A Severe Case of Ulcerating Necrobiosis Lipoidica

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Sir,

Necrobiosis lipoidica (NL) is a chronic granulomatous skin disorder. There is an association with diabetes mellitus. NL most often appears on the pretibial regions as yellowish or brownish, atrophic, telangiectatic plaques surrounded by raised, violaceous borders. Less common locations include the upper extremities, face and scalp. The average age of onset is 30–40 years. The female:male ratio is 3:1. Ulceration occurs in 35% of lesions, usually following minor trauma. Squamous cell carcinomas have rarely been reported to arise in areas of NL (1, 2). Only 4 cases of malignancy have been reported since 1966 (3). The occurrence of NL in diabetic patients is 3 in 1000 (2). NL is not related to the duration or regulation of diabetes (1), but it does appear that diabetic patients with NL have a higher rate of diabetes-related end-organ damage. The exact cause of NL is unknown. One of the leading theories has focused on diabetic microangiopathy. Other theories suggest trauma, inflammatory or metabolic changes and antibody-mediated vasculitis (1). Treatment of NL includes several modalities. Because the underlying pathological mechanism is unknown there is no rational therapy. However, different treatments have been reported to be beneficial. These include potent topical corticosteroids or intralesional corticosteroids injected into the active borders. Other treatments include aspirin plus dipyridamole, cyclosporine, chloroquine, photochemotherapy (psoralen+ultraviolet light A; PUVA), topical tacrolimus, wound healing enhancers, surgery and topical tretinoin (4). No overall treatment for NL has, however, proven to be effective in double-blind placebo-controlled studies (1).

In the present case report, we describe a woman with severe NL, resistant to medical treatment and PUVA who finally underwent surgery with removal of all skin layers followed by split skin grafting.

CASE REPORT

A 47-year-old woman with insulin-dependent diabetes since 1977 was admitted. She had diabetic complications such as microangiopathy, peripheral neuropathy, microalbuminuria, proteinuria and moderately reduced kidney function. In 1999 she was diagnosed with NL. In May 2000 she developed wounds on her left tibia as a result of trauma. She was treated initially in a local hospital, but as of May 2001 further treatment was carried out in a specialized wound centre in the department of dermatology. Topical corticosteroid treatment had no effect. Treatment with methotrexate was tried for 2 months with no effect. Other treatments were considered, including cyclosporine, but this was relatively contraindicated because of her reduced kidney function. From October 2001 a series of 40 PUVA treatments was given. Prior to PUVA treatment there were several ulcerations in the NL area of the left tibia. On commencement of PUVA treatment, the wounds began to show marked healing, but after 40 PUVA treatments they began to deteriorate, increased in number and developed large superficial erosions (Fig. 1). In September 2002, after ending PUVA treatment, ulcerations covered the entire pretibial region. To exclude malignancy 2 punch biopsies were obtained from the left tibia and after preparation stained with haematoxylin and eosin. The histological findings showed fibrosis in the dermis, diffuse chronic inflammatory changes and granulation tissue, but no signs of malignancy and no granulomatous inflammation

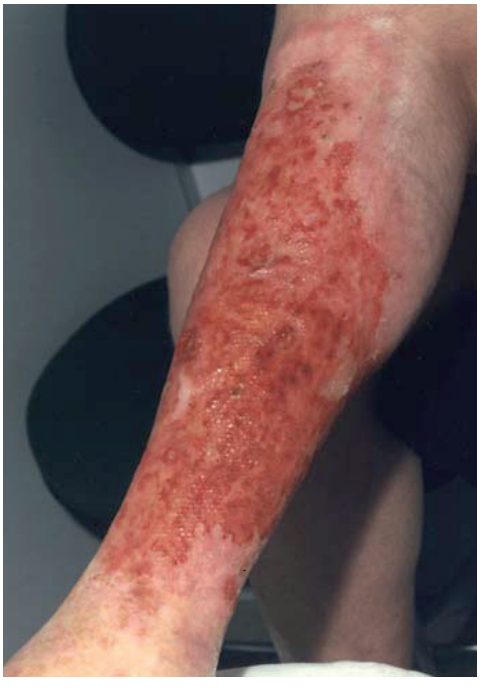

Fig. 1. Deterioration after psoralen + ultraviolet light A (PUVA) treatment (11 October 2002).

