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CASE REPORT

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Periorbital swelling and episcleritis may be a sign of cutaneous lupus erythematosus

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Abstract

It is important that physicians recognize persistent periorbital edema and conjunctival injection as possible disease manifestations of CLE, especially prior to cutaneous involvement. This may lead to more rapid diagnosis and treatment.

KEYWORDS

cutaneous lupus erythematosus, episcleritis, lupus erythematosus tumidus, periorbital edema

1 | CASE REPORT

A 54-year-old female initially presented to the ophthalmology clinic with a 5-day history of right upper eyelid swelling and temporal pain. Her ocular history included nasal pterygia and chronic dry eye. On physical exam, there was tenderness of the lacrimal gland and erythema on eversion of the right upper eyelid. The differential diagnosis included infectious or allergic periorbital edema. Her ESR was normal, and she was prescribed oral azithromycin. Three weeks later, the patient's right periorbital edema worsened despite antibiotics and she had new onset tenderness of the right lacrimal gland. A computerized tomography of the orbits demonstrated right lacrimal gland inflammation and periorbital soft tissue swelling. She was started on prednisone 60 mg daily.

Two weeks later, she presented to the oculoplastics service with persistent symptoms and new onset conjunctival injection of the right eye (Figure 1). On exam, the right conjunctiva blanched with phenylephrine application. A diagnosis of dacryoadenitis and episcleritis was made, and the patient was prescribed clindamycin 300 mg three times daily for 10 days, and she continued her prednisone taper. The patient presented 2 months later with worsening periorbital edema in addition to a sudden eruption on her right periorbital skin, cheek, and chest prompting referral to dermatology.

The dermatology service noted erythematous, smooth, indurated plaques on the right upper eyelid, right cheek, and one plaque on the right chest and right calf (Figure 1). A skin biopsy demonstrated a superficial and deep, dermal perivascular lymphocytic infiltrate with increased dermal mucin. She then developed rapid progression of her rash to the chest, back, and upper extremities within a week. The patient noted tenderness and pruritus of these lesions.

Repeat skin biopsies of the right cheek and calf lesions were characteristic of cutaneous lupus erythematosus (Figure 2). Findings from all other laboratory workups, including

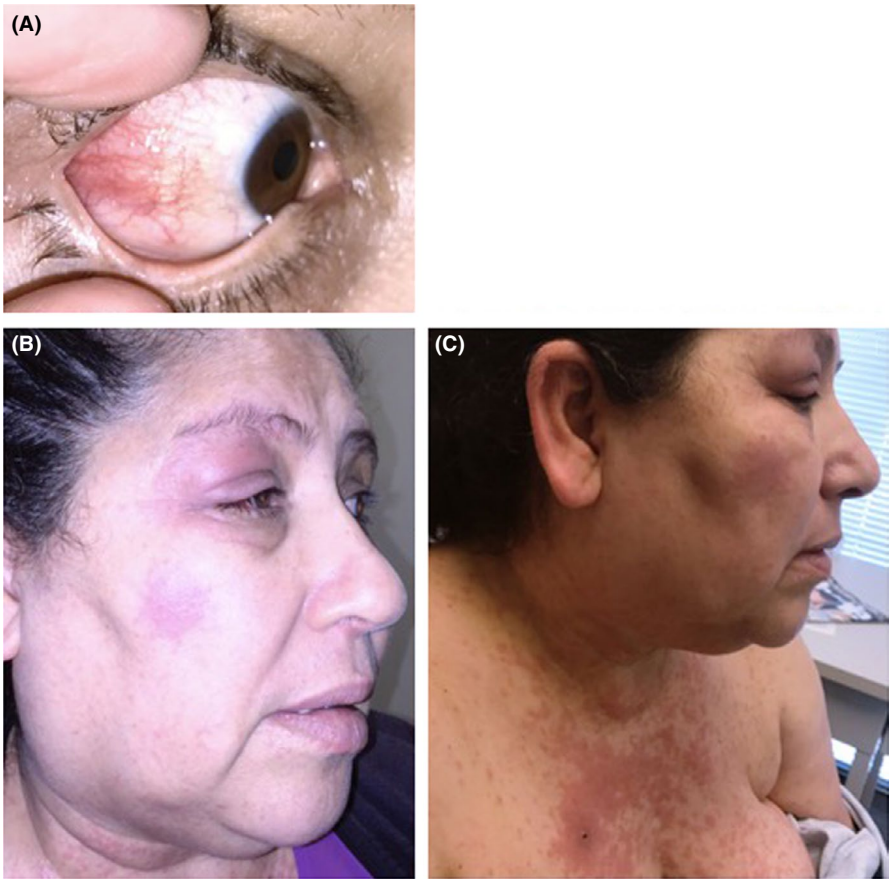

FIGURE 1 A, Episcleritis of the right eye. B, Right periorbital edema and erythema with erythematous, smooth indurated plaque on right cheek. C, Erythematous, smooth indurated plaques on the chest

complete blood count, comprehensive metabolic panel, urinalysis, antinuclear antibody, anti–double-stranded DNA, extractable nuclear antigen, C3 and C4 complements, CK, aldolase, and rheumatoid factor, were unremarkable. A diagnosis of lupus erythematosus tumidus (LET) was made based on lesion morphology, supportive histopathology, and negative laboratory workup. The patient was started on hydroxychloroquine 200 mg twice daily and topical steroids.

