# Scar Sarcoidosis: A Case Report and Brief Review

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GOAL

To understand scar sarcoidosis to better treat patients with the condition

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Describe the clinical presentation of sarcoidosis.
2. Identify modes of diagnosing sarcoidosis.
3. Discuss treatment options for sarcoidosis.

CME Test on page 416.

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*Scar sarcoidosis refers to lesions of cutaneous sarcoidosis that appear in preexisting scars. This condition may be caused by mechanical trauma such as skin cuts or venipuncture, scars caused by infection such as herpes zoster, and tattoos. We present a case of a 34-year-old man who developed scar sarcoidosis following minor trauma to the left calf. We review the epidemiology, clinical presentations, pathophysiology, and treatment options for scar sarcoidosis.*

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Sarcoidosis initially was described by Sir Jonathan Hutchinson in 1875, and cutaneous sarcoidosis (lupus pernio) was described by Besnier [1] in 1899. Sarcoidosis is a multisystem disease that may involve almost any organ system and, therefore, may present with various clinical manifestations.[2] Cutaneous

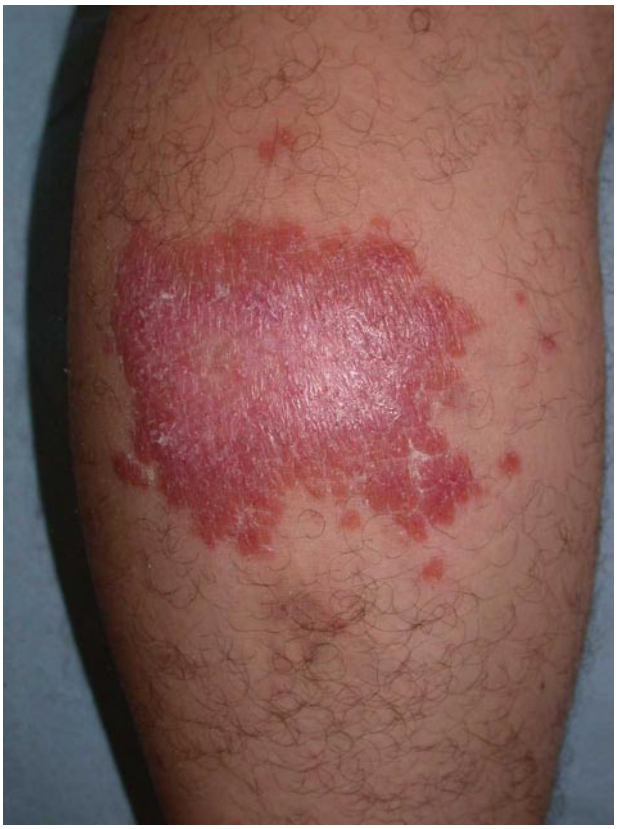

Figure 1. Large erythematous violaceous plaque on the left calf.

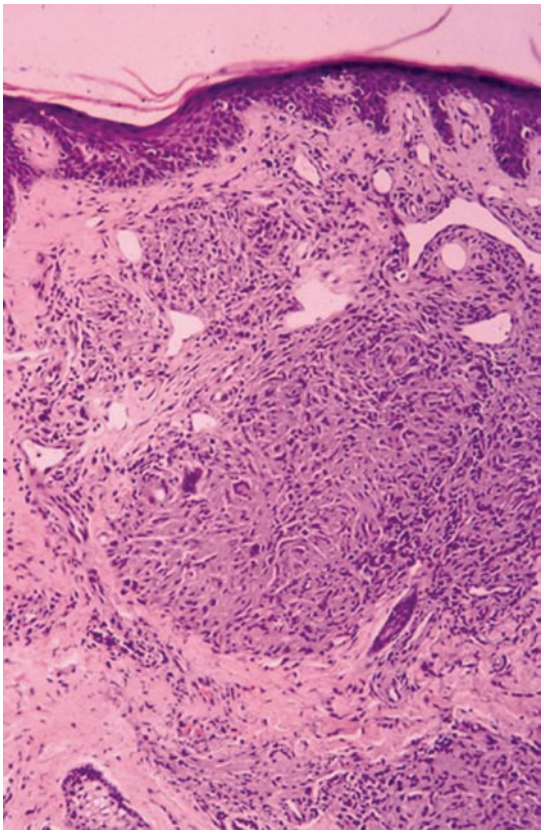

Figure 2. Sarcoid naked granulomas (H&E, original magnification ×40).

