A Rare Cause of Complete Heart Block in a Middle-Aged Indian Woman

Remyasri Nair, Purushotham Ramayya

Department of Medicine, MOSC Medical College, Cochin, Kerala, 'Department of Cardiology, A J Institute of Medical Sciences, Mangalore, Karnataka, India

Abstract

Sarcoidosis is a multisystem disease that might involve the heart during its course. Isolated cardiac involvement can occur in sarcoidosis without pulmonary and other systemic manifestations, and it could be the presenting manifestation. Cardiac involvement is an important cause of death in patients with sarcoidosis. Only 5%–10% cases of cardiac sarcoidosis are symptomatic. However, in autopsy series, cardiac involvement has been found in 50%–85% of cases. Here, we report the case of a middle-aged lady presenting with recurrent presyncope and complete heart block. A final diagnosis of cardiac sarcoidosis was made.

Keywords: Cardiac arrhythmias, complete heart block, sarcoidosis

INTRODUCTION

Immune-mediated diseases are multisystemic in nature. Their clinical presentation might vary from asymptomatic disease to organ failure and death. Lack of a comprehensive approach could result in misdiagnosis and treatment failure. Sarcoidosis is a multisystem disease, characterized by the presence of noncaseating granulomas in various organs. Worldwide incidence of sarcoidosis varies between 4.7 and 64 cases per 100,000 population. The incidence is highest among European and African American individuals, with a female predominance. The incidence is higher among the population in the age group of 15–70 years. Multiple organs such as lung, central nervous system, lymph nodes, skin, and eyes are involved in sarcoidosis. Rarely, sarcoidosis can also involve the heart. Cardiac involvement can manifest in isolation as complete heart block (CHB), ventricular arrhythmias, congestive heart failure, pericardial effusion, pulmonary hypertension, ventricular aneurysms, or sudden cardiac death. However, the diagnosis is made before death in only 40%–50% of patients with autopsy-confirmed cardiac sarcoidosis. The diagnosis of cardiac sarcoidosis remains a challenge, and it is also difficult to assess the disease activity. Here, we are reporting a rare case of cardiac sarcoidosis in a middle-aged woman, who presented with CHB.

CASE REPORT

A 50-year-old lady presented with a history of recurrent presyncope/syncope and exertional dyspnea (New York Heart Association Class III) for 3 months. She was a known case of hypothyroidism since 2 years, presently euthyroid on thyroxine replacement. Otherwise, she had been in good health previously. Her clinical examination was unremarkable. Her previous physician had advised permanent pacemaker implantation (PPI) as her electrocardiogram (ECG) showed CHB [Figure 1]. CHB in a middle-aged woman prompted us to evaluate her for secondary causes. Echocardiography was normal. Chest radiograph showed bilateral diffuse infiltrates [Figure 2]. High-resolution computed tomographic scan of the chest showed bilateral patchy ground glass opacities, subpleural nodules, and peribronchial thickening [Figure 3]; myocardium was looking normal [Figure 4]. Serum angiotensin converting enzyme (ACE) level was 12 U/L (normal 12–68 U/L). Ultrasonography of abdomen...
revealed lymphadenopathy in retroperitoneal region and porta 
hepatis and hepatomegaly with multiple granulomas. Her liver 
function tests were normal (total bilirubin 1 mg/dL, aspartate 
aminotransferase 20 IU/L, alanine aminotransferase 35 IU/L, 
serum albumin 4 g/dL, and alkaline phosphatase 100 IU/L). She 
underwent laparoscopic excision biopsy of lymph nodes and 
liver, which revealed noncaseating granulomas substantiating 
a diagnosis of sarcoidosis [Figures 5 and 6]. Endomyocardial 
biopsy was not done as less invasive alternative investigations

Figure 1: Electrocardiogram showing complete heart block

Figure 2: Initial chest X-ray with pacemaker shows bilateral diffuse 
infiltrates

Figure 3: Computed tomography (lung window) showing bilateral patchy 
ground glass opacities, subpleural nodules, and peribronchial thickening

Figure 4: Computed tomography (cardiac window) showing normal 
myocardium

Figure 5: Photomicrograph of lymph node biopsy showing noncaseating 
granuloma

Figure 6: Photomicrograph of liver biopsy showing noncaseating 
granuloma
were available. She was treated with steroids for 6 weeks. Her CHB persisted despite disease remission. Hence, she underwent PPI (dual chamber) electively. At 1-year follow-up, she was completely asymptomatic, and her chest radiograph was normal [Figure 7].

**Discussion**

Cardiac sarcoidosis has a poor prognosis compared to patients without cardiac involvement. Death might result from heart failure or cardiac arrhythmias. Sudden death might result from CHB or ventricular arrhythmias. In Japan, cardiac sarcoidosis is very common and is responsible for as many as 85% of sarcoidosis-related deaths.

The most common ECG finding in patients with symptomatic cardiac sarcoidosis is CHB. It occurs at a younger age when compared to CHB due to other causes. In a study, 23%–30% and 12%–32% of patients respectively were found to have CHB and bundle branch block. Conduction abnormalities result from involvement of the basal septum by granulomas, scarring, and ischemia caused by involvement of the nodal artery. Due to the associated risk of sudden death, cardiac involvement is considered a definite indication for early initiation of corticosteroids or other immunosuppressive agents. Cardiac disease can progress despite aggressive corticosteroid therapy. Failing which, other treatment modalities such as pacemaker or implantable cardiac defibrillator can be tried to prevent sudden death. Cardiac transplantation may be considered in refractory cases.

In symptomatic cardiac sarcoidosis, 5-year mortality rate exceeds 50%. Not many randomized trials have been performed to establish the efficacy of corticosteroids in the treatment of cardiac sarcoidosis. Case studies have shown short-term efficacy of steroids in the management of cardiac sarcoidosis, but long-term efficacy is yet to be proven. Efficacy of corticosteroids to induce remission in myocardial sarcoidosis remains uncertain due to a lack of controlled studies, diagnostic specificity, and adequate follow-up.

In our case, the patient was initiated on steroids. Her condition improved and symptoms subsided. However, the CHB persisted. Hence, she underwent elective PPI. Currently, she is stable at 1-year follow-up on tapering doses of steroids. Serum ACE levels are often estimated in patients with a suspicion of sarcoidosis. However, the sensitivity and specificity of serum ACE levels are suboptimal. Elevated levels of ACE are seen in only 60% of patients with acute disease and 20% of patients with chronic disease. In our patient, ACE level was normal. A normal ACE level does not exclude sarcoidosis. However, if elevated, it adds to the diagnosis. Endomyocardial biopsy has a high specificity, but it turns out to be negative in many cases of cardiac sarcoidosis due to patchy distribution of granulomas. Magnetic resonance imaging and positron emission tomography are other diagnostic modalities available for assessing cardiac involvement in sarcoidosis.

**Conclusion**

Cardiac involvement in sarcoidosis can occur in isolation. Despite being a well-recognized manifestation, this clinical presentation is rare in India. Long-term prognosis in sarcoidosis is primarily dependent on cardiac involvement. Early diagnosis is essential to avoid sudden death in sarcoidosis.

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**Conflicts of interest**

There are no conflicts of interest.

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