Bilateral periorbital swelling as the initial presentation of cutaneous lupus erythematosus

Valeria Makeeva, BS,[a] Lucia Seminario-Vidal, MD, PhD,[b] Kathleen Beckum, MD,[c] and Naveed Sami, MD[c]
Birmingham, Alabama and Tampa, Florida

Key words: angioedema; lupus erythematosus; periorbital edema.

INTRODUCTION

Cutaneous lupus erythematosus (CLE), defined as isolated cutaneous lesions without significant systemic involvement, has a reported incidence and prevalence of 4.3 and 73.2 per 100,000, respectively.[1] It occurs more commonly among women with a reported age of onset between the second and fourth decades of life.[1] The 3 more common subtypes of lupus are systemic lupus erythematosus (SLE), subacute cutaneous lupus, and discoid lupus erythematosus. A subset of patients shows atypical clinical manifestations of CLE, which are more rare subtypes and pose a diagnostic and therapeutic challenge. Antimalarial drugs are considered the mainstay of systemic therapy. Because it may take 2 to 3 months to note some improvement,[2] oral or topical steroids are used in conjunction to facilitate response. We report a patient with an atypical presentation of CLE who also responded to hydroxychloroquine and review similar cases in the literature.

Abbreviations used:

ANA:	antinuclear antibody
BID:	twice a day
CLE:	cutaneous lupus erythematosus
DIL:	drug-induced lupus
SLE:	systemic lupus erythematosus

CASE REPORT

A man in his 40s presented with persistent periorbital swelling for 7 months. The swelling first appeared 2 days after stripping floors during his work. He denied any associated pruritus, oral symptoms, facial pain, dyspnea, fever, weight changes, myalgias, arthralgias, weakness, or fatigue. He also denied coming into contact with any new products on his face or any new medications. He had a history of asthma but no history of allergies or photosensitivity. He also had eczema, hypertension, and depression well controlled with triamcinolone cream, amlodipine, and doxepin, respectively. He used ibuprofen about 4 times a week for tension headaches for several years without any adverse reactions, but he stopped using it within days of noticing the periorbital swelling. However, the swelling did not improve. A few months after, at an outside hospital, he was given a diagnosis of sinusitis and treated with amoxicillin/clavulanate and systemic steroids. Symptoms initially improved but relapsed when prednisone was tapered to less than 20 mg.

After 7 months of having the above symptoms and lack of improvement of the swelling, he presented to our service. Review of systems yielded normal results other than the indicated above. Physical examination found edema and erythema of the right lower and upper eyelids and on the left upper eyelid (Fig 1).

The following laboratory investigations were within normal limits: complete blood count, complete metabolic profile, C-reactive protein, creatine kinase, IgE levels, C4, C1-INH, C1q, and total hemolytic complement. Results for antinuclear antibody (ANA), anti-Sjögren's syndrome–related antigen A and antigen B, antineutrophil cytoplasmic antibody, antimicrosomal antibody, antihistone antibody, aldolase, anti-Jo1, and anti-Mi2 antibody were normal. Anti-dsDNA level was quantified at 45 IU/mL (nl < 4 IU/mL), and anti-Smith level was 1 AI (nl < 1 AI).

From the University of Alabama School of Medicine[a]; the Department of Dermatology, University of South Florida[b]; and the Department of Dermatology, University of Alabama at Birmingham.[c]

Funding sources: None.
Conflicts of interest: None declared.

Correspondence to: Naveed Sami, MD, Department of Dermatology, EFH 414, 1530 3rd Ave S, Birmingham, AL 35294. E-mail: nsami@uab.edu.

