JAMA Dermatology Clinicopathological Challenge

Persistent Malar Erythema With Atrophy in a Young Woman

Sarah Mattessich, BS; Katalin Ferenczi, MD; Mona Shahriari, MD

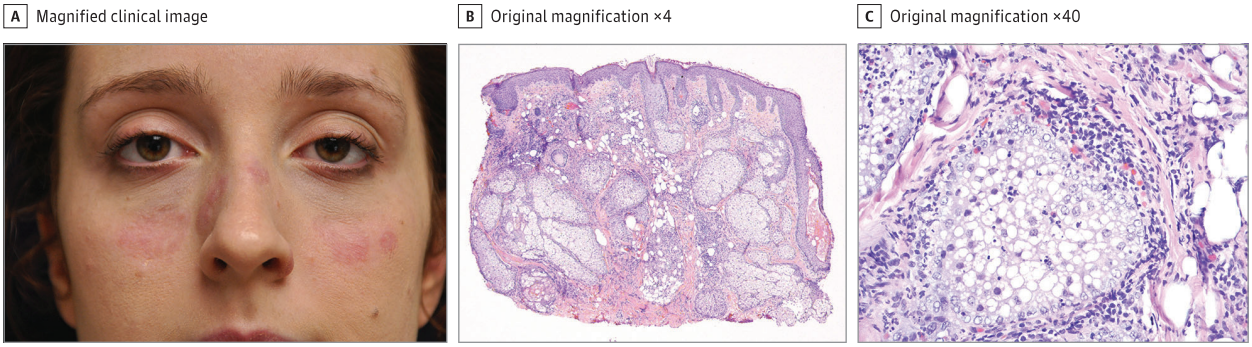

Figure. A, Erythematous, annular plaques on bilateral cheeks and nasal bridge. B and C, Histopathologic images, hematoxylin-eosin.

A 21-year-old woman with no significant medical history presented with a 2-year history of asymptomatic, facial redness that flared in sunlight. She had been previously treated with doxycycline, 100 mg/d, and topical sulfacetamide with no effect. On examination, on the cheeks and nasal bridge there were multiple erythematous, annular plaques with focal areas of atrophy. Her medications included only an oral birth control pill. Serum chemical analyses and complete blood cell count showed no abnormalities, and anti-Ro, anti-La, and qualitative ANA antibodies were negative. A clinical diagnosis was made, and over the next 9 months, treatments with minocycline, desonide, topical metronidazole, 0.75%, cream and azelaic acid, 15%, were unsuccessful, and the plaques developed more scarring and atrophy (**Figure**, A). In addition, the patient began to develop a burning sensation in these areas. A punch biopsy specimen was obtained and submitted for histopathologic review (Figure, B and C).

WHAT IS YOUR DIAGNOSIS?

- **A.** Chronic cutaneous lupus erythematous
- **B.** Granuloma faciale
- **C.** Neutrophilic sebaceous adenitis
- **D.** Rosacea-like dermatitis

Diagnosis

C. Neutrophilic sebaceous adenitis

Microscopic Findings and Clinical Course

Histopathologic examination revealed the presence of neutrophils and eosinophils in the sebaceous glands. There was a superficial and deep perivascular lymphocytic infiltrate in the dermis, and lymphocytes were also noted around the sebaceous lobules (Figure, B). The periodic acid–Schiff (PAS) stain was negative for microorganisms.

Discussion

Neutrophilic sebaceous adenitis (NSA) is a rare dermatosis with unclear etiology and clinical behavior. The term *neutrophilic sebaceous adenitis* was first coined in 1997 by Renfro et al,[1] who discovered unique sebaceous gland histopathologic characeristics of an erythematous eruption on the face of a male patient. Since then, similar presentations of NSA have been described in the literature only 8 times, including 6 reports in men and 2 in women.[2-8]

The erythematous, indurated plaques classically have annular, elevated borders without secondary changes or central clearing. The lesions are typically asymptomatic. Only 1 report[5] describes mild pruritus. The eruptions have a predilection for the face, with 4 cases reporting spread to the trunk, back, or upper extremities. One report[7] describes spread of the lesions from back to face. Hypothesized triggers have included antibiotic use,[1] contact with fiberclass,[2] sunlight,[4,6,7] and febrile illness.[3,6,7] Of the 3 reports that describe exacerbation owing to sun exposure, 2 describe lesions that continuously reoccurred in summer months prior to treatment.[4,6] An association with demodex infection was suspected in 2 cases; however, the burden of these tiny mites histologically was not great enough to diagnose demodicosis.[5,8] More recently, reports of genital NSA have been discovered in women, which are clinically differentiated from cases of facial NSA, being hormone responsive and symptomatically painful.[9,10]

Although its clinical picture is elusive, the histological hallmark of NSA is exclusive inflammation of the sebaceous glands with sparing of the remainder of the hair follicle. Collections of neutrophils gather in sebaceous lobules, along with scattered necrotic sebocytes. The epidermis shows no changes, and no microorganisms are visualized on Gram and PAS stains.[1] This unique histological finding rules out the 3 potential diagnoses for malar erythema listed herein. In addition, features are missing that make an alternative diagnosis more likely. For ex-

ample, the characteristic histological finding of granuloma faciale includes the grenz zone of unaffected dermis above a nodular, polymorphous inflammatory infiltrate containing a significant amount of eosinophils. Rosacea-like dermatitis findings include perifollicular infiltration of neutrophils, plasma cells and lymphocytes, as well as histiocytes in the granulomatous form. The most prominent findings of the discoid variant of chronic cutaneous lupus erythematous include a superficial and deep perivascular and periadnexal lymphocytic infiltrate, interface changes, pilosebaceous atrophy, follicular plugging, and basement membrane thickening. Therefore, histologic results are pivotal for making the diagnosis of NSA.

Because of its rarity, no defined treatment guidelines exist for NSA. Topical steroids and oral prednisone have the most recorded success.[1,3,4,6,7] One case[5] reported resolution with oral isotretinoin, and another case[8] with metronidazole gel. Two cases[2,3] reported spontaneous clearing. Genital NSA in women has been successfully treated with combinations of minocycline, spironolactone, cyproterone acetate, and ethinylestradiol.[9,10] All cases reported eventual complete resolution of the eruptions. Unfortunately, this patient has been lost to follow-up, and her clinical status is currently unknown.

To our knowledge, each reported case of NSA has shown complete resolution without residual changes. Herein, we present the first report of a young woman with chronic, symptomatic facial NSA leading to permanent atrophy and scarring. Additional investigation is necessary to determine the mechanism for the scarring in this particular patient and to fine-tune the pathogenesis and treatment for this rare cutaneous process.

ARTICLE INFORMATION

Author Affiliations: University of Connecticut School of Medicine, Farmington (Mattessich); Department of Dermatology, University of Connecticut School of Medicine, Farmington (Ferenczi, Shahriari).

Corresponding Author: Mona Shahriari, MD, Department of Dermatology, University of Connecticut School of Medicine, 21 South Rd, Farmington, CT 06032 (mshahriari@uchc.edu).

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