

# Asymptomatic annular perianal sarcoidosis



Ijeuru Chikeka, MD,<sup>a</sup> Sameera Husain, MD,<sup>b</sup> and Marc E. Grossman, MD<sup>c,d</sup>  
*New York and New Hyde Park, New York and New Haven, Connecticut*

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## INTRODUCTION

Sarcoidosis is a noncaseating granulomatous disease of unknown cause that can affect multiple organ systems. Cutaneous involvement, with or without systemic manifestation, is seen in about one-third of patients.<sup>1</sup> Perianal sarcoidosis is rare with only 2 reports in the medical literature.<sup>2,3</sup> Often, the dermatologist is consulted to examine a patient with an unknown multisystem disease and to obtain a skin biopsy for histopathologic diagnosis. We report on a woman with a perianal annular yellowish plaque with biopsy-proven sarcoidosis to remind clinicians that a complete skin examination includes this often-overlooked area.

## CASE

A healthy 47-year-old white woman presented with a several-month history of an asymptomatic perianal lesion. She had an annular plaque with a red-brown, raised margin and a slightly firmer, smoother, central area with a yellowish hue compared with her normal skin. The plaque spread from the anus to the gluteal crease with symmetric extension to the opposing medical surfaces of the buttock (Fig 1). A skin biopsy found dermal aggregates of epithelioid histiocytes and multinucleate giant cells surrounded by a sparse lymphocytic infiltrate consistent with a “naked” or sarcoidal granuloma (Fig 2). Periodic acid–Schiff, Grocott-Gomori’s methenamine silver, and acid-fast bacilli histochemical stains performed on formalin-fixed paraffin-embedded tissue were negative for microorganisms. A chest x-ray and routine laboratory values were normal. Eight years later, the plaque increased in size and developed a red border. She



**Fig 1.** Clinical image. Physical examination of the perianal region shows an annular hyperpigmented plaque with a red-brown raised border surrounding an area of slightly firmer central clearing and a yellowish hue compared with normal skin.

also had a group of papules at the base of the neck, a biopsy of which found noncaseating granulomas consistent with sarcoidosis. She was asymptomatic with normal calcium and angiotensin-converting enzyme and unremarkable routine laboratory studies. A purified protein derivative skin test was negative. A computerized tomography scan of the chest found multiple enlarged lymph nodes at both hila and the superior mediastinum. Several lymph nodes were calcified, and there was bilateral apical scarring. The enlarging perianal plaque was treated with intralesional kenalog 4 times over the next 3 years with flattening and no further expansion. Five

From the Departments of Pathology<sup>a</sup> and Dermatology,<sup>b</sup> Columbia University Medical Center; the Department of Dermatology, Yale University School of Medicine<sup>c</sup>; and Department of Dermatology, Hofstra/Northwell Donald and Barbara Zucker School of Medicine, New Hyde Park.<sup>d</sup>

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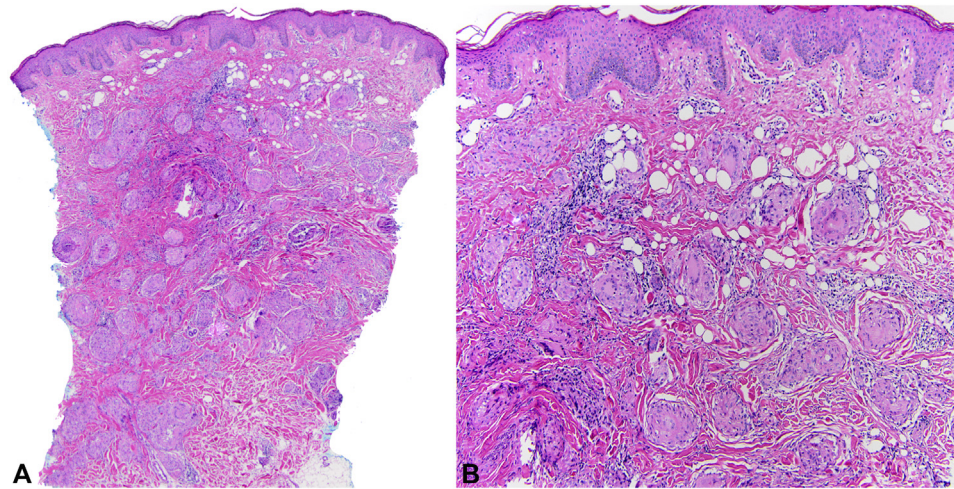
Correspondence to: Marc E. Grossman, MD, Visiting Professor and Lecturer, 12 Greenridge Ave, White Plains, NY 10605. E-mail: [marcmeg9@aol.com](mailto:marcmeg9@aol.com).

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**Fig 2.** **A**, Skin biopsy of the perianal lesion shows noncaseating, naked, epithelioid granulomata spanning the full thickness of the dermis. **B**, Biopsy at higher magnification. (**A** and **B**, Hematoxylin-eosin stain; original magnifications: **A**, ×40; **B**, ×100.)

years later, at age 63, the lesion was not evident, and she remained in good health. Another purified protein derivative skin test performed 2 years after her second presentation was also negative.

## DISCUSSION

Often called the *great imitator* because of its variable clinical morphology, cutaneous sarcoidosis most commonly presents on the face, neck, trunk, and extremities and less so at other sites. It rarely occurs in the perianal area. The 2 previously reported cases of perianal sarcoidosis were both biopsy-proven perianal ulcers in a 34-year-old white woman and a black male in his 50s. Both patients had widespread skin lesions and stable typical thoracic disease. The ulcers healed in the first case with prednisone and hydroxychloroquine and in the second case with adalimumab. Our patient was asymptomatic and responded to intralesional steroid injections initiated because the plaque continued to expand.

The diagnosis of sarcoidosis requires a clinical picture that is consistent, the presence of noncaseating naked granulomas on tissue biopsy, and exclusion of other clinical histopathologic mimics.<sup>4</sup> The differential diagnosis would include necrobiotic xanthogranuloma with paraproteinemia, necrobiosis lipoidica diabetorum, and disseminated granuloma annulare with their associated systemic conditions, multiple myeloma (necrobiotic xanthogranuloma with paraproteinemia) and diabetes mellitus (necrobiosis lipoidica diabetorum and disseminated granuloma annulare).<sup>5</sup> A diagnosis of systemic tuberculosis or tuberculosis and concurrent sarcoidosis was considered.

Although the combinations of pulmonary tuberculosis with pulmonary sarcoidosis and pulmonary tuberculosis with cutaneous sarcoidosis have been rarely reported, the simultaneous occurrence of cutaneous sarcoidosis and tuberculosis has not been found to occur.<sup>6-10</sup> In our patient, the morphologic appearance of the perianal annular centrally yellowish plaque was clinically diagnostic of sarcoidosis. Only a meticulously complete cutaneous examination including the anal region and surrounding skin found the skin lesions characteristic of sarcoidosis. Certainly, a skin biopsy would be the preferred, least-invasive procedure for the pathologic diagnosis of sarcoidosis in a patient with pulmonary, hepatic, or central nervous system disease.

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