Sarcoidosis: An atypical mimicker of acne keloidalis nuchae

Fritzlaine C. Roche, MS,[a,b] Andrew S. Fischer, MD,[b] and Susan C. Taylor, MD[b]
Rochester, New York and Philadelphia, Pennsylvania

Key words: acne keloidalis nuchae; sarcoidosis.

INTRODUCTION

Sarcoidosis is a multisystemic disease of unknown etiology characterized by the formation of non-caseating granulomas in several organs. The skin is the second most commonly involved organ with cutaneous involvement occurring in 25% to 30% of patients.[1] Two types of cutaneous lesions are recognized in sarcoidosis: specific cutaneous sarcoidal lesions and nonspecific reactive lesions.[1] Specific lesions manifest as noncaseating granulomas on skin biopsies; non-specific lesions are reactive to systemic sarcoidosis but do not have granulomas on biopsy.[1] Sarcoidosis of the scalp most commonly manifests as alopecia, which may be scarring or nonscarring, and is primarily seen in women of African descent.[1,2] Sarcoidal scalp morphologies include erythema and indurated plaques and nodules, which may have scale, and all of which demonstrate noncaseating granulomas on biopsy. We present an unusual case of sarcoidosis imitating acne keloidalis nuchae (AKN).

Abbreviation used:

AKN: acne keloidalis nuchae

CASE REPORT

A 47-year-old woman presented with a 2-month history of papules on the occipital scalp without an identifiable precipitating cause. Medical history was significant for a 17-year history of pulmonary sarcoidosis treated with oral corticosteroids and reportedly well controlled. However, review of systems found chronic dyspnea, loss of appetite, malaise, and a 20-pound weight loss in the previous 6 months. Examination was significant for skin-colored papules overlying the occipital scalp, with umbilicated centers noted within a few papules (Fig 1). Additional cutaneous lesions were not identified. Histopathology from a shave biopsy showed a relatively unremarkable epidermis overlying numerous dense, noncaseating granulomas with sparse surrounding lymphocytic inflammation in the dermis (Fig 2). Neither polarizable foreign material nor fungal elements were present. Furthermore, folliculitis, free and broken hair shafts, dense fibrosis, or neutrophilic inflammation were not seen. Histologic features were diagnostic of cutaneous sarcoidosis. The patient started hydroxychloroquine therapy. When the patient was re-evaluated by her pulmonologist, she had 2 peribronchovascular pulmonary nodules consistent with recurrent pulmonary sarcoidosis. Prednisone was added to her treatment regimen.

DISCUSSION

Cutaneous sarcoidosis involving the scalp is rarely reported.[2] Previously reported cases have occurred in African-American women who presented with superficial ulcerations, indurated plaques or nodules, and cicatricial or noncicatricial alopecia.[2] The most frequently reported scalp finding in sarcoidosis is localized cicatricial alopecia.[1-3] Sarcoidal scalp lesions have been reported to resemble discoid lupus erythematous, necrobiosis lipoidica, and lichen planopilaris, with pathology showing noncaseating granulomas.[3-5] This case of scalp

From the University of Rochester School of Medicine and Dentistry[a] and the Department of Dermatology, Perelman School of Medicine.[b]

Funding sources: None.
Conflicts of interest: None disclosed.

Correspondence to: Susan C. Taylor, MD, Associate Professor, Department of Dermatology, Perelman School of Medicine at the University of Pennsylvania, Room 768 South Tower, 3400 Civic Center Blvd, Philadelphia, PA 19104. E-mail: Susan.taylor@pennmedicine.upenn.edu.

JAAD Case Reports 2020;6:397-9.
2352-5126

https://doi.org/10.1016/j.jdcr.2020.02.018

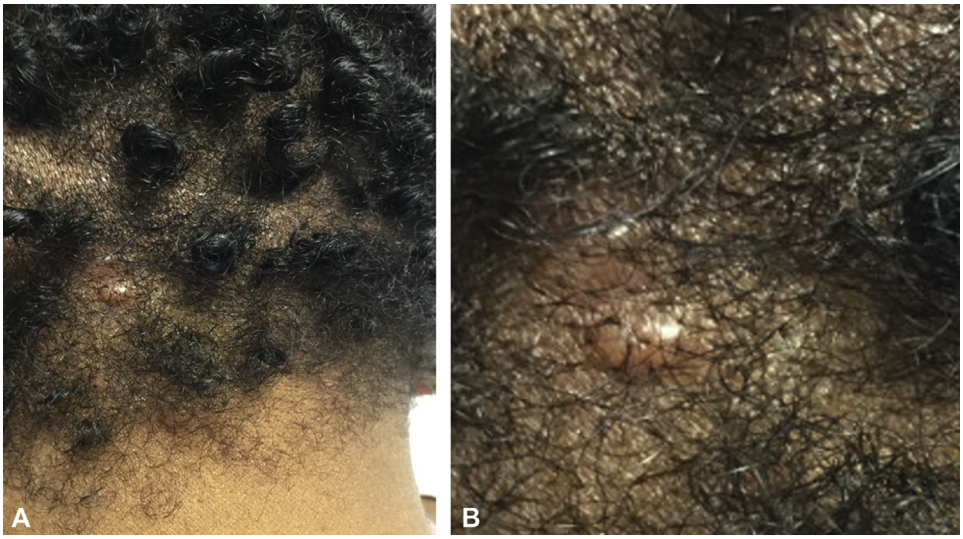

Fig 1. Patient examination finding. **A**, The occipital scalp has 2 firm, skin-colored papules without significant scale, erythema, scarring, dyspigmentation, or alopecia. **B**, Closer view of the clinical lesion.

