

Unilateral Periorbital Edema and Erythema: A Rare Presentation of Chronic Cutaneous Lupus Erythematosus

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ABSTRACT We present a case of a 55-year-old female with unilateral periorbital edema and erythema over the past one year without systemic features or other skin signs, who had photosensitivity and positive antinuclear antibody and also SS-A 60kD antibody. Treatment with topical corticosteroids had remained unsuccessful along one year. We suspected a clinical picture association with cutaneous lupus erythematosus including photosensitivity, chronic cutaneous lesion finding and positive autoantibodies and observed a favorable clinical response to systemic antimalarial therapy within a short time. The observation has confirmed our diagnosis. Similar cases were rarely reported in literature. The persistent edema and erythema involving periorbital area can occur as a rare manifestation of chronic cutaneous lupus erythematosus.

Keywords: Cutaneous lupus erythematosus; erythema; edema; antinuclear antibody; antimalarial treatment

Connective tissue autoimmune diseases such as systemic lupus erythematosus (SLE), dermatomyositis or scleroderma often prominently affect the skin. Specific or nonspecific cutaneous lesions in lupus erythematosus are very common. It is well known that the lupus erythematosus rash shows variable features and is categorised into acute, subacute and chronic varieties according to the clinical and histopathological appearances and duration of symptoms. A proportion of cutaneous lupus erythematosus (CLE) cases can involve only skin or present as part of the symptoms of SLE.¹

Immunologically a positive antinuclear antibody (ANA) is found in 95% of the patients presenting with malar rash with acute cutaneous lupus erythematosus. Antinuclear antibody is positive in 70-80% of the patients showing annular and papulosquamous lesions with subacute cutaneous lupus erythematosus.² Chronic cutaneous lupus (CCLE) includes different lesions such as discoid lupus erythematosus (DLE), lupus erythematosus profundus, chilblain lupus and lupus tumidus. Serologically, DLE patients have a lower incidence of autoantibodies such as ANA ds-DNA and SS-A (Ro) as compared to other CLE subtypes.³

Unilateral or bilateral periorbital skin involvement with or without systemic features or other cutaneous findings in lupus patients is a relatively rare clinical presentation of CLE.⁴

Herein a female patient who presented with unilateral edema and erythema located in periorbital area with the positive autoimmune serology findings including those for antinuclear antibodies and anti-SSA antibody, and also a favorable skin response rate to hydroxychloroquin treatment at 6 weeks is discussed.

CASE REPORT

A 55-year-old female patient was admitted with over one year history with persistent unilateral periorbital erythema and swelling. Her skin lesion has been aggravated by sun exposure. Her past medical and family history were unremarkable. She was followed by another medical center. Treatment with topical corticosteroids failed. On physical examination, the most prominent finding was local periorbital edema and erythema on the right side, she had no pruritis, pain (Figure 1 a), evidence of systemic involvement and other significant skin signs. The general physical examination was otherwise normal. Ophthalmic examination was also normal.

Laboratory findings: were as follows white blood cell count $8 \times 10^3/\mu\text{l}$, neutrophil 68%, Hb 14.7g/L, hematocrit 41.8%, platelet $347 \times 10^3/\mu\text{l}$, erythrocyte sedimentation rate 6 mm/h and C-reactive protein 1.9 mg/dl (normal: > 5 mg/dl). Biochemical tests and urine analysis were normal. Infectious finding was not found. Antinuclear antibody was titers of 1/160, homogeneous and in granular pattern, SS-A 60 kD antibody was also

positive, other ANA subtypes and rheumatoid factor were negative. Serum immunoglobulin and complement 3 and 4 values were within normal limits. Magnetic resonance imaging of the orbita was normal.

Chronic cutaneous lupus erythematosus was suspected due to the presence of photosensitivity, periorbital cutaneous lesion finding, chronic course and positive autoantibodies. An of the disease clinical response to the treatment with systemic anti-malarial drug (hydroxychloroquin sulphate 200 mg /day) was observed (Figure 1 b) and (Figure 1 c). Our clinical observation supported the presence of CCLE. Her skin lesion completely disappeared and she has been followed without symptom without scarring one year.

DISCUSSION

Tufanelli and Dubois reported the incidence of periorbital edema as 4.8% in SLE.⁴ Wu et al enrolled a total of 25 patients with periorbital erythema and swelling as the presenting sign of lupus erythematosus, most of the patients presented with unilateral involvement, all patients had features compatible with CLE on histopathological examination. However autoantibody analysis such as antinuclear antibody showed negative results. During follow-up, six patients developed SLE and two patients developed Sjögren syndrome.⁵

Unilateral or bilateral periorbital or eyelid skin manifestation with or without systemic features or other cutaneous findings in lupus pa-

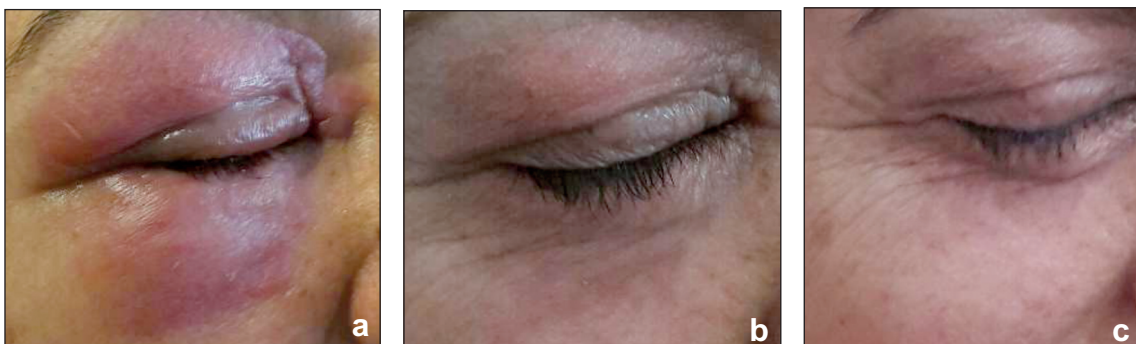


FIGURE 1 a-c: a) Periorbital erythema and edema before treatment, b) Periorbital erythema and edema after three weeks under treatment with hydroxychloroquin, c) Periorbital erythema and edema after six weeks under treatment with hydroxychloroquin.

tients is an unusual condition (Table 1).⁶⁻¹³ The diagnosis can delay because of clinical mimicry to some disorders such as chronic dermatitis, urticaria or similar conditions.

