Ulcerative Lichen Planus of the Feet
– Case Report and Review of the Literature –

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Ulcerative lichen planus of the feet is a rare variant of lichen planus. We report a case presenting with chronic painful ulceration of the feet and cicatricial changes of the toenails. The pathology of the ulceration showed diagnostic of lichen planus. She also had typical lichen planus on the dorsal hands. The ulceration was resistant to many kinds of topical agents, but completely healed within 2 months of dapsone treatment. The clinical manifestations and treatment responses of this disease in the English literature are also reviewed. (Dermatol Sinica 22 : 306-311, 2004)

Key words: Ulcerative lichen planus, Feet, Dapsone

从临床医学角度来看，脚部的溃疡性扁平苔藓是一种罕见的扁平苔藓类型。我们报告了一例伴有脚部慢性、疼痛溃疡及趾甲之瘢痕化，溃疡之病理变化确诊为扁平苔藓，病患于手背处亦有典型的扁平苔藓表现。此一溃疡以多种局部制剂治疗无效，而在以dapsone治疗两个月内完全愈合。英文文献上关于此病之临床表现与治疗反应亦在本文中予以回顾讨论。（中华皮誌22：306-311，2004）

INTRODUCTION

Ulcerative lichen planus of the feet is a rare variant of lichen planus described by Cram et al. in 1966 and is characterized by ulcers of the feet, toes and sometimes hands, permanent loss of the involving nails and cicatricial alopecia of the scalp. The ulcers of the feet and toes are chronic, painful and disabling, and frequently resistant to conventional medical treatment. Several treatment modalities with variable responses and recurrence rates had been reported in the literature. We report here a case in which ulcerative lichen planus of the feet was responsive to dapsone. The clinical manifesta-
tions and treatment responses of this disease in the English literature are also reviewed.1-21

CASE REPORT

A 77-year-old woman complained that erythematous eruptions with occasional bullous formation had occurred on both of her big toes for six years and had resulted in ulceration and nail plate detachment. The ulcers had developed spontaneously and were oozing, painful and produced a burning sensation and caused her great difficulty in walking. Treatment with topical antibiotics and corticosteroids had been unsuccessful.

Physical examination revealed that both of her great toes were blunted with atrophic skin and shallow ulcers (Fig. 1). All toenails, except the one on the fifth toe of the left foot, were completely absent with atrophy of the nail bed. There were two to three pea- to bean-sized, polygonal, violaceous and flat-topped papules on the dorsal aspect of both hands (Fig. 2). Pterygium formation and tenting of the fingernails were also found (Fig. 3). No anomaly was present on the oral and genital mucosae, and no cicatricial alopecia was detected on the scalp.

Biopsy specimens taken from the ulcer of the right big toe and from the papular lesion of the left dorsal hand were both diagnostic of lichen planus. The epidermis showed hyperkeratosis, wedge-shaped hypergranulosis and irregular acanthosis with saw-tooth pattern of the rete ridges. Lichenoid infiltrate composed of lymphocytes was present in the upper dermis with basal vacuolization, pigment incontinence and cytoid bodies at the dermoeidermal junction in both specimens (Fig. 4). There were also collections of neutrophils in the horny layer, and many plasma cells in addition to lymphocytes in the lichenoid infiltrate of the toe lesion (Fig. 4B). Direct immunofluorescence showed sever-
al cytoid bodies coated with IgM along the dermoepidermal junction (Fig. 5).

Results of the following laboratory studies were normal or negative: complete blood count and differential count, serum antinuclear antibody, C3, C4, VDRL and serum chemistry panel (except for elevated liver enzymes). The test for serum surface antigen of hepatitis B was negative but that for anti-HCV antibody was positive. The level of serum glucose-6 phosphate dehydrogenase was normal.

We started treatment with dapsone (diaminodiphenylsulfone) 100 mg per day in September 2001. The ulcers healed completely within two months (Fig. 6). The skin lesions also resolved. The patient was free of pain, and her quality of life and mobility were restored. After seven months of dapsone therapy, we decreased the dosage of dapsone to 100 mg every other day without recurrence after 15 months of follow-up. However, in March 2003, the patient was lost to follow-up and she discontinued dapsone treatment. Painful ulcers recurred on the toes, soles and fingers two months later.

**DISCUSSION**

Ulcerative lichen planus of the feet is a very rare variant of lichen planus. There are 27 reported cases in the English literature from 1960. Among the reported cases, the disease shows a female predominance and a mean duration of 11 years (range: 2 to 49 years). The age at diagnosis is from 42 to 86 years, whereas the
The age of onset is between 26 and 76, but beyond middle age in most cases. The presence of ulcers involving the feet is the unifying feature, with only two cases with involvement of the hands. The oral mucosa is more frequently involved than the genital area. The typical eruptions of lichen planus elsewhere on the body are often presented. Cicatricial changes of the scalp are reported in about a half of the cases and involvement of the nails in four-fifths. Malignant degeneration has been reported, including verrucous carcinoma\(^4\) and squamous cell carcinoma.\(^2\) Association with other autoimmune diseases such as systemic lupus erythematosus,\(^1\) rheumatoid arthritis\(^5\) and sicca syndrome\(^6\) have also been reported. Except for the presence of ulcers on the hands, our patient's characteristics fairly well fitted the average of the reported cases.

