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(CASE REPORT)

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Necrobiosis lipoidica as a cutaneous marker of underlying metabolic and autoimmune disorders: A case report

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Abstract

Introduction: *Necrobiosis lipoidica* is a very rare and chronic disease characterized by yellow-brown, atrophic, telangiectatic changes that occur mostly on the lower extremities. It is characterized by the appearance of collagen necrobiosis and dermal inflammation.

Case report: A 50-year-old patient with changes on both lower legs that date back to a year ago and for which she never sought medical help complains of the appearance of an ulcer two weeks ago on one of the changes after a minor trauma.

Conclusion: *Necrobiosis lipoidica* is a rare granulomatous disease associated with diabetes and autoimmune diseases that patients have already diagnosed, but in our case the skin changes were the first that led us to additional research and diagnosis of diabetes and rheumatoid arthritis.

Keywords: *Necrobiosis lipoidica*; Type 1 diabetes; Rheumatoid arthritis; Autoimmune diseases

1. Introduction

Necrobiosis lipoidica is a very rare and chronic disease characterized by yellow-brown, atrophic, telangiectatic changes that occur mostly on the lower extremities. It is characterized by the appearance of collagen necrobiosis and dermal inflammation. In most cases, patients suffer from type 1 diabetes, and some patients who do not have diabetes have abnormal glucose tolerance or a family history of autoimmune disease. (1)

The disease occurs somewhat more often in women than in men with a ratio of 3:1. (2)

The changes are characterized by a central red-brown color that progresses to yellowish-brown. The central region shows features such as atrophy and erosion of the skin. Telangiectasis occurs as a result of the deterioration of collagen that occurs beneath the epidermal layer.

As the disease progresses, the lesions become less active and more atrophic, making them more susceptible to trauma and ulceration. About 30% of patients develop painful ulcerations. (2) Changes may occur on the face, scalp, torso, groin, and upper extremities, but NL is usually limited to the lower extremities, bilaterally. The diagnosis is established by clinical findings and histological examination.

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Histology reveals a pattern of palisaded and interstitial granulomatous dermatitis and localized connective tissue degeneration and vascular changes involving the dermis and subcutaneous tissue. Described as a "sandwich-like" configuration, palisade granulomas are arranged in a horizontal layer with intermingled layers of altered, homogenized, "necrobiotic" collagen. (2)

The etiology and pathophysiology of this disease are still incompletely clarified. Microangiopathy is thought to play a role because of the presence of the disease in diabetic patients, especially those who require insulin. But this disease can manifest itself even in the absence of diabetes, it is also present in individuals diagnosed with sarcoidosis, chronic inflammatory bowel disease, autoimmune thyroiditis, rheumatoid arthritis, monoclonal gammopathy, as well as in otherwise healthy individuals with normal glucose metabolism and no previous history of autoimmune disorders. (2,3) Another significance of increased GLUT-1 expression may be its involvement in the development of vascular occlusion, leading to glycoprotein adhesion to the receptor and constriction of the vascular lumen, which would lead to impaired perfusion and reduced oxygen metabolism (4). Histologic studies indicate vascular involvement, but this affects less than half of the lesions. The results of flow and oxygenation measurement studies do not give a clear answer about possible tissue hypoxia in the course of NL. (4) There are data that suggest that the lesion is not worse but better oxygenated than healthy skin, which would be the result of the inflammatory process. (4,5)

A positive Konbner phenomenon was observed in these patients, which correlates with autoimmune diseases.

The management of NL presents significant difficulties; even when lesions undergo resolution, post-inflammatory and atrophic alterations might persist. The recalcitrant nature of this disease greatly impacts the quality of life of affected individuals [14]. Steroids, namely topical, intralesional, or sometimes systemic, currently serve as the primary therapeutic approach. Other therapy consists of topical tacrolimus, PUVA, antimalarials, photodynamic therapy, fumaric acid esters, pentoxifylline, ticlopidine, biologics, cyclosporine, and excision surgery followed by skin grafting.

2. Case report

A 50-year-old patient with changes on both lower legs that date back to a year ago and for which she never sought medical help complains of the appearance of an ulcer two weeks ago on one of the changes after a minor trauma. The changes are yellow brown indurated. It does not provide data on other comorbidities, and she insisted that was perfectly healthy. A biopsy was performed on the patient. Epidermis with moderate undulant keratosis and focal discrete spongiosis is observed pathohistologically.

In the papillary and reticular dermis, fibrosis and a moderately abundant chronic inflammatory infiltrate of lymphocytes with a perivascular distribution are seen. The most intense changes are present at the level of the deep reticular dermis and part of the subcutaneous fat tissue, where areas of clearly limited fibrosis and hyalinization of collagen fibrils are seen, between which small granulomas of epithelial cells, histiocytes, macrophages in an attempt to form multinucleated giant cells are located. An intense chronic inflammatory infiltrate in the form of nodular aggregates of mature lymphocytes can be seen peripherally from the granulomas.

Chest X-ray is normal. RF IgM 27.88 IU/ml, glycosylated hemoglobin 12.68, and blood sugar 19.61, urine results show the presence of ketones.

Apart from the elevated rheumatism factor the patient had a long-term problem with pain and swelling of the small joints of the hands, but she never sought help, she was referred to a rheumatologist, where she was prescribed hydroxychloroquinon 200mg twice daily for next 3 months.

In the patient, we started treatment with local clobetasol, pentoxifylline, diclofenac, ascorbic acid 1000mg, acetylsalicylic acid and local treatment with silver sulfadiazine for the ulcer. The ulcer started to show signs of closure after two weeks, while the changes remained the same. The patient was referred to an endocrinologist, where the diabetes was brought under control. After two weeks, the treatment was continued only with local therapy for the ulcer and 0,1% tacrolimus, and ascorbic acid, acetylsalicylic acid and pentoxifylline for the next 4 months. The disease was brought under control without the appearance of new lesions even after 6 months, but old lesions lesions still persist with minimal improvement.

3. Conclusion

Necrobiosis lipoidica is a rare granulomatous disease associated with diabetes and autoimmune diseases that patients have already diagnosed, but in our case the skin changes were the first that led us to additional research and diagnosis of diabetes and rheumatoid arthritis.

No treatment has been proven effective for NLD in large studies, but by bringing the other diseases under control and the skin changes of NL in our case were limited and no progression was observed.

Patients without a known diagnosis of diabetes should have screening with fasting blood glucose concentration or HbA1c level at NL presentation given the well-established association with diabetes. Considering the connection of the disease with other autoimmune diseases, it is necessary to make a screening according to the symptoms that the patients have. NL is a challenging condition to treat, with multiple therapeutic modalities to consider as in our case but without great improvement. Treatment is aimed at addressing signs and symptoms through mitigating the underlying inflammatory processes and reducing the risk of painfull ulceration.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest

Statement of ethical approval

Ethical approval was obtained.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study."

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