Case Report

Ocular Discoid Lupus Erythematosus: More Than what Meets The Eye

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Abstract

Lupus erythematosus is a spectrum that includes auto-immune disorders such as systemic lupus erythematosus (SLE), subacute cutaneous lupus erythematosus (SCLE), and chronic cutaneous lupus erythematosus (CCLE). Discoid lupus erythematosus (DLE) is the most common type of CCLE, and ocular DLE is a rather rare presentation. We report the case of a 42-year-old male patient who had been repeatedly mis-diagnosed and upon detailed dermatological workup was found to possess clinical, dermoscopic, and histopathological features of DLE. Our report focuses on the importance of dermoscopy as a useful point-of-care investigation to avoid delay, and thus related complications. It further suggests how fundus examination could prove helpful in identifying lupus-related fundus changes, and explores how to devise a suitable plan of treatment for such patients.

Keywords: *Dermoscopy, eyelid DLE, lupus erythematosus, ocular DLE*

Introduction

The term lupus erythematosus (LE) is ascribed to a spectrum of auto-immune disorders encompassing entities such as systemic lupus erythematosus, subacute cutaneous lupus erythematosus, and chronic cutaneous lupus erythematosus (CCLE). Discoid lupus erythematosus (DLE) forms the most common variant among CCLE.[1] 60–80% of DLE lesions are localized to sun-exposed sites such as the head, neck, ears, and scalp.[2] It can occasionally involve various mucosae such as the oral, nasal, genital, and ocular mucosa. Ocular involvement is rather rare, and thus often mis-diagnosed. Failure to diagnose and treat in time may result in scarring sequelae. We report one such case of ocular DLE.

Case Report

A 42-year-old male presented with the complaint of the presence of an area of redness along the outer half of his left lower lid margin since 2 years. The patient gave a history of the lesion being asymptomatic initially, but that he had started experiencing excessive watering and irritation in his left eye 4 months back. He then consulted in ophthalmology, where he was given treatment for seborrheic blepharitis, which did not afford the patient any significant improvement. He was later referred to dermatology.

Upon detailed dermatological examination, a well-defined erythematous plaque was found abutting the lateral half of the left lower lid margin. It had a central pinkish-brown area with mild scaling. Ciliary madarosis was also noted in the lesional skin [Figure 1(a)]. The patient had lower lip cheilitis with areas of scaling and atrophy which the patient confirmed to have had since the last 1.5 years [Figure 1(b)]. External ear examination revealed multiple brown atrophic macules in the concha and triangular fossa of the patient's left ear which had escaped the patient's attention [Figure 1(c)]. On dermoscopic examination, follicular keratotic plugs and white perifollicular halo were seen interspersed with speckled brown pigmentation against a pinkish-white background [Figure 2]. Furthermore, in the area of madarosis, intermittent absence of follicular openings was also noted. Towards the lower mid border of the plaque, white structureless areas were seen, suggesting atrophy, and thus poor lesional prognosis. A deep biopsy was taken with the help of an ophthalmologist to confirm the diagnosis [Figure 3(a) and (b)]. The anti-nuclear antibodies (ANA) came out to be positive (titre 1:80), but the rest of the ANA profile was negative. No other

How to cite this article: Pareek S, Mohta A, Mehta RD, Prasad V. Ocular discoid lupus erythematosus: More than what meets the eye. Indian Dermatol Online J 2023;14:399-401.

Received: 11-Sep-2022. **Revised:** 26-Oct-2022. **Accepted:** 05-Nov-2022. **Published:** 04-Apr-2023.

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Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.idoj_482_22

Quick Response Code:

Figure 1: (a) Patient with an erythematous plaque along the lateral half of the left lower lid margin and lower lip cheilitis. (b) Erythematous plaque with a central pinkish-brown area with mild scaling and ciliary madarosis. (c) Brownish macules with atrophy in the triangular fossa and concha of left external ear

systemic findings were present. Thus, the diagnosis of localized DLE was confirmed.

Fundus examination revealed active with healed patches of chorioretinitis in the right eye and one hypo-pigmented macular patch in the left eye, which could be lupus-related ocular changes,[3] suggesting the possibility of progression to SLE in the future.

The patient was prescribed oral corticosteroids and azathioprine along with topical 1% hydrocortisone, and was advised to follow up in the dermatology and ophthalmology out-patient departments regularly.

