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

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 to our diagnosis. The 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 Systemic 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 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 patients presenting malar rash with acute cutaneous lupus erythematosus. ANA is positive in 70-80% of 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 is discussed who presenting unilateral edema and erythema located 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.

CASE REPORT

A 55-year-old female patient who had 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 was unremarkable. She was followed by an other medical center. Treatment with topical corticosteroids failed. On physical examination the most prominent finding was local periorbital edema and erythema on 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. Ophthalmal examination was also normal.

Laboratory findings: white blood cell count 8x10^3/µl, neutrophil 68%, Hb 14.7g/L, Hct 41.8%, platelet 347x10^3 /µl, erythrocyte sedimentation rate 6 mm/h and C reactive protein 1.9 mg/dl (normal: > 5mg/dl). Biochemical tests and urine analysis were normal. Infectious finding was not found. Antinuclear antibody was titers of 1/160, homogeneous and granular pattern, SS-A 60 kD antibody was also positive, other ANA subtypes and rheumato
did factor were negative. Serum immunoglobulin and complement 3 and 4 values were within normal limits. Magnetic resonance imaging of orbits was normal.

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

DISCUSSION

Tufanelli and Dubois reported as 4.8% in incidence of periorbital edema in SLE. 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 autoantibodies analysis such as antinuclear antibody showed negative results. During follow-up, six patients developed SLE and two patients developed Sjögren syndrome.

Unilateral or bilateral periorbital or eyelid skin manifestation with or without systemic features or other cutaneous findings in lupus patients is an unusual condition (Table 1).
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 the other, and used the term “chronic cutaneous lupus erythematosus” for their patients. Both cases responded the therapy with antimalarial drugs. First case of authors with unilateral involvement is similar to our patient.

Silva et al. reported a series of six cases presenting 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.

Ghaninejad H 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.

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

Our patient presented 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 desquamative 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 other medical center, second biopsy was not done in our hospital. Serology are helpful in the diagnosis. In our patient autoantibodies including antinuclear antibody and SS-A (Ro) antibody were positive which supported lupus rash. Serologically, DLE patients have a lower incidence of autoantibodies such as ANA, dsDNA, Sm and Ro/SSA.

<table>
<thead>
<tr>
<th>Author /Ref</th>
<th>Age /Sex</th>
<th>Clinical sign</th>
<th>Histopathology</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cyran et al.</td>
<td>51 F</td>
<td>Unilateral eyelid edema and erythema</td>
<td>CCLE</td>
<td>HQ Response within 5wk (two cases)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Bilateral eyelid edema and erythema</td>
<td>CCLE</td>
<td></td>
</tr>
<tr>
<td>Braun et al.</td>
<td>72 F</td>
<td>Preorbital edema and erythema</td>
<td>DLE</td>
<td>HQ</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Silva et al.</td>
<td>44 F</td>
<td>Unilateral eyelid edema and erythema</td>
<td>C3 deposits by DIF (Skin lesion): + (six cases)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Unilateral eyelid edema and erythema</td>
<td>IgG deposits by DIF (Skin lesion): +</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Unilateral eyelid edema and erythema</td>
<td>IgG deposits by DIF (Skin lesion):</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>SLE + Bilateral eyelids edema &amp; erythema</td>
<td>IgM &amp; IgG deposits by DIF (Skin lesion): +</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>SLE + Unilateral eyelids edema &amp; erythema</td>
<td>IgM deposits by DIF (Skin lesion): +</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>28M</td>
<td>IgG, IgM, IgA deposits by DIF(Skin lesion): +</td>
<td></td>
</tr>
<tr>
<td>Ghaninejad</td>
<td>23M</td>
<td>Bilateral swelling of eyelids and cheeks</td>
<td>CLE</td>
<td>HQ and prednisolone et al.</td>
</tr>
<tr>
<td></td>
<td>36M</td>
<td>Unilateral left side of his face and periorbital swelling</td>
<td>CLE</td>
<td>HQ improvement after 3mo (two cases)</td>
</tr>
<tr>
<td>Erras S et al.</td>
<td>28F</td>
<td>Bilateral swelling &amp; erythema of her eyelids</td>
<td>CCLE</td>
<td>HQ and CS for one mo</td>
</tr>
<tr>
<td>Cakici O et al.</td>
<td>40F</td>
<td>Periorbital erythema &amp; edema</td>
<td>DLE + SLE</td>
<td>HQ and CS for 6 mo</td>
</tr>
<tr>
<td>Wu et al.</td>
<td>41E</td>
<td>Erythema &amp; edema of left upper eyelid</td>
<td>DLE</td>
<td>Topical CS and HQ for 3mo</td>
</tr>
<tr>
<td>Gimenes et al.</td>
<td>23M</td>
<td>Bilateral swelling &amp; erythema of eyelids</td>
<td>DLE</td>
<td>HQ, response over a 6 mo</td>
</tr>
</tbody>
</table>

TABLE 1: Reported cases of periorbital or eyelid erythema and edema manifestation with chronic cutaneous lupus erythematosus.
Ro/SSA antibodies, as compared to other CLE subtypes. Anti-Ro antibodies are particularly common in subacute cutaneous lupus erythematosus, occurring in approximately 60% of pts, these antibodies may be found in about 25% of DLE patients.

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. HQ is recommended as first line treatment for cutaneous lupus patients. Ototake et al. found that the overall skin response rate to hydroxycholoroquin treatment at 16 weeks in patients with cutaneous lupus erythematosus. It was showed improvement of cutaneous lupus erythematosus in 50% of patients treated with HQ after 8 weeks of therapy. We observed an excellent response to hydroxycholoroquin 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.

REFERENCES