

Discoid lupus erythematosus in Iraqi patients: a clinical and histopathological study

Mohammed Kadhim Al-Hattab, MBChB; Makram Al-Waiz, MBChB, DDSc, PhD

Discoid lupus erythematosus (DLE) is a relatively benign disorder of the skin, most frequently involving the face, and characterized by various-sized, reddish, well-defined scaly patches, which tend to heal with atrophy, scarring and pigmentary changes.¹ It has been proposed that genetic factors and somatic mutations are implicated in the pathogenesis of the disease.² Classical DLE lesions, most frequently of the circumscribed or discoid type, occur most commonly on sun-exposed areas of the body,³ particularly on the cheeks, the bridge of the nose, the ears, and the side of the neck and scalp.¹ Early classical DLE lesions typically evolve into sharply demarcated, coin-shaped erythematous plaques covered by a prominent, adherent scale that extends into the orifices of dilated hair follicles.⁴ Early DLE lesions typically expand with erythema and hyperpigmentation at the periphery, leaving atrophic central scarring, telangiectasia and hypopigmentation.⁴ Localized DLE lesions occur only on the head or neck, whereas generalized DLE lesions occur both above and below the neck.⁴ Other variants of DLE include verrucous (hypertrophic) LE, LE profundus, LE panniculitis, mucosal DLE, LE-LP (lupus erythematosus-lichen planus) overlap syndrome, LE telangiectoides and chilblain LE.^{1,4,5} Laboratory abnormalities are found in about 55% of patients.⁵ The presence of persistently abnormal laboratory tests findings and a high titre of ANA in DLE appears to be an indication of the potential for SLE to develop.⁶ The aim of this study was to shed light on the various clinical and histopathological aspects of DLE among Iraqi patients.

Patients and methods

We studied 54 patients with DLE. A full history from each patient was recorded, including age, sex, age of onset and duration of the disease, and precipitating and aggravating factors. A thorough physical examination was conducted to record the site of the lesion, the symmetry and distribution of the lesions, and to determine whether bilateral or unilateral and different clinical variants of the disease were present. Laboratory investigations included ANAs (anti-nuclear antibodies) (with titer), anti-ds-DNA antibodies, a complete blood picture and ESR (erythrocyte sedimentation rate), and a VDRL (Venereal Disease Research Laboratory) test (with titer) for false positive serological tests associated with DLE. A general urine examination was also performed for each patient. Biopsies were taken from 32 patients for

From the Department of Dermatology and Venereology, Baghdad Teaching Hospital, College of Medicine, University of Baghdad Baghdad, Iraq

*Correspondence to:
Professor Makram Al-Waiz
P.O. Box 61269 Medical Collection Post Office
Postal Code Number 12114
Bab Al-Muadham
Baghdad
Iraq
Email: makram55@uruklink.net*

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histopathological examination by hematoxylin and eosin staining.

Results

The 54 patients ranged in age from 13 to 70 years (mean±SD, 36±11.8 years), while age of onset averaged between 7 to 68 years (mean±SD, 30.2±11.4 years). The duration of disease ranged from 1 month to 30 years (median, 3 years). There were 34 females (63%) and 20 males (37%) for a sex ratio of 1.7:1.

The clinical variants of the disease in our patients are shown in Table 1. A variant of disseminated leukoderma or vitiligo-like DLE, which involved the face, most of the trunk and extremities with scarring alopecia of the scalp was seen in one patient (1.8%) (Figure 1). Hypertrophic DLE with a warty-like plaque at the extensor surface of the right forearm was seen in one of the disseminated classical DLE patients. Table 2 shows the sites of involvement. The cheeks and the nose were the commonest sites of involvement (76% and 57.4%, respectively). The scalp was affected in 48% of patients. Scarring alopecia was seen in 42.5% and the vertex was the most commonly affected site (37%). Non-scarring hair loss was seen in 5.5%. The oral cavity was involved in 11% of cases. The lesions were whitish, atrophic patches located on the hard palate and buccal mucosa. Two patients with oral ulcers progressed to SLE. The nasolabial folds were not affected in all patients with DLE. Nails and nail folds were also not affected.

Table 1. Clinical presentation in 54 patients with discoid lupus erythematosus (DLE).

Clinical variant of DLE	No. cases (%)
Localized classical	24 (44.4)
Localized atrophic	12 (22.2)
Disseminated classical	8 (14.8)
Disseminated atrophic	8 (14.8)
Lupus profundus (panniculitis)	6 (11)
Hypertrophic	1 (1.8)
Disseminated leukoderma-like	1 (1.8)

Table 2. Sites of lesions in 54 patients with DLE

Site	No. cases (%)
Cheeks	41 (76)
Nose	31 (57.4)
Ears	25 (46)
Around ears	21 (39)
Forehead	6 (11)
Mandibles	7 (13)
Lips	11 (20.4)
Periorbital	7 (13)
Chin	6 (11)
Oral mucosa	6 (11)
Scalp	26 (48)
Neck	2 (3.7)
Trunk	14 (26)
Arms	11 (20.4)
Legs	1 (1.8)

*Nearly all patients had more than one site of involvement of DLE lesions.

