Multiple Blisters Along the Lip Vermilion are a Clue to Bullous Lupus Erythematosus

Marcello Menta S. Nico¹ and Silvia V. Lourenço²
¹Department of Dermatology, Medical School, and ²Department of Pathology, Dental School, University of São Paulo, Rua Itapeva 590, 3° A, CEP-01332-000, São Paulo, Brazil. E-mail: mentanico@hotmail
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Blistering dermatoses in the setting of lupus erythematosus (LE) have long been recognized. These may include vesiculobullous changes occurring at the active border of advancing of annular LE lesions, discoid cutaneous LE with vesicles, toxic epidermal necrolysis-like LE, and the non-scarring acute eruption with tense, clustered blisters appearing on normal or erythematous skin occurring in patients with active LE, which is characterized microscopically by neutrophilic infiltrates in the dermal papillae (‘bullous LE’) (1, 2). Lee & Ackerman (3) consider bullous LE a caricature of the specific acute LE process on the skin, with massive neutrophil infiltration and mucin production. Sun-exposed skin seems to be more often affected, although several cases without any photosensitivity do exist. Facial lesions are common in bullous LE, as well as intra-oral blisters and erosions. Vesicles occurring on the lip vermilion have been described, but not enough attention has been given to this particular detail (2). We describe here lip lesions occurring in four patients with bullous LE.

CASE REPORTS

Clinical features of the 4 patients are shown in Table I. There were one male and 3 female patients (age range 18–44 years). All patients presented with an acute eruption characterized by a variable number of tense blisters that erupted on normal skin or overlying oedematous plaques, with variable topography. All patients had multiple intact or crusted vesicles on the lip; these tended to assume a linear arrangement along the vermilion in all patients, with almost no visible inflammation (Fig. 1). After crusting, lesions healed quickly. Intraoral blisters and/or erosions were present in all but one patient. Herpes labialis was ruled out by negative Tzanck smears prior to biopsy. Cutaneous, as well as mucosal, lesions were photographed and biopsied for histopathological and immunofluorescence studies. In all cases direct immunofluorescence of oral lesions showed granular IgG and IgM, and linear IgA along the basal membrane zone. For all 4 patients, initial clinical diagnoses included dermatitis herpetiformis, bullous pemphigoid, linear IgA disease, epidermolysis bullosa acquisita, and bullous LE. All patients were finally diagnosed as having bullous LE, based on histopathology, immunofluorescence, and serology (Fig. S1; available from: http://www.medicaljournals.se/acta/content/?doi=10.2340/00015555-1276, Table I).

DISCUSSION

Oral lesions of lupus erythematosus are known to represent the exact mucous counterpart to cutaneous LE lesions; thus, chronic discoid, verrucous, acute and bullous lesions are well-known types of cutaneous LE that can present with analogue oral compromise (4). Oral lesions in subacute cutaneous LE have been observed and characterized (5).

Discoid LE of the lip presents as well-demarcated erythematos, squamous, and atrophic patches that typically spread from the vermilion to the skin of the lip. Subacute LE lip lesions commonly present as a diffuse and poorly demarcated scaly cheilitis. Lip lesions often present in patients with classic butterfly-shaped facial lesions of acute cutaneous LE; an eroded and crusted cheilitis is the more commonly observed aspect. All these presentations represent specific lupus disease occurring on the vermilion; and histopathology is diagnostic in all of them (4, 5).

Oral lesions are believed to occur in 30% of patients with bullous LE (6). The most common presentations include ruptured and intact blisters on the buccal mucosa

Table I. Characteristics of 4 bullous lupus erythematosus patients with lesions on the lip vermilion

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex/age, years</th>
<th>Type and distribution of cutaneous lesions</th>
<th>Intraoral lesions</th>
<th>Lip lesions (vermilion)</th>
<th>Lupus serology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M/18</td>
<td>Discrete blisters on normal skin, without inflammation – face, trunk</td>
<td>Ruptured blisters on buccal mucosa</td>
<td>Intact and crusted vesicles, linearly arranged</td>
<td>ANA-1:1280 Anti DNA- 200 UI/ml</td>
</tr>
<tr>
<td>Fig. 1A</td>
<td></td>
<td></td>
<td></td>
<td>Tiny vesicles and crusts</td>
<td>ANA-1:640</td>
</tr>
<tr>
<td>2</td>
<td>F/40</td>
<td>Large, tense blisters overlying erythematous patches on the face, neck and chest (Fig. S1E)</td>
<td>Intact and ruptured blisters on buccal mucosa</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fig. 1B</td>
<td>and S1E</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>F/22</td>
<td>Tense blisters overlying erythematous patches, widespread on face, trunk and arms</td>
<td>Intact and ruptured blisters on buccal mucosa</td>
<td>Intact and crusted vesicles</td>
<td>ANA-1:1280</td>
</tr>
<tr>
<td>Fig. 1C</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>F/44</td>
<td>Sparse vesicles on normal skin, hands and chest</td>
<td>No</td>
<td>Multiple intact vesicles</td>
<td>ANA-1:1280 Anti DNA- 200 UI/ml</td>
</tr>
<tr>
<td>Fig. 1D</td>
<td></td>
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</table>

DIF: direct immunofluorescence; BMZ: basement membrane zone.
and palate; these represent the LE process on the mucosa (2). The 4 patients described here presented with marked labial compromise besides skin lesions; the pattern of presentation was identical in all of them: tense or crusted, almost linearly arranged small blisters along the vermillion, with virtually no erythema, oedema or scaling.

We believe that this particular distribution of the lesions is not frequently observed in diseases that present clinical resemblance to bullous LE (dermatitis herpetiformis, bullous pemphigoid, linear IgA disease, and epidermolysis bullosa acquisita), but is also very common in pemphigus vulgaris, paraneoplastic pemphigus, and erythema multiforme. However, labial intact blisters are almost never seen in these diseases; instead, one sees irregular painful erosions and hemorrhagic crusts.

Intact and ruptured vesicles are the rule in labial herpes simplex but they present grouped on an erythematous and oedematous base, different to labial bullous LE. We suggest that LE serology should be requested promptly in addition to histopathology and immunofluorescence tests when patients present with lip lesions similar to those we described herein.

Biopsy of a semimucosal lesion was performed only on patient no. 4, since she did not present intra-oral lesions; we preferred to perform intra-oral biopsies in the other three patients, since this procedure may cause less discomfort and is equally diagnostic. Histopathological features were identical in all cases, and are the same as for cutaneous bullous LE: dermo–epidermal cleavage, multiple neutrophils in the dermal papillae, and intense interstitial mucinosis (Fig. S1).

Direct immunofluorescence was positive for IgA, IgM, IgG, and C3 on the dermal–epidermal junction in all specimens, which is typical for bullous LE.

In conclusion, we believe that the presence of several intact or ruptured blisters along the lip vermilion that do not progress to extensive and coalescing erosions or haemorrhage is highly suggestive of bullous LE.

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REFERENCES