Case 2
An 82 year-old Thai woman from Petchaburi
Chief complaint: Multiple erosions for 4 weeks

Present illness: An 82-year-old woman with a medical history of hypertension and DM type II presented with a 4-week history of blisters on her hands and feet that subsequently spread to the trunk and face. Blisters developed on sites of previously uninvolved skin and ruptured into erosions. She also complained of loss of nail on both big toes

Past history:
Hypertension and diabetes type II treated with amlodipine, enalapril and metformin respectively

Physical examination:
Vital signs: normal
A Thai female, not pale, no jaundice
HEENT: not pale, no jaundice, no oral ulcer, no conjunctival injection
Lymph node: no palpable lymph nodes
Breast: no palpable breast masses
Abdomen: no hepatosplenomegaly

Skin examination:
• Multiple, erosions on erythematous to violaceous patches and plaques on face, both ears, right axilla, abdomen, distal fingers and toes with anonychia of both big toes

Histopathology: (S16-2111B, left big toe)
• Subepidermal vesicle with well-preserved dermal papilla
• Dense inflammatory-cell infiltrate of lymphocytes and numerous eosinophils in the papillary dermis

Direct immunofluorescence:
• Continuous linear deposition of IgG and C3 and Shaggy deposition of fibrinogen at the basement membrane zone
• Colloid of IgM
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Diagnosis: Lichen planus pemphigoides

Treatment:
- 0.05% Clobetasol propionate cream apply b.i.d
- 0.1% Momethasone furoate cream apply b.i.d
- Discontinued enalapril

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Discussion: Lichen planus pemphigoides (LPP) is a rare, acquired autoimmune bullous dermatosis which is a combination of lichen planus (LP) and bullous pemphigoid (BP). It was introduced by Kaposi in 1982 as a typical case of lichen planus complicated by a widespread bullous eruption.\(^1\) Later it was defined as lichen planus pemphigoides in 1964.\(^2\) The diagnosis of LPP is made by characteristic clinical, histopathological, and immunologic features.

Clinically, LPP is characterized by an abrupt onset of tense, dome-shaped bullae prior to, during, or after an eruption of lichen planus. The blisters may arise on uninvolved skin, or on preexisting lichenoid lesions.\(^3\) In LPP the lesions most commonly involve the distal extremities,\(^6,\) but they may occur in a generalized form. Oral and conjunctival mucosa involvement has been reported.\(^8,\) LPP more commonly affects males, usually in the fourth or fifth decade of life.\(^6\) Occasionally, childhood onset has been documented.\(^5\)

Histopathologically, LPP is characterized by typical findings of lichenoid tissue reaction with subepidermal bullae and linear deposits of IgG and C3 along the BMZ on DIF of peribullous skin.\(^10\) Our patients fulfilled the clinical, histological, and immunofluorescent criteria of LPP. Furthermore, DIF in our patient revealed classic findings of continuous linear deposition of C3 and IgG and shaggy deposition of fibrinogen along the BMZ.

The pathogenesis of LPP is incompletely elucidated. Some authors hypothesized that a primary inflammatory process by LP causes the release and exposure of hidden antigen leading to a secondary autoimmune response against the BMZ. These circulating autoantibodies induce secondary subepidermal bullous dermatosis. Many cases of LPP reported in the literature have demonstrated IgG antibodies to either one or both, BP180 and BP230 antigens.\(^11\) Zillikens et al. demonstrated antibody reactivity in LPP against a novel epitope of region 4 of the NC16A domain of BP180 antigen besides regions 2 and 3.5. This finding was not demonstrated in sera from patients with BP which indicate that the immunological pattern in LPP differs from that in patients with BP.\(^11\)

We were not able to conduct immunoblotting studies on our patient; however the BP180 ELISA using a commercial kit was positive in this patient.

LPP has been reported to be induced by medications such
In most cases of LPP, low to moderate doses of oral corticosteroid seems to be effective. In some cases, only topical corticosteroid administration was shown to be effective. There is little information about therapy with other immunosuppressant drugs or combination with corticosteroid for LPP.

The prognosis of LPP seems to be better compared to BP and LP. The rate of recurrence was about 20% and the response after treatment was about 1-12 months, which appears to be lower compared with BP and LP.

In our case, LPP was treated with potent topical steroid and enalapril was discontinued. The patient had gradually resolution of the cutaneous lesions with no recurrence.

References


