An atypical case of papular necrobiosis lipoidica masquerading as sarcoidosis

David A. Hashemi, BA, Caroline A. Nelson, MD, Rosalie Elenitsas, MD, and Misha Rosenbach, MD
Philadelphia, Pennsylvania

Key words: cutaneous sarcoidosis; granulomatous skin disease; necrobiosis lipoidica.

INTRODUCTION

Necrobiosis lipoidica (NL) is a rare granulomatous disorder characterized by well-circumscribed yellowish plaques affecting the anterior shins, often in patients with diabetes. Clinical examination is typically sufficient to make the diagnosis, but skin biopsy is sometimes required. Here we present a patient who received a misdiagnosis of sarcoidosis that was unsuccessfully treated who was found on re-examination and biopsy to have an atypical papular presentation of NL.

Abbreviation used:

NL: necrobiosis lipoidica

CASE REPORT

A 52-year-old white woman presented for evaluation and treatment of reddish-brown to orange-yellow firm papules on her anterior shins and lateral calves (Fig 1, *A* and *B*). She carried a presumed diagnosis of sarcoidosis and was treated previously with topical corticosteroids, intralesional corticosteroids, oral antibiotics, and oral methotrexate over 5 years without improvement.

She recently was diagnosed with asthma, but otherwise denied any history of extracutaneous organ involvement. Her previous evaluation included normal ophthalmologic examination, chest radiograph, and pulmonary function testing. She reported a history of prediabetes and had a family history of diabetes mellitus.

A skin biopsy found broad bands of necrobiotic collagen in the dermis, with admixed lymphocytes, histiocytes, plasma cells, and multinucleated giant cells (Fig 2, *A* and *B*). Re-examination of biopsies taken from 3 locations 5 years prior showed granulomatous inflammation throughout the reticular dermis and areas of altered hypocellular collagen with a layered appearance.

The patient's methotrexate was stopped, and pentoxifylline, hydroxychloroquine, and topical clobetasol (0.05% ointment) was initiated. At a 6-month follow-up appointment, a significant improvement in the appearance of the cutaneous lesions was noted, with some residual reddish-brown papules.

DISCUSSION

Necrobiosis lipoidica, first described by Oppenheim in 1929,[1] is a chronic granulomatous disorder. It is isolated to the lower legs in 85% of cases but in 15% of cases may involve other locations such as the abdomen, upper extremities, or scalp.[2] Approximately one-third of patients with NL have or eventually have diabetes mellitus.[3] It more commonly affects females, with a female/male ratio of 3 to 1.[1] Classic lesions are telangiectatic yellow-brown atrophic plaques. The lesions can resolve spontaneously or can persist and ulcerate in 15% of cases.[4] The pathogenesis of NL is not fully understood; hence, there is no consensus on optimal treatment. First-line treatment often consists of topical steroids, intralesional steroids, or topical calcineurin inhibitors, although other commonly used therapies include pentoxifylline, antimalarials, tumor necrosis factor inhibitors, phototherapy, and mycophenolate mofetil, among numerous other options.[5]

From the Department of Dermatology, The University of Pennsylvania School of Medicine.

Funding sources: None.

Conflicts of interest: None disclosed.

Correspondence to: Misha Rosenbach, MD, Department of Dermatology, University of Pennsylvania, Perelman Center for Advanced Medicine, 3400 Civic Center Boulevard, 7th Floor, South Tower, Suite 723, Philadelphia, PA 19104. E-mail: Misha.Rosenbach@uphs.upenn.edu.

JAAD Case Reports 2018;4:802-4.
2352-5126

https://doi.org/10.1016/j.jdcr.2018.07.011

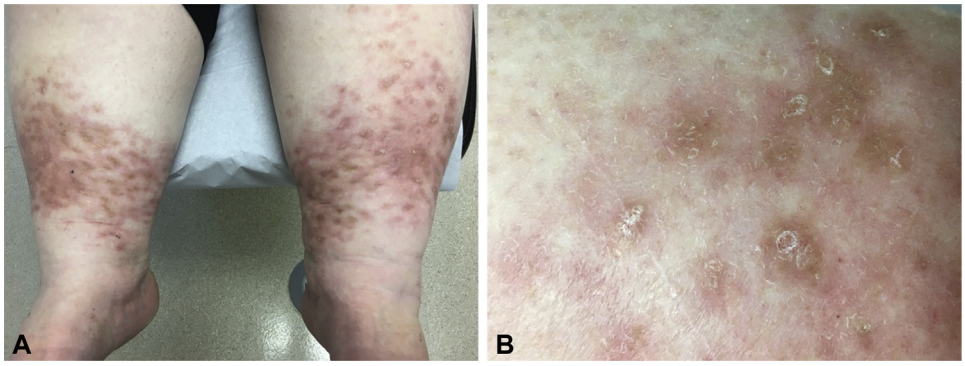

Fig 1. A case of NL in a 52-year-old woman. **A** and **B**, Clinical images of reddish-brown to orange-yellow firm papules on anterior shins and lateral calves at presentation.

