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Cardiac Sarcoidosis

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Authors:

Ryan Rohan Sanni

Medical Student, Windsor University School of Medicine, Cayon, Saint Kitts and Nevis

Frederick M. Tiesenga, MD

Chairman of Department of Surgery, West Suburban Medical Center, Oak Park, Illinois

Charles Bradley Schubert, MD

Vascular Surgeon, West Suburban Medical Center, Oak Park, Illinois

Juaquito M. Jorge, MD

Bariatric Surgeon, West Suburban Medical Center, Oak Park, Illinois

Sandeep Mellacheruvu, MD, MPH

Director of Clinical Research, Loretto Hospital, Chicago, Illinois, and Associate Dean of Clinical Sciences, Windsor University School of Medicine

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A 64-year-old African American woman presented to the emergency department (ED) with constant unremitting sharp pain in the epigastric region, which she rated as 10 of 10 in severity. The pain had been present for the past 24 hours and had progressively become more severe within the past 12 hours, with radiation to the upper thoracic spine. She also described having increasing hemoptysis for the past 24 hours, filling one 12-oz cup in the ED.

She also reported having a history of nonresolving and difficult to manage cardiac arrhythmias, including Mobitz type 1 and type 2 second-degree atrioventricular (AV) block.

Results of laboratory testing in the ED were significant for a critically low hemoglobin level of 6.7 g/dL (reference range, 12.0-16.0 g/dL). Findings of chest radiography performed in the ED showed bilateral hilar lymphadenopathy with dilation of the ascending thoracic aorta (**Figure 1**).

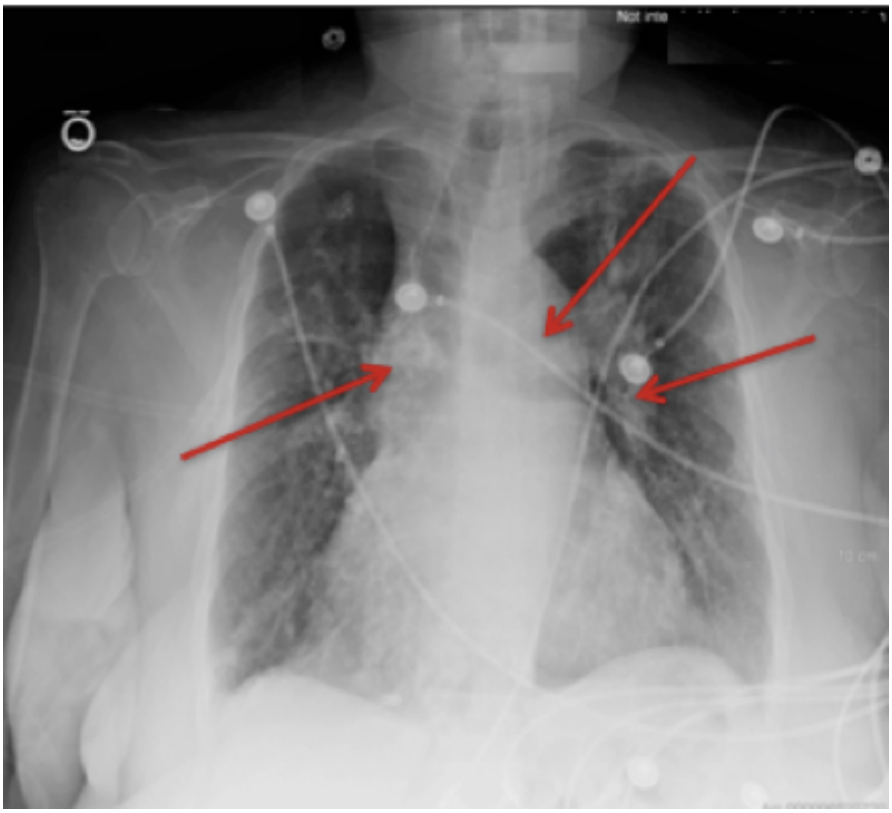

Figure 1. *Chest radiograph with the 2 arrows on either side of the sternum pointing to an enlarged hilar lymph node and the central arrow pointing to aneurysmal dilation of the ascending aorta.*

The patient underwent transfusion with packed red blood cells in the ED as a life-saving measure and was admitted to the internal medicine service for further evaluation.

While further test results, including brain natriuretic peptide (BNP), were pending, electrocardiography (ECG) monitoring on telemetry showed paroxysms of atrial fibrillation with rapid ventricular rate and atrial flutter with 2-to-1 AV block (**Figure 2**).

