Eyelid Discoid Lupus Erythematosus Presenting with Chronic Blepharoconjunctivitis, Mimicking Mucous Membrane Pemphigoid

Sarawin Harnchoowong MD,
Kumutnart Chanprapaph MD.

ABSTRACT:

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DIVISION OF DERMATOLOGY, FACULTY OF MEDICINE, RAMATHIBODI HOSPITAL, MAHIDOL UNIVERSITY, BANGKOK, THAILAND.

Discoid lupus erythematosus (DLE) is the most common form of chronic cutaneous lupus erythematosus. DLE can occur on sun-exposed areas and mucosal surfaces. However, DLE on the ocular mucosa, presenting as chronic blepharoconjunctivitis, is rare and may lead to misdiagnosis with other skin conditions, especially autoimmune diseases like mucous membrane pemphigoid. Moreover, due to the rarity of DLE on the eye, the delay in diagnosis may result in complications and morbidity on the effected eyes. Dermoscopy is a non-invasive tool, commonly used to aid the diagnosis of many skin conditions. Dermoscopic findings in DLE are well-established. However, in this case we used dermoscopy to examine

From: Division of Dermatology, Faculty of Medicine Ramathibodi Hospital, Mahidol University, Bangkok, Thailand

Corresponding author: Kumutnart Chanprapaph MD., email: kumutnartp@hotmail.com

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DLE lesions on the ocular and eyelid skin surface which is rarely reported in the literature. Our dermoscopic evaluation showed telangiectasia, non-atrophic round erythema, brownish pigmentation with adherent scales on skin surface and non-atrophic round erythema with adherent scales on mucosal surface. Antimalarial drugs are the first-line systemic therapy and also most commonly used in the literatures for eyelids DLE. In this case, we also reported that dapsone in addition to hydroxychloroquine were effective treatments for eyelid DLE. Moreover, multidisciplinary care with ophthalmologist is essential to maximize treatment outcome and prevent further complications.

Key words: Blepharoconjunctivitis, dermoscopy, discoid lupus erythematous

Lupus erythematosus (LE) covers a wide range of disorder from localized cutaneous form to life-threatening systemic disease. Chronic cutaneous lupus erythematosus (CCLE) is a chronic benign autoimmune subtype of cutaneous LE that predominantly involves the skin and mucosa. The most common form of CCLE is the classic discoid lupus erythematosus (DLE)[1]. DLE lesions mostly affect sun-exposed skin of the head and neck and also mucosal surfaces (e.g., oral, nasal and genital mucosa)[1]. DLE on the ocular mucosa is exceedingly rare and can mimic the presentation of more common ocular surface disorders such as mucous membrane pemphigoid (MMP).

Case Report

A 40-year-old female first visited the ophthalmic out-patient clinic with chief complaint of stinging pain and pruritus on both lower eyelids for 2 months. There was no epiphora or discharge over her eyes. She had no underlying diseases and denied medications or topical agents used on her eyes. Ophthalmic examination showed follicular conjunctivitis, positive lid notching, eyelid eversion at the mucocutaneous junction and secondary meibomian glands dysfunction. Moreover, there were erosive pinkish plaques on both lower eyelid margin. (Figure 1A-B) Chronic blepharoconjunctivitis was diagnosed. Autoimmune mucocutaneous diseases such as MMP was the primary provisional diagnosis. Topical fluorometholone eye drop thrice daily was commenced without improvement. The ophthalmologist then performed a right lower eyelid pentagonal biopsy. Consequently, she developed another skin lesion on her right ear concha. (Figure 1C) Dermatological consultation was done and skin examination showed solitary atrophic brownish patch on the right concha. No other skin and mucosal involvement was reported. Dermoscopic evaluation on eyelid illustrated telangiectasia, non-atrophic round erythema, brownish pigmentation with adherent

scales on skin surface and non-atrophic round erythema with adherent scales on conjunctival surface. (Figure 2A-B) On right ear, dermoscopy showed follicular keratotic plugs and atrophic brownish pigmentation. (Figure 2C) Histology from eyelid biopsy demonstrated compact hyper-orthokeratosis, vacuolization of basal cell layer with thickened basement membrane, together with dense superficial and deep perivascular, periadnexal lymphocytic infiltration with melanophages and telangiectasia. (Figure 3) Direct immunofluorescence (DIF) on the lesional skin of the eyelid showed homogeneous granular deposition of immunoglobulin G (IgG), IgA, IgM and complement 3 (C3), along the dermo-epidermal junction and as well as the follicular epithelium with few cytoid bodies (IgM, C3) and granular deposition of IgA and IgM along the superficial blood vessels. (Figure 4) Laboratory investigation revealed normal complete blood count, no proteinuria, normal renal function, negative antinuclear antibody (ANA) and negative anti-bullous pemphigoid 180/230 antibodies. Based on all aforementioned findings, the diagnosis of localized CCLE was made. Dapsone (50 mg/day) was commenced from the first visit to the dermatology department due to the initial diagnosis of MMP. Then after the definitive diagnosis of DLE was made, hydroxychloroquine (200mg/day) was additionally prescribed together with 2% hydrocortisone ointment. Her condition gradually improved within 2 months.(Figure 1D-E)

