Isolated Unilateral Periorbital Chronic Cutaneous Lupus Erythematosus: A Case Report and Literature Review

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ABSTRACT:
Chronic cutaneous lupus erythematosus (CCLE) represents the most prevalent subset of cutaneous lupus erythematosus and is well known for its predilection for photo-exposed areas, particularly the face and scalp, as consistently demonstrated in large clinical series and dermatologic epidemiologic studies. Periorbital involvement, particularly as an isolated manifestation, is exceedingly rare and diagnostically challenging. Here, we report a 29-year-old man presenting with a one-year history of recurrent isolated right periorbital swelling. Histopathological examination revealed interface dermatitis with basal vacuolization and lymphoplasmacytic infiltration, while Direct Immunofluorescent (DIF) showed granular deposition of C3 at the dermoepidermal junction. Despite negative antinuclear antibody (ANA) results, the patient responded favorably to hydroxychloroquine and topical corticosteroids. This case highlights the importance of maintaining a high clinical suspicion and utilizing DIF in diagnosing ANA-negative chronic cutaneous lupus erythematosus (CCLE) with atypical presentations.
Key words: Chronic Cutaneous Lupus Erythematosus, Periorbital Swelling, Chronic Cutaneous Lupus, Eyelid Dermatosis, Cutaneous Lupus Erythematosus

Introduction

Cutaneous lupus erythematosus (CLE) represents a broad spectrum of lupus-related skin manifestations and is commonly classified into acute, subacute, and chronic subtypes[1-3]. chronic cutaneous lupus erythematosus (CCLE) includes several variants, such as discoid lupus erythematosus (DLE), tumid lupus, and lupus panniculitis. Because these subtypes may overlap both clinically and histopathologically, careful clinicopathological correlation is essential for accurate classification. Discoid lupus erythematosus (DLE) is the most common form of chronic cutaneous lupus erythematosus (CCLE), classically affecting sun-exposed areas such as the face and scalp[2]. However, atypical presentations involving less common sites, such as the periorbital region, can occur and pose diagnostic challenges, as illustrated in this case.

Case Presentation

A 29-year-old Thai man employed as a civil engineer presented to our dermatology clinic with a one-year history of recurrent right periorbital swelling. The lesion waxed and waned, often worsening after sun exposure or periods of sleep deprivation. There was no associated pruritus, pain, numbness, or visual disturbance. He denied systemic symptoms, including fever, oral ulcers, arthralgia, photosensitivity, alopecia, or weight loss. His past medical history was unremarkable, and there was no family history of autoimmune disease.

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Dermatologic examination revealed a localized, ill-defined erythematous patch with mild edema involving the right upper and lower eyelid, extending to the medial canthus. The lesion was non-scaly, non-ulcerative, and non-tender. The left periorbital area and other parts of the face and body were unaffected. There were no oral ulcers, scalp lesions, or lymphadenopathy.



Figure 1 Clinical presentation of the patient
A: Close-up view, ill-defined erythema and edema of the right upper eyelid
B: Frontal view, isolated right periorbital involvement. No lesions on the contralateral side

The initial differential diagnoses included periorbital eczema, contact dermatitis, dermatomyositis, sarcoidosis, tumid lupus erythematosus, cutaneous pseudolymphoma, mixed connective tissue disease, and discoid lupus erythematosus. Given the chronicity and lack of response to emollients and mild topical corticosteroids, a skin biopsy was performed.

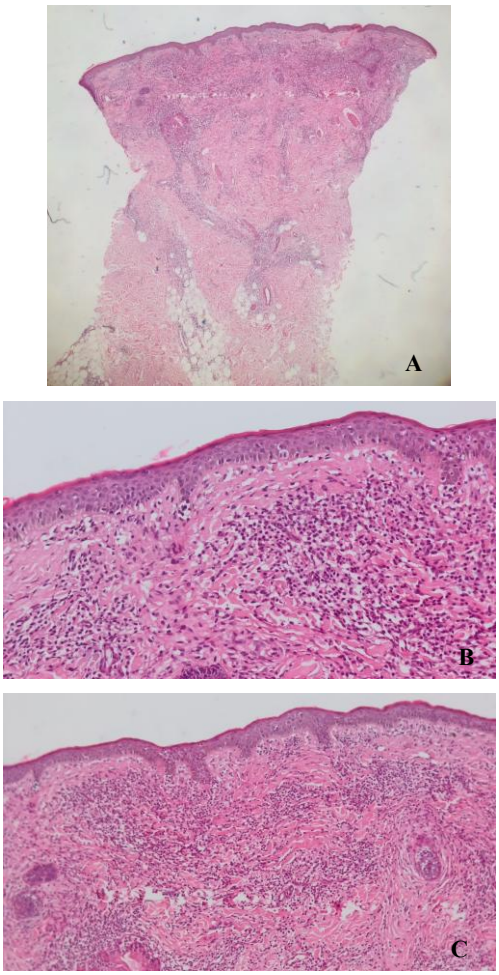


Figure 2 Histopathologic findings of the eyelid lesion
A: Epidermal atrophy, superficial and deep lymphoplasmacytic infiltration (H&E, 40X)
B: Basal cell vacuolization and thickened basement membrane (H&E, 200X)
C: Dense perivascular, periadnexal infiltration without mucin deposition (H&E, 100X)

Histopathologic examination from a 4-mm punch biopsy of the right upper eyelid demonstrated an atrophic epidermis with basal cell vacuolization, thickening of the basement

membrane, and a dense lymphoplasmacytic infiltrate in both superficial and deep perivascular and periadnexal areas. No follicular plugging or necrotic keratinocytes were observed. The Alcian blue stain was negative for dermal mucin. DIF revealed granular C3 deposition at the dermoepidermal junction in a band-like pattern. These findings were consistent with CCLE[4].