sarcoidosis occurs in up to one third of patients with systemic sarcoidosis. Recognition of cutaneous lesions is important because the lesions provide a visible clue to the diagnosis and are an easily accessible source of tissue for histologic examination.[3] Because lesions can exhibit many different morphologies, cutaneous sarcoidosis is known as one of the "great imitators" in dermatology.[4] Lesions of cutaneous sarcoidosis also can appear in preexisting scars, a condition known as scar sarcoidosis.[5] The latter condition may be caused by mechanical trauma such as venipuncture, scars caused by infection such as herpes zoster,[6] and tattoos.[7] Treatment of cutaneous lesions can be frustrating. For patients with widespread disease, the most effective treatment is systemic glucocorticoids. The prognosis of sarcoidosis usually is good, in particular, if the condition predominantly or solely affects the skin.[8]

## Case Report

A 34-year-old man presented with a progressively enlarging lesion on his left calf. He reported that about 3 months prior he had developed a small ulceration at this location following a fall. With local wound care, the ulceration healed with a scar. The scar, however, continued to grow beyond the borders of the previous ulceration and became raised with violaceous discoloration. The patient denied any history of excessive scarring or keloid formation after skin surgeries or trauma. There were no personal or family histories of granulomatous diseases.

Results of a physical examination showed an erythematous-to-dusky plaque measuring approximately 4×3 cm (Figure 1) on the left calf with well-defined irregular borders and discrete papules on the internal aspect of the knee. No tender nodules on the shins were noticed, and no lymphadenopathy was present. Results from a review of systems and a routine chest x-ray were unremarkable. Results of a punch biopsy revealed changes consistent with sarcoid naked granulomas (Figure 2). The patient was started on topical potent corticosteroid tapes and experienced marked improvement.

## Comment

Sarcoidosis occurs more frequently in females than in males, with reported ratios as high as 5:1. In the United States, black individuals are affected 3 to 4 times more often than white individuals.[9] Sarcoidosis is found worldwide and in every race, though the incidence varies dramatically. In Europe, the

disease affects white individuals more commonly than other races, and it affects Western Europeans more than Eastern Europeans. People from Scandinavia have one of the highest incidence rates at 64 cases per 100,000 population; in Poland, the incidence is 3 cases per 100,000 population. The disease is rare in Eskimos, Southeast Asians, New Zealand Maoris, and native Canadian populations.[10,11] The difference in prevalence among certain populations in varying geographic locations suggests that ethnic susceptibility factors, as well as environmental factors, contribute to the etiology of sarcoidosis.[11]

Sarcoidosis is a multisystem disorder characterized by noncaseating, naked, epithelioid granulomas and commonly involves the hilar lymph nodes, lungs, skin, and eyes. The frequency of skin involvement in sarcoidosis is 10% to 30% of all cases, but the prevalence of particular types of cutaneous lesions varies among races, as well as among individual cases.[12]

Clinically, there is spontaneous development of livid or reddish-brown plaques on scars that were previously and mostly atrophic; this phenomenon occurs at varying intervals. Therefore, sarcoidosis should be considered in the differential diagnosis of an enlarging previously inactive scar. Lesions can develop in scars caused by mechanical trauma, such as in Kveim test sites, tuberculin test sites,[5] sites that have received hyaluronic acid injection for wrinkles,[13] sites of cosmetic tattoos,[14] sites of previous laser surgery,[15] and sites used for desensitization injections.[16] Scar sarcoidosis has been reported following herpes zoster infection.[17]

Correctly diagnosing sarcoidosis may be a challenge. Unfortunately, no single test can lead to diagnosis of the condition. Patients are diagnosed with sarcoidosis when a compatible clinical or radiologic picture is present, along with histologic evidence of noncaseating granulomas, and when other potential causes, such as infections, are excluded.

Cutaneous sarcoidosis varies greatly in its clinical presentation and has been labeled as one of the great dermatologic masqueraders.[4,18] Maculopapular lesions can appear as xanthelasma, acne rosea, lupus erythematosus, or adenoma sebaceum. The differential diagnoses of plaques include lupus vulgaris, necrobiosis lipoidica, leprosy, leishmaniasis, psoriasis, and discoid lupus.[4,18]

The etiology of sarcoidosis is unknown, but several immune aberrations have been noted and are thought to play a role in its pathogenesis.[19] Immune dysregulation has been theorized to result from a persistent antigen of low virulence that is poorly cleared by the immune system, leading to a chronic T cell of the $T_H1$ subtype response and causing granuloma formation. Proposed antigens fall into 3 categories that include infectious, environmental, and autoantigens.[20] The most common infectious agents implicated are *Mycobacterium tuberculosis*, *Mycoplasma* species, *Corynebacterium* species, spirochetes, atypical mycobacteria, *Propionibacterium acnes*, *Borrelia burgdorferi*, herpes simplex virus, Epstein-Barr virus, cytomegalovirus, coxsackievirus, rubella virus, *Histoplasma* species, *Cryptococcus* species, coccidioidomycosis, and sporotrichosis.[21] Environmental antigens implicated include metals (eg, zirconium, aluminum, beryllium), organic dusts (eg, pine, pollen), inorganic dusts (eg, clay, soil, talc), and autoantigens (AV 2S3+ and HLA-DR17+).[22]