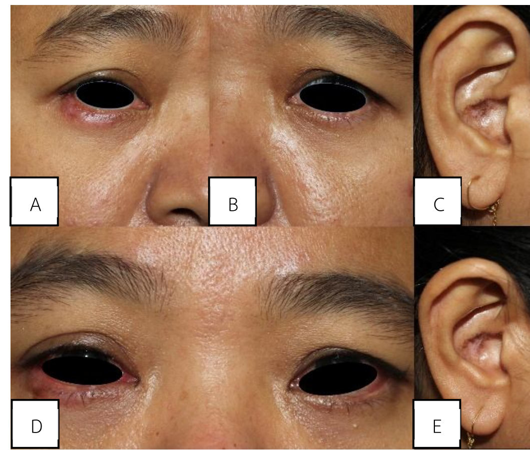

Figure 1 Discoid lupus erythematosus on both eyelids and left ear. A) Right lower eyelid swelling with erosive pinkish plaques on eyelid margin. B) Left lower eyelid swelling with erosive pinking plaques on eyelid margin. C) Solitary atrophic brownish patch on right concha. D,E) Improvement after treatment for 2 months.

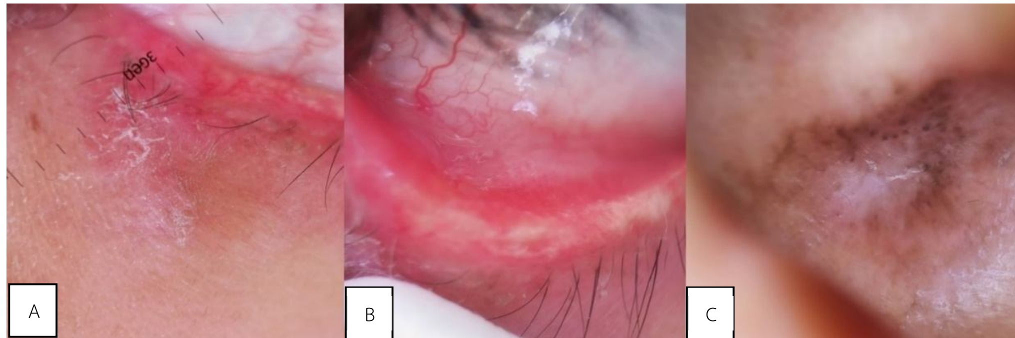

Figure 2 Dermoscopic findings. **A)** Right lower eyelid skin shows telangiectasia, non-atrophic round erythema, brownish pigmentation with adherent scales. **B)** Right lower eyelid conjunctiva shows non-atrophic round erythema with adherent scales. **C)** Right concha shows follicular keratotic plugs and atrophic brownish pigmentation.

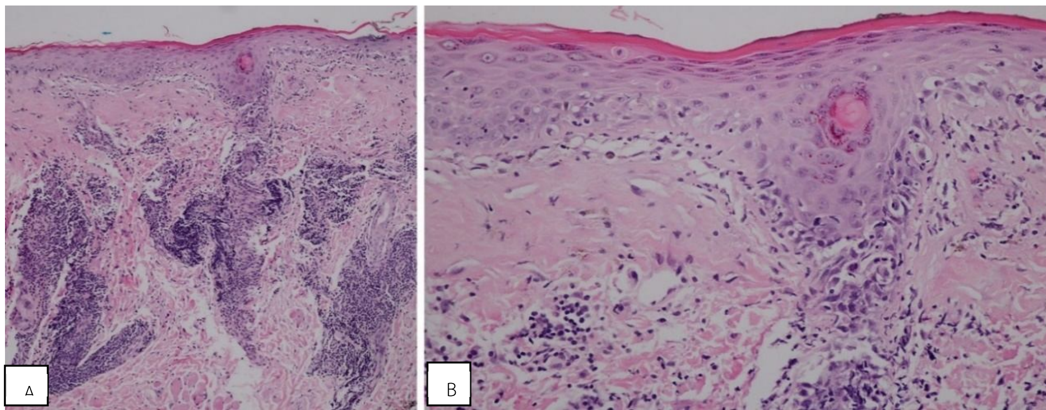

Figure 3 Histopathologic findings. **A)** Dense superficial and deep perivascular infiltration (H&E, X40).
B) Compact hyper-orthokeratosis, epidermal and follicular basal vacuolization and thickened basement membrane, perifollicular cell infiltration of lymphocytes and perivascular infiltration of lymphocytes and melanophages, telangiectasia (H&E, X400).

