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Cutaneous Sarcoidosis in Skin of Color

Jessica R. Williams MS,[a] Cheri Frey MD,[b] George F. Cohen MD[c]

[a]Howard University College of Medicine, Washington DC
[b]Department of Dermatology, Howard University Hospital, Washington DC
[c]Department of Dermatology, University of South Florida, Tampa, FL

ABSTRACT

Cutaneous sarcoidosis presents in 25% of all sarcoidosis cases. African American populations, particularly African American women, are more likely to develop the dermatologic manifestations of the disease. There are several types of skin manifestations of sarcoidosis, which can make it more difficult to diagnose it clinically. Given the higher incidence of sarcoidosis and the poorer outcomes in these populations, it is essential to understand and recognize the variety of dermatologic symptoms associated with sarcoidosis. By doing so, patients can be diagnosed and treated earlier in their disease progression.

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INTRODUCTION

Sarcoidosis is a systemic disease that is identified by the key feature of non-caseating granulomatous inflammation in the affected organ.[11] The diagnostic criteria for sarcoidosis include a clinical and radiologic presentation of sarcoidosis, evidence of non-caseating granulomas, and exclusion of other causes of disease.[11] While the etiology of sarcoidosis is unknown, it is hypothesized that there may be a genetic predisposition or environmental factors that influence the disease progression.[1,10] The non-caseating granulomas develop due to the overstimulation of Th1 cells to secrete interferon gamma (IFN-γ), which activates macrophages. The activated macrophages then promote the secretion of cytokines such as tumor necrosis factor alpha (TNF-α) which leads to the production of the epithelioid histiocytes and the formation of the multinucleated giant cells that make up the composition of the non-caseating granuloma.

Sarcoidosis can affect patients of all ages and racial backgrounds, but it predominantly presents in African American females.[1,4] The pulmonary system is the most affected organ, and the skin is the second most common. Cutaneous sarcoidosis can be found on any skin surface including mucosal layers, and it has a higher propensity to develop in sites of previous skin disruption such as in scarring from injury or tattoos.[4] Clinical presentations of cutaneous sarcoidosis include lupus pernio, papular sarcoidosis, nodular sarcoidosis, plaque sarcoidosis, scar sarcoidosis, and erythema nodosum.[7] Given its high prevalence in African American women, it is essential to be familiar with the various cutaneous presentations of sarcoidosis for early detection and treatment, especially in skin of color.

Types of Cutaneous Sarcoidosis

Diagnosing sarcoidosis can be challenging because of the wide array of presentations associated with the disease. Comprehensive evaluations for sarcoidosis must be done early and include combined evaluations of a patient's clinical exam, blood work, radiologic imaging, and histologic features. The diagnosis of sarcoidosis is confirmed with biopsy indicating the presence of non-caseating granulomas. The absence of non-caseating granulomas in the skin does not rule out the diagnosis but is required in all of the specific skin findings described below.[6]

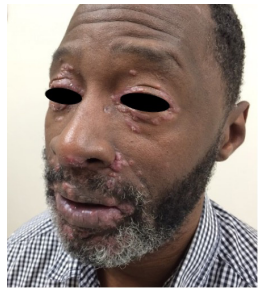
Specific Skin Findings

Papular Sarcoidosis

Papular sarcoidosis is the most common skin manifestation of sarcoidosis.[6] The papules are elevated skin lesions typically seen on the face, but they can be seen in any location on the body (Figure 1). Papular sarcoidosis is <1 mm in size and varies in color from reddish-brown to violaceous.[5] The papules are firm to palpation and have an "apple jelly" appearance when pressure is applied.[6]

Differential diagnoses: Rosacea, Sebaceous hyperplasia, Xanthoma.

FIGURE 1. Papular sarcoidosis.

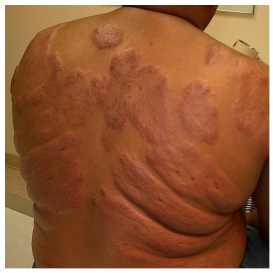


Plaque Sarcoidosis

Plaque sarcoidosis is characterized by elevated lesions >5 mm in size.[6] Plaques can be found on the face, extremities, or trunk, and may occur alone or in multiples. When plaques present in multiples, they are typically seen in a symmetric distribution (Figure 2).[5] Plaques are more likely to develop in deeper skin layers than papules.[6]

Differential diagnoses: Lichen planus, Psoriasis, Cutaneous T-cell lymphoma.

FIGURE 2. Plaque sarcoidosis.



Scar Sarcoidosis

Scar sarcoidosis involves patches that appear in areas of previous scarring. The patches may present as erythematous or violaceous in color and will affect areas such as the face, trunk, scalp, and extremities. The initial scarring can be caused by any mechanical trauma to the skin including venipunctures, previous infections, and tattoos (Figure 3). These lesions themselves are often asymptomatic and can be an indication of a sarcoidosis exacerbation.[6]

Differential diagnoses: Keloids, Hypertrophic scar.

FIGURE 3. Scar sarcoidosis.

Lupus Pernio

Lupus pernio more commonly affects women with skin of color.[6,9] It presents as indurated papules or plaques that vary in color from red to purple.[9] Lupus pernio is seen predominantly on the skin over the cheeks, nose, lips, and ears.[6]

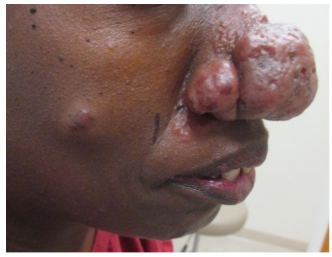
Differential diagnoses: Lupus erythematosus, Lupus vulgaris, Leprosy.

