Transient blindness with periorbital erythema and swelling: Manifestations of recurrent systemic lupus erythematosus

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Key words: cutaneous lupus erythematosus; lupus erythematosus; perineuritis; periorbital lupus erythematosus; transient visual blindness.

INTRODUCTION

Lupus erythematosus (LE) is a multisystem disorder that can prominently affect the skin and cause significant internal abnormalities.[1] According to the most widely used classification of the cutaneous manifestations of LE by Gilliam and Sontheimer,[2] the skin lesions can be segregated into those that are LE specific and LE nonspecific based on whether the histology shows the presence or absence of interface dermatitis, respectively; the category of LE-specific cutaneous lesions can be further classified into acute, subacute, or chronic LE.[2] Periorbital swelling and erythema is an uncommon, poorly characterized, frequently misdiagnosed, and often treatment-resistant cutaneous manifestation of LE.[3,4] This case describes the exceedingly rare phenomenon of recrudescence of previously quiescent systemic lupus erythematosus (SLE) as periorbital cutaneous lupus erythematosus, posterior scleritis, and optic perineuritis.

CASE REPORT

A 42-year-old woman with a medical history significant for SLE complicated by biopsy-proven class IV lupus nephritis in 2011 (treated with prednisone and hydroxychloroquine, subsequently in remission on no maintenance medications), psoriasis, and a year of persistent left periorbital rash of unclear etiology presented to the emergency room with 3 episodes of transient left eye blindness occurring over the preceding 2 days.

Abbreviations used:

LE: lupus erythematosus
SLE: systemic lupus erythematosus

Each successive episode of vision loss occurred for longer periods (range, 1-10 minutes) and resolved spontaneously. She denied oral ulcers; hair loss; joint pain, swelling, or stiffness; chest pain; shortness of breath; muscle weakness; or hematuria. Computed tomography scan of the face with contrast found increased soft tissue edema around the left eye, and MRI of the face and orbits with and without contrast found enhancement of the left optic nerve sheath and posterior scleritis. Magnetic resonance angiography of the head and neck showed widely patent vessels with no abnormalities. Both the neurology and ophthalmology departments were consulted, and no abnormalities were detected during the neurologic and ophthalmologic physical examinations. Subsequent cardiovascular workup to investigate for vascular etiologies of amaurosis fugax, including echocardiography and mobile outpatient cardiac telemetry, were unrevealing.

The dermatology service was consulted for the patient's ongoing, intermittent left periorbital rash, which began as an erythematous papule that then spread to involve her left upper and lower eyelids over the course of approximately 1 year. She had no

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Funding sources: None.
Conflicts of interest: None disclosed.
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JAAD Case Reports 2019;5:1088-90.
2352-5126

https://doi.org/10.1016/j.jdcr.2019.10.009

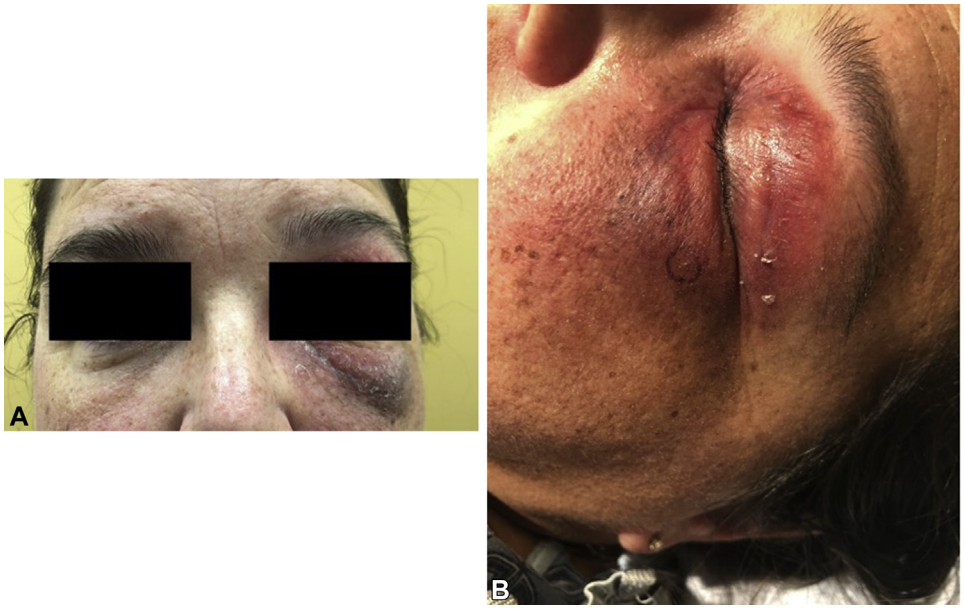

Fig 1. A, Left periorbital swelling and erythema with overlying scale. **B**, Well-demarcated edematous, erythematous plaque on the left upper and lower eyelids. Site of punch biopsy is indicated by circle in marker ink.

