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Case Report

Cardiac sarcoidosis as initial presentation of multisystemic sarcoidosis

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ABSTRACT

Sarcoidosis is a multisystemic inflammatory disorder whose clinical presentation varies from asymptomatic disease to organ failure and death. Isolated cardiac sarcoidosis (CS) often escapes detection in absence of clinically apparent disease in other organs. Our case illustrates the evaluation of a young female presenting with syncopal attack with complete heart block which was later diagnosed being due to sarcoidosis. This is a rare case of sarcoidosis presenting with cardiac symptoms as its initial presentation. Our study emphasizes that the differential diagnosis of sarcoidosis should be kept in an advanced atrioventricular block especially in a young patient with no predisposing factors. Establishing the diagnosis of CS enables benefit from immunosuppressant therapy and prevents morbidity as well as mortality.

Keywords: Cardiac sarcoidosis, Complete heart block, Multisystemic involvement

INTRODUCTION

Sarcoidosis is a chronic inflammatory granulomatous disease affecting multiple organ systems, including the lung, skin, eye, liver, peripheral lymph node, nervous system, the heart, and the musculoskeletal system.[1] Cardiac manifestations though infrequent are associated with increased mortality rates.[1,2] Sarcoidosis presenting as a cardiac disease is rare and found in around 5% of cases.[3] We present a case of symptomatic complete heart block (CHB) in a young female where underlying etiology was diagnosed as sarcoidosis. Early diagnosis is essential to avoid sudden death and long-term organ affection associated with this presentation.[3,4]

CASE REPORT

A 23-year-old lady presented to the emergency with complaints of syncope. She was conscious and oriented, her blood pressure was 110/70 mmHg, and she had a regular pulse rate of 40 beats/min and her electrocardiogram revealed CHB. During evaluation, she developed polymorphic ventricular tachycardia (VT) which was terminated by electric cardioversion. A temporary pacemaker was inserted. Review of the case revealed that she had similar syncopal attack while doing household work 3 weeks before. She gave no history of fever, cough, shortness of breath, night sweats, weight loss, tuberculosis or contact with tuberculosis case or history suggestive of any connective tissue disorder such as joint pain, oral ulceration, alopecia, rash, photosensitivity, dry eyes or dry mouth, abdominal pain or neck swelling, or any prior history of cardiovascular disease. General physical and systemic examination was unremarkable. Her blood counts, renal and hepatic function tests, serum electrolytes, thyroid hormone levels, C-reactive protein, erythrocyte sedimentation rate and cardiac enzyme levels were within normal range. Ultrasonography of whole abdomen was normal and so was the chest radiograph. Anti-nuclear antibody profile and anti-phospholipid antibodies were negative, levels of complements 3 and 4, serum angiotensin converting enzyme, 25 hydroxy D3, and calcium were normal. Tests for lyme, cytomegalovirus, hepatitis B and C, and Brucella were negative. Mantoux and interferon gamma assay were negative. Amyloidosis workup was negative. However, 2D echocardiography revealed septal dyskinesia with normal chamber dimensions and ejection fraction of 60%. In view of echocardiographic findings, cardiac magnetic resonance imaging (MRI) was carried out which revealed focal septal thickening with delayed mid myocardial and epicardial enhancement in the left ventricular (LV) wall suggestive of cardiac sarcoidosis [Figure 1, Video 1]. Contrast-enhanced computed tomography chest and abdomen revealed patchy nodular opacities with bronchial wall thickening and mucus plugs within the bronchi in lung fields with multiple enlarged heterogeneously enhancing non-necrotic mediastinal lymphadenopathy [Figure 2]. Non-contrast computed tomography head showed gliotic changes in the left periventricular region. Contrast-enhanced MRI brain showed faint leptomeningeal enhancement in bilateral occipital region,

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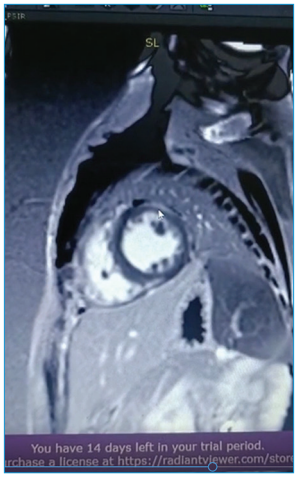

Video 1: Video of cardiac MR showing subepicardial late gadolinium enhancement in anterior and superolateral segment.

along with lacunar infarcts in bilateral gangliocapsular region and left centrum semiovale. Histopathologic study of the mediastinal lymph nodes revealed non-caseating granulomas without acid-fast bacilli. The patient refused endomyocardial biopsy. The final diagnosis was kept as multisystem sarcoidosis with cardiac presentation.

