BRIEF ARTICLE

A Case of Cutaneous Sarcoidosis with Direct Osseous Involvement

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ABSTRACT

Sarcoidosis is a granulomatous disorder that presents with cutaneous manifestations in one-third of patients, often as an initial symptom prompting interaction with the healthcare system. Here, we report a case of cutaneous sarcoidosis on the forehead with directly underlying erosive osseous disease. The patient was imaged further, uncovering pulmonary involvement. The lesion was treated with topical and intralesional corticosteroids with significant resolution. Though there exist a range of classic eruptions associated with sarcoidosis, skin involvement can present variably and should prompt additional imaging, particularly to assess for osseous and pulmonary involvement. Topical and intralesional corticosteroids can be effective first-line therapy for cutaneous sarcoidosis.

INTRODUCTION

Sarcoidosis is a multisystem autoinflammatory granulomatous disorder that primarily involves the lungs.[1] In approximately 25-30% of cases, cutaneous manifestations are noted, and often correlate with systemic inflammation.[2] Although skin involvement can manifest variably and may even involve the mucosa, lesions often have a predilection to sites of previous injury, including scars and tattoos.[2] Here, we present a case of cutaneous sarcoidosis as the first sign of systemic disease, located at the site of a previous injury, with direct underlying osseous involvement.

CASE REPORT

A Black male in his 50s presented with a firm, fixed nodule on the forehead at the site of a scar from a childhood injury. The nodule was previously treated with ten days of trimethoprim-sulfamethoxazole for a presumed diagnosis of skin abscess, but continued to enlarge despite antibiotic therapy. The lesion was asymptomatic and had not drained. Physical examination demonstrated a 3 cm firm, fixed subcutaneous nodule without significant epidermal change or warmth on the left forehead (Figure 1).

Review of systems was negative for fevers, chills, cough, shortness of breath, weight loss, dizziness, headaches, vision changes. Ultrasound demonstrated a heterogenous soft tissue mass overlying the left frontal bone with internal vascularity and microcalcifications. CT of the head and neck demonstrated a 0.8 x 3.0 x 4.2 cm mass, lymphadenopathy in the right lower neck and mediastinum, and, notably, subtle areas of irregularity involving the underlying bone,

concerning for osseous erosion secondary to an aggressive neoplasm (Figure 2).

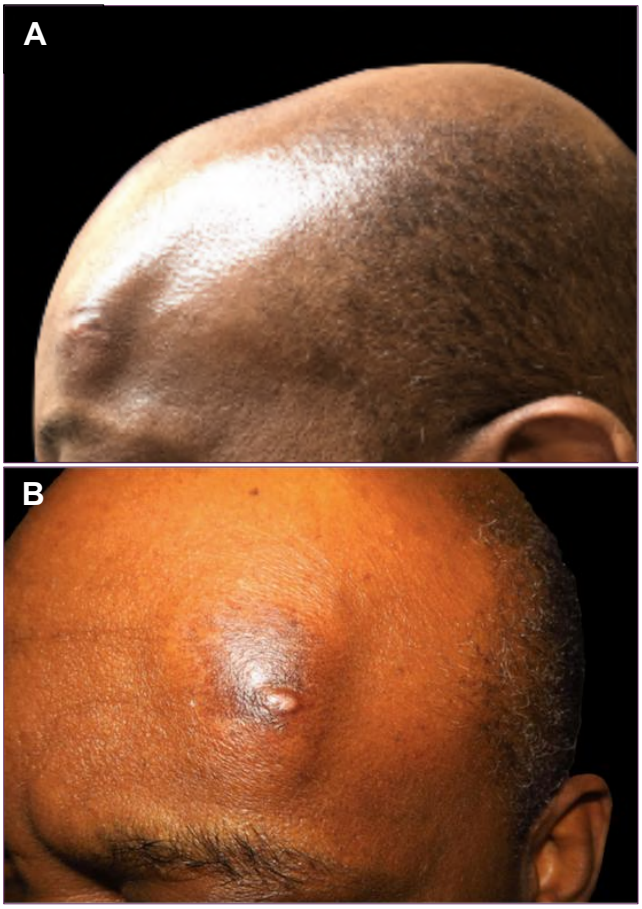

Figure 1. A) 1 cm horizontal scar from childhood **B)** Fixed subcutaneous mass on left forehead

A fine needle aspiration was performed, and pathology demonstrated nonspecific rare fibroblasts in a background of debris. Thus, an excisional biopsy was performed; histopathologic analysis demonstrated aggregates of epithelioid histiocytes forming multiple non-caseating granulomas in the dermis (Figure 3). Acid-fast bacilli stains were negative for mycobacteria, and polarizable material was not identified. Granulomatous disease became the leading diagnosis and a CT of the chest demonstrated bilateral hilar lymphadenopathy and lung nodules in a perilymphatic distribution. The histopathologic and radiographic findings