Cyran et al. reported two unusual cases with eyelid edema and erythema which was unilateral in one case and bilateral in another, and used the term “chronic cutaneous lupus erythematosus” for their patients. Both cases responded the therapy with antimalarial drugs.⁶ The first case of the authors with unilateral involvement is similar to our patient.

Silva et al. reported a series of six cases presenting with persistent eyelid edema and erythema with or without other manifestations of lupus erythematosus. The presence of limited lesions was described as a specific cutaneous manifestation of lupus erythematosus.⁷

Ghaninejad et al. described two patients with severe periorbital edema and erythema as the sole manifestation of cutaneous lupus erythematosus. In their cases, the disease was limited to the skin but the lesions lacked the appearance of discoid lupus erythematosus, including atrophy, scarring, and follicular plugging.⁸

Discoid rash is the prototype of specific chronic skin lesions in lupus erythematosus, can occur as a localized process which usually involves head and neck area in photo-exposed areas. The lesions can be erythematous, raised, indurated papules or plaques and lead to scar. Serologically, DLE patients have a lower incidence of autoantibodies such as ANA, dsDNA, Sm and Ro/SSA antibodies, as compared to the other CLE subtypes.³ Periorbital or eyelid erythema and edema were reported in patients with discoid lupus erythematosus as a rare presentation of cutaneous manifestation.¹⁰⁻¹²

Our patient presented with persistent unilateral periorbital edema and erythema without systemic involvement. Her skin lesions were not similar to the appearance of typical discoid lupus erythematosus. There was no desquamatic plaques leading atrophic scars, alopecia, or permanent pigmentary changes which is the morphologic signs of discoid rash. The diagnosis of CLE requires also histologic findings, her skin histopathological examination had been performed at another medical center, so a second biopsy was not obtained in our hospital. Serology is helpful for making a diagnosis. In our patient, autoantibodies including anti-nuclear antibody and SS-A (Ro) antibody were

TABLE 1: Reported cases of periorbital or eyelid erythema and edema manifestaion with chronic cutaneous lupus erythematosus.

Author/Ref	Age /Sex	Clinical sign	Histopathology	Treatment
Cyran et al.	51 F	Unilateral eyelid edema and erythema	CCLE	HQ Response within 5wk (two cases)
		Bilateral eyelid edema and erythema	CCLE	HQ
Braun et al.	72F	Preorbital edema and erythema	DLE	HQ
Silva et al.	44F	Unilateral eyelid edema erythema	C3 deposits by DIF (Skin lesion): + (six cases)	
	47F	Unilateral eyelid edema& erythema	IgG deposits by DIF (Skin lesion): +	
	31F	Unilateral eyelid edema & erythema	IgG deposits by DIF (Skin lesion): +	
	47F	SLE +Bilateral eyelids edema&erythema	IgM & IgG deposits by DIF (Skin lesion): +	
	56F	SLE +Unilateral eyelids edema&erythema	IgM deposits by DIF (Skin lesion):+	
	26M	SLE+ Bilateral eyelids edema&erythema	IgG,IgM& IgA deposits by DIF(Skin lesion):+	
Ghaninejad	23M	Bilateral swelling of eyelids and cheeks	CLE	HQ and prednisolone et al.
	36M	Unilateral left side of his face and periorbital swelling	CLE	HQ improvment after 3mo (two cases)
Erras et al.	26F	Bilateral swelling & erythema of her eyelids	CCLE	HQ and CS for one mo
Cakıcı et al.	40F	Periorbital erythema & edema	DLE + SLE	HQ and CS for 6 mo
Wu et al.	41E	Erythema & edema of left upper eyelid	DLE	Topical CS and HQ for 3mo
Gimenes et al.	23M	Bilateral swelling & erythema of eyelids	DLE	HQ, response over a 6 mo

positive which supported lupus rash. Serologically, DLE patients have a lower incidence of autoantibodies such as ANA, dsDNA, Sm and Ro/SSA antibodies, as compared to the other CLE subtypes.³ Anti-Ro antibodies are particularly common in subacute cutaneous lupus erythematosus, occurring in approximately 60% of patients, these antibodies may be found in about 25% of DLE patients.²

Hydroxychloroquin (HQ) has been used for a long time as disease-modifying anti-rheumatic agents in the autoimmune disease such as rheumatoid arthritis, Sjögren syndrome, systemic lupus erythematosus, chronic cutaneous lupus erythematosus and recommended as first line treatment for cutaneous lupus patients. Ootake et al. found the overall skin response rate to hydroxychloroquin treatment at 16 weeks in patients with cutaneous lupus erythematosus.¹⁴ Improvement was shown in cutaneous lupus erythematosus as 50% of the patients treated with HQ after 8 weeks of therapy.¹ We observed an excellent response to hydroxychloroquin treatment at 6 weeks.

In clinical practice, the connective tissue disease with unusual presentation should be suspected and the presence of autoantibodies supports diagnosis in the patients.

Informed Consent

Written informed consent was obtained from the patient who participated in this study.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution.

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