice as many females as males. The salient histopathological features of classic DLE lesion are hyperkeratosis with follicular plugging, liquefaction degeneration of the basal cell layer of the epidermis, degenerative changes in the connective tissue, and patchy dermal lymphocytic infiltrate around the appendages. Immunohistology shows the presence of immunoglobulins IgG, IgA, IgM, and complement at the dermo-epidermal junction in the skin lesions. To study the relevant serological abnormalities. The study was Department of Rheumatology, Government Kilpauk Medical College, and Hospital, Chennai, Tamil Nadu, India in the year 2017-2018. 51 patients were clinically diagnosed as Discoid Lupus Erythematosus out of 30,056 patients attending Dermatology OP during the study period. A detailed history as given in the proforma was elicited. Various presenting complaints like photosensitivity, Raynaud's phenomenon, joint pain, and loss of hair were also recorded. Biopsy was done on all patients. The incidence was 4.79 per 10000 cases (51 of 106368 dermatology patients) showing a female-to-male ratio of 4.1:1. Localized type was more common than the disseminated type. Few lesions (less than five) in a localized area without head and neck involvement were also classified as localized type in this study. Mucosal, verrucous, tumid, and lupus panniculitis were the variants of DLE encountered. The sites involved were the face, scalp, trunk, and upper and lower limbs in descending order of frequency. Antinuclear antibody (ANA) was positive in 22 of 30 cases (73%). The systemic involvement was seen in 15 patients all of whom were diagnosed as systemic lupus erythematosus (SLE). Squamous cell carcinoma was seen in 2 cases of disseminated DLE. The majority of patients had disease onset in the 3rd to 5th decade showing female predominance. When compared to the localized type, the disseminated type was found more frequently in males. Early onset and severe disease were noted among offspring born to a patient suffering from disseminated DLE. Serious morbidity like lupus nephritis was observed only in 1 case. The occurrence of DLE over the herpes zoster scar was an interesting observation.

Keywords: DLE, Lupus nephritis, SLE, Squamous cell carcinoma

https://doi.org/10.33887/rjpbcs/2024.15.1.1

Corresponding author

ISSN: 0975-8585

INTRODUCTION

Discoid lupus erythematosus is a benign disorder of the skin characterized by well-defined, erythematous atrophic plaques covered by a prominent, adherent scale that extends into the orifices of dilated hair follicles. The disease affects twice as many females as males [1]. The salient histopathological features of classic DLE lesion are hyperkeratosis with follicular plugging, liquefaction degeneration of the basal cell layer of the epidermis, degenerative changes in the connective tissue, and patchy dermal lymphocytic infiltrate around the appendages [2]. Immunohistology shows the presence of immunoglobulins IgG, IgA, IgM, and complement at the dermo-epidermal junction in the skin lesions. In DLE, sections reveal the dermis contains a perivascular and peri adnexal lymph histiocytic infiltrate under an interface dermatitis [3].

The epidermal interface activity shows degeneration of the basal layer, apoptotic keratinocytes, and a marked thickening of the basement membrane (figures 2, 3). In well-established lesions, there may be marked follicular plugging and sometimes an epidermal reaction which may mimic a squamous cell carcinoma (verrucous lupus erythematosus) [4].

There is a characteristic lymph histiocytic infiltrate surrounding appendages and vessels. Deposition of dermal mucin may be impressive. The majority of researchers consider DLE to be part of a spectrum of lupus erythematosus diseases (LE) [5]. Accordingly, the clinical expression of LE varies from DLE, a benign and strictly cutaneous form, to a systemic form with an unfavorable prognosis, known as Systemic Lupus Erythematosus (SLE). The risk of developing overt SLE is only approximately 6.5%. The risk is higher in patients with Disseminated DLE (22%) than in DLE confined to the head and neck (1.2%) [6, 7].

MATERIALS AND METHODS

The study was Department of Rheumatology, Government Kilpauk Medical College, and Hospital, Chennai, Tamil Nadu, India in the year 2017-2018. 51 patients were clinically diagnosed as Discoid Lupus Erythematosus out of 30,056 patients attending Dermatology OP during the study period. There were **51** patients who were clinically diagnosed as Discoid Lupus Erythematosus out of 30,056 patients attending Dermatology OP during the study period. A detailed history as given in the proforma was elicited. Various presenting complaints like photosensitivity, Raynaud's phenomenon, joint pain, and loss of hair were also recorded. Biopsy was done on all patients. All patients were subjected to various investigations like Total count, differential count, hemoglobin, erythrocyte sedimentation rate, blood sugar, blood urea, liver function test, Antinuclear antibody titer, Rheumatoid factor, 'C' reactive proteins, and anti-double-stranded DNA antibody. Direct Immunofluorescence was done for a few patients. Opinions of Rheumatologists were obtained in patients with joint pain.