JAAD Case Reports 2016;2:72-6.
2352-5126

http://dx.doi.org/10.1016/j.jdcr.2015.12.009

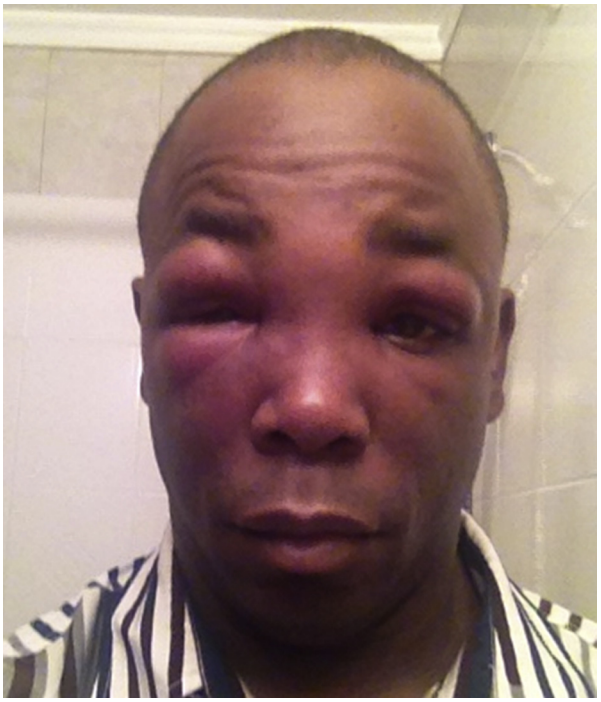

Fig 1. Clinical presentation. Marked edema and erythema of the right and left eyelids, with diminished palpebral aperture.

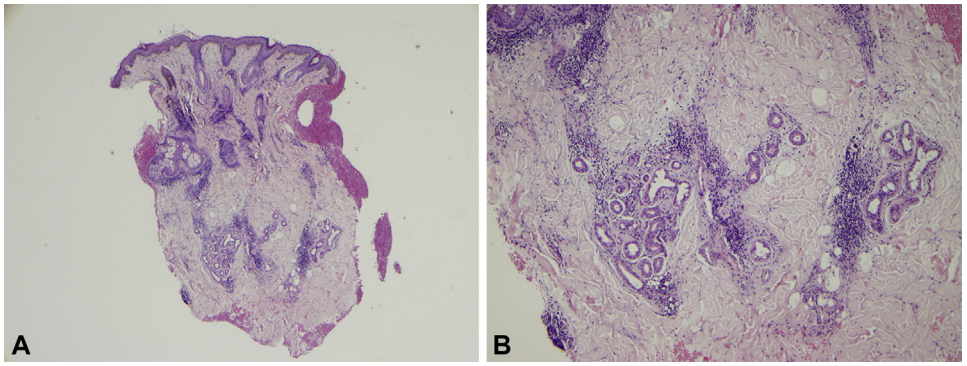

Fig 2. **A**, Superficial and deep lymphocytic infiltrate perivascular and periadnexal infiltrate. **B**, Deep periadnexal lymphocytic infiltrate with increased dermal mucin. (Original magnifications: A, ×40; B, ×100.)

Angiotensin-converting enzyme levels, chest radiograph, and urinalysis were normal. A maxillofacial computed tomography scan found soft tissue swelling in the right lacrimal fossa with no focal abscess. Biopsy results of the right lower eyelid are shown in Fig 2.

He was further evaluated by the rheumatology department, and no systemic findings were noted. Our patient's clinical presentation and histopathologic findings were consistent with a diagnosis of tumid lupus, a rare variant of CLE. Therefore, he was initially treated with oral prednisone (40 mg/d) and hydroxychloroquine (400 mg/d). The prednisone dose was tapered and discontinued over 2 months with complete resolution of the periorbital edema. Our patient remains under control and without a relapse of cutaneous disease or development of systemic symptoms on hydroxychloroquine 400 mg/d at his 1-year follow-up.

DISCUSSION

Periorbital edema is a rare and nonspecific presentation for several conditions including SLE, dermatomyositis, solid facial edema, drug reactions (including drug-induced lupus [DIL]), infections, angioedema, systemic contact dermatitis, superior vena cava syndrome, hypersensitivity reactions, sarcoidosis, and others.[3] Laboratory, histopathology, and imaging studies ruled out these diagnoses in our patient.