Dermatology Online Journal

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Letter: Vesico-bullous subacute cutaneous lupus erythematosus – An uncommon entity successfully treated with dapsone and hydroxychloroquine

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Abstract

Vesico-bullous subacute cutaneous lupus erythematosus is an uncommon and severe presentation. The authors report an exuberant case of vesico-bullous subacute cutaneous lupus erythematosus successfully treated with dapsone and hydroxychloroquine.

Introduction

Subacute cutaneous lupus erythematosus (SCLE) has several clinical variants, with bulla formation being an uncommon feature. We report an exuberant case of vesico-bullous SCLE.

Case report

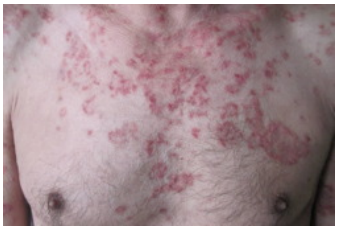


Figure 1

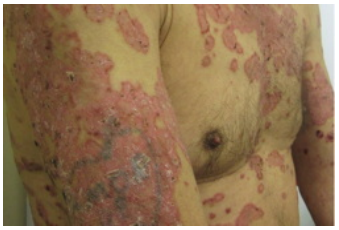


Figure 2

Figure 1. Clinical features of the erythematous annular plaques on the thorax.

Figure 2. Cutaneous lesions on the trunk and upper limbs, 2 weeks after the previous photo, with more pronounced scaling and infiltration.

A 49-year-old man presented with a 4-month history of progressive pruritic erythematous lesions with blisters, in a photodistribution, that did not respond to a 20-day course of oral prednisone and hydroxyzine. His past medical history included a traumatic amputation of the second and third left fingers. He was otherwise healthy and was on no regular medication.

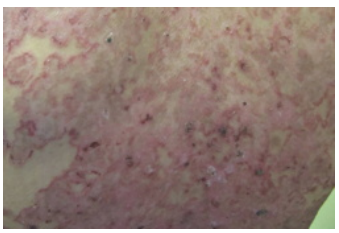


Figure 3

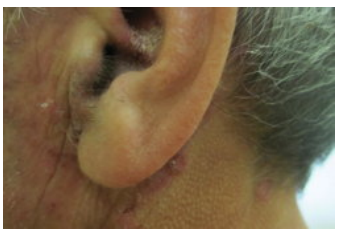


Figure 4

Figure 3. Clinical features of the erythematous annular scaling plaques on the back.

Figure 4. Erythematous scaling plaques on the lateral aspects of the face and ears.

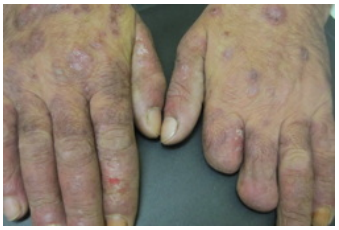


Figure 5

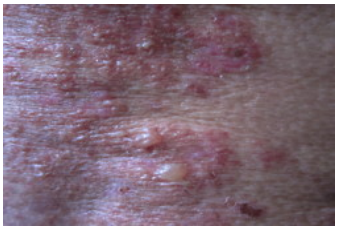


Figure 6

Figure 5. Cutaneous lesions on the dorsum of the hands.

Figure 6. Highlight of the vesico-bullous lesions at the edge of the cutaneous lesions on the abdomen.

Physical examination revealed well-defined erythematous scaling annular plaques, with raised border and central clearing, on the face (lateral aspects and ears), scalp, trunk, and upper limbs, with more pronounced scaling.

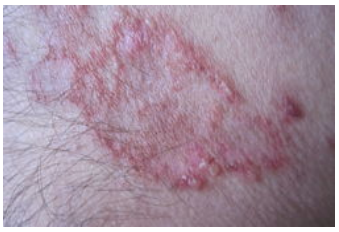


Figure 7

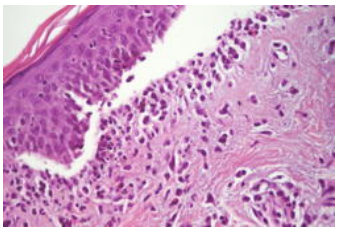


Figure 8

Figure 7. Highlight of the vesico-bullous lesions at the edge of the cutaneous plaques on the thorax.

Figure 8. Histopathologic examination showing a subepidermal blister and an inflammatory infiltrate on the dermis composed of neutrophils and rare eosinophils (H&E x40).

At the edge of these lesions tense vesicles/bullae developed, 2-15 mm in diameter, with negative Nikolsky sign.