RESULTS

Of the total 30,056 patients attending Skin OP, Government General Hospital, Chennai during the period between July 2006 to September 2008, the total number of patients with Discoid Lupus Erythematosus was 51. The incidence of Discoid Lupus Erythematosus was 1.7 per 1000 which accounts for 0.17%. Youngest age at the presentation was a 16-year-old female. The oldest age at the presentation was in a 65-year-old woman. The incidence of DLE peaked between 31-50 years. DLE was more common in females (69%) than males (31%). The sex ratio between females and males was 2.3: 1 in this study. Peak incidence was between 31-40 years. Peak incidence among males was between 41-60 years & among females was between 31-50 years. Localized DLE was more common in females (77%) than in males (23%). The sex ratio between females and males was 3.5: 1 in this study.

ISSN: 0975-8585

Table 1: Localized DLE

S. No	Clinical Presentation	No. of Cases	Percentage
1	Localized Discoid lesions	22	56%
2	Localized Discoid lesions + Mucosal lesions	6	15%
3	Localized Discoid lesions + Follicular pits in ears	7	18%
4	Hypertrophic / Verrucous DLE	1	3%
5	Mucosal lesions alone	2	5%
6	Eye lesions	1	3%
	Total	39	76%

The commonest clinical presentation of Localized DLE was Localized circumscribed or discoid type of lesions (56%).

Table 2: Disseminated DLE

S. No	Clinical Presentation	No. of Cases	Percentage
1	Disseminated Discoid lesions	6	49%
2	Disseminated Discoid lesions + Mucosal lesions	4	33%
3	Lupus Erythematosus - Lichen Planus overlap syndrome	1	9%
4	LE gyratus repens	1	9%
	Total	12	24%

Incidence of Mucosal lesions in DLE: 27%.Incidence of involvement of only the Mucosa: 4%. Incidence of Mucosal lesions with cutaneous discoid lesions: 23%.Mucosal involvement in females (28%) was more common than in males (25%). The sex ratio between females and males was 1.1: 1 . Patients with Disseminated DLE had a higher incidence (42%) of mucosal lesions than those with localized DLE (18%).

Table 3: Clinical Presentation

Clinical Presentation	Total No. of Cases	Total	
	Male	Female	
Mucosal lesions alone	0	2	2
Mucosal lesions along with Localized DLE	1	6	7
Mucosal lesions along with Disseminated DLE	3	2	5
Total	4	10	14

Table 4: Types Of Mucosal Lesions

S. No	Mucosal Lesion	No. of Cases
1	Erosions and crusting of Lip	8
2	Hyperpigmented patches on the Buccal Mucosa	3
3	Erosions on the Palate	2
4	Erosions over the Lips and Palate	1
Total		14

Table 5: Presenting Complaints

S. No	Presenting Complaint	No. of Patients
1	Itching	15
2	Burning Sensation	20
3	Loss of hair	5
4	Cosmetic disfigurement	5
5	Scaling	3
6	Pain	3

ISSN: 0975-8585

The most common presenting complaint was a Burning Sensation (39%) on exposure to sunlight.

Table 6: Precipitating Or Exacerbating Factors

S. No	Precipitating or Exacerbating Factor	No. of Cases
1	Sunlight	29
2	Trauma	1
3	Mental Stress	2
4	Burns	1
5	Pregnancy	1
6	Drugs	2
7	Premenstrual Flare	1

Table 7: Site Of Involvement Localised DLE

S.No	Site	No. of Cases
1.	Scalp	15
2.	Face	4
3.	Lips	2
4.	Ears	3
5.	Multiple Sites (Scalp, Face, Ears, Lips)	15

Total number of cases: 39
The most common site involved in Localized DLE - scalp (38%)

Table 8: Disseminated DLE

S.No.	Site	No. of cases
1.	Head, Trunk & Upper limbs	2
2.	Head, Trunk, Upper & lower limbs	2
3.	Head & Trunk	6
4.	Head and upper limb	1
5.	Trunk alone	1

The trunk was involved in 92% of Disseminated DLE Cases along with lesions on the head. Trunk alone was involved in a single case.

Table 9: Raynaud's Phenomenon

Type	Raynaud's phenomenon+	Total no. of cases	%
Localized DLE	1	39	2.5 %
Disseminated DLE	2	12	17 %
Total	3	51	6 %

Table 10: Joint Pain

Type	No. of Cases with joint pain	Total no. of cases	%
Localized DLE	2	39	5 %
Disseminated DLE	4	12	33 %
Total	6	51	12 %

ISSN: 0975-8585

12% of Cases (6) had complaints of joint pain. Joint pain was more common in Disseminated DLE than in Localized DLE.

