CASE REPORT

Cutaneous lupus erythematosus manifesting as unilateral eyelid erythema and swelling

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A R T I C L E   I N F O

Article history:
Received: Feb 1, 2011
Revised: Feb 21, 2011
Accepted: Mar 7, 2011

Keywords:
discoid lupus erythematosus
eyelid

A B S T R A C T

Unilateral eyelid involvement, as the only manifestation of discoid lupus erythematosus (DLE), is very rare. Here, we report the case of a 41-year-old woman who presented with erythema and swelling of her left upper eyelid, which was initially misdiagnosed as cutaneous lymphoid hyperplasia. She had initially received radiotherapy at another institute for treatment. Our examinations established the diagnosis of DLE, and direct immunofluorescence confirmed this diagnosis. The skin lesion improved after treatment with topical steroids and oral hydroxychloroquine.

Introduction

Chronic cutaneous lupus erythematosus (LE) is the most common type of cutaneous LE.1 According to the classification scheme developed by Gilliam and Sontheimer,2 there are many subtypes of chronic cutaneous LE, including classic discoid lupus erythematosus (DLE), hypertrophic LE, LE panniculitis/profundus, mucusal LE, LE tumidus, chilblain LE, and discoid LE-lichen planus overlap. Classical DLE is the most common clinical subtype of chronic cutaneous LE, and the localized form is far more common than the generalized form.3 Classic DLE lesions are characterized by erythematous and infiltrated plaques with hyperkeratosis. The most commonly involved areas are the face, V-region of the neck, and extensor surface of the arms.1 Patients with chronic cutaneous LE and palpebral involvement have been estimated to occur in up to 5% of the population, and these patients typically present with bilateral eyelid involvement and associated mucocutaneous lesions.4 Here, we report a case of DLE that manifested as unilateral upper eyelid erythema and swelling, which was initially misdiagnosed as cutaneous lymphoid hyperplasia at another institute and subsequently treated using radiotherapy.

A 41-year-old woman presented with erythema and swelling of her left upper eyelid that had initially presented more than 20 years previously. She had a past history of allergic rhinitis. She was initially treated with oral Chinese herb medicines, and the skin lesion subsided. However, the same lesion recurred about 3 years ago. There were no other mucocutaneous lesions or associated symptoms. Physical examination revealed erythematous swelling of her left upper eyelid with mild scaling and faint telangiectasia (Figure 1A). Orbital computed tomography revealed a diffuse infiltrative lesion in the left upper orbital area (Figure 2). A lacrimal gland biopsy performed at a medical center in the United States reported the lesion as a lymphoid hyperplasia with mild scaling and faint telangiectasia. Alcian blue stain, with and without hyaluronidase digestion, also detected the presence of mucin in the skin (Figure 3). These microscopic
findings were suggestive of LE. The direct immunofluorescence study revealed the granular deposition of IgG, IgM, IgA, C3, and C1q in the dermoepidermal junction, dermal vessels, and adnexal structures (Figure 4), which also supported the diagnosis of LE. The gene-rearrangement study, performed using fresh skin tissue, was polyclonal, thereby excluding the possibility of malignant lymphoma. Laboratory examinations, including complete blood cell and differential counts and analysis of anti-nuclear antibody anti-phospholipid antibody, complement 3 and 4 levels, anti-double stranded DNA, anti-RNA, anti-Sm, anti-Ro/La, and urinalysis were negative or within normal limits. The patient was diagnosed with localized DLE without systemic involvement. She received 400 mg/day of oral hydroxychloroquine and topical steroid cream. After 3 months of treatment, the amount of infiltrating plaque on her left upper eyelid had decreased (Figure 1A).

Discussion
DLE with unilateral eyelid involvement as the only manifestation is extremely rare and the manifestation could be atypical,1,4 which makes diagnosis difficult and may lead to delayed or inappropriate treatment. Ophthalmic complications of DLE include madarosis,
ectropion, scarring, synechiae, and blindness. Therefore, making the correct diagnosis is important for the prevention of further ophthalmic damage. However, due to its unusual presentation, many differential diagnoses of eyelid erythema and/or edema should be considered, such as blepharitis, contact dermatitis, seborrheic dermatitis, chronic eczema, ophthalmic rosacea, dermatomyositis, ocular trauma, cellulitis, angioedema, lymphedema, myxedema, lymphocytic infiltration, lymphoma, polymorphic light eruption, facial granuloma, sarcoidosis, and tinea. Diagnostic decisions can be challenging. Despite the fact that many diseases have similar clinical presentations, DLE should always be kept in mind in order to avoid inappropriate treatment. Only five similar cases have been reported (Table 1). The case described here and four other cases presented as erythema and/or edema of one eyelid, either the upper or lower part. One case presented as periorbital erythema and edema. Another case presented with ophthalmic symptoms (epiphora), while other cases and the case described here were asymptomatic. All cases were treated using antimalarial agents as monotherapy. One regimen of combination therapy showed a therapeutic response.

