

Perforating necrobiosis lipoidica

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Perforating necrobiosis lipoidica (PNL) is a very rare variant of necrobiosis lipoidica (NL), almost always associated with diabetes. A non-diabetic 43-year-old man referred to us with a gradually enlarging plaque on his forehead. The patient was initially diagnosed with cutaneous tuberculosis based on the histopathologic findings, but failed to respond to anti-tuberculosis therapy. Upon further evaluation and observation of the transepidermal elimination of degenerated collagen in the pathology slides, we rendered a diagnosis of PNL. The patient underwent successful treatment with topical tacrolimus and systemic hydroxychloroquine.

Keywords: necrobiosis lipoidica, perforating disorders, transepidermal elimination

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INTRODUCTION

Necrobiosis lipoidica (NL) is a rare idiopathic dermatological condition frequently associated with diabetes. Perforating necrobiosis lipoidica (PNL) is a very rare variant of NL characterized by transepidermal collagen elimination. We report a rare case of PNL located on the forehead of a non-diabetic man.

CASE REPORT

A 43-year-old man presented with an asymptomatic, slowly enlarging plaque on his forehead that had been present for approximately one year. At 6 months prior to presentation, another plaque began to develop on his trunk. There was no history of trauma or injections to the involved areas. The patient had a history of migraine headaches treated with ergotamine.

On physical examination, we observed a well-defined annular plaque with an atrophic center and raised borders located on the frontoparietal scalp. Telangiectasias in the center and comedo-like papules on the periphery were also appreciated (Figure 1). There was another plaque on the patient's back, which was smaller but similar in appearance (Figure 2).

Laboratory investigations showed a hemoglobin of 13.2 g/dL and tuberculin test with induration of 16 mm. Routine biochemistry tests, antinuclear antibodies, serum protein electrophoresis, and complement levels were all within normal limits. Fasting blood sugar and a glucose tolerance test showed no evidences of diabetes.

The patient had 3 skin biopsies prior to presentation. The first biopsy was performed with clinical suspicion for basal cell carcinoma and lymphocytoma cutis. However, the histopathology was non-specific and suboptimal, and revealed



Figure 1. Well-defined annular plaque with atrophic center, raised borders, and comedo-like structures.



Figure 2. An atrophic plaque with comedo-like lesions on the back.

only a relatively dense lymphocytic infiltration plus a few neutrophils, mast cells, and plasma cells. In the second and the third biopsies, the main histopathological findings were palisading granuloma with focal hyaline necrobiosis; numerous giant cells and foamy histiocytes; cholesterol clefts; and follicular plugging with spongiosis surrounded

by a dense lymphocytic infiltration. Ziehl-Neelsen staining failed to show any acid-fast bacilli and lupus vulgaris was considered the most probable diagnosis. The patient was previously treated with multiple drug anti-tuberculosis therapy by a dermatologist because of the positive tuberculin test. He failed to respond to anti-tuberculosis therapy and was referred to us. We performed another biopsy at the time of presentation to our clinic. This biopsy demonstrated dermal histiocytic infiltration with a few multinucleated Langerhans cells and foreign body giant cells in association with necrobiosis, cholesterol clefts, and mild plasma cell infiltration (Figure 3). The overlying epidermis showed comedo-like invaginations with transepidermal elimination of degenerated collagen and inflammatory cells (Figure 4).

The patient was treated with hydroxychloroquine, 200 mg twice daily, and topical tacrolimus. He responded nicely to the treatment and has had no new lesions during a one-year follow up (Figure 5).