The biopsy specimens of ulcerative and papular lesions on our patient both revealed diagnostic features of lichen planus. In the specimen from the toe ulcer, the lichenoid inflammatory infiltrate contained lymphocytes and many plasma cells. Plasma cells are not usually found in lichen planus, but were reported as constituting up to 60 to 70% of the cells in the inflammatory infiltrate in one case with characteristic ulcerative lichen planus of the feet; no explanation for this phenomenon was offered.\(^7\)

The main differential diagnosis is dystrophic epidermolysis bullosa, which also presents with chronic ulceration, blistering, scarring and nail dystrophy on the acral part. Ulcerative lichen planus of the feet differs from this condition in adult onset and the absence of milium formation.

Since there were many plasma cells in the lichenoid infiltrate in the specimen from the toe ulcer, secondary syphilis should also be considered, but the negative VDRL test in this case excluded this possibility.

Dapsone therapy resulted in complete healing of the ulcers and remission of the disease in our patient. We decreased the dosage to 100 mg every other day without a recurrence after 15 months of follow-up. New ulcers recurred on the toes, heels and fingernails two months after discontinuance of dapsone treatment. The clinical course suggests that dapsone does not eradicate or cure the disease but only induces and maintains remission.

Dapsone therapy was previously reported to be useful in one case, who was disease free after 15 months of follow-up.\(^8\) Although most of the diseases responsive to dapsone are neutrophil-related, the effectiveness of dapsone in this lymphocyte-rich and neutrophil-poor disease could be explained by its inhibition of the myeloperoxidase-hydrogen peroxide-halide systems, which is found both in neutrophils and mononuclear cells.\(^8\) Since mononuclear cells also contain myeloperoxidase, dapsone may have changed the inflammatory response in our patient's lichenoid infiltrates by its inhibition of mononuclear cell-mediated myeloperoxidase.

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**Table I. Comparison of the therapeutic results (case number) of the previously reported effective treatment**

<table>
<thead>
<tr>
<th>STSG</th>
<th>Dapsone</th>
<th>Etretinate</th>
<th>Cyc</th>
<th>Cyc</th>
<th>Thalidomide</th>
<th>PDGF-BB gel</th>
<th>Tacrolimus ointment 0.1%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Successful</td>
<td>1,2,5,11,12,13,16</td>
<td>2,9*</td>
<td>3,14,15</td>
<td>1,10</td>
<td>0</td>
<td>2,11</td>
<td>1,12</td>
</tr>
<tr>
<td>Responsive</td>
<td>1,21</td>
<td>0</td>
<td>0</td>
<td>1,19</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ineffective</td>
<td>1,16</td>
<td>4,10,12,16</td>
<td>4,10,16</td>
<td>1,11</td>
<td>2,10,11</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

*Including the present case
**Ulcers worsened with local application of cyclosporine solution.

STSG: split-thickness skin graft
Cyc: cyclosporine
PDGF: platelet-derived growth factor
However, four other cases were reported to be unresponsive to dapsone treatment and one case showed only slight improvement (Table I).9, 11

Ulcerative lichen planus of the feet is known to be frequently resistant to medical therapy and the main reliable treatment to date is excision with autologous skin grafting.2, 13 Treatment modalities and clinical responses reported in the English literature since 1960 are summarized in Table I. Many agents have been used systemically, including steroids, azathioprine, hydroxychloroquine, isoniazide, dapsone, etretinate, cyclosporine and thalidomide. Success has not been reported for treatment with any of the first four agents. Dapsone achieved remissions in two cases (including the present case) but failed in four.9, 10, 11, 16 Etretinate achieved complete remission in one case,15 90% healing in another5 and some improvement in one patient who discontinued therapy because of cheilitis and headache.14 However, it also failed in four cases.6, 11, 16 Cyclosporine has been used in 3 cases with one success,16 one initial improvement but recurrence after complete withdrawal of cyclosporine,19 and one failure.11 Thalidomide, in a regimen of 150-mg per day initially and 50-mg every other day as maintenance dose, has been reported to be effective in 2 cases refractory to many medical treatments.11

Few topical agents have been reported to be effective. Topical high strength steroid was ineffective in all of the reported cases except in one patient whose ulcer healed with betamethasone valerate ointment and triamcinolone intralesional injection.3 Cyclosporine in an oily solution four times daily exacerbated the ulceration in two cases.10 Platelet-derived growth factor (PDGF)-BB gel and tacrolimus ointment 0.1% were each reported to be effective in one case.12, 18 In one case, treatment of ulcers with topical PDGF-BB gel was successful when mycophenolate mofetil and UV A1 therapy had failed.

In most cases, autologous skin grafting with or without excision resulted in complete healing and long-term remission, with recurrence in only two of thirteen patients.

In summary, ulcerative lichen planus of the feet is an unusual variant of lichen planus characterized by painful ulceration of the feet and toes, permanent loss of the involving nails and cicatricial alopecia. It is notoriously unresponsive to conventional medical treatment. We report a case in which ulcerative lichen planus of the feet is characteristic and responsive to dapsone.

REFERENCES