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CASE REPORT

Sarcoidal alopecia mimicking discoid lupus erythematosus: Report of a case and review of the literature

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ABSTRACT

Sarcoidal alopecia is a subtype of plaque-forming cutaneous sarcoidosis that may resemble discoid lupus erythematosus (DLE). Because the clinical appearance of the two lesions is similar, the correct diagnosis may be missed. The systemic involvement and progressive nature of sarcoidosis, make it important to differentiate sarcoidal alopecia from DLE, so that proper treatment can be initiated and potential long-term sequelae avoided. We present the case of a 57-year-old Taiwanese woman with sarcoidal alopecia of the scalp that mimicked DLE.

Introduction

Sarcoidosis is an idiopathic systemic granulomatous disease, in which noncaseating granulomatous inflammation can occur in any organ. The skin is affected in about 25% of cases, and the majority of cases occur in black women.[1] The scalp is rarely involved, and the inflammation may lead to cicatricial alopecia as a result of the destruction of hair follicles by the granulomatous formation.[1] Clinically, sarcoidosis may present as papules, nodules, or plaques, and in some cases may resemble discoid lupus erythematosus (DLE) or necrobiosis lipoidica.[2–6]

Case report

A 57-year-old Taiwanese female had been very healthy most of her life. However, over the past 4–5 years, she had developed painful ulcerative wounds with alopecia on her frontoparietal scalp (Figure 1). She had not been treated for the wounds or alopecia prior to visiting our hospital. On examination, we saw several bean- to coin-sized depressed ulcers surrounded by a violaceous to erythematous hue, and telangiectasia on the right frontoparietal scalp. No other skin lesions were noted.

DLE was suspected, and an incisional biopsy was performed. The pathology report showed noncaseating granulomatous inflammation involving the superficial and deep dermis (Figure 2). The granulomas were composed of epithelioid cells and occasional giant cells, rich in asteroid bodies. Results of direct immunofluorescence tests for immunoglobulin A (IgA), IgG, IgM, complement component 3 (C3), fibrinogen, and complement C1q were all negative. Periodic acid Schiff, Fite, and acid-fast stains did not reveal any pathogens, and the Venereal Disease Research Laboratory (VDRL) test was negative. Therefore, the diagnosis of sarcoidal alopecia was confirmed.

The immunologic profiles, including antinuclear antibody, anti-extracted nuclear antigen (ENA) antibody, C3, and C4 were all within normal ranges. However, plain chest films and computed tomography (CT) scans revealed pulmonary sarcoidosis with lymphadenopathy (Figure 3). Lung function tests showed normal spirometric values and diffusing capacity. Ophthalmologic examination ruled out any ocular involvement. The results of other blood tests, such as complete blood cell counts, and measurement of serum aspartate aminotransferase, alanine aminotransferase, creatinine, blood urea nitrogen, sodium, potassium and calcium levels, were all within normal limits.

Because the patient refused treatment with systemic corticosteroids, she was treated with oral hydroxychloroquine, 400 mg daily, and intralesional injections of triamcinolone, 10 mg once a month. She received a total of nine intralesional triamcinolone injections from October 2009 to December 2011; these injections were given on a monthly basis, except during the 18 months from April 2010 to September 2011.

Because the patient had significant overall improvement and regrowth of hair, we discontinued local corticosteroid treatment in January 2012. The dosage of oral hydroxychloroquine was also tapered to 200 mg daily (beginning February 2012) and the disease

Conflicts of interest: The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in this article.

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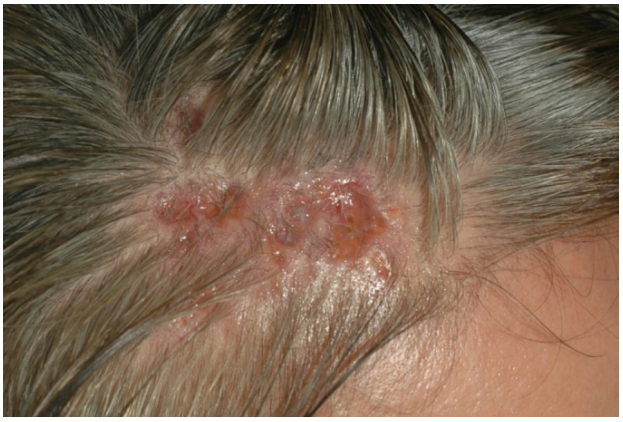

Figure 1 Cicatricial alopecia with several bean- to coin-sized depressed ulcers. A surrounding violaceous to erythematous hue can be seen on the patient's right frontoparietal scalp.