Table 11: Associated Conditions

Associated Conditions	No. of Patients
Polymorphic Light Eruption	1
Diffuse Hair Loss	1
Urticaria	1
Prurigo nodularis	1
Macular Amyloid	1
Hypothyroidism	2
Diabetes Mellitus	2
Hypertension	1
Pterygium of Eye	1

Table 12: Laboratory Abnormalities In Dle Anaemia:

Type	No. of Patients with anemia	Total no. of Pts	%
Localized DLE	10	39	26 %
Disseminated DLE	3	12	25%
Total	13	51	25%

Table 13: Erythrocyte Sedimentation Rate (ESR)

ESR >20mm/hr	No. of Patients
Total no. of patients investigated	51
Total no. of patients with raised ESR	29
Percentage of patients with raised ESR	57%

Table 14: Antinuclear Antibody Positivity

Types of DLE	ANA Positivity	Total No. of Cases	Percentage
Localized DLE	10	39	26%
Disseminated DLE	5	12	42%
Total	15	51	29%

The percentage of patients with ANA positivity was found to be 29%

Table 15: Rheumatoid Factor

Type	RF Positive	Total No. of Patients	Percentage
Localized DLE	11	39	28%
Disseminated DLE	6	12	50%
Total	17	51	33%

Table 16: C-Reactive Protein (CRP)

Type	CRP Positive	Total No. of Patients	Percentage
Localized DLE	9	39	23%
Disseminated DLE	8	12	67%
Total	17	51	33%

Anti – Double Stranded DNA Antibody

Was found to be negative in all patients.

Histopathology

Skin biopsy was done in all the cases. Histopathological study showed compatibility with the features of DLE in all cases, except one. One case showed features suggestive of Lichen planus. Biopsy of the verrucous growth from the lesional skin on the right elbow in this case showed features of well-differentiated squamous cell carcinoma. Another case showed features suggestive of Verrucous DLE.DIF was done in a few cases. The biopsy was taken from the lesional skin, exposed nonlesional skin, and unexposed non-lesional skin. Most of the cases showed moderately strong homogenous IgG, IgM, IgA, and C3 at the basement membrane zone band in the lesional skin and negative in the uninvolved skin, features suggestive of Discoid Lupus Erythematosus. One case showed a Discontinuous ragged fibrinogen band in the basement membrane zone and IgG, IgM, and C3 colloid bodies in the lesional skin. These features were suggestive of Lichen planus.

DISCUSSION

Clinically and histopathologically it is difficult to discern the discoid lesions from others. The classical discoid lupus erythematosus generally shows a central erythematous area, ulcerated or atrophic, surrounded by fine white, irradiating striae.2similar features are seen in the case of classic Lichen planus but in lupus erythematosus, the irradiated striae are more delicate than the Wickham striae of lichen planus [7]. PAS staining for the mucopolysaccharide appears to be of great value in diagnosing the oral lesions of chronic lupus erythematosus. The characteristic finding is an extremely intense reaction beneath the stratum germinativum of the epithelium and around small blood vessels. WHO(1978) reported PAS-positive deposits juxtaepithelially, which resembles a thickening of the basement membrane.4PAS Staining in the above-reported case also demonstrated a thickened basement membrane [8]. The direct immunofluorescent(DIF) studies of oral DLE lesions in the literature revealed three major classes of immunoglobulins IgA, IgM, and IgG as well as different complement components found in the basement membrane zone in a linear and/or granular pattern [9]**.** Herein we present the initial validation of classification criteria for DLE. Based on our proposed model, a score of at least 5 yields classification as DLE with sensitivity of 84.1% and specificity of 75.9%, with increasing points yielding higher specificity [10]. These classification criteria can be applied to both localized (lesions above the neck) and generalized (lesions above and below the neck) diseases and do not require a biopsy to apply these classification criteria successfully [11]. A few items, such as scarring alopecia, were not included in the criteria because their inclusion did not substantially change our test characteristics or receiver operating characteristic curve [12].Furthermore, dermatopathology was not included in the final model. There are several reasons for this. First, only 86 patients (40.0%) had pathological findings available, which limited our ability to meaningfully incorporate these items into classification criteria models [13]. Characteristic features of cutaneous lupus erythematosus also are present, which include a thick basement membrane, dermal mucin deposition, vacuolar interface changes, and superficial and deep, perivascular and peri adnexal, lymphocytic inflammatory infiltrates [14]. Transepidermal elimination of elastic fibers as well as features of both keratoacanthoma and hypertrophic lichen planus have been reported. An association between smoking and the development of lupus erythematosus has been reported. In addition to an increased incidence of lupus erythematosus observed in current smokers in several studies recent research has suggested a role in contributing to disease activity as well Furthermore, smokers may be less likely to respond to antimalarial therapy. Treatments reported to be efficacious for hypertrophic discoid lupus erythematosus include intralesional triamcinolone acetonide, hydroxychloroquine, acitretin, thalidomide, and isotretinoin [15].

