The Management of Sarcoidosis

A Primary Care Approach

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KEYWORDS

• Sarcoidosis • Treatment • Management • Corticosteroids • Primary care
• Bilateral hilar adenopathy • Löfgren syndrome

KEY POINTS

- Sarcoidosis is an idiopathic, granulomatous inflammatory disorder, which can affect any organ system.
- Hypercalcemia and hypercalciuria in patients with sarcoid is usually caused by an increased 1,25-hydroxyvitamin D level.
- Syncope and palpitations must be aggressively evaluated in patients with sarcoid, because they may be a sign of cardiac involvement.
- Steroids are the mainstay of treatment, but they should be tapered to their lowest effective dose.
- Steroid-sparing adjuvant therapy for sarcoidosis is available, depending on the organ system involved, most commonly methotrexate, azathioprine, and occasionally biological agents.
- Death from sarcoidosis most commonly originates from involvement of the pulmonary, cardiac, and nervous system.

INTRODUCTION

Sarcoidosis is an idiopathic granulomatous disease that can affect any organ system. Commonly, diagnosis requires histologic examination with evidence of noncaseating granulomas. The clinical picture is often complex and, combined with the rarity of the disease, can make diagnosis difficult for any health care provider. Despite these challenges, it is possible to successfully manage patients with sarcoidosis in a primary care setting.

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NATURAL HISTORY OF SARCOIDOSIS

In counseling patients with sarcoidosis, it is important to understand the disease manifestation as well as its possible progression. Certain factors, organ involvement, and demographic information may give insight into the possibility of good clinical outcome. Most patients experience spontaneous remission within months to years after onset, with no long-term sequelae.

Table 1 shows risk factors for sarcoidosis. In the United States, African Americans are at increased risk of suffering from sarcoidosis at 2.4% lifetime risk and also have increased severity of disease.[1] Whites are less commonly affected, at 0.85% lifetime risk, typically have more limited disease severity, and are more likely to experience disease remission.[1] The classic presentation of sarcoidosis of asymptomatic bilateral hilar adenopathy, present in 75% of those with sarcoidosis, is also more likely to undergo spontaneous regression (70%–80% within 1 year), whereas pulmonary fibrosis, certain cutaneous manifestations, and neurologic problems are more likely to be associated with increased chronicity of disease.[1,2] Cardiac involvement and pulmonary fibrosis are the most common causes of mortality in sarcoidosis. In counseling patients, this information can be used to offer hope or to begin to prepare the patient for a more insidious or severe clinical course. **Box 1** lists indicators of good prognostic versus more worrisome clinical findings.

Sarcoidosis is a disease with an unknown cause and nonspecific treatment options. Corticosteroids are the treatment of choice, but limited data have shown that treatment does not affect disease progression but may only manage symptoms and prevent irreversible changes.

INITIAL EVALUATION

A 27-year-old African American woman presented to her primary care provider with exertional dyspnea. By report, her mother had lung problems and eventually died of sudden death. Initial examination showed no other complaints, but a chest radiograph showed bilateral hilar adenopathy. A complete blood count (CBC), electrolytes and calcium, aspartate aminotransferase, alanine transaminase, and bilirubin tests and urinalysis are obtained. An electrocardiogram (EKG), echocardiogram, and tuberculin skin test are also ordered. The laboratory values and other tests are all normal except with a mild increase of liver function test (LFT) results. Not surprisingly, the tuberculin skin test is also within normal limits.

This is a classic description of a patient with sarcoidosis, more often occurring in African Americans and females. Eighty percent of cases occur in individuals 20 to 40 years old.[1] The tuberculin skin test helps rule out other granulomatous diseases; however, 60% of patients with sarcoidosis also experience skin anergy, which

Table 1
Risk factors for sarcoidosis

Risk Factors	Comments
Ethnicity/nationality	More common in African American, Scandinavian, Irish, German, and West Indian individuals. Rare in Japanese, Spanish, and Portuguese individuals
Genetic	Idiopathic complex, non-Mendelian
Age	80% of sarcoidosis occurs in individuals aged 20–40 y
Women	Marginal risk factor