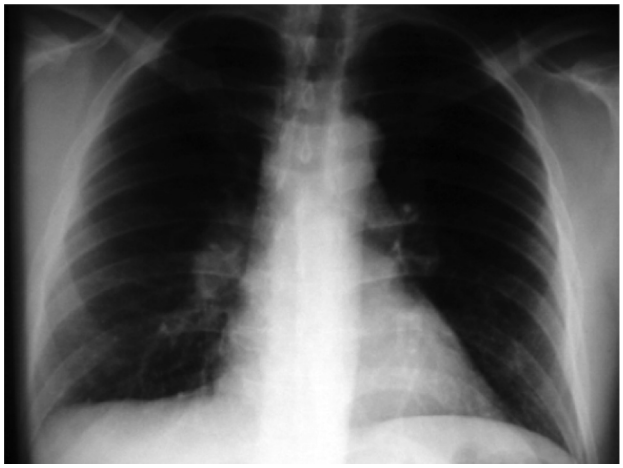

Figure 3 Plain chest film shows bilateral hilar enlargement.

showed no deterioration after 3 months (Figure 4). Serial CT examinations of the chest from 2009 to 2012 showed that the disease had stabilized, and lung function tests remained normal.

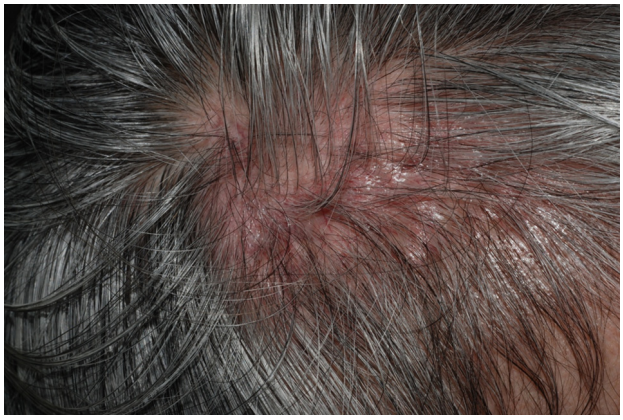

Figure 4 After treatment, the alopecia was nearly resolved.

Discussion

Sarcoidal alopecia is a rare manifestation of cutaneous sarcoidosis that predominantly affects black women.[1] It is a form of secondary cicatricial alopecia and can have variable morphologies. Most commonly, cutaneous sarcoidosis on the frontoparietal facial region may extend into the scalp, which may lead to hair loss. Such lesions are the type of sarcoidal alopecia that most mimics DLE clinically.[1–4]

On the scalp, sarcoidosis can manifest as atrophic, erythematous, scaly, or ulcerative areas of alopecia.[1] The typical appearance of DLE is of well-circumscribed, erythematous, scaly, atrophic plaques, that may occasionally also be ulcerative.[2] In both situations, follicular plugging can be observed on dermoscopy.[5] The differential diagnosis of sarcoidal alopecia and DLE can be made by histopathologic examination. Sarcoidal alopecia shows classic naked granulomas in the dermis.[1–4] In contrast, DLE is characterized by follicular plugging, epidermal atrophy, vacuolar degeneration of basal keratinocytes, and basement membrane thickening, as well as superficial and deep perivascular and periadnexal lymphocytic infiltrates.[2]

Patients with sarcoidal alopecia almost always have other cutaneous lesions, and the vast majority of cases will also demonstrate systemic involvement.[1] About 30% of patients with the initial form of cutaneous sarcoidosis will develop its systemic form within months to several years of diagnosis.[6] Therefore, it is recommended that any patient with cutaneous sarcoidosis be screened for systemic lesions, even if there are no clinical complaints of systemic involvement at initial visits. Several diagnostic studies can be performed during the workup of sarcoidosis, including chest films, chest CT scans, and pulmonary function tests.

A review of the English literature revealed 47 reported cases of scalp sarcoidosis, including ours.[1,3,7–16] In these studies, patients were predominantly female (35/40). Where race was mentioned,

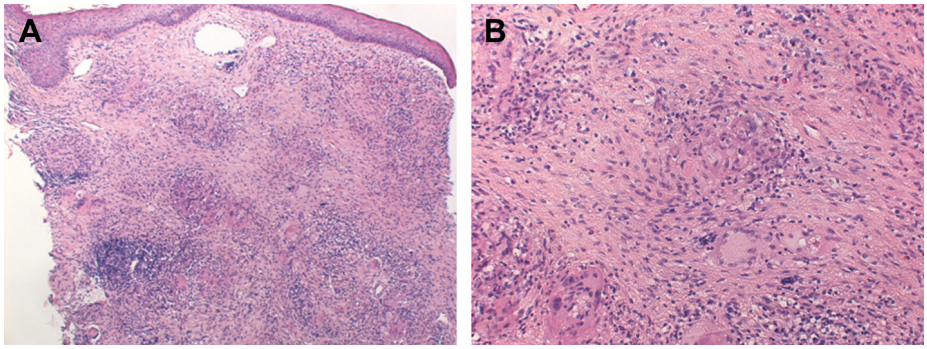

Figure 2 Pathological examination revealed noncaseating granulomatous inflammation involving superficial and deep dermis. Many asteroid bodies were found. (A) Original magnification 40×, hematoxylin and eosin stain; (B) original magnification 100×, hematoxylin and eosin stain.