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Authors
Tong, Lana X
Penn, Lauren
Meehan, Shane A
et al.

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Necrobiosis lipoidica

Lana X Tong MD MPH, Lauren Penn MD, Shane A Meehan MD, Randie H Kim MD PhD

Affiliations: The Ronald O. Perelman Department of Dermatology, New York University School of Medicine, New York, New York, USA

Corresponding Author: Randie H Kim MD PhD, 240 East 38th Street, 11th Floor, New York, NY 10016, Tel: 212-263-5250, Fax: 212-263-1683, Email: Randie.Kim@nyulangone.org

Abstract

We present a case of necrobiosis lipoidica (NL) of the right abdomen in a 75-year-old man. A skin biopsy performed showed a layered infiltrate of mono and multinucleated histiocytes palisaded around degenerated collagen bundles. Laboratory workup was unremarkable. The patient was treated with topical corticosteroids with cessation of progression of his disease, although the eruption did not resolve. There are a number of treatments for NL reported in the literature, all with varying efficacy. Although NL lesions are usually asymptomatic, patients with NL must be monitored closely for signs of ulceration or malignant transformation, in which case more aggressive treatment options may be warranted.

Keywords: necrobiosis lipoidica, granulomatous disease, diabetes

Introduction

A 75-year-old man previously followed in the NYU Skin and Cancer unit for seborrheic and eczematous dermatitis presented with a 10-day history of a rash on the right abdomen. The patient stated the rash was asymptomatic and initially began as red bumps which then darkened in color. He denied pain, pruritus, or burning. Owing to the dermatomal distribution of the lesions, he was presumed to have herpes zoster. Antiviral medication was not prescribed as the lesions had been present for over 72 hours. Of note, he had not previously received the shingles vaccine.

One month later, the patient reported the continued appearance of new lesions on the right flank, although the eruption remained asymptomatic. He denied any blistering. He was started on valacyclovir 1g three times daily for seven days and a biopsy was performed. After the biopsy, he was started on triamcinolone 0.1% ointment twice daily. Although the eruption did not resolve, no further lesions appeared and his eruption remained stable and asymptomatic. His review of systems was positive for pruritus of the scalp and back. Sensation was decreased below the waist.

His past medical history was notable for multiple sclerosis, atrial fibrillation, coronary artery disease, pressure ulcers, methicillin-resistant Staphylococcus aureus and Pseudomonas aeruginosa colonization, Barrett esophagus, migraines, congestive heart failure, and peptic ulcers. The patient was bedbound owing to his multiple sclerosis. His medications included amiodarone, metoprolol, nitrofurantoin, pantoprazole, fexofenadine, gabapentin, nystatin, rivaroxaban, baclofen, topiramate, fluocinolone, and triamcinolone. He had not recently started any new systemic medications.

On examination, there were several erythematous and hyperpigmented papules scattered in a dermatomal distribution on the right flank, some with central pallor or atrophy (Figure 1). No vesicles or bulla were observed.

The complete blood count was notable for a hemoglobin of 10.9g/dL (reference range 13.7-16.5g/dL). The comprehensive metabolic panel was within normal limits. A lipid panel was unremarkable. Hemoglobin A1c was 5.3% (reference range 5.7-6.4%). Thyroid function tests were within normal limits.
Necrobiosis lipoidica (NL) is a rare chronic granulomatous condition that classically presents on the lower extremities [1]. The etiology is unclear and may be a result of immunoglobulin deposition, trauma, microangiopathy, or abnormal collagen synthesis [2]. NL typically presents in females in the third or fourth decade of life as erythematous papules that coalesce into yellow-brown plaques, often with an erythematous rim, and ultimately develop atrophy, hairpin vessels, and telangiectasias. Disease progression can be difficult to predict and although there are many potential therapeutic options, treatment is generally unsatisfactory [3]. Systemic associations have been debated in the literature, with the most common condition thought to be diabetes mellitus (DM). NL was previously termed necrobiosis lipoidica diabeticorum. However, as the incidence of NL is estimated to only be 0.3-1.2% in individuals with DM, it is now referred to as simply NL [4, 5]. Glucose control does not appear to be correlated with disease activity [6]. Other reported associated conditions include thyroid disorders, inflammatory bowel disease, sarcoidosis, and rheumatoid arthritis.

The differential diagnosis of NL includes other granulomatous diseases such as necrobiosis xanthogranuloma, granuloma annulare, sarcoidosis, venous stasis ulcers, or erythema nodosum. In our patient, the distribution of his eruption was initially suggestive of herpes zoster or segmental granuloma annulare. There has only been one previously reported case of NL located on the abdomen [7]. Definitive diagnosis of NL is based on histopathology. Palisading granulomatous inflammation is observed in the dermis with multinucleated giant cells located between collagen and without mucin [8], as was seen on biopsy in our patient.

Most lesions are asymptomatic owing to nerve damage, but up to 35% may experience tenderness and ulceration [5]. Despite being asymptomatic, close monitoring of these lesions is recommended owing to risk of ulceration as well as potential for malignant transformation. Ulcerations may be particularly difficult to manage when they are on the...
lower extremities and they require appropriate wound care and monitoring for signs of infection. It is unknown if the increased risk of squamous cell carcinoma is simply related to chronic inflammation or if NL independently leads to malignant transformation [9]. Numerous options for management have been widely reported in the literature with varying efficacy and no randomized control trials have been published regarding treatment outcomes. First line treatments include topical, intralesional, and systemic corticosteroids [6], although these should be used with caution in diabetic patients. Other reported treatment options include topical retinoids [10], topical calcineurin inhibitors [11], tumor necrosis factor inhibitors [12], cyclosporine, pioglitazone [13], hyperbaric oxygen [14], clofazimine [15], pentoxifylline [6] and psoralen plus ultraviolet A phototherapy [16], although recurrence rates are high upon cessation of therapy. As our patient did not find his eruption bothersome and did not develop ulcerations, he was treated with topical corticosteroids alone, which prevented further progression but did not resolve his lesions. After biopsy, his hemoglobin A1c level was drawn and was within normal range. Although the patient did not have any systemic conditions classically associated with NL, he did have multiple sclerosis, which is considered to be an autoimmune disease. Close clinical observation will be continued to monitor for progression of disease, ulcerations, and possibility of malignant transformation.

References