Over the next 3 months, patient continued to have skin involvement, right eyelid swelling, and erythema with new onset of polyarticular joint pain. Joint pain was present in bilateral knees, wrists, and fingers without swelling or erythema. She was prescribed methotrexate 7.5 mg and folic acid 1 mg, and continued on hydroxychloroquine. Patient was referred to rheumatology who determined that patient did not meet ACR criteria for SLE,[1,2] and joint pain was consistent with osteoarthritis.

She began to improve with resolution of skin lesions with no scarring after 8 months on methotrexate 10 mg and hydroxychloroquine 200 mg twice daily. The episcleritis and dacryoadenitis of the right lacrimal gland also eventually resolved.

Informed consent for publication of photographs was obtained from the patient.

2 | DISCUSSION

Cutaneous lupus erythematosus (CLE) can be divided into three forms: acute cutaneous lupus erythematosus (ACLE), subacute cutaneous lupus erythematosus (SCLE), and chronic cutaneous lupus erythematosus (CCLE). Lupus erythematosus tumidus (LET) is a less common variant of chronic cutaneous lupus erythematosus. The prevalence and incidence of LET is not well known but affects females more than males with onset mostly in the 3rd to 4th decade. Patients usually do not have systemic involvement and LET rarely coincides with SLE. The cutaneous lesions of LET are described as erythematous, succulent, smooth, urticarial-like plaques located in sun-exposed areas. In contrast to other types of CLE, LET lesions do not demonstrate scarring, atrophy, dyspigmentation, or follicular plugging.[3] Histologically, LET has a superficial and deep perivascular and periadnexal infiltrate with abundant dermal mucin deposition.[3,4] Some cases of LET have also shown vacuolar interface alterations.[4]

Periorbital edema is a rare presentation of CLE.[5-9] A review of 77 patients with CLE and ocular findings by Arrico et al[10] revealed that the main ocular site involved

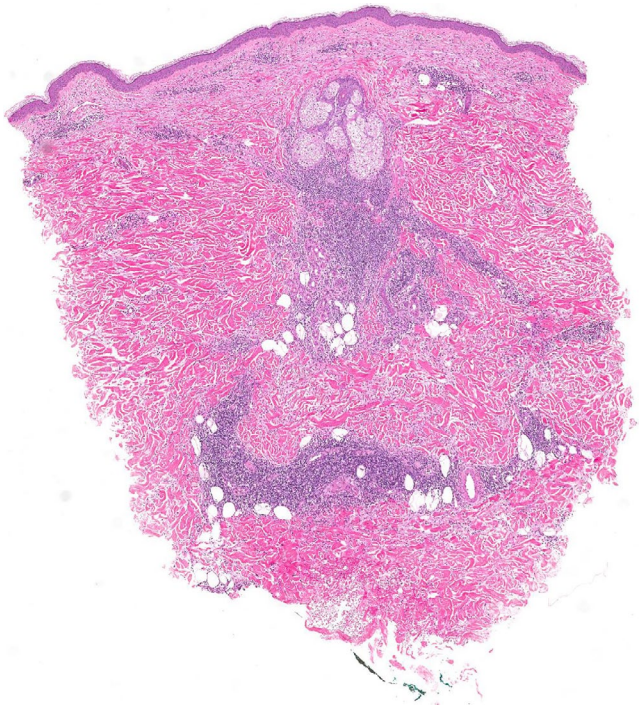

FIGURE 2 Histopathologic Findings From Lesional Specimens (hematoxylin and eosin, 1.4 X): Vacuolar interface alterations with superficial and deep dermal perivascular perifollicular lymphocytic infiltrate and increase in reticular dermal mucin

is the eyelids in 63 patients (88.7%) followed by orbit and periorbital tissues in six (8.4%) and cornea in two (2.6%). These findings were most common in discoid lupus erythematosus (DLE) (92.2%) followed by lupus erythematosus profundus (7.8%).[10]

Episcleritis and scleritis of the eye can be a feature of systemic lupus erythematosus (SLE) and rheumatoid arthritis.[11] Our patient did not have any other local or systemic clinical features and failed to meet the ACR criteria for SLE. Burge et al[12] reported that episcleritis occurred in nine percent of patients with SLE and not seen in chronic cutaneous lupus erythematosus (CCLE). Several cases have documented eyelid involvement in CLE, but there is no report of conjunctival involvement. Therefore, our patient is a unique case of episcleritis in CLE.

Cutaneous lupus erythematosus typically responds to antimalarial drugs, topical and oral corticosteroids, and photoprotection. Our patient had complete resolution of skin, without scarring or atrophy, and ocular findings while on hydroxychloroquine, topical steroids, an oral prednisone taper, and methotrexate. She is currently on hydroxychloroquine and methotrexate and remains in remission.

This report demonstrates the difficulty in making the diagnosis of CLE when the presenting features are periorbital edema and episcleritis in the absence of cutaneous findings. In our case, the diagnosis of CLE was made only after the patient presented with widespread cutaneous lesions. Therefore, it is important that physicians recognize persistent periorbital edema and conjunctival injection as possible disease manifestations of CLE. This may lead to more rapid diagnosis and treatment.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

TZ: contributed to the conception and design, collected and analyzed the data, and drafted and revised the manuscript. MP: involved in clinical care of patient, contributed to drafting the manuscript, and critically revised the manuscript. CH, KE, and AL-S: involved in clinical care of patient, contributed important intellectual content during manuscript revision, and critically revised the manuscript. PG: provided histopathology image and interpretation, and revised the manuscript. All authors approved the revised and final version of the manuscript.

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