SKIN

BRIEF ARTICLE

A Case of Atypical Presentation of Morpheaform Verrucous Sarcoidosis

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ABSTRACT

Morpheaform verrucous sarcoidosis is a rare variation of cutaneous sarcoidosis that is characterized by prominent, localized keratotic plaques on the skin. The etiology of this condition is unknown, and the current literature is limited. Sarcoidosis is typically a systemic condition with high incidence in the African-American population, and cutaneous presentation without systemic manifestation is exceedingly rare. This report details the challenging identification of this condition given the patient's history of multiple concurrent autoimmune and inflammatory conditions. Only nine cases of morpheaform sarcoidosis have been reported in the literature. This case demonstrates the complexity of diagnosing cutaneous sarcoidosis if the clinical systemic symptoms have not yet fully developed. It is important to take a multi-disciplinary approach with dermatology, rheumatology, and pulmonology to arrive at the correct diagnosis as cutaneous lesions can mimic several different cutaneous dermatoses. Therefore, physicians should consider morpheaform sarcoidosis on the differential in patients who present with indurated and eroded verruciform plaques.

INTRODUCTION

Morpheaform verrucous sarcoidosis is a rare variation of cutaneous sarcoidosis that is characterized by prominent, localized keratotic plaques on the skin.[1] The etiology of this condition is unknown, and the current literature is limited. Sarcoidosis is typically a systemic condition with high incidence in the African-American population, and cutaneous presentation without systemic manifestation is exceedingly rare.[1] Here we present a case of atypical morpheaform verrucous sarcoidosis.

CASE REPORT

A 59-year-old African American female with a history of speculated Sjogren's syndrome and pulmonary fibrosis presented with an asymptomatic lesion on her scalp in the fall of 2019. Over the next year, these lesions gradually spread to her legs and lesions would come and go. On exam, there were verrucous, hyperkeratotic plaques on the scalp, arms, upper chest, and upper back (**Figure 1**). A punch biopsy taken from her abdomen displayed prominent hyperparakeratosis with extensive inflammatory exudate and neutrophilic debris. The superficial to mid dermis was

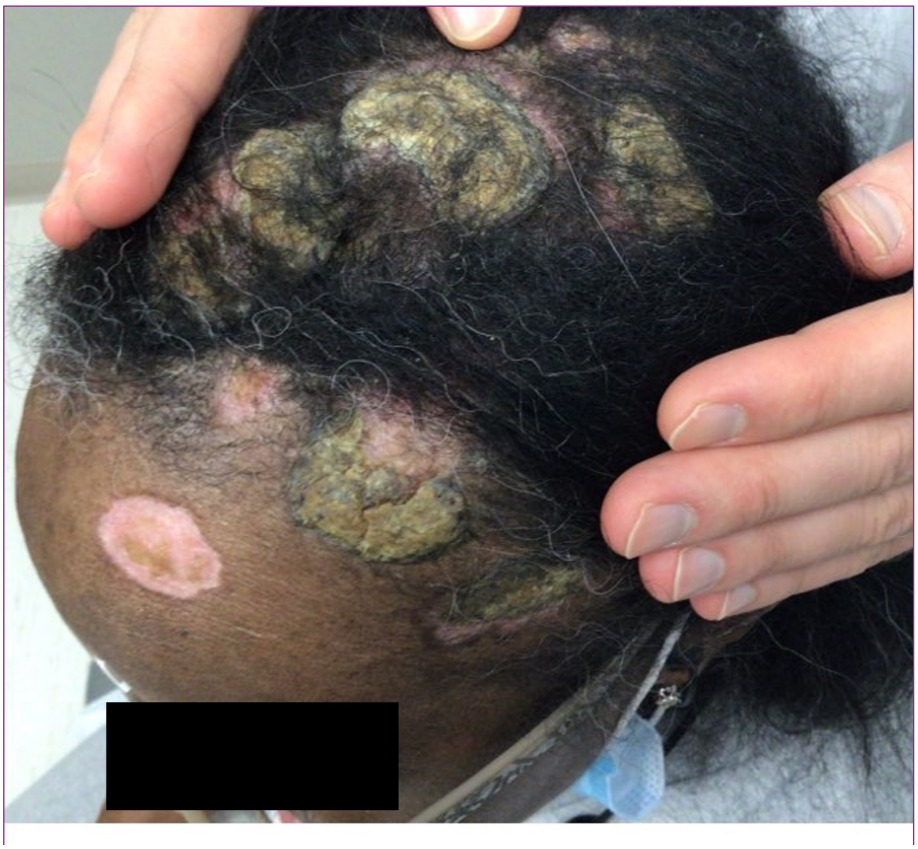

Figure 1. Verrucous, hyperkeratotic plaques identified on the scalp.

