Purplish Papules and Plaques with Blisters on Lower Lip

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CASE REPORT

A 47-year-old woman came to our dermatology clinic for longstanding erosive and crusting vesicles on her lower lip. The erosions began three years earlier and responded poorly to topical steroids. Dermatologic examination revealed rupturing vesicles arising from violaceous polygonal plaques over her lower lip with erosions, blood clots and crust formations. The lesions bled easily while we pulled the lower lip down for recording photos (Fig.1a). Other parts of the oral mucosa were intact and the skin had no similar lesions.

A skin biopsy taken from the edge of the vesicle and the underlying plaque over the lower lip showed a subepidermal blister, vacuolization of the basal cell layer, and Civatte bodies. (Fig. 2 and Fig. 3)
DIAGNOSIS
Bullous Lichen Planus on Lower Lip

DISCUSSION
Lichen planus (LP) is a chronic mucocutaneous disease of unknown causes that is relatively commonly seen in the dermatological clinics.1 LP lesions often affect flexions, lower limbs, genitalia and oral mucosa.1 By comparison, the incidence of bullous lichen planus (BLP) is much lower.1, 2 Reviewing the literature, the cases of BLP that have been reported were most often involving lower limbs and trunk.1 BLP on oral mucosa is rarely seen.

The oral BLP are commonly seen on the buccal mucosa and less commonly on gingival and inner aspect of the lips.2 The bullae are generally short-lived and leave ulcerative lesions on rupturing.2 The lacking of underlying polygonal violaceous plaques of lichen planus delimits pemphigus, cicatricial pemphigoid and other mimicking disease from BLP. More then clinical appearances pathology findings also offer striking values for diagnosis.

The histopathologic hallmarks of BLP are subepidermal blisters along with typical findings of LP.1 It should be distinguished from other subepidermal bullous disorders such as bullous pemphigoid (BP) and lichen planus pemphigoides (LPP), an entity of co-existing of LP and BP.1 BP typically contains eosinophils, neutrophils and predominant lymphocytes; relative normal epidermis and absence of the dense band-like infiltration along dermo-epidermal junction.3 LPP possesses histopathologic features of BP and LP.3 Immunopathology provides additional clues. Direct immunofluorescence (DIF) tests show linear deposition of C3 or immunoglobulins along basement membrane zone in BP and LPP, but usually not in BLP.3

The causes of BLP are not well known. Immunologic processes, exogenous chemical substances, and physical agents are all considered to play roles in the pathogenesis of BLP. These events are supposed to trigger the destruction of the basal keratinocytes through host immune responses to the wide range of various antigens.4

The Langerhans cells presumably act as antigen presenting cells which process the antigens and present antigens to the lymphocytes which in turn destroy keratinocytes causing cytolysis.4 It then follows that the "subepidermal clefts" (Max-Joseph spaces) results from the cytolysis of increasing numbers of cells along the basal layer of the epidermis.4 With further extremely destruction course, the clinically rarely seen blisters of BLP eventually developed.4

The treatment modalities of OLP include topical corticosteroids, topical treinoin, topical cyclosporine, intralesional corticosteroids, systemic corticosteroids, retinoids, antimalarials, griseofulvin, dapsone, oral PUVA, and surgical techniques.5 Recently, topical tacrolimus has been reported showing promising effects for OLP.5 Our case is the first reported case of oral BLP that has been effectively treated with topical tacrolimus in Taiwan. (Fig. 1b-d) Tacrolimus inhibits calcineurin and thus interferes with the synthesis of cytokines such as IL-2, IL-3, IL-4, IL-12, TNF and INF-γ. Tacrolimus also inhibits T-cell proliferation,5 and we suppose this may prevent further destruction of keratinocytes by infiltrated lymphocytes in BLP lesions because of the recruitment of cytotoxic lymphocytes is disrupted.

REFERENCES