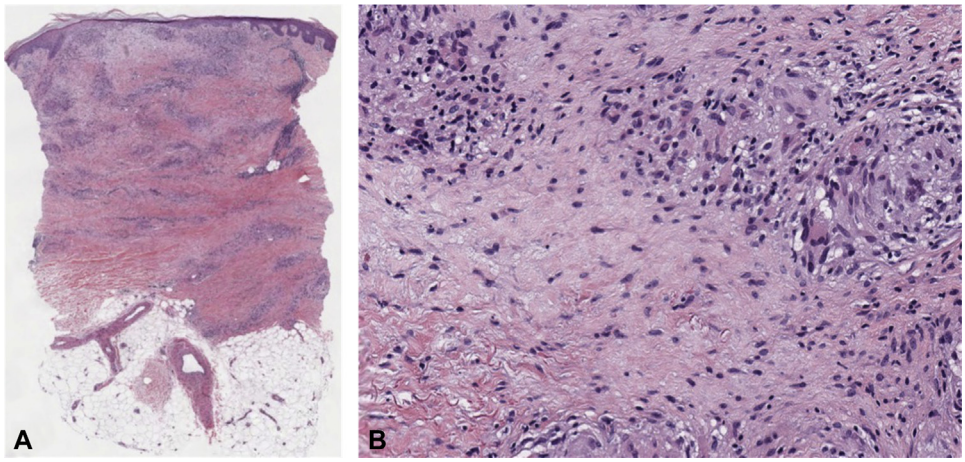

Fig 2. Histopathologic images from skin biopsy specimen before treatment. **A**, Broad bands of necrobiotic collagen with a layered appearance. **B**, Granulomatous inflammation with admixed lymphocytes, histiocytes, plasma cells, and multinucleated giant cells can be seen at higher power. (Hematoxylin-eosin stain; original magnifications: **A**, ×40; **B**, ×200.)

A diagnosis of NL can be confirmed via histopathology, particularly if the lesions do not have the classic appearance. Biopsies of NL find layers of horizontal granulomatous inflammation in the dermis, in an arrangement that is layer cake—like in appearance.[5] The granulomas are primarily composed of histiocytes and multinucleated giant cells, whereas the intervening layers of inflammatory infiltrate are predominantly lymphocytic with some plasma cells and eosinophils. The presence of plasma cells and an absence of mucin help differentiate NL from granuloma annulare. The alternating areas of necrobiotic collagen may help differentiate NL from sarcoidosis.[6]

This case represents an atypical variant of NL in which it both presented and persisted with a purely papular morphology, giving it a sarcoidlike clinical appearance. Previously there were reported cases of sarcoid skin lesions with histology resembling NL,[7] but these cases exhibited symptoms of systemic sarcoidosis and presented with ulceration. There also are reports of papules developing within—or in conjunction with—the classic atrophic plaques of NL.[1] This case illustrates that dermatologists should consider this atypical variant of NL when faced with papular lesions localized to the anterior shins without systemic manifestations. If suspected, histopathologic confirmation should be sought, as early diagnosis can impact both prognosis and optimal treatment.

REFERENCES

1. Khan F, Ramirez-Fort MK, Chan CS, Rosen T. Hyperkeratotic necrobiosis lipoidica. *Dermatol Pract Concept*. 2013;3(1):13-15.
2. Muller SA, Winkelmann RK. Necrobiosis lipoidica diabeticorum. A clinical and pathological investigation of 171 cases. *Arch Dermatol*. 1966;93(3):272-281.
3. Reid SD, Ladizinski B, Lee K, Baibergenova A, Alavi A. Update on necrobiosis lipoidica: a review of etiology, diagnosis, and treatment options. *J Am Acad Dermatol*. 2013;69(5):783-791.

4. Markey AC, Tidman MJ, Rowe PH, Missen GA, Macdonald DM. Aggressive ulcerative necrobiosis lipoidica associated with venous insufficiency, giant-cell phlebitis and arteritis. *Clin Exp Dermatol*. 1988;13:183-186.
5. Sibbald C, Reid S, Alavi A. Necrobiosis lipoidica. *Dermatol Clin*. 2015;33:343-360.
6. Yen PS, Wang KH, Chen WY, Yang YW, Ho WT. The many faces of necrobiosis lipoidica: a report of three cases with histologic variations. *Dermatologica Sinica*. 2011;29(2):67-71.
7. Nappi F, Laurain D, Jalalat S, Saco M, Fenske N. Cutaneous sarcoid with necrobiosis lipoidica—like histology. *J Am Acad Dermatol*. 2016;74(5 Suppl1):AB97.