

Necrobiosis Lipoidica Diabeticorum of the Scalp: A Case Report

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Abstract

Necrobiosis lipoidica diabeticorum (NLD) is a rare chronic and granulomatous skin disorder that affects 0.3% of diabetic patients. Although the etiology and pathogenesis of NLD is still controversial, it is thought that microangiopathy has an important role. The legs are the most common site for NLD, but involvement of other areas such as the abdomen, upper extremities and scalp have been reported. We present a case of scalp and leg NLD which was diagnosed on the clinical and histopathological basis. (*Iran J Dermatol* 2009; 12: 93-95)

Keywords: necrobiosis lipoidica, necrobiosis lipoidica diabeticorum, diabetes mellitus

Introduction

Necrobiosis lipoidica diabeticorum (NLD) is a rare, chronic and granulomatous disorder that affects 0.3% of diabetic patients¹⁻⁴. Typical lesions of NLD occur as irregular, ovoid plaques with a violaceous, indurated periphery and a yellow central atrophic area. In the later stage of development, visible telangiectasia on the surface is common. The most commonly affected site is the leg. Eighty-five percent of cases involve the legs exclusively and only 2% have no leg involvement⁵.

The lesions of NLD may appear on other parts of the body including scalp⁶, face, abdomen and upper extremities^{3,7}.

One-third of the lesions ulcerate, sometimes spontaneously or after trauma⁸. Hypohidrosis, partial alopecia and skin anesthesia within the lesions have been reported by others⁵. The clinical features on the scalp vary from large plaques of cicatricial alopecia to multiple small areas of scarring⁴. An atrophic form affecting predominantly the forehead and the scalp has been described⁶. In general, the differential diagnosis is



Figure 1. Yellowish coloured, annular plaque of cicatricial alopecia with scaly indurated margin associated with central atrophy and telangiectasia

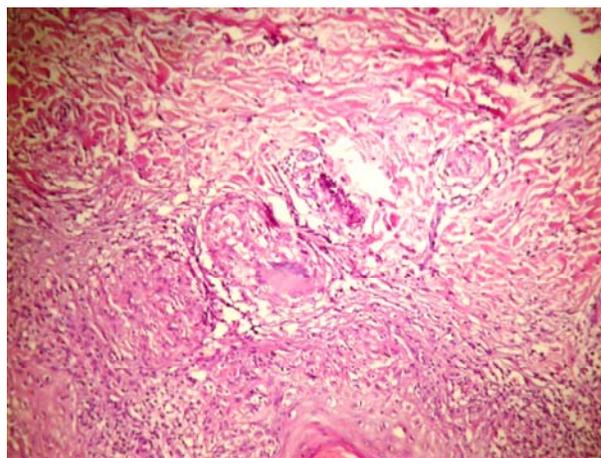


Figure 2. Degenerated collagen, surrounded by a palisade of histocytes, lymphocytes and fibroblasts, with many giant cells (H&E*10).

sarcoidosis and granuloma annulare^{6,9,10}.

The pathogenesis of NLD is unknown. Among proposed causative factors. Microangiopathy and alteration of microcirculation have an important role in the pathogenesis^{4, 11}. The majority of NLD patients are diabetics (both insulin and non- insulin dependent) but progression of the lesions dose not correlate with the control of the hyperglycemia¹¹.

Case Report

A 61-year-old man presented with an approximately one year history of a painless, slowly enlarging annular hairless lesion on scalp. He had a history of diabetes mellitus and hypertension of 15 and 5 years ago, respectively. There was no personal or family history of any other diseases. Physical examination revealed a well – margined 5X5 cm, yellowish coloured, annular plaque of cicatricial alopecia with scaly indurated margin associated with central atrophy and telangiectasia on the scalp (Figure1). There was also a well-marginated, 3X4 cm purple, hyperkeratotic and indurated plaque on the anterior part of right leg. Ichthyotic dry skin and few shin spots were conspicuous on both legs. At the time of physical examination, patient was received glibenclamid 2.5 mg/daily, Atenolol 50 mg/daily and Amlodipine 5mg/daily. Systemic examination revealed no abnormality and blood counts, liver function tests, blood urea, creatinine, triglyceride (TG) and cholesterol were all normal. Blood glucose level was 122 mg/dl, and urine analysis showed one plus of glucose.

A 5-mm scalpel biopsy taken from the scalp lesion showed intact epidermis with some keratin plug in the epidermis layer, the dermis showed degenerated collagen, surrounded by a palisade of histocytes, lymphocytes and fibroblasts, with many giant cells. Mucin stain was negative (Figure2).

A 4-mm biopsy taken from the right leg lesion showed multiple epithelioid cells granuloma with foreign body and langhans giant cells. Granulomas were bare or surrounded by lymphocytes. They were located mostly in the upper and mid dermis and occasionally in the deep dermis. Mucin stain was negative.

Discussion

NLD is a chronic granulomatous dermatitis that is usually associated with type-I diabetes mellitus (DM) and less frequently with type II¹². The characteristic lesions occur on the pretibial skin and begin as a firm, dull-red papule or plaque which

slowly enlarges to become a yellowish, atrophic plaque with an erythematous edge. The surface is often glazed in appearance and telangiectatic vessels may be prominent. The lesions of NLD may appear elsewhere on the body including the trunk, penis, upper extremities and scalp^{4,13}.

Despite extensive investigations, the pathogenesis and cause of NLD is still unknown. Among the several theories, it is thought that microangiopathy has an important role^{1,4,11}. Diabetic microangiopathy, immune-complex vasculitis, and collagen abnormalities have been considered as possible underlying causes in recent publications¹⁴.

Usually the entire thickness of the dermis or its lower third is affected by a process that exhibits a variable degree of granulomatous inflammation, obliterative endarteritis, degeneration of collagen (necrobiosis) and sclerosis.

The granulomatous component is usually conspicuous and there are histiocytes, a few scattered epithelioid histiocytes and giant cell (usually of langhans or foreign- body type). Older lesions, particularly in the middle and lower dermis often show thickening of their walls with proliferation of their endothelial cells. The process may lead to partial and rarely to complete occlusion of lumen^{15,16}.

Necrobiosis of the collagen is thought to develop as the aftermath of the resultant ischemia, which may also be expected to lead to epidermal ulceration and/or atrophy. The granulomatous reaction is believed to occur after the attempt by macrophages to remove the necrobiotic collagen. The differential diagnosis for NLD includes various forms of granulomatous dermatitis. The list comprises sarcoidosis, granuloma annulare, necrobiotic xanthogranuloma with paraproteinemia, annular elastolytic giant-cell granuloma and mycobacterial infections¹⁶.

Scalp involvement by NLD is rare and has been reported in few cases¹⁷. We wanted hereby suggest that NLD should be considered in any patient presenting with hairless granulomatous lesions on the scalp and granulomatous lesion on the leg.

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