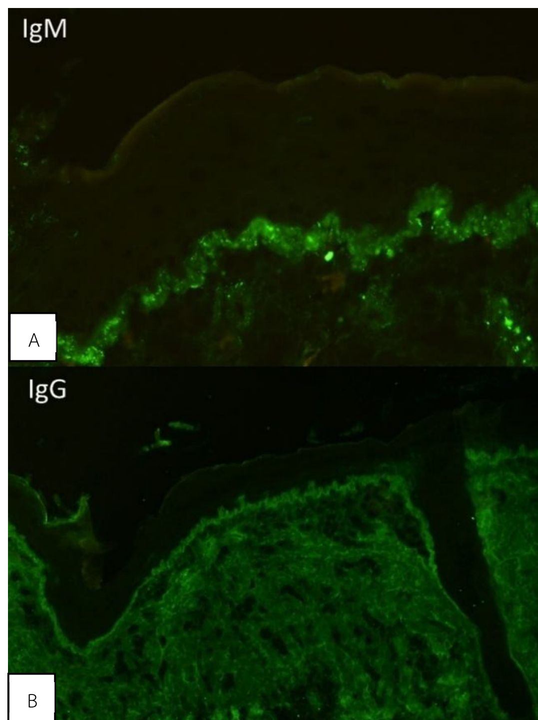

Figure 4 Direct immunofluorescent findings.
A,B) Homogenous granular IgM and IgG deposition along dermo-epidermal junction, superficial blood vessels and follicular epithelium.

Discussion

Our patient first presented with erosive lesions on both lid margins, consistent with chronic blepharitis and conjunctivitis. While the initial sign and symptoms were suggestive of autoimmune blistering disease such as MMP, dermoscopic, histopathologic and DIF were compatible with DLE.

Many dermatologic conditions can present with chronic blepharitis and conjunctivitis. MMP is one of the most common autoimmune disease presenting with cicatricial conjunctivitis[2]. Other autoimmune conditions comprise of epidermolysis bullosa acquisita, linear immunoglobulin A bullous dermatosis, pemphigus vulgaris and paraneoplastic pemphigus[2]. Drug-related blepharoconjunctivitis like Stevens-Johnson syndrome/toxic epidermal necrolysis could be excluded with absence of recent medication[3]. Other inflammatory diseases such as atopic/contact dermatitis, seborrheic dermatitis, connective tissue diseases, psoriasis, sarcoidosis, ichthyosis, infection and malignancy can also cause chronic conjunctivitis[3].

DLE of eyelids is an uncommon form of LE, described in <100 cases in the English literature. Because of its rarity, misdiagnosis with other dermatoses could delay treatment resulting in sequelae compromising the eyesight[4]. Time from the onset of symptoms to the diagnosis of eyelids DLE was approximately 41.40 months. Blepharitis is the most common finding (53.5%), especially on lower eyelid. Other presents are madarosis (28.2%), lid plaques (22.5%), lid edema (12.7%), erythematous macules and papules (11.3%) and less commonly hypertrophic or verrucous lesions. Most reports showed DLE of eyelid involved unilateral eye[5]. Other complications of periorbital DLE include lid scarring, ectropion, entropion and trichiasis[6]. Cornea involvement, such as stromal keratitis and punctate keratopathy, are less

frequent complications[7]. Wu et al. reported that 28% of periorbital cutaneous LE patients subsequently developed systemic disease[8]. Lupus-related serologies are often negative in patients with localized DLE. For DLE of the eyelids, ANA was negative in 58.33% of the patients. The speckled pattern is most prevalent, followed by nucleolar and diffuse patterns[7].

Due to the scarring nature of DLE, delay in diagnosis may lead to ocular complications. Dermoscopy may help accerate the diagnosis process with better precision. We reported the use of dermoscopy in examining DLE lesions specifically on eyelid skin and conjunctival surface. As reported in previous literature, the most frequent dermoscopic findings of DLE is perifollicular whitish halo, followed by follicular keratotic plugs, telangiectasia, white scales, pigmentation, structureless whitish areas and follicular red dots[9]. A previous case report described dermoscopic features on eyelid skin surface as erythematous background, telangiectatic vessels, whitish structureless areas and whitish scales. Salah et al, reported that the most common dermoscopic features of lip lesion was telangiectasia, followed by brownish pigmentation and scales, hairpin vessels and bleeding spots, storiform vessels and ulceration/erosions. Our dermoscopic features of DLE on the eyelid are somewhat comparable to the findings of previous eyelid and perioral DLE[10,11].

Treatment of eyelids DLE remains similar other areas. However, optimum care must be giving to prevent eye complications. Administration of systemic treatment is advised to eyelid DLE to prevent further damage compromising the eye sight. Antimalarial drugs are the first-line and the most commonly used systemic therapy for eyelids DLE. Hydroxychloroquine at a dose up to 6.5 mg/kg/day is considered the drug of choice. Short course of prednisolone (0.5-1 mg/kg/day) can be used in severe cutaneous LE with recommended tapering over 2-4 weeks[1]. Other drugs reported beneficial for general CCLE are oral mycophenolate, retinoids, methotrexate, azathioprine and cyclosporine[1,7]. In our case, dapsone at the dose of 50 mg/day in addition to hydroxychloroquine 200 mg/day were effective treatments for eyelid DLE.

In conclusion, blepharoconjunctivitis is an uncommon presentation of DLE and can often lead to misdiagnosis. Detailed examination and skin biopsy should be promptly done to aid early diagnosis, as delayed treatment may lead to further complications and serious morbidity of the effected eye. We emphasis that dermoscopy can be a valuable tool to support the diagnosis of DLE on the eyelid.

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