

Rosacea-like Cutaneous Lupus Erythematosus: An Atypical Presentation Responding to Antimalarials

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Lupus erythematosus (LE) with predominant skin involvement comprises chronic cutaneous LE (CCLE) and subacute cutaneous LE (SCLE). The first includes the so-called discoid CCLE (1), which is the most common form, most frequently involving the face, and characterized by well-defined, red scaly patches healing with atrophy, scarring and pigmentary changes (1). More uncommon subsets of CCLE are lupus tumidus (2, 3) and lupus panniculitis (4, 5). SCLE typically presents with non-scarring annular or papulosquamous eruptions on sun-exposed skin (6, 7), although a number of unusual variants have been described (8–11). The present report describes the clinical findings and course as well as the histopathological, immunofluorescence and laboratory characteristics of 4 patients with cutaneous LE limited to the central face mimicking rosacea. Rosacea is characterized by erythema and telangiectasia and is punctuated by episodes of inflammation manifesting as papules, pustules and swelling (12).

CASE REPORTS

The 4 patients with rosacea-like cutaneous LE were 3 women and 1 man (F/M ratio 3:1), with a mean age at disease onset of 54 years (range 41–69). The mean duration of disease was

13 months (range 6–24). On admission to our hospital, all 4 patients presented with erythema that was localized to the central face and was associated with a few raised, smooth round erythematous or erythematous-violaceous papules ranging from 2 to 3 mm in diameter over the malar areas and forehead (Fig. 1a); the skin lesions were accompanied by intense burning and, occasionally, by slight to moderate pruritus. In all 4 cases the onset of the above cutaneous picture had been sudden, and the patients had noticed aggravation of the rash after sun exposure. Before our observation, all 4 patients had been given treatments for rosacea in other institutions, including tetracyclines, azithromycin or metronidazole orally, in combination with topical metronidazole, with no benefit. The patients were not treated for other concomitant diseases. After LE was diagnosed, we evaluated the patients for systemic symptoms and signs associated with LE, which were lacking in all 4 cases; moreover, none of them fulfilled the American College of Rheumatology criteria for the diagnosis of systemic LE (SLE) (13). The possible association with Sjögren's syndrome or other autoimmune disorders was excluded. The 4 patients were treated with oral hydroxychloroquine 400 mg/daily, which induced a complete clearing of the skin lesions (Fig. 1b) with a mean resolution time of 7 weeks (range 5–8 weeks). Hydroxychloroquine was discontinued 1 month following complete resolution. The patients were warned against exposure to sunlight and they were advised to apply total-block sunscreens in bright sunlight. Currently, all patients are alive and free of disease, with a mean time of follow-up of 4 years (range 2–7 years), with neither relapses nor evolution into SLE.