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were demonstrated. Topical tacrolimus (Protopic, Astellas Pharma, Glostrup, Denmark) and pimecrolimus (Elidel, Novartis, Copenhagen, Denmark) treatment were tried for a period of 2 months, but with no effect. Because conventional as well as experimental medical treatments had no effect, the patient was referred to the department of plastic surgery. Excision of the lesion and the surrounding skin, including the deep fascia, which was also affected, was performed on the left tibia, followed by split skin grafts to cover the defect (Fig. 2). Complete take was observed and at 4 months follow-up the wound was fully healed (Fig. 3). In December 2005 the patient had a trauma on the anterior aspect of her right tibia resulting in an ulcer measuring 5×3 cm. The surrounding skin showed clinical signs of NL. This ulcer, however, healed during the following 5 months by means of moist wound treatment and compression. At follow-up in June 2006, both legs were healed and no signs of ulceration were observed.

DISCUSSION

In the present case a 47-year-old woman was treated for NL during a period of more than 2 years. She was diagnosed with NL in 1999. A trauma to her leg in 2000 led to non-healing wounds and treatment was required.

The response to all the different treatments was variable. Initially, the wounds were treated with topical steroids followed by methotrexate, but with no effect. A few reports have indicated a beneficial effect of PUVA on NL (5, 6). Therefore, PUVA was tried in our patient. Tacrolimus and pimecrolimus were also used topically. None of all these treatments were effective overall. Surgery has been reported previously to be beneficial in NL (7). Seven patients were treated successfully with surgical excision of the lesions followed by split skin grafting (7). Therefore, the patient was referred to the department of plastic surgery for excision of all skin layers followed by split skin grafting. Full take was observed and the patient healed completely within 3 months. As a consequence of surgery our patient needed to use compression stockings for the treatment of lymphoedema of the forefoot. In our patient histological examination was performed from skin biopsy specimens from skin within the NL area. Histological examination did not show the classical findings for NL, which is granulomatous inflammation together with a lymphocytic infiltrate or necrobiosis with focal loss of elastic tissue (1). In our case the histology was non-specific, showing fibrosis in the dermis and diffuse chronic inflammatory changes. These findings could indicate that NL was at the end stage with no specific granulomatous inflammatory changes. This may explain why our patient did not respond to the different treatments given. The NL was not a classical NL but an ulcerating NL and the only treatment that was successful and stopped the progression of the ulceration was plastic surgery with excision of all inflamed tissue followed by split skin grafting. The patient recently had a trauma to her right tibia followed by ulceration in a skin area that clinically had changes consistent with NL. This ulcer, however, healed by means of conventional local moist wound treatment combined with compression without the need for medical or surgical treatment. This new ulcer underscores the relation between ulcerating NL and trauma.

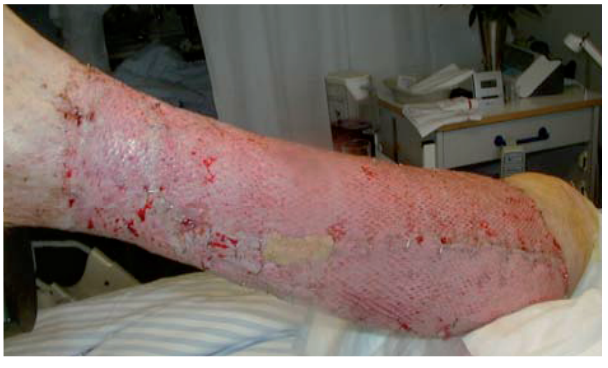

Fig. 2. Clinical presentation after grafting (10 January 2003).

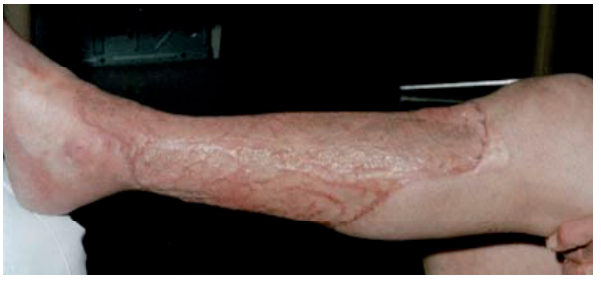

Fig. 3. Clinical presentation 4 months after grafting (14 May 2003).

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