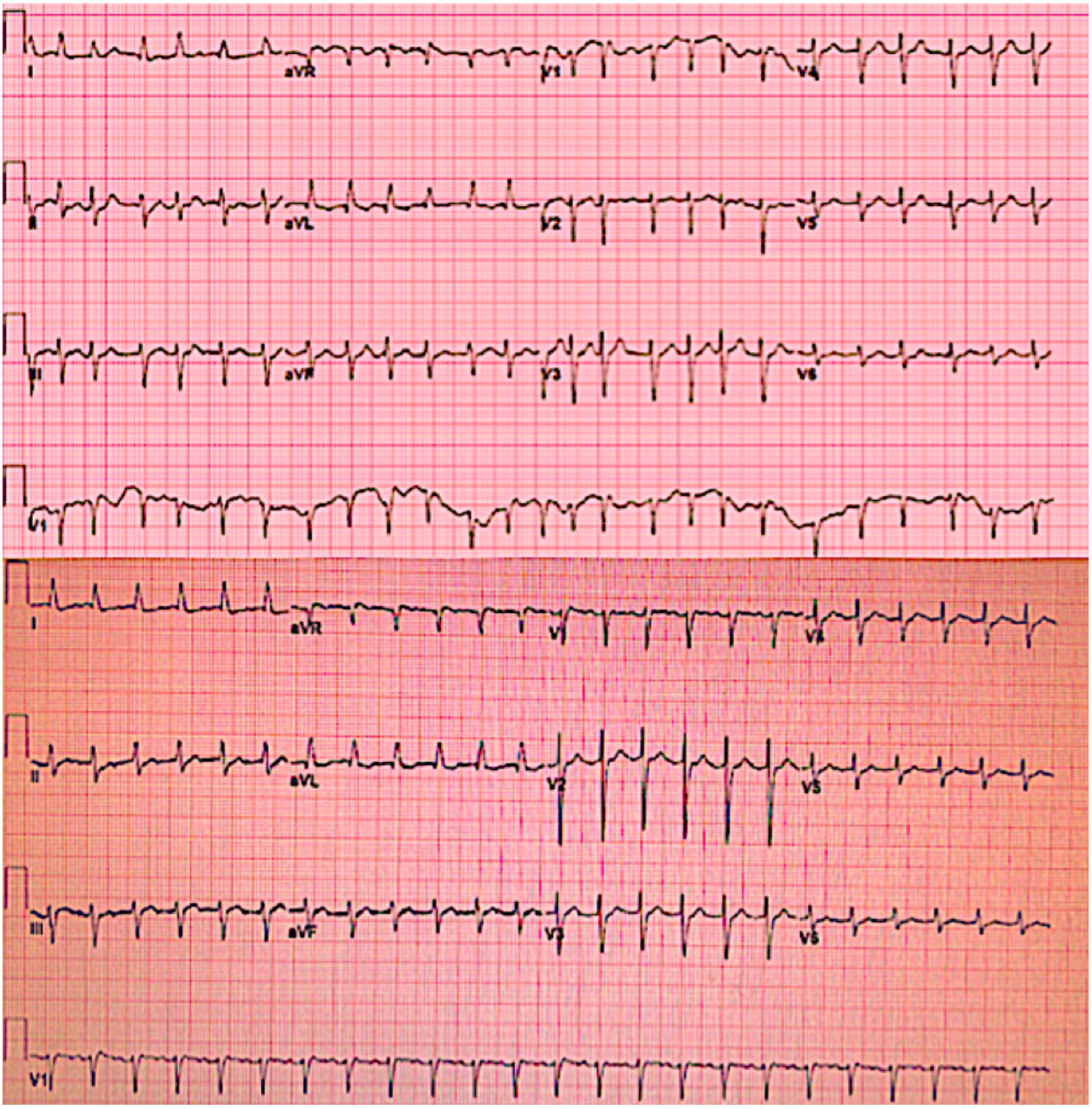

Figure 2. *The ECG tracing on the top shows atrial fibrillation with a rapid ventricular rate. The ECG tracing on the bottom shows fluctuating atrial flutter with 2-to-1 block.*

BNP test results came back elevated at 328 pg/mL (reference value, <100 pg/mL), indicating heart failure (HF) secondary to cardiac dysfunction.

Computed tomography (CT) imaging of the chest was done to further investigate the source of the hemoptysis. The results showed a cavity in the left upper lobe with a superimposed fungus ball inside. Additional findings included multiple bilateral ground-glass and nodular opacities, cardiac enlargement, and coronary artery calcification, suggesting spread of sarcoidosis to the great vessels. The ascending aorta measured 4.0 × 4.1 cm, highly suggestive of imminent dissection or enlarging aneurysmal dilation (**Figure 3**).