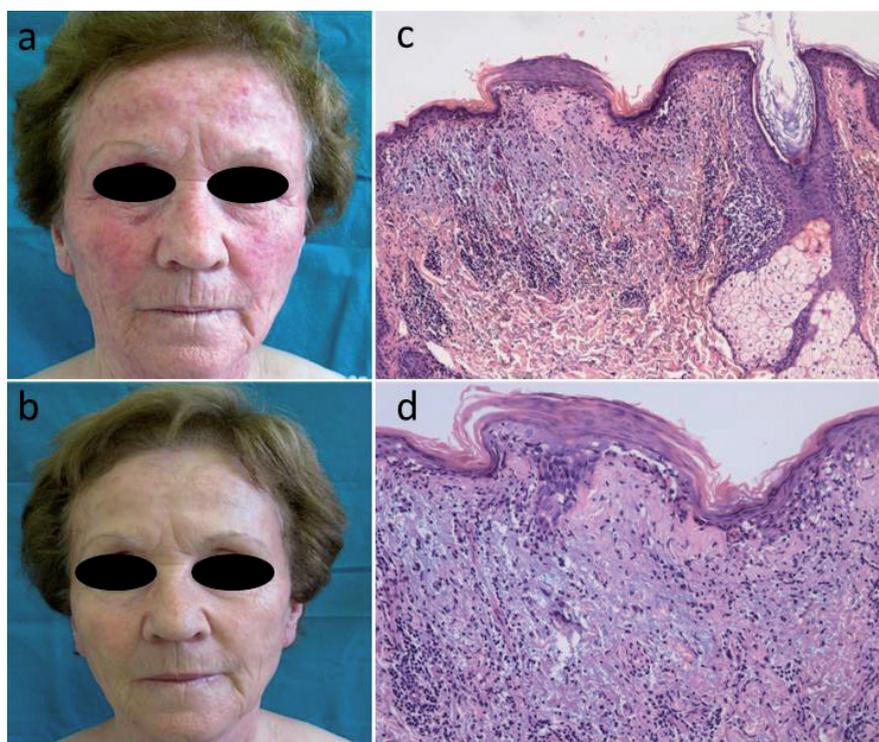


Fig. 1. (a) Redness with small erythematous papules involving the central face in patient 1. (b) Complete resolution after hydroxychloroquine therapy. (c) Histology showing a pattern of lichenoid interface dermatitis (haematoxylin and eosin (H&E) stain; original magnification, $\times 100$). (d) Medium-power view demonstrating the hydropic degeneration of the epidermal basal cell layer; in the dermis, a mixed inflammatory infiltrate associated with mucin deposition is evident (H&E stain; original magnification, $\times 200$).

The erythrocyte sedimentation rate was moderately elevated in only one patient (case 3) at disease onset (53 mm in the first hour; normal <20), and reverted to normal after resolution. Antinuclear antibodies (ANA) were present, up to 1/640 with a fine speckled pattern, in all 4 cases. Anti-Ro/SSA antibodies were also found in all 4 patients. All the other immunological parameters evaluated, notably anti-double stranded-DNA antibodies, were normal or negative. Anti-Ro/SSA antibodies, re-evaluated in clinical remission at the time of writing this paper, remained positive in all 4 patients.

The 4 patients demonstrated similar histological changes in biopsy specimens taken from facial papular lesions surrounded by erythema. These changes include epidermal atrophy, hydropic degeneration of the epidermal basal cell layer, and a superficial perivascular and periappendageal lymphohistiocytic infiltrate (Fig. 1c). Abundant dermal deposition of mucin was seen (Fig. 1d).

In all 4 patients, direct immunofluorescence performed on biopsy specimens taken from lesional skin revealed granular deposits of immunoglobulin (Ig)M and IgG (case 3) or IgM alone (remaining cases) at the dermoepidermal junction; dermoepidermal granular deposition of C3 component of complement was also demonstrated in 2 patients (cases 1 and 4).

DISCUSSION

Although an erythematous eruption involving the face with sun exposure as triggering event is a classical cutaneous finding within the spectrum of LE, the presentation in our 4 cases is unique in that it resembled acne rosacea (12). However, the absence of pustules, telangiectasia, flushes and ocular signs, in addition to the lack of response to classical therapies for rosacea led us to test the patients for ANA and other autoantibodies, allowing us to diagnose LE. The 4 patients responded dramatically to hydroxychloroquine and there were neither relapses nor evolution into SLE after a mean follow-up of 4 years. The very rapid response to antimalarials may be explained by the fact that more superficial LE skin lesions, including erythema and papules, as in our patients, usually respond more rapidly than scaly, atrophic and scarring lesions. The absence of recurrence after treatment withdrawal may be due to the less aggressive nature of this atypical presentation of cutaneous LE. Anti-Ro/SSA antibodies, which are closely related to photosensitivity, are possibly the laboratory hallmark for this presentation, as for SCLE (7). However, our patients were unlikely to have SCLE due to the absence of typical annular or psoriasiform lesions. While classic discoid lupus was easily ruled on the basis of clinicopathological criteria, the tumidus variant should also be considered. However, the typical skin lesions in LE tumidus are erythematous, urticaria-like, non-scarring plaques, and its histology lacks changes of interface dermatitis as seen in our cases and typically shows a dermal infiltrate (3, 14). The strong mucin deposits found in our cases might suggest reticular erythematous mucinosis (REM) (15). Based on its possible association with autoimmune diseases, notably LE, the commonly observed photosensitivity, the deposition of IgM at the dermoepidermal junction

found in some patients with REM and the good response to antimalarials, it has been classified among the specific cutaneous lesions of LE (14). However, similarly to lupus tumidus, REM is regarded as an example of dermal cutaneous LE and anti-Ro/SSA antibodies are usually lacking in this subset (15).

The authors declare no conflicts of interest.

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