Genetic factors also are thought to play a role in the disease process.[23] Familial clustering of cases has been reported. Monozygotic twins are 2 to 4 times more likely to have the disease than dizygotic twins.[23] Certain HLA associations have been demonstrated; the most common allele found in sarcoidosis is HLA-B8. Other associated alleles include HLA-A1 and HLA-DR3.[24]

Most authors divide cutaneous lesions into specific and nonspecific categories.[25] Specific skin lesions display noncaseating granulomas on biopsy. Nonspecific skin lesions display no granulomas on biopsy. Scar sarcoidosis is a specific form of cutaneous sarcoidosis in which old scars become infiltrated with noncaseating epithelioid cell granulomas. Typical sarcoid lesions are characterized by the presence of circumscribed granulomas of epithelioid cells with little or no necrosis. Granulomas usually are in the superficial dermis but may involve the full thickness of the dermis and extend to the subcutaneous tissue. Islands of epithelioid cells may have a few Langerhans giant cells.[25] Giant cells may contain asteroid or Schaumann bodies; asteroid bodies are star-shaped eosinophilic structures; and Schaumann bodies are round or oval laminated structures that usually are calcified at the periphery.[26] Granulomas are referred to as naked because they have only a sparse lymphocytic infiltrate at the margins of the granulomas. Fibrosis, if present, usually starts at the periphery and advances toward the center.[26]

The treatment of cutaneous sarcoidosis often is frustrating, and the condition often is refractory to therapy or recurs following successful treatment. Therapeutic approaches range from topical, intralesional, and systemic use of corticosteroids to systemic medications such as cytostatic drugs, chloroquine,[27] allopurinol (300 mg/d),[28] and thalidomide.[29]

For localized involvement of cutaneous sarcoidosis, topical or intralesional steroids are used. Physicians frequently use superpotent topical corticosteroids because the drugs occasionally are effective.[30-32] However, the corticosteroid often does not adequately

penetrate the skin lesion. Intralesional corticosteroids (eg, triamcinolone acetonide in a dose of 5 mg/mL) typically are more effective, with injections repeated at 2- to 3-week intervals.[30,31]

Alternative therapies include oral psoralen plus UVA, surgical excision, and laser treatment.[32] The Q-switched ruby laser appears to be a rapid and effective means of treating scar sarcoidosis in traumatic tattoos without adverse effects.[33] Surgical excision of small lesions or excision of larger lesions with skin grafting can be attempted but may cause the recurrence of hypertrophic and keloidal scarring.[34]

Systemic agents are reserved for widespread progressive lesions or for lesions that impair function. Systemic glucocorticoids are the most effective agents and are commonly used at slow tapering dosages, starting at 20 to 60 mg/d of oral prednisone for 4 to 5 weeks. However, there are many drawbacks to this therapy. Aside from the well-known complications of chronic steroid use, not all patients respond to systemic steroids.[35] Patients who do respond frequently experience disease flare-ups after cessation of therapy. Many other medications may be used in refractory cases, including agents such as hydroxychloroquine sulfate,[36] methotrexate,[37] and thalidomide.[29] Although randomized controlled trials are lacking, multiple anecdotal reports suggest the efficacy of these agents.

The course and prognosis of sarcoidosis correlates with the mode of onset of the disease, the patient's race, and the presenting stage. In general, the prognosis of cutaneous sarcoidosis depends on systemic involvement. The course is variable, ranging from self-limited acute episodes to a chronic debilitating disease that may result in death.[38] Spontaneous remissions occur in nearly two thirds of patients, but 10% to 30% of patients have a more chronic or progressive course. The mortality rate is 1% to 6%. Sarcoidosis can lead to death either from severe involvement of lung parenchyma, which leads to pulmonary fibrosis and respiratory failure,[38,39] or from myocardial involvement, which leads to arrhythmias and cardiac failure.[39] Other causes of significant morbidity and mortality include central nervous system involvement, blindness, pulmonary hemorrhage, renal insufficiency, hypopituitarism, and liver disease.[35]

Cutaneous sarcoidosis usually has a prolonged course. Papules and nodules tend to resolve over months or years, though plaques may be more resistant.[19] As treatment is withdrawn, relapses are frequent, especially in black patients who tend to have more severe and prolonged symptoms.[11]

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