

DISCOID LUPUS ERYTHEMATOSUS – A REPORT OF TWO CASES**Dr. Y. Shreya*¹, Dr. Tirumala Kanakadurga Sripathi², Dr. P. Karkuzhali³, Dr. Hemalatha Ganapathy⁴**¹I Year Post-Graduate, Department of Pathology, Sree Balaji Medical College, Chrompet, Chennai-44.²II Year Post-Graduate, Department of Pathology, Sree Balaji Medical College, Chrompet, Chennai-44.³Professor & HOD, Department of Pathology, Sree Balaji Medical College, Chrompet, Chennai-44.⁴Professor Department of Pathology, Sree Balaji Medical College, Chrompet, Chennai-44.***Corresponding Author: Dr. Y. Shreya**

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ABSTRACT

Lupus Erythematosus is a chronic, multi-system, auto-immune disease. It manifests with diverse signs and symptoms; with localized cutaneous LE (CLE) on one end of the spectrum and severe systemic LE (SLE) on the other end. Cutaneous lupus is categorized as: Chronic Cutaneous Lupus (CCLE), Sub-acute Cutaneous Lupus (SCLE) and Acute Cutaneous Lupus (ACLE). Discoid Lupus Erythematosus (DLE) is the most common of the CCLEs. Most patients with untreated classic DLE lesions suffer indolent progression, resulting in large areas of cutaneous dystrophy and scarring alopecia, which can be psychosocially devastating. Progression to systemic lupus erythematosus (SLE) is a possibility, necessitating early diagnosis and prompt treatment with regular follow-up.

KEYWORDS: Cutaneous Lupus Erythematosus, Chronic Cutaneous Lupus Erythematosus, Discoid Lupus Erythematosus.

INTRODUCTION

Discoid Lupus Erythematosus (DLE) is the most common form of chronic cutaneous lupus erythematosus according to the classification proposed by Gilliam^[12,13] Classic DLE lesions present as well-defined reddish-purple macules, papules or small plaques and rapidly develop a hyperkeratotic surface, often seen in sun-exposed areas. The disease often has a chronic and relapsing course, which can be induced or aggravated by UV light. Histopathological examination by means of a biopsy is essential to confirm the diagnosis. Regular follow-up is essential in view of its chronic course and systemic treatment is sometimes indicated. It is important to screen a patient with CLE for LE-non-specific symptoms since their presence can imply systemic involvement and progression to SLE.^[11]

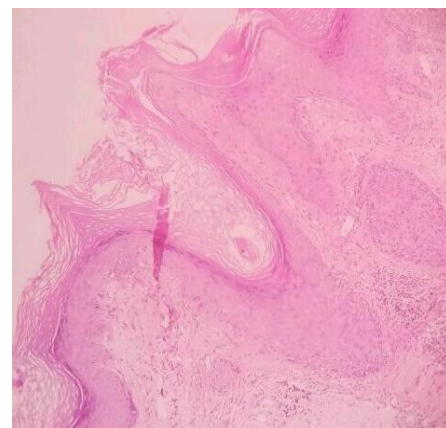
CASE REPORTS

1. A 72-year-old female came with complaints of discoloration of the skin over the scalp for the past 2 years. On examination, a depigmented plaque with peripheral pigmentation and scaling was noted over the vertex of scalp. A Provisional diagnosis of DLE was made.
2. A 34-year-old female presented with complaints of raised, dark skin lesions over her nose and chin for 10 years. On examination, well-defined pigmented plaques with central scaling were seen over the tip

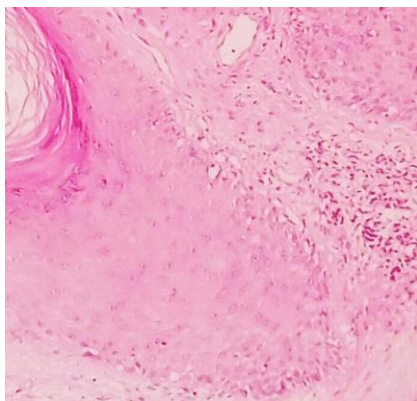
and sides of nose and chin. A Provisional diagnosis of? Discoid Lupus Erythematosus? Porokeratosis? Bullous Lupus Erythematosus was made.

RESULT

Histopathological examination revealed hyperkeratotic and acanthotic epidermis with follicular plugging and prominent granular layer overlying fibro-collagenous dermis. There was thickening of the basement membrane, focal basal cell vacuolar degeneration and lymphocytic infiltration in the superficial dermis and surrounding adnexal structures. A diagnosis of "Features consistent with discoid lupus erythematosus" was made.



Low power view: Shows hyperkeratosis and acanthosis with follicular plugging.



High power View: Shows basal cell vacuolar degeneration.

DISCUSSION

The purpose of this report is to study the clinicopathological features of discoid lupus erythematosus.