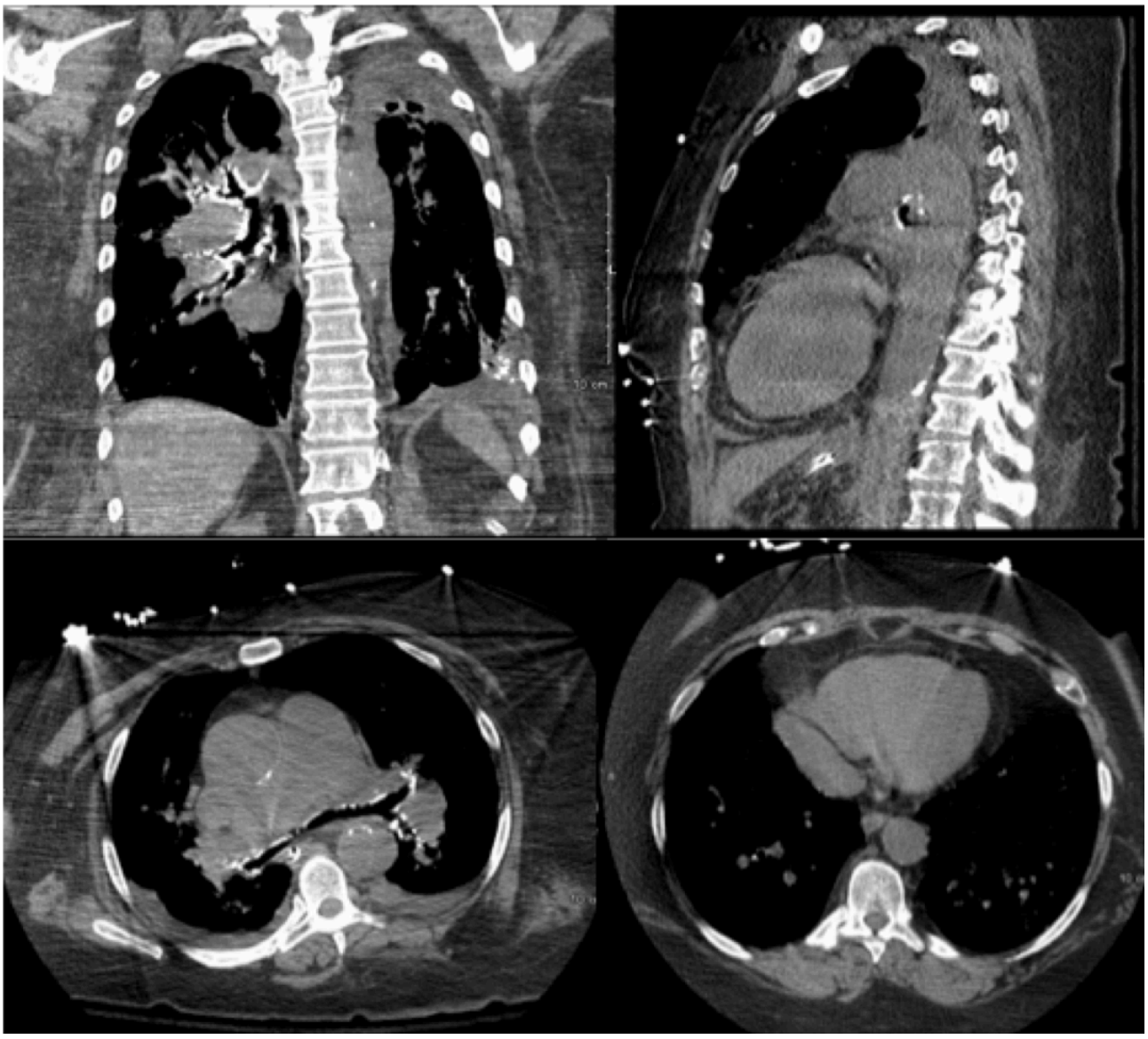

Figure 3. *Top left: A fungus ball located in a pulmonary sarcoid cavity, with a prominent aorta. Top right: A 4.0 × 4.1-cm dilation of the aortic root, and calcifications. Bottom left: Areas of calcification indicative of spread of sarcoid to great vessels. Bottom right: Bilateral pathognomonic ground-glass opacities seen with sarcoidosis.*

The suspicious enlarged hilar lymph nodes on chest radiographs prompted a pulmonologist to perform bronchoscopy with bronchial biopsy. The pathologic specimens demonstrated multiple noncaseating peribronchial granulomas. Given these findings, a diagnosis of sarcoidosis was made.