Anti-nuclear antibodies (ANA) were positive in 13 patients (24%) with a titre ranging between >1:40 to >1:80. The patterns of ANA reactions were speckled in 9 patients (16.6%), homogenous in 3 patients (5.6%) and nucleolar in 1 patient (1.8%). Anti-ds-DNA antibodies were positive in 6 patients (11%). Two of the 3 patients who started as DLE and changed to SLE had positive anti-ds-DNA antibodies. The VDRL test was false positive in 5 patients with a titer ranging between 1:8 to 1:16 (9.2%). Hemoglobin levels ranged between 9 to 10 g/dL in 9 patients (16.6%). Leukopenia (< 3000/mm³) was found in 5 patients (9.2%); 2 of 3 patients who changed to SLE had leukopenia in serial measurements. ESR levels were high, ranging from 45 to 115 mm/hour in 10 patients (18.5%). GUE showed positive granular and epithelial casts in 2 patients (3.7%) (Table 3). The histopathological findings in 32 patients with different clinical variants of the disease are shown in Table

Table 3. Laboratory abnormalities in 54 patients with DLE

Parameter	No. cases (%)
Positive ANA	13 (24)
Positive anti-ds-DNA antibody	6 (11)
False positive VDRL test	5 (9.2)
Anemia	9 (16.6)
Leukopenia	5 (9.2)
High ESR levels	10 (18.5)
Granular or epithelial cases in GUE	2 (3.7)

Table 4. Histopathological findings in 32 patients with DLE lesions.

Feature	No. cases (%)
Hyperkeratosis	26 (81.2)
Follicular plugging	28 (87.5)
Parakeratosis	4 (12.5)
Atrophy of epidermis	21 (65.6)
Irregular acanthosis	4 (12.5)
Normal thickness epidermis	7 (21.8)
Papillomatosis	2 (6.2)
Focal basal liquefaction	23 (71.8)
Diffuse basal liquefaction	2 (6.2)
Pigmentary incontinence	11 (34.3)
Telangiectasia	11 (34.3)
Colloid bodies	5 (15.6)
Patchy, periappendigeal and perivascular inflammatory infiltrate	24 (75)
Sparse upper dermal infiltrate	3 (9.4)
Granuloma-like inflammatory infiltrate	1 (3)
Panniculitis	2 (6.2)

4. Figure 2 shows the histopathological features of classical DLE. A 50 year-old female patient with a scar due to previous localized atrophic DLE in a butterfly distribution on the face of 20 years duration developed squamous cell carcinoma in the previous year on the part of the scar located on the upper lip. No metastasis to regional lymph nodes was noticed (Figure 3).

Discussion

DLE is primarily a disease of adult life with the average time of onset in the fourth decade, with a female:male ratio of about 2:1.³ We found that the disease occurred commonly in adults, mainly in the fourth decade of life with a female:male ratio about 1.7:1. This finding is comparable to those reported in the literature.³

The clinical variants and the morphology of skin lesions of the patients in the present study were comparable to what



Figure 1. Disseminated leukopenia or vitiligo-like discoid lupus erythematosus (DLE). Multiple and extensive hypopigmented atrophic patches affecting the back and extensors of the forearm.



Figure 3. Squamous cell carcinoma complicating the scar of localized atrophic DLE involving the upper lip.

