

Case Report

## Pyoderma Gangrenosum-like Presentation of Ulcerative Necrobiosis Lipoidica: Diagnostic and Therapeutic Challenge

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### Abstract

**Observations:** Necrobiosis lipoidica is an uncommon granulomatous disease. Other than the classic form of the disease, ulcerative necrobiosis lipoidica is rarely seen. It is one of the differential diagnosis of leg ulcers. We report a case of 76-year-old woman with severe, chronic, treatment resistant ulcerative necrobiosis lipoidica of the legs. The ulcers were similar to pyoderma gangrenosum. The patient was treated with oral cyclosporine and marked improvement was seen after three months.

### Introduction

Necrobiosis lipoidica (NL) is an uncommon granulomatous skin disease. Classic form of NL has a very characteristic clinical appearance, with yellow-brown, atrophic, telangiectatic plaques surrounded by raised, violaceous rim in the legs [1, 2]. Ulcerative form of NL has not typically clinical morphology and it may be misdiagnosed by the dermatologists. Ulceration occurs in 15-35% of cases of NL and it is usually resistant to therapy. Hence, multiple treatment modalities that are including corticosteroids, fibrinolytic agents, psoralen plus ultraviolet A (PUVA), UVA1, etanercept, mycophenolate mofetil and cyclosporine have been tried [3]. We report a case of 76-year-old woman with severe ulcerative NL who was successfully treated with cyclosporine.

### Case Report

A 76-year-old woman was referred from plastic surgery clinic with gradually enlarging painful

ulcerated lesions on both lower legs. She had two giant ulceration for two years. The ulcer on the medial malleol of right leg was 3x4 cm in diameter, with necrotic material and cribriform base. The other ulcer on the pretibial region of the left leg was 4x6 cm in diameter, with erythematous base by raised border (Figure 1a and 1b). She had rheumatoid arthritis for 20 years and she had characteristics hand deformities.

Multiple biopsies were taken from both ulcers. Histopathological examination showed necrobiotic collagen in the mid-dermis. A cellular infiltrate of histiocytes, polymorphic nuclear leukocytes and palisaded granulomatous inflammation surrounded the necrobiotic area. Mucine deposition in focal areas were seen with Alcian Blue stain. Necrobiosis lipoidica was diagnosed. The biochemical test including blood glucose were all in normal ranges.

Topical mometazone furoate, intralesional triamcinolone and oral prednisolon had been tried during one year but, without benefit. Cyclosporine was given 4 mg/kg/d (100 mg b.i.d). After one month period, little improvement with healing from the edges was seen in the lesions. After three months, marked improvement was



**Figure 1a.** The ulcer on the medial malleol of right leg.



**Figure 1a.** The ulcer on the left leg, with erythematous base by raised border like pyoderma gangrenosum.

seen in all of the ulcers (**Figures 2a** and **2b**). The patient tolerated cyclosporine well and no side effect was seen. The therapy was stopped after three months and during the one year the follow-up period, there was no evidence of relapse.

### Discussion

Necrobiosis lipoidica is a relatively rare cutaneous condition which is closely associated with diabetes mellitus. It is a degenerative disease of collagen and its etiology is still unknown. However many theories have been proposed, including a diabetic microangiopathy, immune complex disease and defective collagen synthesis [4, 5]. A T-cell mediated hypersensitive immune reaction has been suggested in the pathogenesis of NL in the recent studies [6, 7, 8]. The immune reaction precedes and initiates the release of cytokins that finally cause the degeneration of collagen synthesis and granulomatous alterations [2, 8].

It is not clear what causes NL to ulcerate. Trauma, the poor circulation over the pretibial area and an exaggerated immune response have been suggested as the cause of ulceration. This form is more resistant to therapy than classic variant. However, these patients need to be treated since squamous cell carcinoma occurring in ulcerative NL has been reported in literature [3, 4].

In our case, the patient had giant ulcers on both legs. She had no chronic venous insufficiency. The lesions were similar to pyoderma gangrenosum. Histopathologic findings were consistent with NL. The patient had rheumatoid arthritis for 20 years and ulcerative necrobiotic lesions. Similar lesions as a variant of rheumatoid nodule have been reported in these patients in the literature [3]. However rheumatoid nodule was excluded by histologic changes and clinical appearance for our patient.



**Figure 2a.** Marked improvement of the ulcers on the right leg after treatment.



**Figure 2b.** Marked improvement of the ulcers on the left leg after treatment.

The treatment of ulcerative NL is problematic. Multiple therapeutic approaches have been tried, with varying degrees of success [2, 6, 7, 8, 9]. The lesions of our patient did not respond to topical and systemic steroids. The lesions were so painful and the quality of life of our patient was negatively affected. Thus, cyclosporine was started and it was well tolerated by the patient. After one month therapy, improvement was seen. The beneficial effect of cyclosporine detected in our patient is consistent with the results from other reports suggesting that cyclosporine worked effectively in the therapy of NL. Cyclosporine acts as a very specific anti-inflammatory agent, selectively inhibiting T-cell proliferation. The response to cyclosporine may support the hypothesis that the cell-mediated immunity is involved in the pathogenesis of ulcerative NL [3, 9].

Ulcerative NL must be considered in the differential diagnosis of leg ulcers [10, 11]. Clinical appearance of ulcers may be variable. Cyclosporine may be considered as an alternative agent for the therapy resistant ulcerative NL.

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