Nodular (Subcutaneous) Sarcoidosis

Nodular sarcoidosis, also known as Darier-Roussy sarcoidosis, involves non-tender firm subcutaneous nodules that are mobile and 0.5 - 2 cm in size (Figure 4).[6]

Differential diagnoses: Granuloma annulare, Lipomas.

FIGURE 4. Nodular sarcoidosis.



Ulcerative Sarcoidosis

Ulcerative sarcoidosis may arise with or without the presence of a pre-existing lesion on the lower extremities.[3] Ulcerative lesions are twice as likely to develop in women and individuals with darker skin tones.[6]

Differential diagnoses: Ulceration from stasis dermatitis, Cutaneous tuberculosis.

Hypopigmented Sarcoidosis

Hypopigmented sarcoidosis typically presents in patients with darker skin tones as well-demarcated hypopigmented macules and can also present as papules or nodules.[6] The papules are erythematous or skin-colored and may develop at the center of a hypopigmented lesion, giving the appearance of a fried egg.[8]

Differential diagnoses: Seborrheic dermatitis, Pityriasis alba, Vitiligo.

Ichthyosiform Sarcoidosis

Ichthyosiform sarcoidosis is rare and presents as scaly hyperpigmented plaques that are polygonal in shape and vary in color from gray to brown.[8] These plaques are commonly found on the lower extremities and are nontender and nonpruritic.[6] Approximately 95% of patients with ichthyosiform sarcoidosis will develop systemic sarcoidosis.[6]

Differential diagnoses: Eczema, Ichthyosis vulgaris.

Nonspecific Skin Findings

Erythema Nodosum

Erythema nodosum is caused by inflammation of subcutaneous fat (panniculitis) and is characterized as tender erythematous nodules that typically present on the shins anteriorly. It more commonly presents in patients of European, Puerto Rican, and Mexican descent, and often remits without treatment.[6] Erythema

nodosum is the most common nonspecific skin finding in patients. However, it can have other causes such as fungal and bacterial infections, leprosy, and inflammatory bowel disease.

Differential diagnoses: Erysipelas, Thrombophlebitis, Nodular Vasculitis.

Lofgren Syndrome

Lofgren syndrome includes erythema nodosum in addition to bilateral hilar lymphadenopathy, symmetric polyarthralgia, anterior uveitis, and fever. It predominantly affects patients of African, Puerto Rican, and Scandinavian descent and has a favorable prognosis with resolution of all symptoms within 2 years of the initial diagnosis.[6]

Differential diagnoses: Infectious (Coccidioidomycosis, Histoplasmosis, Tuberculosis), Inflammatory bowel disease.

Dermatologic Manifestations in Skin of Color

Sarcoidosis is a multisystemic disease and cutaneous manifestations occur in approximately 25% of cases, with some patients only manifesting cutaneous symptoms.[6] Sarcoidosis affects all races, but in the United States, African Americans have a higher prevalence of disease.[6] Additionally, due to the variety of skin manifestations, African American patients are more likely to be diagnosed when they are already in an advanced stage of the disease.[2] Factors influencing poorer prognosis in African Americans include access to healthcare, income, and level of education.[2] African American patients present earlier in life with more advanced disease, and have higher rates of hospitalization and higher rates of mortality.[1] These trends all lead to a poorer prognosis for African American patients with sarcoidosis.[6]

Treatment

The majority of cutaneous manifestations associated with sarcoidosis resolve without treatment. The prognosis of the disease is determined by the systemic symptoms and cannot be determined by the skin manifestations alone.[6] While the dermatologic changes are not an indication of disease severity, they can help clinicians diagnose and treat the disease earlier. Since sarcoidosis cannot be cured, treatment is based on providing symptomatic relief and preventing disease progression.

First-line agents used for cutaneous sarcoidosis are corticosteroids, which work by inhibiting the inflammatory response in the production of non-caseating granulomas.[6] If the cutaneous sarcoidosis is localized to a distinct area on the skin, then topical treatments such as clobetasol may be used. If the skin lesions include plaques and papules, then injections of triamcinolone every 4 weeks may be more effective.[6] If the cutaneous sarcoidosis does not respond to initial topical corticosteroid treatment, there is extensive skin involvement, or there is a potential for scarring, then systemic corticosteroids such as prednisone are employed.[6]

Antimalarials such as chloroquine and hydroxychloroquine may also be used for cutaneous sarcoidosis and work similarly to prevent granuloma formation.[6] Second-line treatments include immunosuppressive therapies such as methotrexate and cyclosporine. Additional options for refractory cutaneous sarcoidosis are monoclonal antibodies (infliximab), thalidomide, and isotretinoin.[6]

CONCLUSIONS

Cutaneous sarcoidosis can present in multiple forms and locations on the body. While the cutaneous manifestations may not be an indication of the severity of the disease, they can be an important clue for the prompt diagnosis of sarcoidosis. African American females are disproportionately affected by sarcoidosis. These populations are more likely to not only develop the disease but also have higher rates of hospitalization and worse outcomes. Given these higher incidence rates, it is critical to recognize the potential cutaneous manifestations that can be seen in African American patients.

DISCLOSURES

The authors have no conflicts of interest to declare.

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AUTHOR CORRESPONDENCE

Jessica R. Williams MS
E-mail:..... jessica.williams1@bison.howard.edu