The patient was treated with prednisolone at 1 mg/kg bodyweight and azathioprine up to 2 mg/kg body weight for 6 weeks after which the prednisolone was tapered gradually to a daily dose of 5 mg. The patient has had no further syncope.

DISCUSSION

Worldwide incidence of sarcoidosis varies between 4.9 and 71 cases/100,000 population[5] with the highest incidence among European and African American individuals, and a female predominance.[3,6]

Clinical features of CS depend on the location, extent, and activity of the disease with the principal manifestations being conduction abnormalities, sudden death, and heart failure.[3,7] The extent of LV dysfunction seems to be the most important predictor of prognosis in CS.[3] Among conduction disturbances, CHB is most common at 23–30% of patients, followed by VT in 23% and supraventricular arrhythmias in 15%.[8,9] Rarer manifestations include features similar to arrhythmogenic right ventricular cardiomyopathy.[10]

Outcome of patients with clinically silent CS is controversial, while therapy with immunosuppression is advocated for the treatment of clinically manifest CS. Device therapy, primarily with implantable cardioverter-defibrillators, is often recommended in those patients with arrhythmias.[2,4]

It is being slowly realized that CS can be the first manifestation of sarcoidosis. Between 16% and 35% of patients under 60 years of age presenting with complete

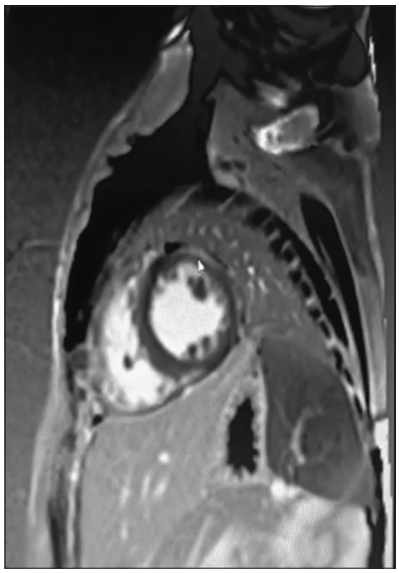

Figure 1: A 23-year-old lady who presented with syncope and bradycardia underwent cardiac MRI (short axis late gadolinium-enhanced pulse saturation inversion recovery sequence) which revealed focal septal thickening with delayed mid myocardial and epicardial enhancement in the left ventricular wall suggestive of cardiac sarcoidosis.

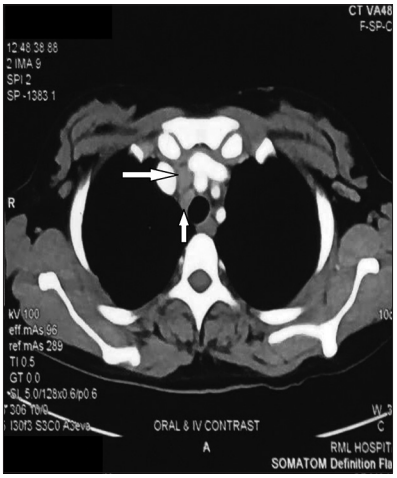

Figure 2: Contrast-enhanced computed tomography chest axial window revealed multiple enlarged heterogeneously enhancing non-necrotic mediastinal lymphadenopathy (white arrow).

atrioventricular (AV) block or VT of unknown etiology have previously undiagnosed CS as the underlying etiology.[11] Findings of sarcoidosis in 3.4% of LV biopsies at the time of LV assist device implantation[3] attest to the fact that CS as the underlying cause of heart failure is often missed.

The absence of histopathologic confirmation of sarcoidosis in the heart is a limitation in our study. Although the definitive diagnosis of CS can be made from endomyocardial biopsy, its sensitivity is low due to patchy myocardial involvement

usually <20%.[12] Cardiac MRI with gadolinium contrast can detect area of myocardial scar and/or edema in active stage of sarcoidosis.[13]

Our case stresses the importance of considering sarcoidosis as a differential diagnosis in cases of higher degree AV blockade particularly in younger patients because it gives the unique opportunity of starting immunosuppressants to arrest the progressive nature of disease and prevent sudden cardiac death.

CONCLUSION

Our study highlights that complete heart block may be an initial presentation of cardiac sarcoidosis especially in a young patient with no predisposing factors. It is necessary for the clinicians to be aware of the presentation so that early diagnosis is not missed. This is important as cardiac sarcoidosis is amenable to corticosteroids and this helps in preventing further morbidity and mortality.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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