Histopatological examination showed a subepidermal blister and an inflammatory infiltrate in the dermis composed of neutrophils and rare eosinophils.

Direct immunofluorescence of perilesional skin revealed a continuous band of granular deposits of IgG and C3 at the dermal-epidermal junction. Laboratory workup disclosed positivity of antinuclear antibodies (1/160, normal <1/80) and rheumatoid factor (48.0 UI/mL, normal <14). The remainder of the immunology, complete blood count, chemistry profile, serologies, and peripheral blood flow cytometry were unremarkable. Altogether these findings were compatible with the diagnosis of vesico-bullous SCLE. To exclude paraneoplasic disease, chest X-ray, abdominal ultrasound, and colonoscopy were performed, all normal. The patient started dapsone 100 mg qd, hydroxychloroquine 400 mg qd, hydroxyzine 25 mg tid, and sunscreens. Five days later, development of new lesions had stopped and 3 weeks later clinical improvement was evident. At this time, dapsone was withdrawn.

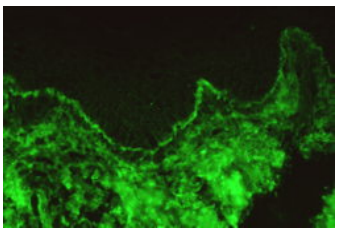


Figure 9

Figure 9. Direct imunofluorescence of perilesional skin showing continuous granular deposit of IgG at the dermal-epidermal junction (x40).

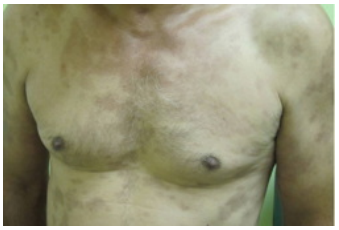


Figure 10

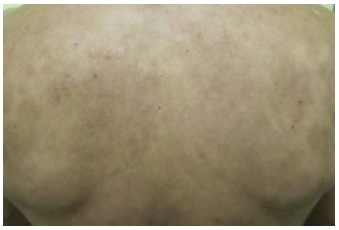
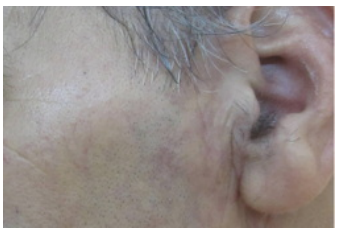


Figure 11

Figure 10. Complete resolution of the cutaneous lesions on the thorax and abdomen after treatment.

Figure 11. Complete clearing of the lesions on the back.



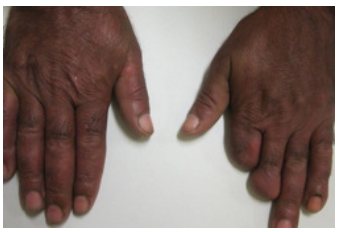


Figure 12	Figure 13

Figure 12. Resolution of the cutaneous lesions on the face with treatment.

Figure 13. Complete resolution of the lesions on the dorsum of the hands.

Clearance of the lesions continued, with discrete post-inflammatory hyperpigmentation. Hydroxychloroquine was reduced and stopped 3 months later. The patient remains free of lesions after 1 year of follow-up.

Discussion

Bullae formation in lupus erythematosus (LE) occurs more frequently in the systemic or drug-induced forms [**1**, **2**, **3**]. Vesico-bullous SCLE is rare and the unusual presentation may delay the diagnosis, requiring a high index of clinical suspicion [**4**, **5**]. Its diagnosis is established by the combination of clinical, immunologic, histopathologic, and immunofluorescence findings [**1**, **4**, **5**]. SSA and SSB antibodies, frequently associated with SCLE, were negative in our patient but their presence is not mandatory for the diagnosis [**1**, **4**, **5**]. The formation of blisters is caused by the severe vacuolar degeneration in the basal cell layer, leading to dermal-epidermal cleavage [**1**, **4**]. There is no standard treatment and results are variable. Hydroxychloroquine's efficacy in cutaneous LE is well known, but the clinical response is slow (generally longer than 6 weeks) and unpredictable [**6**, **7**]. Dapsone is used in numerous dermatologic diseases, with higher success rates in those with neutrophilic infiltrates [**6**, **7**]. Consistent responses have been documented in bullous systemic LE, with dramatic clinical improvement in 24-48 hours [**6**, **7**, **8**]. With these facts in mind we decided to combine hydroxychloroquine and dapsone in our patient, with an excellent response in only 5 days and so far without relapses.

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