Discussion

A limited number of cases of eyelid DLE have been described in the literature.[4] Owing to the paucity of available literature, it often escapes the clinicians' attention. Delay in diagnosis and treatment may result in irreversible loss of sight.[5] Lower lid blepharitis is the most common finding, with most reports citing the involvement of unilateral eyelid.[6] Other ocular manifestations may include proptosis, periorbital edema, madarosis, trichiasis, ectropion, entropion, lid scarring, hypertrophic/verrucous lesions, conjunctivitis, conjunctival scarring, symblepharon, and stromal keratitis.[7-9]

Although the exact etiology of eyelid DLE remains elusive, exposure to ultraviolet rays, drugs, heavy metals, pesticides, and silica dust may contribute to its development in susceptible individuals.[10] The differential diagnoses include chronic blepharoconjunctivitis, seborrheic blepharitis,

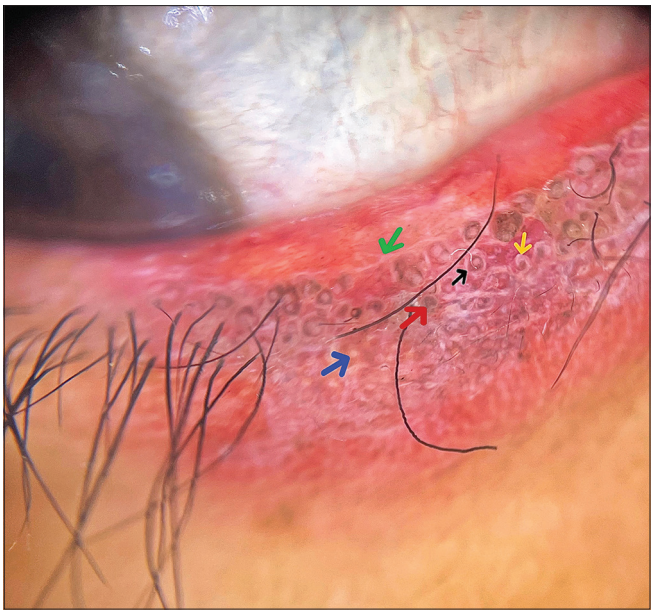

Figure 2: Demoscopic examination shows follicular keratotic plugs (red arrow), perifollicular halo (black arrow), patchy whitish scales (yellow arrow), follicular white dots indicating fibrosed follicles (blue arrow), and polarization-specific shiny white lines indicative of scarring (green arrow) (Dermlite DL4, ×10)

eyelid eczema, psoriasis, sebaceous cell carcinoma, sarcoidosis, and eyelid lichen planus.[7] A strong clinical suspicion should be corroborated by serological tests and biopsy. Typical dermoscopic findings as described in our case report may also aid the clinicians in making timely diagnosis in the case of confusing clinical presentations.

It is an entity which requires dermatologists to work in conjunction with ophthalmologists to avoid any possible morbidities. Fundus examination is advisable as it may reveal lupus-related fundus changes and thus suggest the need for a closer follow-up of the patient. Furthermore, relevant fundus abnormalities, if present, preclude the use of hydroxychloroquine, which remains the most widely prescribed drug for ocular DLE.

Conclusion

Ocular DLE is an uncommon clinical entity with little literature available on it. It may mimic many dermatological and ophthalmological conditions which run a rather benign course. Prompt diagnosis is of paramount importance as delay may lead to disfiguring and visually impairing sequelae. Clinical suspicion should never be ignored, and biopsy should always be taken. Dermoscopy may also prove to be a useful tool. Fundus examination may reveal other relevant ocular findings and thus should be advised in addition to other investigations. With timely treatment, the lesions may heal with little or no scarring and/or pigmentary changes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have

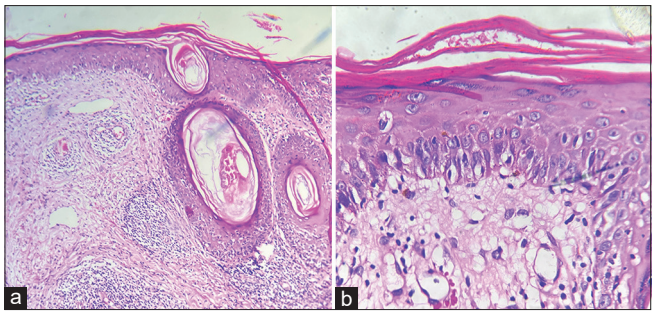

Figure 3: (a) Focal epidermal thinning and basal cell vacuolization; smudged dermoepidermal junction; moderately dense perivascular and periappendageal lymphocytic infiltrate; follicular plugging (H and E, ×100). (b) Vacuolar degeneration of the basal layer with scattered colloid bodies at the dermoepidermal junction (H and E, ×400)

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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