

BRIEF ARTICLES

Unusual presentation and successful treatment of necrobiosis lipoidica in a 15-year-old girlAllison L Limmer, BS, BA¹, Levi C Holland, BS¹, Annabelle L Garcia, MD²¹McGovern Medical School at UTHealth, Houston, TX²Independent Researcher, San Antonio, TX**ABSTRACT**

Necrobiosis lipoidica (NL) is a rare, granulomatous skin disease with a predilection for diabetic patients. NL can present both histopathologically and clinically in a similar fashion to other granulomatous dermatoses such as granuloma annulare, necrobiotic xanthogranuloma, and cutaneous sarcoidosis. When NL is suspected in patients without glucose intolerance, affirmative diagnosis can be difficult, and other comorbidities may be probed, as NL has been associated with conditions such as hypertension, dyslipidemia, and thyroid disorders. Familial disease has also been reported. In the following case report, we discuss a 15-year-old girl who presented with a new, single, pink atrophic plaque of the right pretibial region. Biopsy showed palisaded granulomatous inflammation within the dermis, absent dermal mucin, and rare multinucleated giant cells. This histopathologic description combined with the clinical context led to the diagnosis of necrobiosis lipoidica. The patient developed another similar lesion on the left anterior shin over the next 1.5 years which was also diagnosed as necrobiosis lipoidica. The lesions were treated with intralesional triamcinolone acetonide and topical tacrolimus. Both lesions are now resolved with only mild atrophy of the affected areas. To date, the patient's labs have shown no evidence of glucose intolerance, hyperlipidemia, or thyroid disorder.

INTRODUCTION

Necrobiosis lipoidica (NL) is a rare, disfiguring, granulomatous skin condition commonly affecting the pretibial region. Classically, histopathologic analysis reveals granulomatous inflammation, sclerosis, and altered connective tissue spanning the dermis with or without palisading histiocytes and giant cells. In fact, the term “necrobiosis” refers to the connective tissue changes, as it appears more pale, gray, compact, and disorganized. Occasionally perivascular lymphocytic infiltrates or deep dermal plasma

cells are noted.¹ The differential diagnosis of NL includes granuloma annulare, necrobiotic xanthogranuloma, and cutaneous sarcoidosis. Granuloma annulare is distinguished by interstitial histiocytes with or without perivascular lymphocytic infiltrate and/or focal organization about dermal mucin. Biopsy of necrobiotic xanthogranuloma may reveal dermal necrosis surrounded by palisading histiocytes with giant cells. Finally, cutaneous sarcoidosis should be suspected if circumscribed islands of epithelioid cells with rare giant cells and minimal to absent fibrin

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Figure 1: Necrobiosis lipoidica. Patient's right anterior tibia at presentation (2014).

are noted, especially in the context of asteroid or Schaumann bodies.¹

NL affects women twice as often as men and frequently presents in the sixth decade of life.^{2,3} Up to 65% of NL is associated with diabetes mellitus, and the lesions can either indicate or precede the existence of disease, with 0.3% of diabetics affected in their lifetime.⁴ However, diabetes is not necessary for the existence of NL, as the disease has also been associated with other conditions such as hypertension and dyslipidemia.² Higher rates of thyroid disorders have also been noted in patients with NL than in the general population.⁵ Lastly, NL can present in siblings with no evidence of diabetes, termed familial nondiabetic necrobiosis lipoidica.⁶

CASE REPORT

Here, we detail the unusual case of a 15-year-old girl who presented in 2014 with a new rash of the right anterior shin. The pretibial lesion was a single, unilateral, pink atrophic plaque with orange-brown red rolled borders (Figure 1). Biopsy of the lesion revealed palisaded granulomatous inflammation within the dermis, absent

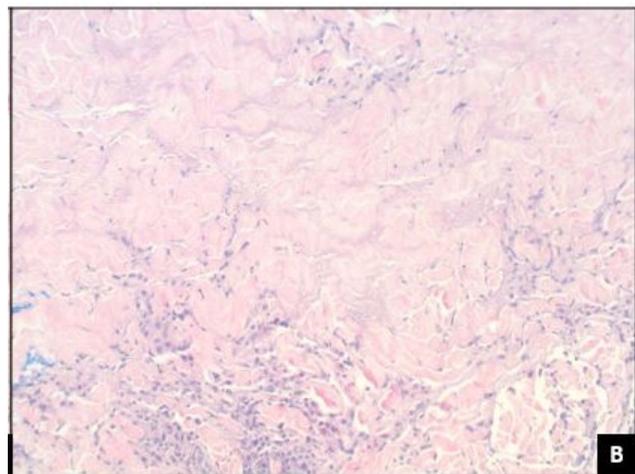
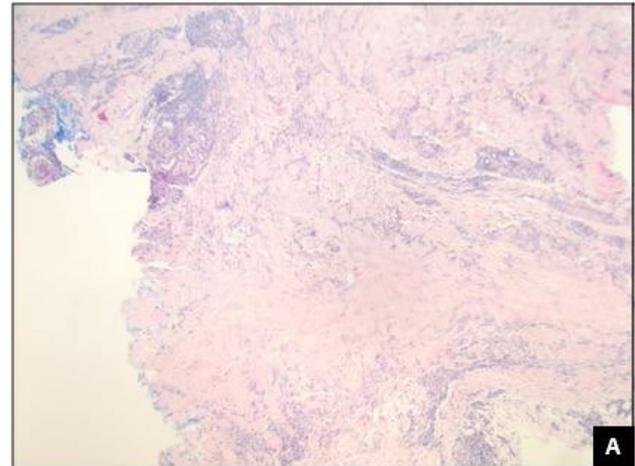