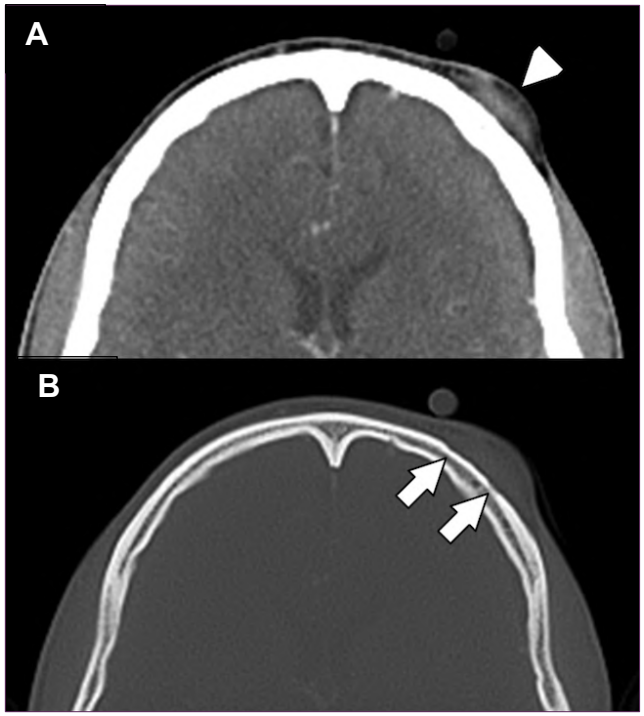

Figure 2. **A)** Contrast enhanced axial CT images in soft tissue and bone **B)** algorithms show a soft tissue mass in the left forehead (arrowhead) and focal areas of subtle erosion in the adjacent frontal bone (arrows).

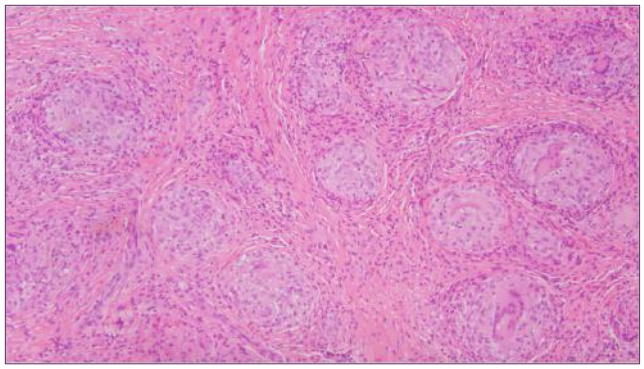

Figure 3. Section of left forehead excisional biopsy showed numerous dermal aggregates of epithelioid histiocytes and multinucleated giant cells with fibroplasia and a perivascular lymphohistiocytic infiltrate. (H&E, x10)

were consistent with a diagnosis of sarcoidosis.

The patient was initially treated with topical mometasone cream while establishing a diagnosis of cutaneous sarcoidosis. The patient was then transitioned to minocycline

and hydroxychloroquine, and later methotrexate. The skin nodule was treated with intralesional triamcinolone injection, with significant softening of the mass after the first injection. A total of three rounds of intralesional steroids were administered with marked clinical improvement.

DISCUSSION

The prevalence of sarcoidosis in the United States is 10 to 40 per 100,000 persons, with a higher incidence in African Americans and women.[3] The classic skin manifestations of sarcoidosis include red-brown to violaceous papules and plaques, generally symmetric in distribution and favoring the nose, periocular, perioral face.[4] However, cutaneous sarcoidosis can present variably and is considered an imitator of other skin diseases.[4] Overarchingly, manifestations are classified as either specific – containing non-caseating granulomas on pathology – or nonspecific – without granulomas on pathology but present in the context of systemic disease.[1] This patient notes a specific, albeit atypical, manifestation with direct extension of granulomatous disease to deeper structures.

Bony involvement of sarcoidosis is reported in 5 to 10% of patients and has been correlated with skin involvement – that is, it is rare to have osseous manifestations without cutaneous manifestations.[5] Despite this, there is a paucity of case reports documenting direct bony invasion of a cutaneous sarcoidosis lesion.[6, 7] To the authors' knowledge, only one previous case of direct invasion has been reported, involving the nasal bone.[7] In both this case and the previous report, overlying skin involvement caused a significant burden of symptoms; however, isolated osseous sarcoidosis is often asymptomatic and incidentally detected on imaging.[7, 8] Sarcoidosis involving the small bones most commonly affects the hands and feet and, radiographically, has a lace-like, lytic appearance. Sarcoidosis of the large bones can be lytic or sclerotic and may present as focal lesions or diffuse marrow infiltration.[7, 9, 10] Among cutaneous manifestations, lupus pernio may be more associated with bone cysts, though uncommonly through direct extension.[1]

The diagnosis of cutaneous sarcoidosis requires skin biopsy and histopathological confirmation illustrating non-caseating granulomas composed of epithelioid histiocytes. Confirmation of diagnosis should prompt a thorough systemic evaluation including history, physical examination, laboratory work-up (complete blood count, comprehensive metabolic panel, thyroid function tests), and chest radiography.[2] Additional imaging may be indicated to characterize extrapulmonary involvement, particularly in the presence of correlating clinical symptoms. Osseous disease is best characterized with MRI or PET/CT and may or may not prompt need for pathologic confirmation of diagnosis.[8]

CONCLUSION

Topical and intralesional steroids are first line in the treatment of cutaneous sarcoidosis. Depending on severity of skin disease and involvement of other organ systems, other treatment modalities include anti-malarials, minocycline, methotrexate, thalidomide, and TNF alpha inhibitors.[11] Such systemic therapies, with the addition of oral or intravenous glucocorticoids, also effectively treat symptoms associated with osseous sarcoidosis. However, in the absence of symptoms, there are no clear treatment guidelines for bone involvement.[8] Finally,

while certain cutaneous morphologies of sarcoidosis portend a worse prognosis – for example, lupus pernio – osseous disease has a more benign natural history.[2, 12] Previous studies have linked osseous sarcoidosis to favorable outcomes and the vast majority of cases remain stable or improved with treatment.[12] Thus, this case demonstrates an unusual presentation of cutaneous sarcoidosis in which imaging suggested direct osseous involvement, with excellent response to intralesional steroid injection.

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