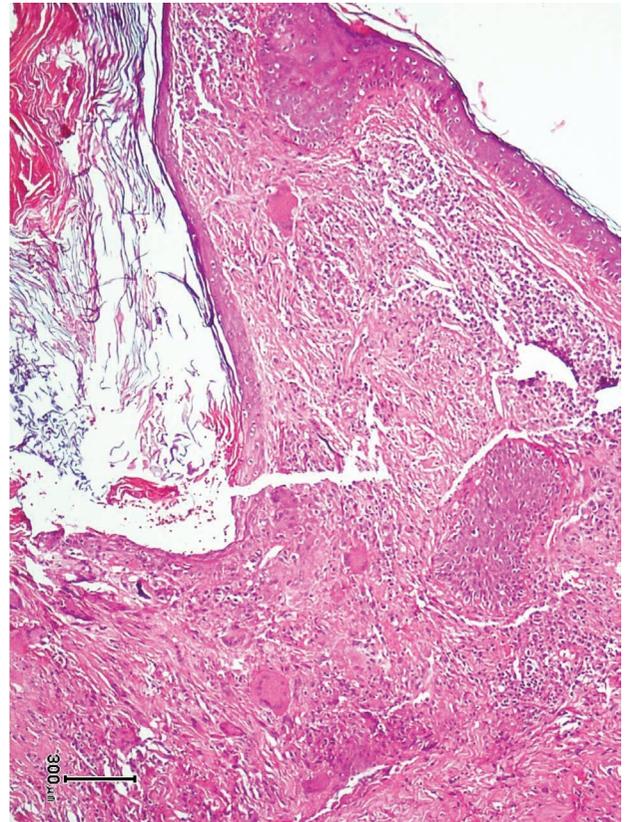


Figure 3. Histiocytic infiltration in the dermis with a few multinucleated Langerhans and foreign body giant cells associated with necrobiosis, cholesterol clefts, and mild plasma cell infiltration (H&E, 100 \times).

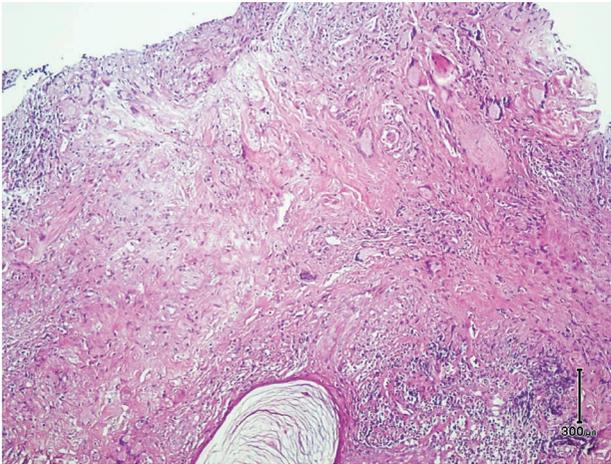


Figure 4. Trans-epidermal elimination of degenerated collagen (H&E, 400×).



Figure 5. Noticeable improvement after treatment with topical tacrolimus and systemic hydroxychloroquine.

DISCUSSION

NL is an idiopathic dermatological condition strongly associated with diabetes mellitus. However, it has been described in patients with sarcoidosis, inflammatory bowel disease, autoimmune thyroiditis, rheumatoid arthritis, monoclonal gammopathy, and otherwise healthy patients with

normal glucose metabolism^{1,2}.

NL often presents as slowly expanding violaceous patches located mostly on the lower legs. The advancing border is usually red with a yellow-brown central area. The central areas are atrophic and have a waxy surface with telangiectasias. In approximately 85% of cases, the legs are exclusively involved; however, NL can occur in other locations such as the hands, fingers, forearms, face, and scalp¹. The presence of multiple lesions is rare³.

NL is usually a clinical diagnosis, but if the clinical suspicion is uncertain, a skin biopsy assists in differentiation from sarcoidosis, necrobiotic xanthogranuloma, and granuloma annulare¹. Pathologically, the lesions show degeneration of collagen, granulomatous inflammation of subcutaneous tissues and blood vessels, capillary basement membrane thickening, and obliteration of vessel lumina.

Acquired perforating dermatosis is a rare group of skin disorders characterized by transepidermal elimination of dermal tissue material. These disorders are classified histopathologically according to the type of epidermal disruption and the nature of the eliminated material. They include Kyrle's disease, perforating folliculitis, reactive perforating collagenosis, and elastosis perforans serpiginosa⁴.

PNL is a rare variant of NL that is almost always associated with diabetes. Parra first noted transepidermal elimination in NL in 1977⁵. This phenomenon is very rare and clinically manifests as multiple hyperkeratotic plugs on the surface of the lesions. Less than 10 cases of PNL have been reported in the literature⁶⁻⁹. Scalp involvement has not been reported previously in PNL.

In summary, this was an exceedingly rare case of PNL in a non-diabetic patient who responded to topical tacrolimus and systemic hydroxychloroquine.

Conflict of Interest: None declared.

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