Periorbital discoid lupus erythematosus: A retrospective study

Erin Theisen, MD, PhD,[a,b] Janice Tiao, MD,[a,b] and Flavia Fedeles, MD[a,b]
Boston, Massachusetts

Key words: discoid lupus erythematosus; eyelid lupus; lupus; medical dermatology; periorbital discoid; periorbital lupus.

INTRODUCTION

Discoid lupus erythematosus (DLE) is a form of chronic cutaneous lupus that typically presents on the head and neck and results in scarring.[1] DLE may rarely present as eyelid edema and erythema (periorbital DLE), a variant reported rarely in the literature.[2]

METHODS

To better characterize the disease and clinical course of this rare entity, we searched the patient research database at the Mass General Brigham institution for patients with a diagnosis of DLE (International Classification of Diseases 9/10 code) and the keywords "periorbital," "eyelids," or "eyelid", as well as "periocular". We screened 154 charts from 01/01/2000 through 10/31/2021 that met the specified search criteria and identified 9 patients with a diagnosis consistent with periorbital DLE (Table I). The diagnosis of periorbital DLE was confirmed by review of available clinical notes, documented physical examination findings, patient photographs (when available), and histopathology (available in 7 of 9 cases).

RESULTS

Seventy-eight percent (7/9) of the subjects were women. Six out of 9 subjects self-identified as white. The average age at diagnosis was 52.7 years (range, 19-71 years). Forty-four percent of the subjects (4/9) had cutaneous involvement of only the periorbital area, whereas the others had additional cutaneous involvement, most often elsewhere on the face. In subjects with only periorbital involvement, diagnosis was delayed an average of 32 months from symptom onset (range, 11-60 months). Common initial diagnoses included contact dermatitis or eyelid dermatitis. The most common physical examination finding was swelling and erythema of a single upper eyelid (4/9), followed by lower eyelid lesions (4/9), and bilateral upper eyelid involvement (3/9) (Fig 1, *A* and *B*).

Abbreviation used:

DLE: discoid lupus erythematosus

Eight of 9 subjects had documented low titers of antinuclear antibody, typically in a speckled pattern. Two subjects were diagnosed with systemic lupus erythematous. Most subjects were treated with hydroxychloroquine and topical steroids or calcineurin inhibitors. After diagnosis, more than half of subjects had periorbital lesions that persisted for more than 1 year; however, all eventually healed with scarring or hypopigmentation. Seventy-eight percent of subjects (7/9) experienced repeated flares of their periorbital DLE, suggesting that periorbital DLE is a chronic, relapsing process. Time to disease flare ranged from 3 months to more than 20 years.

DISCUSSION

Eyelid involvement in DLE has been reported to occur in 5%-6% of patients with DLE, often in

From the Department of Dermatology, Massachusetts General Hospital, Boston, Massachusetts [a]; and Harvard Medical School, Boston, Massachusetts.[b]

Funding sources: None.

IRB approval status: The study was approved by the Mass General Brigham IRB (Protocol No. 2021P002880).

Correspondence to: Flavia Fedeles, MD, Department of Dermatology, Massachusetts General Hospital, Harvard Medical School, 50 Staniford Street, Suite 200, Boston, MA 02114. E-mail: ffedeles@mgh.harvard.edu.

JAAD Case Reports 2022;25:78-82.
2352-5126

https://doi.org/10.1016/j.jdcr.2022.05.024

Table I. Characteristics of patients with periorbital discoid lupus erythematosus

Patient	Sex	Race	Age at diagnosis or involvement of periorbital region (if known DLE)	Distribution of cutaneous lesions	Symptom onset to time of diagnosis	Time to flare following initial clearance	Laboratory workup	Pathology results	Treatment
1	M	Other	45	Right upper eyelid	11 mo	3 mo	ANA 1:40, speckled Negative for anti-dsDNA, Ro, La, Smith, and RNP antibodies	Interface, superficial, and deep, perivascular and periadnexal dermatitis with dermal mucin deposition	Topical: pimecrolimus, tacrolimus, and mometasone
2	F	White	56	Bilateral upper eyelids and right lower eyelid	60 mo	3 mo 9 mo 6 mo	ANA 1:40, speckled Negative for anti-Ro, La, Smith, and RNP antibodies	Interface, perivascular, and periadnexal dermatitis with dermal mucin deposition	Topical: tacrolimus, hydrocortisone, and valerate Prescribed HCQ but did not take
3	M	Black	19	Left lower eyelid	24 mo	6 mo 13 mo 10 mo	ANA 1:40, speckled Negative for anti-dsDNA, Ro, La, Smith, and RNP antibodies	Interface dermatitis with marked pigment incontinence and papillary dermal edema, superficial perivascular and periadnexal lymphocytic infiltrate, and increased dermal mucin deposition	Topical: tacrolimus and mometasone Prescribed HCQ but did not take
4	F	White	50	Right upper eyelid	Data not available	6 y	ANA 1:80, pattern not described	Data not available	Topical: pimecrolimus, desonide HCQ

