Severe scalp sarcoidosis in an unlikely patient

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INTRODUCTION

Sarcoidosis is an idiopathic, multisystemic disease with variable presentations. Specifically, sarcoidosis most commonly involves the lungs, affecting at least 90% of patients.[1] Bone and skin involvement have been reported in this disease as well, with the scalp rarely being an affected cutaneous site.[2] The characteristic histopathologic findings in sarcoidosis are noncaseating epithelioid granulomas. Because alopecia secondary to sarcoidosis can mimic other diseases such as lupus or lichen planopilaris, dermatopathology examination of tissue is key in determining the underlying etiology. Here we present a case of a Caucasian woman with sarcoidosis whose presenting symptom was alopecia of the vertex scalp.

CASE REPORT

A 73-year-old woman presented to the emergency department with a closed intertrochanteric right femur fracture after a fall. Initial trauma workup found lytic bone lesions of the calvarium and mediastinal lymphadenopathy concerning for metastatic malignancy or multiple myeloma. The dermatology department was consulted for a 2-month history of a peculiar scaly scalp rash and alopecia previously diagnosed as psoriasis by the patient's primary care provider. Physical examination of the vertex and bilateral parietal scalp found 2 coalescing patches of cicatricial alopecia with peripheral scale and erythema, central pallor, and an apple jelly—like appearance (Fig 1). A 4-mm punch biopsy of the vertex scalp showed an attenuated epidermis overlying a dense dermal granulomatous infiltrate of lymphocytes, histiocytes, multinucleated histiocytes, and scattered neutrophils with few terminal hairs and significant dermal atrophy (Fig 2, *A* and *B*). Acid-fast bacillus, Gomori methenamine silver, and Fite stains of the skin biopsy specimen were negative, and a diagnosis of scalp sarcoidosis was made. Further workup found multiple liver nodules and a cervical mass. Liver biopsy found noncaseating granulomas and fibrosis, and cervical biopsy was negative for malignancy. A mammogram was negative. An ophthalmologic examination found no uveitis or any other ocular abnormalities. Bone marrow aspiration showed normocytic, normochromic anemia, lymphopenia, thrombocytosis, hypercellular marrow with fibrosis, and noncaseating granulomas.

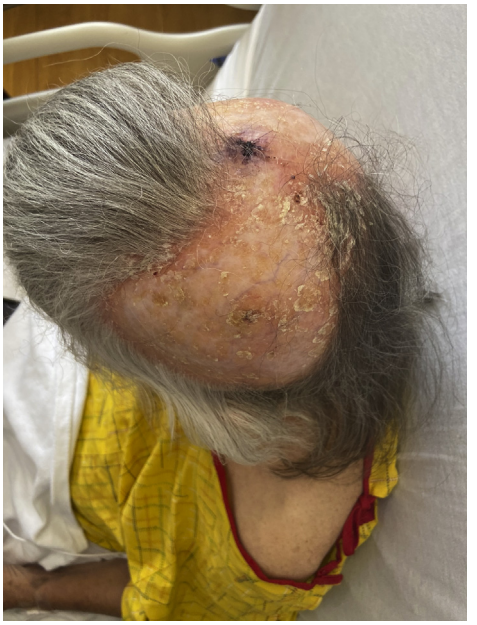

Fig 1. Vertex and bilateral parietal scalp shows 2 coalescing plaques of cicatricial alopecia with peripheral scale and erythema, central pallor, and an apple jelly—like appearance.

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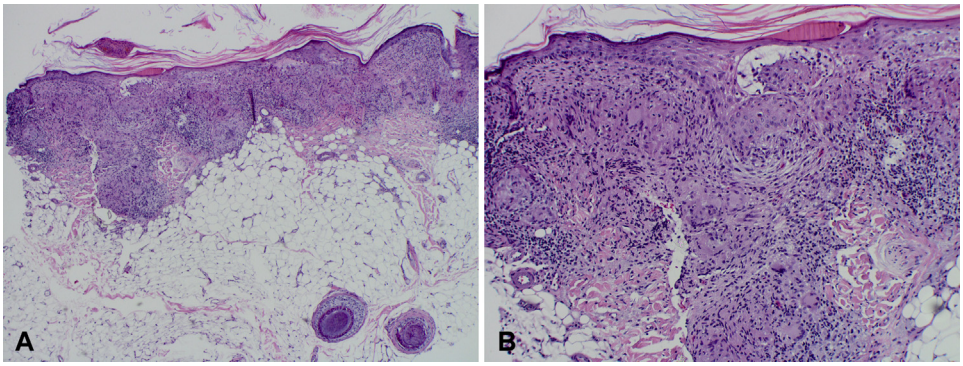

Fig 2. Attenuated epidermis overlying a dense dermal granulomatous infiltrate of lymphocytes, histiocytes, multinucleated histiocytes, and scattered neutrophils with few terminal hairs and significant dermal atrophy. **A** and **B**, Hematoxylin-eosin stain; original magnifications: **A**, ×40; **B**, ×100.

Acid-fast bacillus and Gomori methenamine silver stains of bone marrow aspirate were negative. Laboratory studies were significant for angiotensin-converting enzyme level of 57 U/L and serum calcium of 10.7 mg/dL; blood cultures, β-glucan, quantiferon gold tuberculosis, antinuclear antibody, and hepatitis panel were negative. A diagnosis of disseminated sarcoidosis was made, and the patient was started on hydroxychloroquine, 200 mg by mouth twice a day, and methotrexate, 15 mg by mouth weekly. Unfortunately, the patient was lost to follow-up.

DISCUSSION

Scalp sarcoidosis is a rare disease presentation and often an accompanying sign of additional cutaneous or systemic disease.[3] The case presented here represents an atypical presentation in an unlikely patient. A literature review found that 87% of patients were African American with only 2 cases reported in Caucasians.[3,4] Additionally, our patient's presentation was further complicated by sarcoidal bone lesions. Bone involvement in sarcoidosis has been described in a case-control study as affecting only 4% of patients, with a predilection for African-American females.[5] Most commonly, pulmonary findings account for 90% of systemic involvement.[6] Interestingly, our patient denied any history of pulmonary sarcoidosis, was not considered to be an at-risk demographic, and experienced overt alopecia compared with previous case reports.

Scalp sarcoidosis continues its reign as the "great imitator" with reported clinical morphologies including localized cicatricial alopecia, erythematous scaling, superficial ulcerations, nonscarring alopecia, and discoid lupus erythematosus–like presentations.[3,7] Because of its clinical mimicry, scalp sarcoidosis should remain on the alopecia differential until it can be histologically ruled out.

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