CONCLUSIONS

CLE is a multifactorial autoimmune disease, resulting from the interaction of environmental, genetic, and immunological factors, which presents varied dermatological manifestations. The identification of the clinical subtype is important for the diagnostic approach, therapeutic decision, and determining the prognosis, both in exclusively cutaneous disease and in the context of SLE. Diagnostic criteria for defining the different subtypes of CLE are still incipient. More assertive criteria are expected, which may be incorporated into clinical practice and therapeutic trials in the future, helping to assess the cutaneous manifestations of LE. Photoprotection, topical corticosteroids, and antimalarials are still the first lines of treatment for CLE. Alternative medications for systemic use include methotrexate, oral retinoids, dapsone, and thalidomide, among others. With advances in knowledge of disease pathogenesis, new therapeutic strategies have been developed, targeting the different immune activation pathways that

have been identified. Photosensitivity was the most common clinical manifestation, whereas ANA was the most frequent autoantibody of the LE patients of this region. Patients with different subtypes of CLE have distinct clinical and pathological characteristics. In the absence of consensus on a definition that makes it possible to differentiate cutaneous forms of LE from SLE, the dermatologist's role in the correct diagnosis and classification of such patients is fundamental.

REFERENCES

[1] Talbott JH: Historical background of discoid and Systemic Lupus Erythematosus in Lupus Erythematosus, Ed. Dubois EL, McGraw Hill book company, New York and London, 1966: 1-9.

[2] Walter JB and Israel MS: The collagen diseases in General pathology, Ed. Walter JB and Israel MS Edn-5, Churchill, Livingstone Publications, Edinburgh, London and New York, 1979: 205.

[3] Harry MA, Shulman LE, Tumultz PC, et al: Systemic Lupus Erythematosus; Review of literature and clinical analysis of 138 cases, Medicine; 1954; 33: 291-437.

[4] Cazenare PLA, Chausit M. Du lupus. Ann Malad Peau Syph 1852; 4: 113- 117.

[5] Dubois EL, Tuffanelli DL – Clinical manifestation of Systemic Lupus Erythematosus.

[6] Sontheimer RD. A review and personal perspective on the nomenclature and classification of the cutaneous manifestation of lupus erythematosus. Lupus 6:84,1997.

[7] Damm J, Sonnischsen N. Clinical examinations of chronic lupus erythematosus. Dermatol Wochenschr 1964; 150: 268.

[8] Cummer CL Actiology of lupus erythematosus. Arch. Dermatol Syphilol 1936; 33: 434-45.

[9] Damm J, Sonnischsen N. Clinical examinations of Chronic Lupus erythematosus. Dermatol Wochenschr 1964; 150: 268.

[10] Bielsa I, Herrero C, Ercilla G et al. Immuno-genetic findings in cutaneous Lupus erythematosus. J Am Acad Dermatology 1991; 25: 251- 7.

[11] Costner MI, Sontheimer RD. Lupus erythematosus.: Freedberg IM, Eisen AZ, Wolff K, Austen KF, Goldsmith LA, Katz SI. Editors: Fitzpatrick' Dermatology in general medicine. 6th ed. New York: McGraw-Hill; 2003.p.1677-93

[12] Paterson KS, Winchester RJ. Systemic Lupus Erythematosus: Pathogenesis In Kooper WJ, editor. Arthritis and allied condition. Philadelphia: Lippincott Williams & Wilkins;2000.p.1.

[13] Costner MI, Sontheimer RD, Provest TT. Lupus erythematosus In Sontheimer, Prost TT, editors. Cutaneous manifestation of rheumatic diseases. 2nd ed.Philadelphia: Lippincott Williams & Wilkins: 2004.p.15- 64.

[14] Knop J, Bonsman G, King P et al. Antigen of the major histocompatibility complex in patients with chronic Discoid Lupus erythematosus. Br. J Dermatol 1990; 122: 723-8.

[15] Lodiri A Discoid lupus erythematosus and trauma. Acta Derm Venereol (Stockh) 1963: 43: 142.