

2020 MDS Annual Meeting - Clinical Case with Advanced Teaching Points

Title: Unilateral periorbital eruption as a manifestation of systemic lupus erythematosus

Authors: Sepideh Ashrafzadeh¹, BS, Cristina Thomas¹, MD, Julia A Benedetti¹, MD
1-Harvard

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Learning points:

- Cutaneous manifestations of systemic lupus erythematosus (SLE) are varied. A rare presentation of SLE, as described here, is unilateral periorbital erythema and edema.
- Because the differential of unilateral periorbital erythema and edema is broad, histopathology and laboratory testing can assist in the diagnosis of this unusual manifestation of SLE.
- Early recognition of this rare presentation is critical to the timely initiation of appropriate treatment and prevention of potential organ damage.
- Recurrent SLE may present as only a periorbital eruption even though prior SLE episodes may not have had skin involvement.

Abstract:

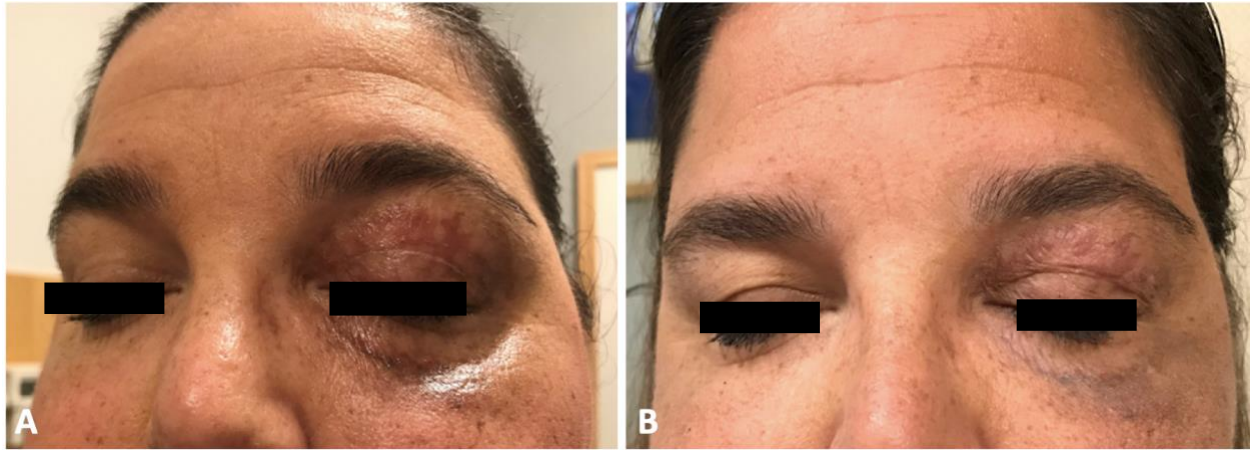
A 43-year-old woman with systemic lupus erythematosus (SLE), psoriasis, and Ehlers-Danlos syndrome presented with a one-month history of a left periorbital erythematous plaque associated with a burning sensation. The eruption started as an erythematous papule that spread to involve her left periorbital region. She denied other skin lesions, oral ulcers, pruritus, fever, arthralgia, or weakness. Her SLE had been in remission for the past seven years without treatment. On examination, a well-demarcated, scaly, erythematous plaque with associated underlying edema was present on the left upper and lower eyelids. Topical tacrolimus was trialed for treatment of a suspected allergic contact dermatitis without improvement of her symptoms. Patch testing at that time was negative for any contact allergens. Several months later, she was hospitalized for transient left eye vision loss. Orbital magnetic resonance imaging showed asymmetric enhancement of the posterior left globe and optic nerve sheath, suggestive of scleritis and perineuritis. Given her persistent rash, a biopsy was performed and revealed a lichenoid interface dermatitis with superficial and deep perivascular and periadnexal lymphocytic infiltrate, basement membrane zone thickening, and follicular plugging consistent with involvement of a connective tissue disorder. Further work-up revealed elevated ESR (26 mm/h) and CRP (14.9 mg/L), ANA 1:160, and anti-dsDNA antibodies 1:80. She was started on 200 mg hydroxychloroquine twice daily and 20 mg prednisone daily for a suspected SLE flare with marked improvement in the rash and resolution of her vision loss. Over the following eleven months, prednisone was tapered to 5 mg daily with only residual post-inflammatory hyperpigmentation, potentially complicated by her underlying Ehlers-Danlos syndrome.

This case presented a diagnostic dilemma as the lesion mimicked allergic contact dermatitis given its unilateral presentation. While a few reports document eyelid edema and erythema secondary to cutaneous lupus erythematosus, their average times to diagnosis range from two to three years because of their diagnostically challenging presentations.¹⁻⁵ Therefore, this case highlights the utilization of a patient's past medical history, histopathology, and laboratory findings to facilitate diagnosis and treatment of this rare manifestation of SLE in an attempt to minimize scarring, atrophy, and long-term organ damage.^{6,7}

Figure 1. The patient's initial presentation with an erythematous, scaly plaque associated with edema of the left upper and lower eyelids. A purple patch with scale is located on the zygomatic arch.



Figure 2. The patient's periorbital eruption A) four months after initiating treatment and B) seven months after initiating treatment.



References

1. Smith CA, Pinals RS. Periorbital edema in systemic lupus erythematosus. *Arch Intern Med*. 1982;142(9):1711-1712.
2. Cyran S, Douglass MC, Silverstein JL. Chronic cutaneous lupus erythematosus presenting as periorbital edema and erythema. *J Am Acad Dermatol*. 1992;26(2 Pt 2):334-338. doi:10.1016/0190-9622(92)70049-I
3. Silva LL da C, Romiti R, Nico MMS. Persistent eyelid edema and erythema as manifestation of lupus erythematosus: a series of six cases. *J Dtsch Dermatol Ges J Ger Soc Dermatol JDDG*. 2015;13(9):917-920. doi:10.1111/ddg.12632
4. Arrico L, Abbouda A, Abicca I, Malagola R. Ocular Complications in Cutaneous Lupus Erythematosus: A Systematic Review with a Meta-Analysis of Reported Cases. *J Ophthalmol*. 2015;2015. doi:10.1155/2015/254260
5. Wu MY, Wang CH, Ng CY, et al. Periorbital erythema and swelling as a presenting sign of lupus erythematosus in tertiary referral centers and literature review. *Lupus*. 2018;27(11):1828-1837. doi:10.1177/0961203318792358
6. Castro LA, Davis DMR, Davis MDP, Bruce AJ, Pittelkow MR. Facial edema and crusted patches: a precursor to life-threatening acute systemic lupus erythematosus. *J Am Acad Dermatol*. 2007;56(5 Suppl):S126-127. doi:10.1016/j.jaad.2006.02.055
7. Santoro FA, Huang J. Ocular involvement in cutaneous connective tissue disease. *Clin Dermatol*. 2016;34(2):138-145. doi:10.1016/j.clindermatol.2015.11.011