Figure 2: Necrobiosis lipoidica. Histologic section of skin from right pretibial region at 40x (A) and 100x (B) stained by H&E. Sections demonstrate a tiered focus of lymphohistiocytic inflammation within the mid to deep dermis. Rare multinucleated giant cells are seen surrounding this area, which did not demonstrate mucin deposition on colloidal iron stain (not shown).

interstitial mucin evaluated by colloidal iron stain, and rare multinucleated giant cells in the deeper aspects (Figure 2). This histopathologic description combined with the clinical context, especially the lesion's pretibial location, was consistent with necrobiosis lipoidica. Other diagnoses such as granuloma annulare and cutaneous sarcoidosis were considered less suspect secondary to absent dermal mucin, present giant cells, lack of classic island formation, and no asteroid bodies.

Subsequent to a diagnosis of necrobiosis lipoidica in the absence of preexisting diabetes, the patient was evaluated for other comorbidities, especially those associated with NL. The patient had no evidence of hypertension, obesity, or hyperlipidemia. Pertinent lab values were: hemoglobin A1c 4.8% (nl 4.0-5.6), fasting glucose 71 mg/dL (nl 70-99), insulin 11.9 aIU/mL (nl 29.2), TSH 1.263 uIU/mL (nl 0.340-4.410), free T4 0.60 ng/dL (nl 0.61-1.12), free T3 3.2 pg/mL (nl 2.3-4.2), sedimentation rate 18 mm/hr (nl <20), and globulin gap 2.7 g/dL (nl 1.8-4.0). Although the free thyroxine value was 0.01 ng/dL below the normal range for the test used, the patient had no clinical symptoms of hypothyroidism, had normal thyroid stimulating hormone and free triiodothyronine levels, and lacked thyroglobulin or thyroid peroxidase antibodies. Thus, the finding was considered clinically insignificant, and the patient is not on thyroid hormone replacement therapy. Additionally, the patient had no family history of NL, diabetes, or other autoimmune diseases like celiac disease, rheumatoid arthritis, or systemic lupus erythematosus.

Over the next 1.5 years, the patient developed an additional small NL lesion on the left anterior shin. Both lesions were treated with intralesional triamcinolone acetonide in clinic, and the patient was instructed to apply tacrolimus 0.1% topical ointment twice daily at the initiation of treatment, tapered to twice weekly by the end. The right shin lesion was treated at three separate visits with intralesional triamcinolone acetonide. Doses varied to maximize therapeutic value while minimizing atrophy as lesions were responding to treatment. At ten days after presentation, the lesion was treated with 4.8 mg triamcinolone acetonide, at one month with 9.0 mg, and at four months with 1.5 mg. The left shin was treated once with 1.5 mg triamcinolone

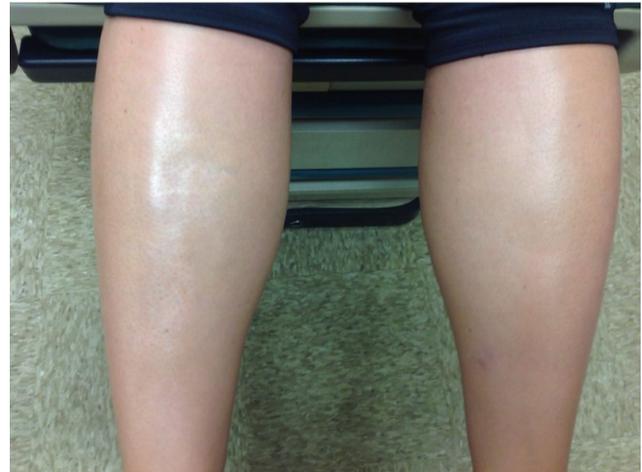


Figure 3: Necrobiosis lipoidica. Patient's legs 2.5 years after diagnosis (2016).

acetonide. Both lesions are now resolved with only mild atrophy of the affected areas (Figure 3). The patient is pleased with the cosmetic result.

DISCUSSION

This case demonstrates the appropriateness of keeping necrobiosis lipoidica in the differential diagnosis for patients presenting with plaques or ulcerating lesions, especially of the pretibial area, even with no associated comorbidities and/or uncommon histopathology. Treatment regimens for NL are largely based on case reports and uncontrolled studies, and published epidemiologic statistics vary widely, suggesting avenues for future research. While some patients forego medical therapy, the patient described here elected to pursue treatment with intralesional corticosteroids and topical tacrolimus. We believe prompt biopsy and diagnosis, close follow-up, and targeted intralesional therapy allowed the patient to experience minimal atrophy and complete resolution of her lesions.

Conflict of Interest Disclosures: None.

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