Given the patient's recurrent unresolving hemoptysis and the additional complicated pulmonary and cardiac problems, she underwent surgical tracheostomy placement. The patient returned from surgery in stable condition, and the hemoptysis was noticed to have been reduced.

Later in the patient's hospital course, repeated chest CT scans demonstrated findings concerning for a possible type A aortic dissection. The patient underwent urgent

transesophageal echocardiography, which revealed a type A ascending and descending aortic dissection (**Figure 4**).

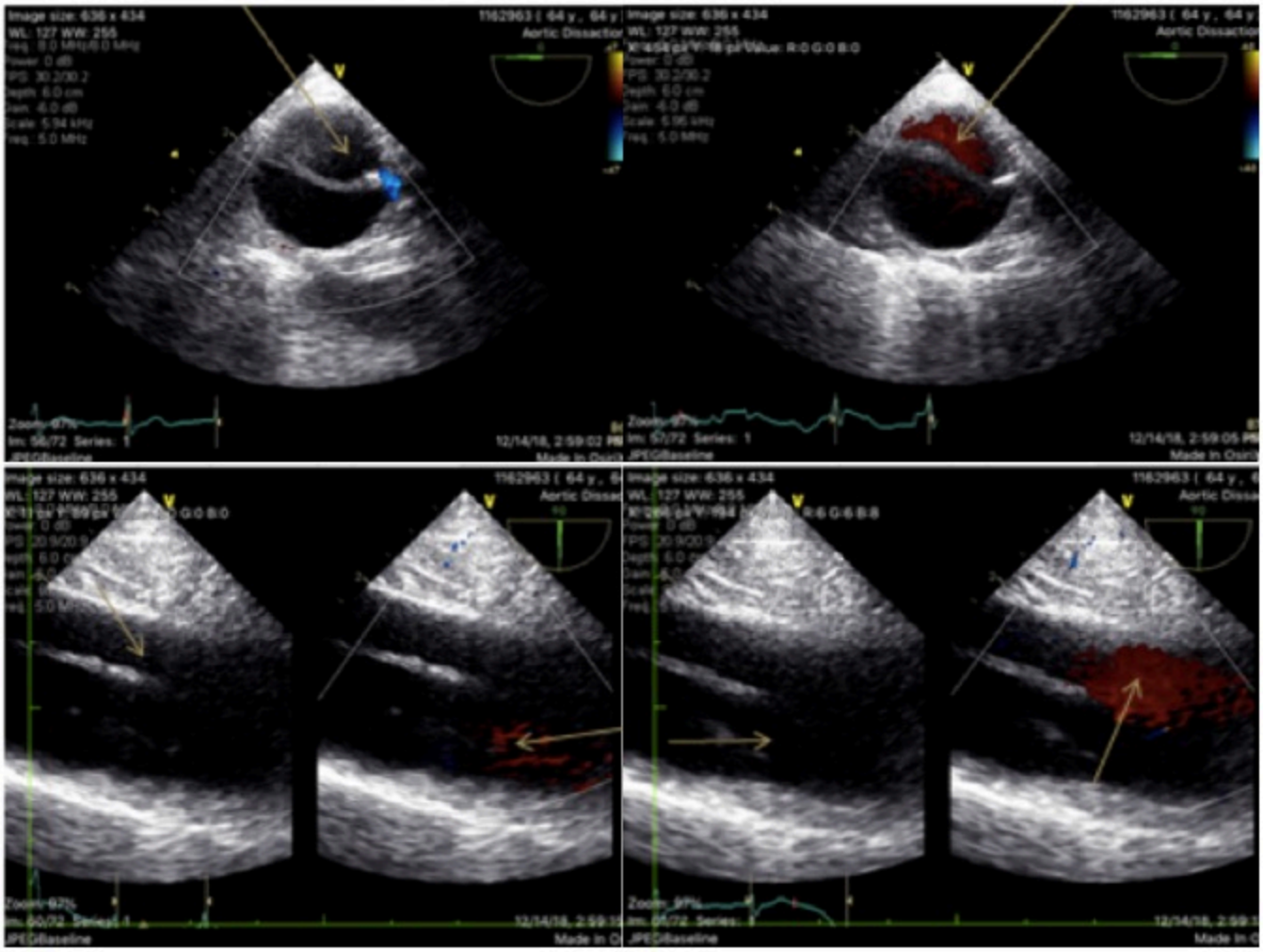

Figure 4. *In these transverse echocardiographic views of the aorta, the ascending aorta appears dilated. A linear structure is visualized extending from the aortic root into the aortic arch and descending aorta. True and false lumens were visualized. These findings are suggestive of a type A aortic dissection. The arrows in the top left and right images denote the false lumen. The arrows in bottom left and right images highlight the exposure of a dissection flap with the false lumen exposed.*

Ultimately, after extensive discussion with the family, the decision was made to pursue palliative care; she was discharged to hospice care and died from sudden cardiac death (SCD) shortly thereafter.

