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LETTERS TO THE EDITOR

Ulcerative Lichen Planus: A Case Responding to Recombinant Platelet-derived Growth Factor BB and Immunosuppression

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Sir,

Lichen planus ulcerosus, a rare variant in the clinical spectrum of lichen planus, leads to limited mobility, pain and disability. The first description of ulcerative lichen planus was written by Friedman (1), who already mentioned the combination with secondary anonychia (1, cf. 2). Since then, several case reports and small series have been published (2–8).

The association of ulcerative plantar lichen planus with onychoatrophy and a variety of other autoimmune features has been classified as an autoimmune syndrome (7, 8).

We describe here a female patient with a combination of ulcerative and hypertrophic lichen planus of the skin, oral lichen planus and secondary anonychia treated with systemic mycophenolate mofetil and UVA1. The plantar ulcers, however, were recalcitrant. Ulcer healing was induced by topical treatment with recombinant platelet-derived growth factor (PDGF)-BB.

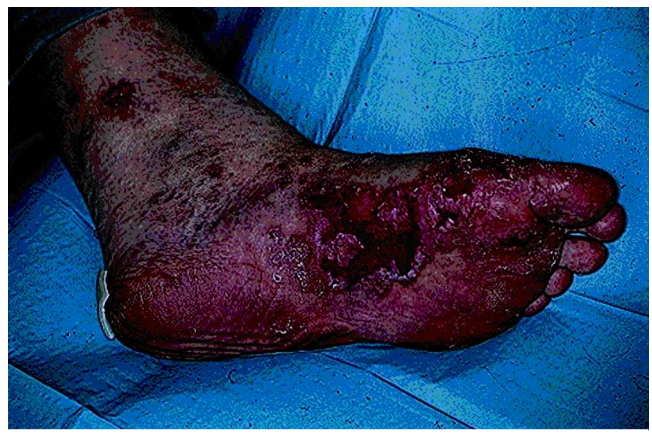
CASE REPORT

A 42-year-old otherwise healthy woman with a 10-year history of chronic inflammatory skin and oral lesions was referred to our department. She already had undergone diagnostic skin biopsies that confirmed the diagnosis of lichen planus, for which she was receiving treatment with topical corticosteroids, PUVA-bath and chloroquine, but without success. She developed secondary anonychia of the toenails and painful plantar erosions and ulcers that had been covered with hydrocolloids but did not heal.

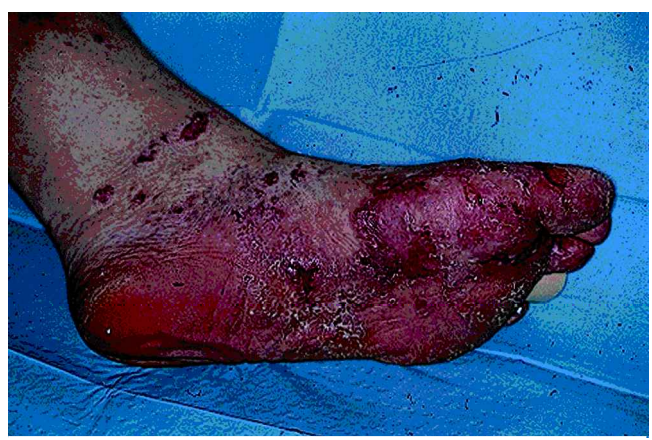
On clinical examination, typical lesions were found on the buccal mucosa and the lips, showing a Wickham striae pattern; the tongue epithelium was atrophic. The patient had multiple inflammatory plaques on the trunk and limbs. On the lower legs the lesions were hypertrophic, and partially excoriated. On the palms painful erosive and ulcerating lesions were combined with anonychia of the toenails (Fig. 1*a*).

Laboratory investigations

Normal values were obtained for differential blood count, serum enzymes, creatinine, urine analysis, protein electrophoresis, zinc, serum iron, and blood sedimentation rate. We detected autoantibodies against endomysium (IgA-type, 1:320), reticulin (IgG 1:160, IgA 1:160), thyroideal microsomes (MAK, 1:1600), cardiac muscle (1.40) and antinuclear antibodies, speckled type, 1:100. Antibodies against pemphigus and pemphigoid antigen and syphilis serology were negative.



(a)



(b)

Fig. 1. Ulcerating plantar lichen planus. (*a*) before and (*b*) at the end of topical treatment with platelet-derived growth factor BB. Marked improvement with an almost complete wound closure.

Treatment and course

We started treatment with mycophenolate mofetil (CellCept, Hoffmann-La Roche, Germany) 2 g/day in July 1999 in combination with topical bethametasone ointment and achieved a partial improvement, except on the soles of the feet. From August to October 2000, we combined the treatment with UVA1 low-dose irradiation (30 times, last single dose 6 Jm^{-2}, total dose 180 $J\,m^{-2}$), achieving a remarkable improvement.

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The remaining plantar ulcers were covered with hydroactive wound dressings (Askina transorbent; Braun-Petzold, Germany). The patient noted some alleviation of pain. Secondary infections were prevented but the ulcers failed to heal. In October 2000, topical PDGF-BB gel was applied (Regranex, Janssen-Cilag, Germany), which caused an almost complete remission within 5 months (Fig. 1*b*).

The treatment was well tolerated. No systemic or topical side effects were noted, and quality of life and mobility were enhanced remarkably. At the end of treatment, the patient was completely free of pain.

DISCUSSION

Lichen planus ulcerosus is frequently resistant to medical systemic treatment including chloroquine, corticosteroids, PUVA, and cyclosporin A (4–6). Surgical excision and grafting are considered to be the treatment of choice (7, 9–11).

The present patient was characterized by a combination of ulcerative lichen planus and secondary anonychia that has been identified as an autoimmune syndrome (7, 8). Indeed, she showed a broad range of autoantibodies including antinuclear antibodies and organ or tissue specific antibodies. In patients with ulcerative lichen planus autoimmune diseases should be investigated.

Medical treatment is difficult but we, as well as others (12), observed improvement of disseminated lesions in the present patient after mycophenolate mofetil and UVA1 low-dose therapy. Plantar ulcers, however, did not respond to medical therapies and hydroactive dressings despite the fact that inflammation and pain were reduced.

PDGF is released from alpha granules of platelets during the acute phase of wound healing. Three different isoforms have been detected, but PDGF-BB is the most potent. In addition to autocrine and paracrine stimulation of its own synthesis, PDGF is mitogenic and chemotactic for connective tissue cells and stimulates the production of other cytokines and growth factors important in wound healing (13). Recombinant PDGF-BB gel has been shown to improve wound healing in difficult-to-treat neuropathic diabetic foot ulcers (14) and full thickness pressure ulcers (15). The present case seems to be the first with ulcerative lichen planus treated with PDGF-BB gel that resulted in an almost complete response, thus making grafting unnecessary.

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