Continued

Table I. Cont'd

Patient	Sex	Race	Age at diagnosis or involvement of periorbital region (if known DLE)	Distribution of cutaneous lesions	Symptom onset to time of diagnosis	Time to flare following initial clearance	Laboratory workup	Pathology results	Treatment
5	F	White	59	Right upper eyelid, scalp, forehead, nose, right eyebrow, and conchal bowl	DLE 4 y prior to diagnosis	No documented periorbital flares	ANA 1:160, speckled *diagnosed with SLE	Documentation of biopsy performed and consistent with DLE (copy of pathology record not available)	Topical: tacrolimus, Vytone MTX, leflunomide, HCQ, and quinacrine
6	F	White	58	Bilateral lower eyelids, neck, and cheeks	DLE 7 y prior to diagnosis	21 mo	ANA 1:40, diffuse	Interface dermatitis with sparse superficial perivascular lymphocytic infiltrate and focal mild dermal mucin deposition dermatitis	Topical: tacrolimus and HCQ
7	F	White	59	Bilateral upper eyelids, conchal bowls, eyebrows, temples, and cheeks	Data not available	22 y	ANA 1:640, speckled Positive for anti-Ro antibodies Negative for anti-Smith, RNP, dsDNA antibodies	Interface and a focal vacuolar dermatitis with extensive lichenoid infiltrate and pigment incontinence. Effacement of the rete ridges, molecular plugging, and thickening of the basement membrane	Topical: tacrolimus, triamcinolone, clobetasol, and HCQ

8	F	Black	57	Left upper eyelid, scalp, cheeks, and nose	DLE 14 y prior to diagnosis	No documented periorbital lesion flares	ANA negative	Interface dermatitis, vacuolar type, associated with perivascular and periadrexal mononuclear cell infiltrates.	Topical: pimecrolimus and halobetasol
9	F	White	71	Bilateral upper and lower eyelids, cheeks, nose, and temples	DLE 3 y prior to diagnosis	7 mo	ANA 1:320, speckled *diagnosed with SLE	Vacuolar interface dermatitis with follicular involvement	Topical: tacrolimus, desonide, tofacitinib HCQ, lenalidomide, and prednisone,

ANA, Antinuclear antibody; *DLE*, discoid lupus erythematosus; *dsDNA*, double-stranded DNA; *F*, female; *HCQ*, hydroxychloroquine; *M*, male; *MTX*, methotrexate; *RNP*, ribonuclear protein; *SLE*, systemic lupus erythematosus.

Fig 1. Images of clinical findings in periorbital discoid lupus erythematosus. **A,** Bilateral upper eyelids with scaly, pink plaques and papules of the right lower eyelid. **B,** Erythema and swelling of an upper eyelid.

conjunction with other cutaneous lesions.[2] Involvement limited to periorbital skin is rare. Unfortunately, the diagnosis of periorbital DLE is often delayed by years,[3] a finding that was observed in our cohort. In addition to cutaneous scarring and hypopigmentation, untreated periorbital DLE has been reported to result in conjunctival scarring, symblepharon formation, trichiasis, and ectropion,[4] making prompt diagnosis crucial. Limited systemic involvement was observed in our cohort, a finding consistent with the reported low risk of developing systemic lupus erythematous with limited DLE lesions.[5]

In conclusion, periorbital DLE is a rare manifestation of chronic cutaneous lupus that is rarely associated with systemic disease. Diagnosis is often delayed and results in chronic recurrent lesions that can scar and cause pigmentation changes. Providers should be aware of this rare entity to ensure prompt treatment to limit scarring and potential visual sequelae.

Conflicts of interest

None disclosed.

REFERENCES

1. Okon LG, Werth VP. Cutaneous lupus erythematosus: diagnosis and treatment. *Best Pract Res Clin Rheumatol*. 2013;27(3):391-404. https://doi.org/10.1016/j.berh.2013.07.008
2. Wang X, Wang P, Wang M, et al. Discoid lupus erythematosus solely involving the eyelids: case report and literature review. *Int J Dermatol*. 2021;60(10):1190-1198. https://doi.org/10.1111/ijd.15397
3. Gupta T, Beaconsfield M, Rose GE, Verity DH. Discoid lupus erythematosus of the periorbita: clinical dilemmas, diagnostic delays. *Eye (Lond)*. 2012;26(4):609-612. https://doi.org/10.1038/eye.2011.340

4. Frith P, Burge SM, Millard PR, Wojnarowska F. External ocular findings in lupus erythematosus: a clinical and immunopathological study. *Br J Ophthalmol*. 1990;74(3):163-167. https://doi.org/10.1136/bjo.74.3.163

5. Zhang YP, Wu J, Han YF, Shi ZR, Wang L. Pathogenesis of cutaneous lupus erythema associated with and without systemic lupus erythema. *Autoimmun Rev*. 2017;16(7):735-742. https://doi.org/10.1016/j.autrev.2017.05.009