DISCUSSION

In the United States, the prevalence of sarcoidosis ranges from 5 to 40 cases per 100,000 population.[1] The prevalence in the white population is 11 cases per 100,000 population and is higher in the African American population, at 34 cases per 100,000 population.[1] Furthermore, approximately 20% of African American patients have a family member with sarcoidosis

compared with only 5% of white patients.[1] African Americans reportedly experience more severe and chronic disease.[1] Women are more often affected by extrapulmonary involvement of sarcoidosis. The incidence of sarcoidosis has been shown to peak once at 25 to 35 years of age and again for women only at 45 to 65 years of age.[1] These data directly correlate with our patient, a 64-year-old woman.

Approximately 5% of cases of sarcoidosis are asymptomatic and are incidentally detected via chest radiography; 45% of cases are associated with systemic symptoms of fever, anorexia, and arthralgia; and 50% of cases are associated with pulmonary symptoms such as dyspnea on exertion, chest pain, and cough. Hemoptysis, as seen in our patient's case is a very rare pulmonary symptom. It is also important to note that chest radiography findings show bilateral hilar lymphadenopathy with infiltrates depending on severity. Ocular manifestations, specifically granulomatous uveitis, also can be present in 30% to 60% of sarcoidosis cases.[1]

The prevalence of cardiac sarcoidosis remains unknown and is potentially underestimated, since many individuals with cardiac sarcoidosis have nonspecific symptoms or subclinical disease. Cardiac sarcoidosis can affect patients of all backgrounds and ages.[2] Some reports estimate the prevalence of cardiac sarcoidosis among US patients with systemic sarcoidosis to be 25%.[3] The most frequent clinical manifestations of cardiac sarcoidosis are AV block, HF, and SCD. Arrhythmias, including AV block, are the most common clinical presentation; supraventricular or ventricular tachyarrhythmia are also common. SCD due to ventricular tachyarrhythmias or conduction blocks account for 25% to 65% of deaths caused by cardiac sarcoidosis.[2] The presence of HF, with laboratory test results indicating elevation of BNP, in a patient with extracardiac sarcoidosis such as in our patient's case should prompt immediate suspicion for cardiac sarcoidosis.

There are several challenges related to the management of patients with cardiac sarcoidosis. First, data regarding the benefit of treatment are limited to small observational studies. Second, most treatments such as immunosuppressive drugs and implantable cardioverter defibrillators have a high likelihood of adverse effects. For a patient with cardiac sarcoidosis, starting with prednisone at a dose of 60 mg/day of is suggested, followed by a gradual reduction to a maintenance dose of 10 to 15 mg/day over 1 year.[2] Alternative agents such as methotrexate, azathioprine, infliximab, or mycophenolate mofetil may be given to patients who cannot tolerate the adverse effects of glucocorticoids or who have inadequate response to them.[2]

Tumor necrosis factor (TNF) antagonists such as infliximab can be used, but these agents may worsen HF and thus should be used with great caution in patients who have volume overload or other signs or symptoms of HF.[2] Given the increased toxicities and cost associated with anti-TNF agents, they should be used as second- or third-line options in patients who have an inadequate response to or intolerance of the other agents listed above.[2]

Drugs such as quinidine, mexiletine, and amiodarone may be used for patients who experience acute rapid ventricular tachyarrhythmias requiring immediate medical cardioversion. Long-term permanent pacemakers and catheter ablation have also shown promise as possible methods of treatment, but results are mixed due to the complexity of the disease.[2]

Further study is needed to better define the role of immunosuppressive therapies, including the ideal regimen, dose, and duration of therapy, as well as whether glucocorticoid-sparing agents can be used as first-line therapy, thus avoiding glucocorticoids. However, given that cardiac sarcoidosis remains relatively rare, and given the wide variability in severity of disease and clinical symptoms at presentation, such studies are difficult to conduct and require multicenter collaborations.[2] With emerging new monoclonal antibodies such as interleukin 17– and 23–targeted immunotherapy, studies should be conducted to assess the efficacy of these agents in the treatment of cardiac sarcoidosis.

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