

# Fifty Shades of Sarcoidosis: A Case Report of Löfgren Syndrome

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## INTRODUCTION

Sarcoidosis is a multi-organ disorder that is characterized by the presence of noncaseating granulomas in involved organs. It commonly affects young and middle-aged individuals of all races, but is 3-4 times more common in African Americans and typically presents earlier with more severe symptoms.<sup>1</sup> The lungs are affected in 90% of patients and pulmonary disease accounts for the majority of the morbidity and mortality associated with this disease. However, approximately 30% of patients can present with extrapulmonary findings and can have involvement of other organs such as the skin and eyes.<sup>1</sup> The various presentations of sarcoidosis can make it challenging to diagnose and can lead to delays in treatment. Therefore, it is important for clinicians to recognize the wide variety of manifestations of sarcoidosis. In this case report, we present a case of sarcoidosis in a young man with arthralgias and skin lesions.

## CASE PRESENTATION

A 44-year-old African American male with a past medical history of hypertension, prior treated syphilis and anal warts presented with a three-week history of a constant, throbbing headache at the top of his head and night sweats. His headache was worse in the morning and improved with nonsteroidal anti-inflammatory drugs (NSAIDs). He noted a month prior that his eyes were red and that his vision had become blurry and he was prescribed glasses. Around that time he noticed a pruritic, tender rash on the ink lines of his tattoos on his upper extremities bilaterally. His tattoos were five years old and he had not had any issues with them prior to this episode. He also noticed tender, hard bumps on his lower extremities bilaterally. He complained of night sweats, chills, rigors, bilateral ankle pain, and weakness but denied any neck pain, photophobia, confusion, chest pain, shortness of breath, and cough. Of note, the patient had been diagnosed with syphilis four years ago when he presented with weakness in his lower extremities. His symptoms reportedly improved after a gluteal injection of an unknown medication. The patient's social history was positive for anal intercourse with his husband but he denied any new partners in the last six months. His husband was HIV positive but the patient reported that they used condoms consistently. On exam, the patient had a temperature of 100.9F, heart rate of 106 bpm, blood



**Figure 1.** Skin findings of tender firm nodules along the ink-lines of the tattoo on the patient's upper extremity.

pressure of 166/96 mmHg, and oxygen saturation 100% on room air. The only positive findings on physical exam were multiple raised, firm, and tender nodules along the ink-lines of both tattoos on bilateral upper extremities (**Figure 1**) and three firm, erythematous, non-fluctuating nodules on his lower extremities.

## DIFFERENTIAL DIAGNOSIS

In a middle-age, African American male with fevers, ankle pain, and new skin findings consistent for erythema nodosum (EN) on his lower extremities, sarcoidosis remained high on the differential despite the lack of pulmonary symptoms. Based on his past medical and social history, human immunodeficiency virus (HIV) and neurosyphilis were also on the differential. Given the patient's symptoms at the time of syphilis diagnosis (lower extremity weakness), the story seemed consistent for neurosyphilis. However, given the fact that he received only an intramuscular injection, it was unclear if he had been treated properly for neurosyphilis which would require IV Penicillin. Given his fever and tachycardia, infection was on the differential as well. During the work-up, the only abnormal labs were an elevated serum erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and angiotensin-converting enzyme (ACE) level. Blood cultures, HIV, rapid plasma reagin (RPR) test, and sexually transmitted disease (STD) testing were negative. A chest x-ray was unremarkable but a subsequent chest

CT showed bilateral multifocal pneumonia and possible mild hilar lymphadenopathy. A punch biopsy of the tattoo was performed which confirmed the presence of a sarcoidal granulomatous tattoo reaction with negative cultures. Given his clinical presentation of EN, hilar lymphadenopathy, arthralgias and fever the patient was diagnosed with Löfgren Syndrome.

## OUTCOME & FOLLOW UP

The patient was discharged the following day with a course of moxifloxacin for pneumonia and prednisone 40 mg daily with a plan to slowly taper. At his one-month dermatology follow up, there was improvement of his skin lesions and headaches. As his prednisone was tapered during the next four months, he reported a return of his headaches and new dyspnea on exertion. He was started on hydroxychloroquine and told to follow up with ophthalmology for clearance to continue this medication. Unfortunately, the patient did not follow up for clearance and hydroxychloroquine was discontinued.

## DISCUSSION

This case emphasizes that sarcoidosis is a multisystem disease with a variety of clinical presentations.<sup>2</sup> About 50% of patients diagnosed with sarcoidosis are asymptomatic with a normal lung exam at the time of diagnosis. However, sarcoidosis may present with pulmonary symptoms including non-productive cough, dyspnea, and chest pain along with fatigue, fever, joint pain, and weight loss.<sup>3</sup> The occurrence of sarcoid granulomas on old scars or tattoos is a rare but well-recognized phenomenon and can be the initial presentation of sarcoidosis in some patients.<sup>4</sup> A common presentation of extrapulmonary sarcoidosis is the combination of hilar lymphadenopathy, migratory polyarthralgia, and fever with or without EN, also known as Löfgren syndrome. Sarcoidosis is a diagnosis of exclusion that relies on a combination of symptoms, radiologic findings, histologic evidence of noncaseating granulomas in involved organs, and exclusion of other known causes of granulomatous inflammation such as lymphoma and tuberculosis. In the absence of Löfgren's syndrome, a tissue biopsy of the most accessible lesion is needed for diagnosis. Chest x-ray is the only routine imaging recommended for suspected sarcoidosis and can show bilateral hilar lymphadenopathy and pulmonary infiltrates. Chest CT scans can be used in more atypical presentations. In Löfgren syndrome, EN and arthralgias are managed with NSAIDs. Based on consensus recommendations, only patients with severe pulmonary symptoms are managed with prednisone 20-40 mg/day that is slowly tapered.<sup>3</sup> Since 60% of patients have recurrent symptoms, a maintenance dose of prednisone 10-15 mg/day for 3-6 months is recommended. Extrapulmonary treatment is

determined individually and may be managed with a similar steroid taper. In cutaneous disease, topical high-potency steroids are the first-line treatment. Additional systemic therapies include methotrexate and hydroxychloroquine.<sup>3</sup> Patients should follow up with a rheumatologist or pulmonologist. Recognizing that sarcoidosis can affect many organs and present with non-specific symptoms is essential in order to diagnose and treat this disease in a timely fashion.

## KEY POINTS

- Common organs affected in sarcoidosis include the lungs, skin and eyes.
- Extrapulmonary sarcoidosis can manifest as Löfgren syndrome which is a constellation of symptoms and objective findings including hilar lymphadenopathy, migratory polyarthralgia, and fever with or without erythema nodosum.
- Diagnosis of sarcoidosis is made from a combination of symptoms, radiologic findings, evidence of noncaseating granulomas in involved organs and exclusion of other known causes of granulomatous inflammation.
- Standard treatment of sarcoidosis is prednisone 20-40 mg/day that is tapered every 4-12 weeks and follow-up with a rheumatologist or pulmonologist.
- If symptoms reoccur, maintenance therapy with prednisone 10-15 mg/day can be continued for 3-6 months.
- In cutaneous disease, topical high-potency steroids, methotrexate or hydroxychloroquine can be used.

## REFERENCES

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