Laboratory investigations, including complete blood count, liver and renal function tests, erythrocyte sedimentation rate, and C-reactive protein, were within normal limits. The ANA test was negative. Anti-dsDNA, anti-Ro/SSA, anti-La/SSB, anti-Sm, and antiphospholipid antibodies were also negative. Serologies for hepatitis B, C, and HIV were unremarkable.

Based on the clinical and histopathological findings, a diagnosis of ANA-negative CLE limited to the right periorbital area was made. The patient was initiated on oral hydroxychloroquine 200 mg twice daily and prescribed 0.1% mometasone furoate cream for topical application. He was advised on strict photoprotection and scheduled for regular follow-up.

At the 6-week follow-up, moderate improvement in erythema and swelling was noted. By 3 months, the lesion improves and gradually subsides. The patient continued hydroxychloroquine treatment with regular ophthalmologic monitoring and showed continuous improvement of the lesions at the 3-month follow-up.

Discussion

Periorbital lupus erythematosus is an infrequent presentation of CLE and can be easily overlooked. Eyelid involvement is observed in approximately 6% of cutaneous lupus cases; however, isolated periorbital CLE is exceedingly rare, with fewer than 15 cases documented to date[5,6]. Makeeva et al. reported six such instances in which periorbital edema was the sole presenting feature, frequently misdiagnosed as angioedema or dermatitis[5]. Most of these patients were ANA-negative, and all responded favorably to hydroxychloroquine.

The pathogenesis of CLE is complex, involving genetic susceptibility, environmental triggers, and immune dysregulation. Ultraviolet radiation is a known inciting factor that leads to the apoptosis of keratinocytes, releasing nuclear antigens into the extracellular space. Impaired clearance of these apoptotic bodies, especially in individuals with defective complement or phagocytic pathways, promotes the formation of immune complexes. These complexes can activate plasmacytoid dendritic cells (pDCs) through toll-like receptor (TLR) pathways, leading to robust type I interferon (IFN) production and perpetuation of local inflammation[5-7].

Histologically, CCLE exhibits interface dermatitis with basal vacuolar changes, a thickened basement membrane zone, and dense perivascular and periadnexal lymphoplasmacytic infiltrates. Mucin deposition may be variably present in the dermis. DIF often reveals the granular deposition of immunoglobulins and complement at the dermoepidermal junction, termed the lupus band test[8,9]. This is especially helpful in cases with negative serologic markers.

Histopathologically, differentiation from other conditions such as tumid lupus, dermatomyositis, or cutaneous sarcoidosis is essential. Tumid lupus, for instance, shows abundant mucin deposition, absence of epidermal changes, and perivascular/periadnexal infiltrate without significant basal vacuolar change[2,6]. Dermatomyositis can involve the eyelids, presenting as heliotrope rash, but it typically presents with muscle symptoms and positive myositis-specific antibodies. Sarcoidosis demonstrates non-caseating granulomas, which were absent in our case[8].

The pathophysiology of CLE involves the interplay of ultraviolet light-induced keratinocyte apoptosis, release of nuclear antigens, pDC activation, and IFN-driven inflammation[10,11]. Autoantibodies and immune complexes enhance tissue damage, although ANA negativity does not exclude the disease. According to recent international guidelines on cutaneous lupus erythematosus, skin biopsy and DIF are recommended in atypical or isolated presentations to help establish an accurate diagnosis[12].

Therapeutic strategies aim to suppress inflammation and prevent scarring. Hydroxychloroquine remains the first-line agent due to its immunomodulatory and photoprotective effects[11]. Topical corticosteroids or calcineurin inhibitors are effective adjuncts to treatment. Methotrexate, mycophenolate mofetil, or dapsone may be considered for refractory disease. Belimumab and anifrolumab, which target B-cell activation and type I IFN pathways, respectively, have shown promise in systemic lupus and are under study in CLE[13,14].

In our patient, the combination of oral hydroxychloroquine and topical mometasone furoate led to improvement of the lesion within three months. This clinical course is consistent with previously published case reports. Makeeva et al. described six patients with isolated periorbital CLE who were mostly ANA-negative and initially misdiagnosed as having allergic or eczematous conditions but responded well to hydroxychloroquine after histopathologic confirmation[5]. Similarly, Ghaninejad et al. reported a case of persistent periorbital edema as the sole manifestation of CLE, which showed dramatic improvement following antimalarial therapy[6].

This case reinforces the importance of maintaining a high clinical suspicion in atypical cases. It highlights the pivotal role of skin biopsy and DIF in guiding early diagnosis and effective treatment. Early initiation of hydroxychloroquine, combined with topical corticosteroids, should be considered to prevent disease progression. The absence of systemic symptoms or serologic markers such as ANA underscores the need for clinicians to consider CCLE in the differential diagnosis of chronic periorbital swelling. In such presentations, histopathologic examination and DIF are essential not only for diagnosis but also for avoiding treatment delays and minimizing long-term morbidity.

Conclusion

Periorbital CLE is an uncommon and often misdiagnosed presentation of chronic cutaneous lupus erythematosus. This case highlights the diagnostic challenges and emphasizes the utility of histopathology and DIF in establishing a diagnosis, whether ANA is positive or not. Prompt recognition and appropriate therapy with hydroxychloroquine and topical corticosteroids can result in complete remission and prevent long-term sequelae.

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