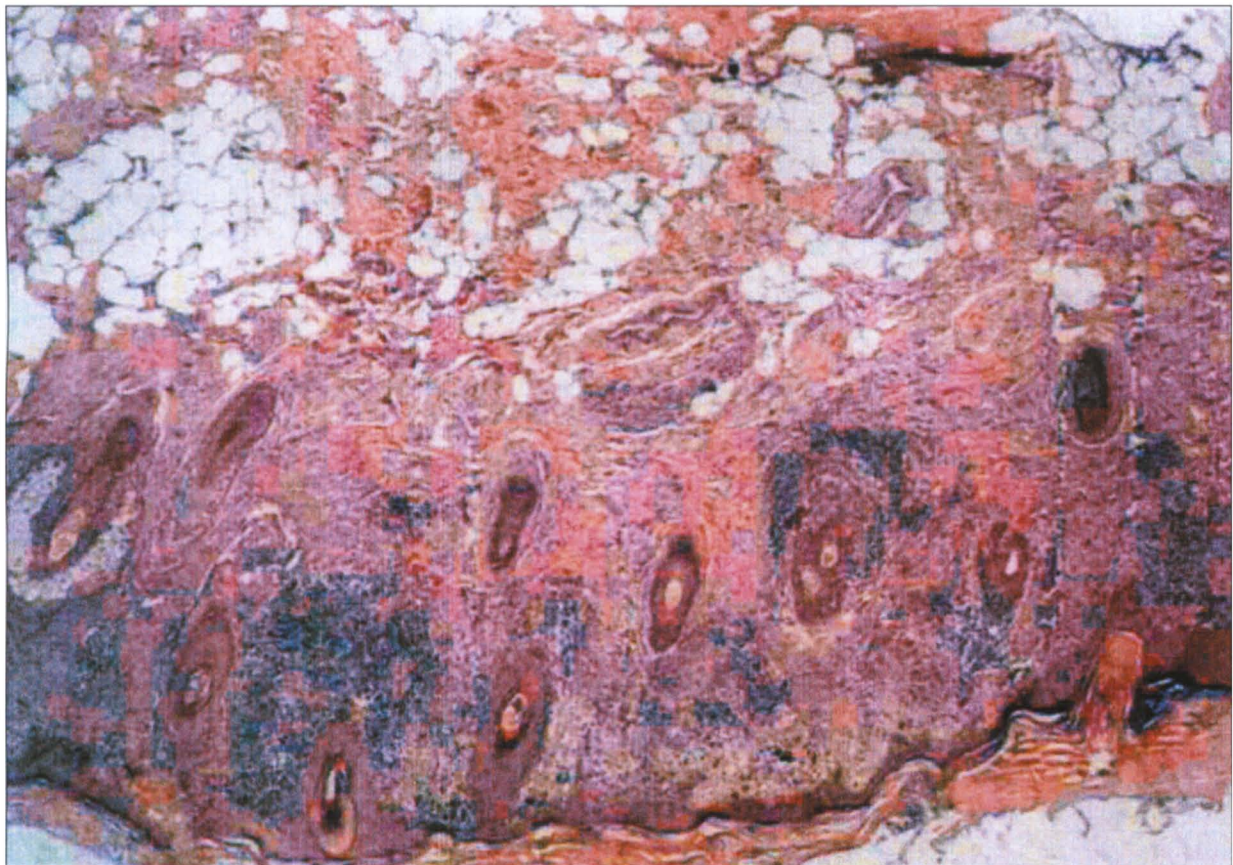


Figure 2. Histopathology of localized classical DLE showing the typical histopathological features of DLE: hyperkeratosis, follicular plugging, atrophic epidermis, and patchy and periappendageal lymphoid inflammatory cells, mainly in the upper dermis (H&E, x40).

of the patients in the present study were comparable to what has been published.^{1,4} However, a variant of disseminated DLE was found. This variant, a leukoderma or vitiligo-like form, misdiagnosed earlier as a case of vitiligo, was seen in one patient (1.8%). We found that the scalp was involved in 48% of patients with scarring hair loss in 42.5% and non-scarring hair loss in 5.5%. In the literature, scalp involvement has been similarly affected.^{3,4} In this study, as well as in a previous Iraqi study concerning scarring alopecia in Iraqi patients,⁷ active DLE lesions on the face associated with scarring alopecia of the scalp suggest that the scalp could be involved early in the disease process. This finding has not been reported before. The present work showed that oral mucosa was involved in 11% with oral ulceration in 3.7% of patients. This was comparable to what has been described in the literature.³ Ears are commonly affected in DLE. Either one ear or bilateral involvement was seen in 46% of patients. Lesions were located on the rim or the external auditory meatus of the ear. Ear involvement was seen even in those patients who covered their ears continuously when outdoors (e.g., wearing a veil in females), an observation that may reinforce the autoimmune pathogenesis of the disease. Therefore, patients with DLE on the face should have their ears examined.

Previous studies have reported that ANAs were found in 30% to 35% of patients with DLE.^{1,3} The homogenous type of antinuclear factor was twice as frequent as the 'speckled' type.^{1,3} We showed that ANAs were positive in 24%, with the 'speckled' pattern of antinuclear factor being three times as frequent as the 'homogenous' type. One patient had a

nucleolar pattern. This study demonstrated that ANAs were more common in the disseminated type, in disease with oral involvement, in patients with scarring alopecia, and in patients who had the disease for an extended period of time. Anti-ds-DNA antibodies were positive in 11% of patients. This was comparable to what has been reported in the literature.^{8,9} These antibodies were found to be positive especially in those patients who started with DLE and transformed to SLE. The risk of a patient with DLE developing overt SLE is small. Transformation varies from 1.3% to 6.5%.^{5,6,10} The risk is higher in patients with disseminated DLE than in DLE confined to the head and neck.^{5,6,10} We found that the percentage of patients transforming to SLE was 5.5%, which is comparable to rates reported in the literature.^{5,6,10}

The incidence of neoplastic changes in DLE was about 1.8%, which is comparable to the incidence of 3.3% noticed in a series of 120 white patients with DLE.¹¹ The squamous cell carcinoma in a scar on the upper lip one patient had been present for about 3 months with no metastasis to regional lymph nodes. The patient had a duration of DLE of about 20 years. The histopathological features of different clinical variants of DLE were similar to what has been reported.^{1,12} Although it is well known that parakeratosis is not conspicuous and may be absent,¹² we showed that parakeratosis was present in 12.5% of specimens. This discrepancy could be attributed to superimposition of other inflammatory disorders such as dermatitis or malignancy. DLE seems not uncommon in Iraq. No previous study has evaluated the disease in our country so our report is a first step towards shedding light on this important subject.

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