

Vesiculobullous Subacute Cutaneous Lupus Erythematosus: A Rare Presentation

Sir,

A 53-year-old female presented with a 2-month history of an asymptomatic dermatosis beginning on the anterior chest, with progression to the back and arms. Large, easily rupturing blisters subsequently developed in those areas. Her medical history was positive for hypothyroidism and arterial hypertension, for which she was taking levothyroxine and indapamide. She denied commencing any new medication prior to the lesions.

Examination revealed extensive, symmetric, crusted erosions on the anterior chest, dorsum, and extensor aspects of the arms, as well as scattered erythematous papules and vesiculopustules [Figure 1a-c]. A marked demarcation from photoprotected skin was observed. Nikolsky's sign was negative. The involvement of the hands relatively spared to the interphalangeal joints [Figure 2a] and pustules and target-like lesions were noted on the wrists and palms [Figure 2b]. Painful oral erosions were present without other mucosal lesions.

Laboratory tests revealed a decreased lymphocyte count (870 cells/mm^3) and elevated inflammatory parameters. Immunology panel showed positivity for antinuclear antibodies (1/640, normal $<1/160$) and anti-dsDNA antibodies (136 U/mL, normal <30). Complement fractions C3c and C4 were diminished. The remaining laboratory and radiological studies, including renal function and urinalysis, were unremarkable.

Histopathological examination showed a subepidermal blister containing neutrophils, epidermal atrophy, hydropic degeneration of the basal layer and numerous cytoid bodies [Figure 3a and b] and [Figure 4]. Direct immunofluorescence performed on perilesional skin revealed IgG and C3c deposition at the dermal-epidermal junction [Figure 5a and b].

A diagnosis of vesiculobullous subacute cutaneous lupus erythematosus (SCLE) was made. The patient was started on oral prednisolone 1 mg/kg/day, with remarkable improvement within 3 weeks. She remains disease-free and without evidence of systemic involvement after 14 months of follow-up.

In the setting of lupus erythematosus (LE), the presence of bullae or its equivalents poses a significant diagnostic challenge. Different pathogenic mechanisms underlie the formation of such lesions. When the vacuolar degeneration of the epidermal basal layer is particularly intense, it can induce a vesiculobullous change of acute or subacute LE-specific cutaneous lesions, as seen in our patient.^[1,2] In extreme cases, large areas of epidermal detachment may



Figure 1: (a-c) Extensive symmetric erosions on the anterior chest, shoulders, dorsum, and arms with scattered erythematous papules and vesiculopustules



Figure 2: (a) Involvement of the dorsum of the hands sparing the interphalangeal joints. (b) Pustules and target-like lesions on the wrists and palms

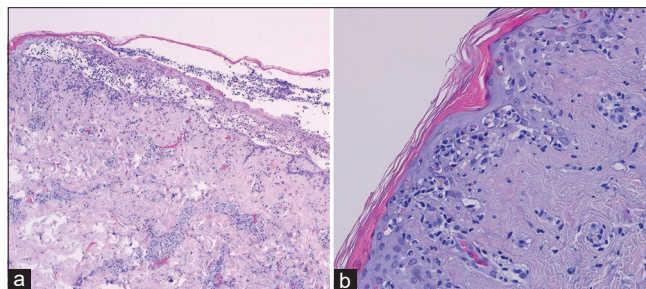


Figure 3: (a) Subepidermal blister containing neutrophils (H and E, 100 \times). (b) Evidence of epidermal atrophy, vacuolar degeneration of the basal layer, and presence of numerous cytoid bodies (H and E, 200 \times)

develop, simulating toxic epidermal necrolysis (TEN). As such, a designation of “acute syndrome of apoptotic pan-epidermolysis” was proposed to unify all clinical settings in which massive epidermal cleavage is due to a hyperacute apoptotic injury, including drug-induced TEN and other TEN-like conditions (namely, LE or pseudoporphyria).^[1-3]

This scenario must be differentiated from classic bullous LE, a rare acquired blistering disorder that accompanies acute cutaneous LE with flares of systemic activity.^[3] Histologically, a neutrophilic infiltrate without the evidence of interface dermatitis is observed. In this case, criteria for a defined autoimmune blistering disease, such as bullous

