

Severe Generalized Ulcerative Lichen Planus

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Sir,

Ulcerative lichen planus (ULP), a rare variant of lichen planus, is characterized by chronic painful ulcers on the feet (1). We report a patient who presented after PUVA therapy with generalized ulcerations on an erythematous base, and with natural remission within a month.

CASE REPORT

A 67-year-old man was referred to our hospital in March 2000 for evaluation of poikiloderma on the entire body surface. The lesions started as an itching rash on the buttock in 1965 and had gradually spread, especially during summer months. Erosions and/or ulcers had formed on the trunk and limbs a few times per year since 1968. These lesions healed within a month without treatment, leaving either hypo- or hyperpigmented macules, and developed to poikiloderma when he was 64 years of age. Simultaneously, he gradually lost the hair on his scalp. His medical history included atrial fibrillation without medication since the age of 50. He had not used any kinds of drugs habitually.

Physical examination revealed diffuse erythema with fine scales, based on reticulate pigmentation and teleangiectasia on the trunk, limbs and head. Hyperkeratosis was seen on the palms and soles. The scalp hair was sparse (Fig. 1). The finger and toe nails showed pachyonychia with yellowish discoloration. There were no lesions on the mucous membranes.

The results of routine laboratory examinations, including liver function tests, were within normal limits. The results of the following laboratory studies were within normal limits or negative: anti-HTLV-1 antibody, anti-hepatitis C virus antibody, hepatitis B surface antigen, LE test and uroporphyrine.

A skin biopsy from the erythema on the thigh showed wedge-shaped hypergranulosis, irregular acanthosis, damage to the basal cell layer, and band-like dermal lymphocytes. Interestingly, fatty tissues, reaching to the upper dermis, were seen beneath the band-like zone of inflammation. Below the fatty tissue there were thick collagen deposits through the deep dermis and a few hair follicles.

Direct immunofluorescence showed no specific deposits. The findings were highly suggestive of lichen planus, but a provisional diagnosis of parapsoriasis lichenoides was made because of the generalized lesions. Topical applications of diflucortolone valerate and tacalcitol, and topical PUVA therapy to the back, were initiated in March 2000 and had a moderate effect.

In April, the patient had a sudden onset of high fever with numerous small vesicles and erosions on the back. These symptoms led us to discontinuation of PUVA therapy. Aciclovir was given without effect and antibodies

to herpes simplex and varicella zoster viruses were negative. Direct immunofluorescence of the vesicular skin showed no linear deposits of IgG or C3 in the basal membrane zone, with positive immunostaining for laminin on the floor of the cleft (Fig. 2). Furthermore, immunoblot analysis demonstrated no specific antibodies. Ulcerative lichen planus was diagnosed on the basis of these data.

A spiking fever continued and the erosions developed rapidly into irregular-shaped ulcers, widespread over the whole body. The patient was treated with symptomatic therapies and the ulcers began to re-epithelize on the upper trunk. In mid-May, the high fever gradually subsided and the ulcers cleared almost completely. Several erosions and vesicles were noted occasionally, but they did not spread widely during the subsequent 2 months. The patient suffered from recurrent episodes of widespread vesicular lesions in mid-July. In this episode, the patient became bed-ridden due to pain from the ulcers, and had worsening decubitus on the sacral region. The ulcers and general condition improved with unchanged symptomatic therapies in September. However, his condition progressively deteriorated due to



Fig. 1. Poikiloderma of the back.

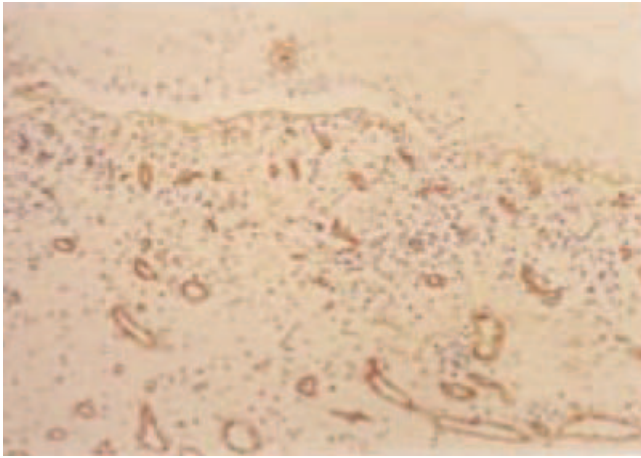


Fig. 2. Laminin at the floor of the fissure, demonstrated by immunostaining. No deposits of IgG or C3.

bacterial infection on the decubitus, causing septicaemia, and he died on 29 September. Autopsy was refused.

DISCUSSION

Lichen planus is a subacute or chronic dermatitis that may involve the skin, mucous membranes, hair follicles and nails (1). Variations in the morphological appearance of lichen planus have been noted for both skin and mucosal lesions. The lesions may be annular, vesicular and bullous, hypertrophic, linear, erosive and ulcerative, atrophic and follicular, or actinic and pigmented (2, 3). This disease may also be classified into localized, disseminated and generalized types. In generalized and disseminated cases the tendency is for a more acute onset and spread of the lesions and, usually, for a short duration. The localized forms tend to become chronic (2).

The clinical characteristics of our patient were as follows: chronicity, generalized lesions, periodic episodes of ulceration and cicatricial alopecia but no involvement of mucous membranes or nails. The differential diagnosis includes lichen planus pemphigoides, bullous pemphigoid, epidermolysis bullosa acquisita, bullous lupus erythematosus, porphyria cutanea tarda and parapsoriasis lichenoides. Our patient's clinical and histological features and the results of immunofluorescence,

immunostaining and immunoblotting studies are compatible with ULP, a rare variant of lichen planus.

ULP is an entity characterized by chronic, painful, cicatricial bullae and ulceration of the sole of the foot, and often of the mouth and other mucosa, together with scalp and nail lesions (4). The ulcers are resistant to medical therapy. Surgical excision and grafting of ULP has been reported in individual cases since 1951 (4, 5). Compared with classical ULP, our case showed a great difference in the anatomical region of ulceration and in resistance to therapy. Shared features are chronicity of the lesions, cicatricial alopecia and painful ulcers.

Interestingly, our patient's histological characteristics were scar formation in the deep dermis and a lichenoid tissue reaction in the upper dermis. In addition, an erosive lesion showed a cleft between the basal cells and the basal membrane zone. These findings supported the clinical characteristics of relatively shallow ulcer formation and severe pain, and the recurrent episodes of ulceration.

There was a summer-time exacerbation in our case. In fact, PUVA therapy induced an ulcerative condition. Lichen planus actinicus is a variant of lichen planus worsened by exposure to the sun. Accordingly, our case seemed unlikely to be of this type. Although PUVA therapy is a regimen advocated for various types of dermatitis, including lichen planus, it should be kept in mind that PUVA therapy may possibly exacerbate lichen planus, as well as increased risks for skin cancer (6).

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