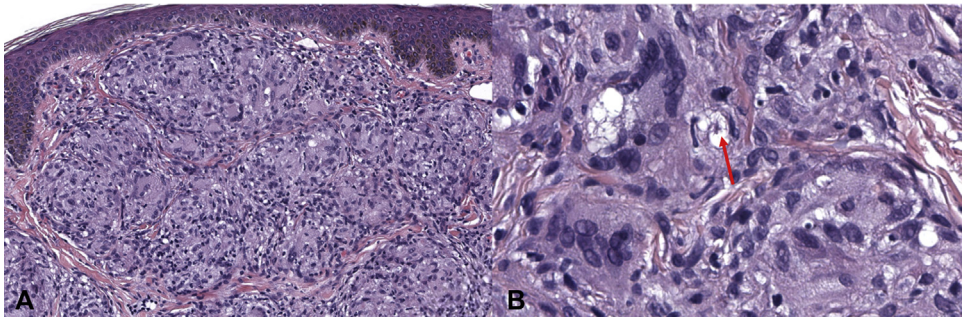

Fig 2. Histopathologic findings. **A**, Numerous dense sarcoidal-type granulomas with sparse surrounding lymphocytic inflammation in the papillary to mid dermis. No polarizable foreign material was present. **B**, High-power magnification of multinucleated giant cells and an asteroid body (*red arrow*) within a giant cell of a noncaseating granuloma. (**A** and **B**, Hematoxylin-eosin stain; original magnifications: **A**, ×200; **B**, ×400.)

sarcoidosis deviates from those previously reported, as it clinically mimicked AKN.[6]

AKN, a disorder of African-American men and women with coarse, curly hair, is characterized by dome-shaped papules on the occipital scalp or posterior neck that may coalesce to form keloid-like plaques or scarring alopecia with disease progression.[6,7] Histologically, AKN is characterized in the early stage by follicular and perifollicular infiltration of neutrophils and lymphocytes with a plasma cell predominance as the fibrosis progresses.[6] In later stages, free and broken hair shafts can be seen enmeshed in fibrosis and surrounded by a foreign body giant cell reaction, but the typical sarcoidal-type granulomas are not a feature.

The clinical finding of dome-shaped papules involving the occipital scalp in this patient were characteristic of AKN, but the histologic findings of sarcoidal-type granulomas with sparse surrounding lymphocytic inflammation revealed the correct diagnosis. Furthermore, the subsequent discovery of pulmonary nodules in this patient heightens the importance of systemic evaluation for sarcoidosis when a skin biopsy, including the scalp, is diagnostic for sarcoidosis.[7]

Sarcoidosis remains an enigmatic process, but trauma can be a precipitating factor in susceptible individuals, and sarcoidal lesions can develop within preexisting scars from various cases.[8] In these cases, the clinical appearance of the lesion may be slightly altered because of the presence of dermal fibrosis; therefore, a range of appearances has been reported including brownish-red papules, skin-colored subcutaneous nodules, and perilesional erythema.[8] We did consider the possibility that the clinical lesion in our patient arose within a scar given that the clinical

appearance was skin colored as opposed to the typical violaceous hue of cutaneous sarcoidosis; however, the patient did not report prior trauma at this site, and dense fibrosis was not noted on pathology findings. The possibility of minor trauma at this site, though, cannot be entirely excluded.

Cutaneous sarcoidosis can have a range of clinical morphologies and has been referred to as the *great imitator*.[9] We present this case as a reminder to clinicians to consider a biopsy in patients with systemic sarcoidosis who present with new scalp lesions.

REFERENCES

1. Wanat KA, Rosenbach M. Cutaneous Sarcoidosis. *Clin Chest Med*. 2015;36(4):685-702.
2. Katta R, Nelson B, Chen D, Roenigk H. Sarcoidosis of the scalp: a case series and review of the literature. *J Am Acad Dermatol*. 2000;42(4):690-692.
3. House NS, Welsh JP, English JC 3rd. Sarcoidosis-induced alopecia. *Dermatol Online J*. 2012;18(8):4.
4. Harman KE, Calonje E, Robson A, Black MM. Case 1. Sarcoidosis presenting as a scarring alopecia resembling necrosis lipoidica. *Clin Exp Dermatol*. 2003;28(5):565-566.
5. La Placa M, Vincenzi C, Misciali C, Tosti A. Scalp sarcoidosis with systemic involvement. *J Am Acad Dermatol*. 2008;59(5 Suppl): S126-S127.
6. Kelly AP, Bayat A. *Acne Keloidalis Nuchae*. 2 ed. McGraw Hill; 2016.
7. Mana J, Marcoval J, Graells J, Salazar A, Peyri J, Pujol R. Cutaneous involvement in sarcoidosis. Relationship to systemic disease. *Arch Dermatol*. 1997;133(7):882-888.
8. Atci T, Baykal C, Kaya Bingol Z, Polat Ekinci A, Kilicaslan Z. Scar sarcoidosis: 11 patients with variable clinical features and invariable pulmonary involvement. *Clin Exp Dermatol*. 2019; 44(7):826-828.
9. Tchernev G. Cutaneous sarcoidosis: the "great imitator": etiopathogenesis, morphology, differential diagnosis, and clinical management. *Am J Clin Dermatol*. 2006;7(6): 375-382.