A diagnosis of discoid lupus erythematosus can be made with the characteristic clinical appearance, location of the cutaneous lesions along with the histopathological examination. Typically, discoid lupus is characterized by erythematous macules, papules, and plaques with telangiectases, scales, and follicular plugs, which results in a scarring process with atrophy and dyspigmentation.^[13] Lesions are often localized to the face, nose and ears; may be generalized; and may or may not be restricted to sun-exposed skin. It is a photosensitive lesion; Photosensitivity is observed in 50% of patients.^[12,14] However, no clear temporal association has been established between sun exposure and its development. Patients with DLE have predisposition for developing arthralgia.

Discoid lupus erythematosus can be limited, involving only the head and neck, or more extensive and diffuse. Those patients with diffuse lesions below the head and neck are more likely to develop systemic lupus erythematosus (SLE). Small subsets (5-10%) of patients with DLE have co-existent systemic disease. Progression to systemic lupus erythematosus (SLE) occurs with greater frequency in the setting of disseminated or generalized discoid lesions and often occurs within one to three years after diagnosis.^[12,14] Patients with localized disease tend to have fewer systemic manifestations and have a 50% chance of remission. Conversely, fewer than 10% of patients with generalized DLE experience remission.^[12]

Histopathological features include interface dermatitis with superficial and deep, perivascular and periadnexal infiltrate that is composed primarily of lymphocytes. Liquefaction degeneration of basal keratinocytes with melanin incontinence, increased dermal deposition of mucin, and diffuse thickening of the basement membrane are observed. Follicular plugs may be prominent in discoid lesions.^[5] Immunoglobulins and complement

proteins are deposited in a granular distribution along the dermal-epidermal junction of lesional skin in up to 90% of cases of chronic cutaneous lupus erythematosus.^[6]

Many treatment options have been used successfully to treat skin lesions of CLE. Sun avoidance and high SPF sunscreen are highly effective preventative measures. Topical therapy is the staple of CLE treatment. Systemic interventions range from older, clinically proven treatments, such as anti-malarial therapy, to newer, cutting edge immunological and biological drugs with novel mechanisms of action. The concept of combination therapy is also on the horizon for the treatment of CLE.^[17]

CONCLUSION

DLE is a chronic dermatological disease that can lead to scarring, hair loss and hyperpigmentation of the skin; the healing of a lesion takes place in the center, producing atrophy, scarring, telangiectases, pigmentation changes and scarring alopecia. The DLE-associated cutaneous lesions thus have a significant impact on the skin. This highlights the importance of early diagnosis and prompt treatment. Also, since patients with DLE might develop SLE in the future, it is important to screen for systemic manifestations at regular intervals. Appropriate treatment of DLE ranges from topical steroids to potent systemic immunosuppressive therapy.

REFERENCES

1. Levers histopathology, 10th edition.
2. WHO classification of skin tumours.
3. Journal of Cutaneous Pathology.
4. Dirk Elston Dermatopathology, 2nd Edition.
5. Rook's Textbook of Dermatology Ninth Edition.
6. Evidence-Based Dermatology, 3rd Edition.
7. Winkelmann, R. K.: diagnosis and treatment of lupus erythematosus, dermatomyositis, and scleroderma, with emphasis on cutaneous findings. *J. Chron. Dis*, 1961; 13: 401.
8. Dubois, E. L., and Martel, S.: discoid lupus erythematosus: an. Analysis of its systemic manifestations. *Ann. Int. Med*, 1956; 44: 482.
9. Scott, A., and Rees, E. G.: the relationship of systemic lupus erythematosus and discoid lupus erythematosus. *A. M. A. Arch. Dermat*, 1959; 79: 422.
10. Jacob S. Lo, Robert. E. Berg, Kenneth J. Tomecki: Treatment of discoid lupus erythematosus. *International journal of dermatology*, 1989.
11. 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.
12. Gilliam JN, et al. Distinctive cutaneous subsets in the spectrum of lupus erythematosus. *J am acad dermatol*, 1981; 4: 471.

13. Callen JP. Cutaneous lupus erythematosus: A personal approach to management. *australas j dermatol*, 2006; 47: 13.
14. Lin JH, et al. pathophysiology of cutaneous lupus erythematosus. *clinic rev allerg immunol*, 2007; 33: 85.
15. Lee La. Lupus erythematosus. in: bolognia j, et al, eds. *dermatology*, 2nd edition, london: mosby, 2008; 561.
16. David-Bajar KM, et al. pathology, immunopathology, and immunohistochemistry in cutaneous lupus erythematosus. *Lupus*, 1997; 6: 145.
17. R. R. Winkelmann, Grace K. Kim and James Q. Del Rosso. treatment of cutaneous lupus erythematosus- review and assessment of treatment benefits based on oxford centre for evidence-based medicine criteria. *j clin aesthet dermatol*, 2013 Jan; 6(1): 27–38.