pruritus or pain but reported a pins and needles–like sensation in the area of the rash. Over the prior year, she had several evaluations for the left periorbital rash, and the differential diagnosis included contact dermatitis and psoriasis. Patch testing was negative, and she denied taking any new medications prior to the onset of the rash. She was not applying topical medications at the time of presentation, although she reported the rash worsened when she was previously treated with hydrocortisone cream and tacrolimus ointment by local dermatologists. Physical examination found a well-demarcated, scaly, edematous purple-gray plaque centered on the left upper and lower eyelids (Fig 1). A skin biopsy was performed, and histologic sections showed interface dermatitis of the epidermis and follicular epithelium with dyskeratotic keratinocytes, follicular plugging, pigment incontinence, and perivascular chronic inflammation (Fig 2, *A*). A periodic acid–Schiff–diastase stain found focal basement membrane zone thickening, whereas a CD123 immunostain highlighted plasmacytoid dendritic cell aggregates (Fig 2, *B*). Laboratory analysis at the time of presentation also found an elevated antinuclear antibody titer of 1:160 and a positive test for anti–double-stranded DNA antibodies. Otherwise, the patient's complete blood count, comprehensive metabolic panel, complement levels, lupus anticoagulant, and urinalysis with microscopy were within normal limits, and the patient had negative results for anti-Smith, anti-U1RNP, anti-Ro/SSA, anti-La/SSB, anti-cardiolipin, and anti–β-2-glycoprotein antibodies.

Given the histologic findings present in the biopsy, the patient's periorbital rash was thought to be consistent with cutaneous lupus. Her transient vision loss was attributed to optic perineuritis and posterior scleritis associated with SLE. She was placed on a prednisone taper, azathioprine, and hydroxychloroquine. Because of her prior intolerance to high-dose prednisone including significant mood changes and weight gain, she was started on a relatively low dose of prednisone, 20 mg daily, and tapered slowly over 6 months. During this time, her periorbital rash improved, and she had no further episodes of vision loss.

DISCUSSION

In this case, the patient had previous systemic manifestations (nephritis, arthritis, pleurisy) of SLE, but she did not have previous skin symptoms prior to her periorbital rash. In fact, because she was otherwise symptom free, her SLE was thought to be in remission, and she had discontinued her medications several years prior. To the best of our knowledge, this case may represent the first instance of recrudescence of previously quiescent SLE as periorbital LE. Moreover, posterior scleritis and optic perineuritis, which is a rare inflammatory disorder characterized by optic nerve sheath inflammation and visual loss, have only occasionally been linked

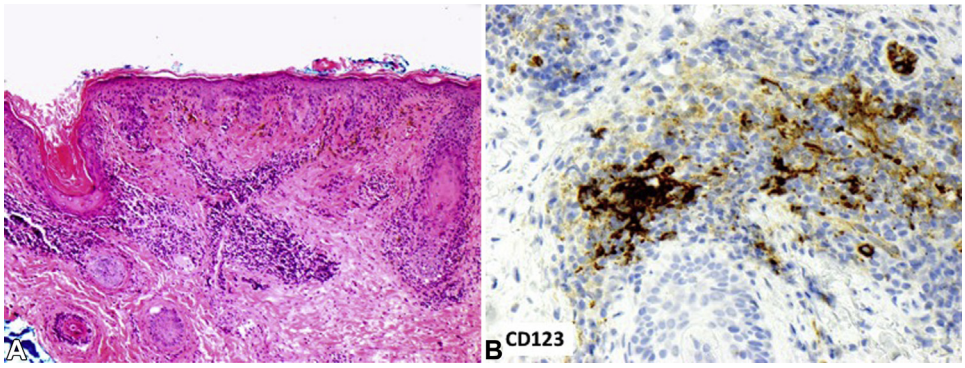

Fig 2. **A**, The left periorbital skin biopsy found epidermal and follicular interface dermatitis with follicular plugging, perivascular lymphocytic infiltrate, and pigment incontinence. **B**, Plasmacytoid dendritic cell aggregates were identified. (**A**, Hematoxylin-eosin stain; **B**, CD123 immunohistochemistry. Original magnifications: **A**, ×100; **B**, ×400.)

to SLE.[5] Unlike other cases of gradual onset of visual symptoms, this case may represent the first report of sudden optic perineuritis preceded by longstanding periorbital LE.

In a retrospective analysis of 553 patients with cutaneous manifestations of LE from 3 tertiary referral medical centers, 25 patients had periorbital erythema and swelling as a cutaneous manifestation of LE.[3] Of these 25 patients, all had histopathologic features of LE, 68% were women, mean age was 46.7 years, and 84% had unilateral periorbital involvement.[3] Periorbital LE was refractory to treatment in 63.6% of the 25 patients after an average follow-up period of 219.3 weeks. In addition, 27.3% of the patients eventually had SLE,[3] which is a higher conversion rate than the reported rate of 10% to 20% for localized discoid lupus erythematosus.[1,4]

Periorbital LE is a skin manifestation of either acute, subacute, or chronic cutaneous LE given the consistent presence of interface dermatitis, as documented in several reports.[3,6,7] Heightened awareness of periorbital LE is important because it is misdiagnosed in 76% of patients,[3] as was the case in this patient. Although the clinical and histologic features overlap with other forms of chronic cutaneous LE, periorbital LE is more frequently associated with SLE and is more refractory to topical treatments, warranting systemic antimalarial and immunosuppressive medications in most instances to achieve clinical resolution.

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