The treatment of cutaneous LE includes both topical and systemic treatments. The use of topical treatments, such as corticosteroids, calcineurin inhibitors, retinoids, imiquimod, R-salbutamol cream, laser, cryotherapy, and UVA-1 therapy, have been reported. Commonly prescribed systemic agents include antimalarials (quinacrine, chloroquine, and hydroxychloroquine), corticosteroids, methotrexate, retinoids, dapsone, thalidomide, intravenous immunoglobulins, and biological agents. At first, the patient's lesion was treated as cutaneous lymphoid hyperplasia with systemic prednisone and localized radiotherapy. The initial improper diagnosis and the use of radiotherapy, or an insufficient dose of systemic prednisone, may explain the poor treatment response. In addition, according to previous reports, topical corticosteroids and antimalarial agents are the treatment of choice for cutaneous LE, and topical corticosteroids, when used as a mono-therapy, are not adequate. We used both topical corticosteroid and

| Case Age Sex Clinical Pathology Treatment and Follow-up |
|-----------------|-----------------|---------------------------------|--------------------------------------------------|
| Donzis et al7 42 M Periorbital edema and violaceous discoloration of left eye Vacular degeneration of basal layer, perivascular and periappendageal lymphocyte infiltration IgG, IgM, and deposition of complement at D-E junction Edema and erythema subsided after 4 weeks of oral hydroxychloroquine |
| Tosti et al8 45 F Purple-red, slightly scaly lesion on the upper eyelid of right eye Epidermal atrophy, focal vacuolization of the basal layer of the epidermis, and infundibular follicular epithelium. Edema, telangiectases, and superficial and deep perivascular and focal perifollicular lymphohistiocytic infiltrate Granular deposition of IgG, IgM, IgA, and C3 along the basement membrane zone Response to chloroquine phosphate after treatment for 1 mo Treatment with chloroquine phosphate resulted in complete regression within 2 mo |
| Cryan et al9 53 F A small erythematos and scaly patch on the upper eyelid of left eye No detailed description of the histological findings or lupus band test were reported Granular deposition of IgG, IgM, IgA, C3 and C1q along the D-E junction Improved after treatment with systemic prednisolone, mycophenolate mofetil, and hydroxychloroquine |
| Ricotti et al9 38 F Asymptomatic left-sided lower eyelid edema and erythema Interface dermatitis associated with follicular involvement, superficial and deep perivascular lymphocytic infiltrate, increased dermal mucin deposition Dense granular IgM, C3, IgG, and C5b-9 deposits along the epidermal and adnexal basement membrane zone Response to systemic prednisolone, mycophenolate mofetil, and hydroxychloroquine |
| Present case 41 F Erythema and edema of left upper eyelid was the only manifestation Follicular plugging, basal layer vacuolar degeneration, perivascular and perifollicular lymphocytic infiltration, and dermal mucin deposition Granular deposition of IgG, IgM, IgA, C3, and C1q along the D-E junction Improved after treatment with topical steroids and oral hydroxychloroquine for 3 mo |

Table 1 Reported cases of discoid lupus erythematosus with unilateral eyelid involvement as the only symptom.

M, male; F, female; D-E junction, dermoepidermal junction.

Figure 4 Direct immunofluorescent images showing granular deposits of (A) IgG and (B) C3 at the dermoepidermal junction and around a hair follicle.
antimalarial agents and obtained a good treatment response. The patient did not have any apparent ophthalmic complications except mild lacrimal gland destruction. She did not have any clinical eye symptoms. The patient needed continuous ophthalmologic follow-up due to the destruction of her lacrimal gland and the use of hydroxychloroquine.

In conclusion, here we report a patient who presented with DLE as unilateral eyelid involvement as the only manifestation. Suspicion regarding this unusual presentation, along with the results of the histopathological and immunofluorescence studies, helped to establish the proper diagnosis and avoid unnecessary treatments.

References