filled with dense non-necrotizing, non-polarizing granulomatous inflammation, with brisk lymphocytic infiltrate and occasional neutrophils. Foci of interface damage was noted in the epidermis and more evident on deeper sections. In the deeper dermis, there was sclerotic homogenized collagen extending to the base of the biopsy with patchy foci of plastic cells and giant cells. The findings of the biopsy were consistent with sarcoid, scleroderma, and an interface dermatitis seen in cutaneous lupus.

Due to the patient's worsening shortness of breath, a high-resolution CT scan was performed and revealed hilar lymphadenopathy. A pulmonary lymph node biopsy that demonstrated non-caseating granulomas and no concern for malignancy. She also recorded an unintentional 85 pounds weight loss over the past year. Additionally, she developed panuveitis and Bell's palsy suggestive of sarcoid.

Based on the clinical and histopathological findings, a diagnosis of morpheaform sarcoidosis was made. Soon after biopsy, the patient was started on mycophenolate mofetil 500mg twice daily and prednisone 60 mg daily in addition to topical clobetasol 0.05% ointment. The patient failed to improve on mycophenolate mofetil so the decision to switch to weekly subcutaneous methotrexate 25mg/mL was made. Every three months, intralesional injections of triamcinolone were also performed which led to significant improvement in the induration of some of her plaques on the scalp. However, over time the patient continued to develop worsening systemic symptoms and new cutaneous lesions, so she was started on Infliximab 5mg/kg infusions every six weeks in addition to subcutaneous methotrexate. This combination therapy ultimately led to stability in her disease status.

DISCUSSION

This report details the challenging identification of this condition given the patient's history of multiple concurrent autoimmune and inflammatory conditions. Only nine cases of morpheaform sarcoidosis have been reported in the literature.[2] Of these cases, the majority were women, about half were African American, and time of diagnosis was 58 years old on average.[2] Our patient seems to match the majority of the demographic data of others with this condition. The feature of dermal sclerosis in cutaneous sarcoidosis is seen more commonly in lupus pernio, however this can be a finding in specific cutaneous sarcoidosis as seen in two cases.[3]

Sarcoidosis can present with weight loss and respiratory symptoms. Dyspnea is present in 18-51% of patients.[4] Although rare, about 30% of cases of sarcoidosis present with a dermatological aspect.[4] This condition can be confused with several other conditions such as psoriasis, lichen planus, and lupus erythematous which can lead to misdiagnosis.[4] Involvement of the eyes is also a common occurrence, particularly uveitis.[4] All of these aspects of sarcoidosis were present in our patient which allowed determination of diagnosis. There are also several occupational and environmental triggers associated with sarcoidosis. These include inhaled bioaerosols, products of combustion, and metal dusts.[5]

Treatment can be difficult, as highlighted in our case, as there is a high rate of recurrence. Standard therapies include oral corticosteroids, antimalarials, and methotrexate.[6] Pentoxifylline, infliximab, and etanercept have also been used with some success.[6] New potential therapeutics on the horizon include combined levofloxacin,

ethambutol, azithromycin, and rifampin (CLEAR) and apremilast for cutaneous sarcoidosis.[7]

CONCLUSION

This case demonstrates the complexity of diagnosing cutaneous sarcoidosis if the clinical systemic symptoms have not yet fully developed. It is important to take a multi-disciplinary approach with dermatology, rheumatology, and pulmonology to arrive at the correct diagnosis as cutaneous lesions can mimic several different cutaneous dermatoses. Therefore, physicians should consider morpheaform sarcoidosis on the differential in patients who present with indurated and eroded verruciform plaques.

Consent: Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available

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