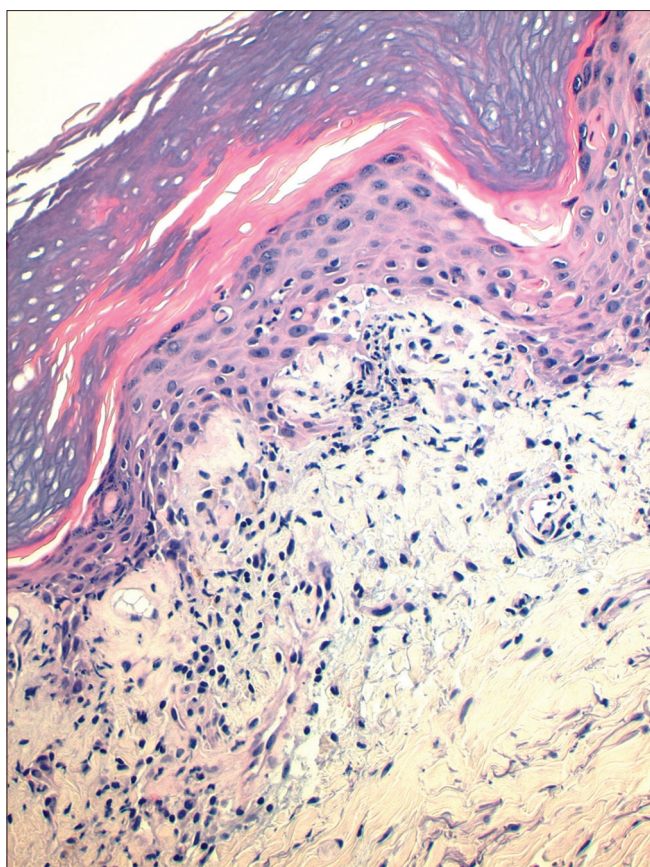


Figure 4: Extensive basal cell vacuolization adjacent to the bulla (H and E, 200×)

pemphigoid or epidermolysis bullosa acquisita must be searched, as these may be concurrent with LE.^[1,2] In our patient, those LE-nonspecific disorders were excluded on the basis of clinical presentation and histopathological features, that is, the absence of eosinophils. Furthermore, they do not occur as an extension of the LE-specific interface dermatitis.^[1,2] A significant neutrophilic cutaneous infiltration may also occasionally be seen, with evidence suggesting that a subgroup of patients may be more prone to develop nonspecific neutrophilic dermatoses in association with LE.^[1]

Vesiculobullous SCLE is rare and a combination of clinical, laboratorial, and histopathological features is crucial for diagnostic confirmation.^[4,5] The presence of anti-Ro/SS-A antibodies is frequently reported, although its absence does not exclude the diagnosis.^[5] Half of the patients fulfill the criteria for systemic LE, however, the majority have only mild disease.^[3,4] Our patient fulfilled 5 American College of Rheumatology (ACR) criteria (1997) but remained without the evidence of systemic

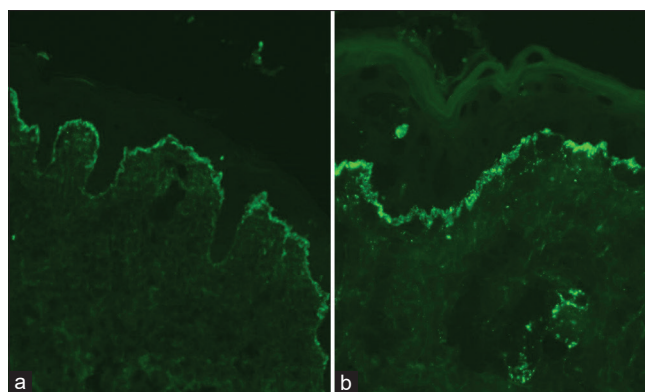


Figure 5: Direct immunofluorescence confirming IgG (a) and C3c (b) deposition at the dermal-epidermal junction

involvement throughout the course of the dermatosis and during follow-up.

Herein, we report a rare presentation of SCLE, reinforcing the need for a high suspicion index, particularly in the absence of previously known disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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