Cardiac sarcoidosis presenting as complete heart block and heart failure: Importance of multidisciplinary approach

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SUMMARY

Introduction: Cardiac sarcoidosis is uncommon but causes significant morbidity and mortality. Unfortunately, achieving a definitive diagnosis is challenging. Case presentation: Two gentlemen are presented who complained of syncope due to complete heart block and heart failure. Blood investigations were non-diagnostic, while echocardiography showed poor ejection fraction. Cardiac MRI showed infiltrative heart disease with mediastinal lymphadenopathies. Biopsies of the lymph nodes and lung parenchyma were done through a flexible bronchoscope with endobronchial ultrasound (EBUS). Result: Histopathology showed non-caseating granulomas suggestive of sarcoidosis. Treatment with corticosteroid therapy and dual-chamber pacemaker insertion resulted in resolution of symptoms. Conclusion: Cardiac sarcoidosis should be suspected in patients with conduction problems or non-ischaemic cardiomyopathy. A multidisciplinary team approach is important for definitive diagnosis and proper management. EBUS-FNA provides a minimally invasive and effective method for tissue confirmation.

INTRODUCTION

Sarcoidosis is a systemic inflammatory disease caused by the accumulation of non-caseating granulomas that can occur in various organs. The lung is the most common organ affected, but approximately 5% of patients may have cardiac involvement. Cardiac sarcoidosis is uncommon but deadly. Non-caseating granulomas of sarcoidosis lead to inflammation of the pericardium, endocardium and, most commonly, the myocardium. Progression from focal inflammation to scar formation in the left ventricular free wall and the papillary muscles may result in the development of cardiomyopathies, arrhythmias, and even sudden cardiac death.[1]

Two cases of cardiac sarcoidosis are presented to highlight the importance of multimodal imaging together with biopsy to obtain the diagnosis in highly suspicious patients.

CASE PRESENTATION

Case 1

A 69-year-old Indian gentleman with underlying type 2 diabetes mellitus, hypertension and dyslipidaemia presented to the cardiology team with multiple syncopal attacks in 2018. Full blood counts, renal profiles, electrolyte levels and thyroid function tests were normal. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) of the brain and coronary angiogram were normal. Electrocardiogram (ECG) revealed complete heart block (Figure 1A), and echocardiogram (ECHO) showed dilated right atrium, right ventricle, and left atrium with reduced ejection fraction of 40%. He underwent dual-chamber pacemaker insertion for symptomatic complete heart block. The patient has been well since then with no subsequent syncopal episodes.

However, the patient developed worsening failure symptoms in 2020. Repeated ECHO demonstrated a worsening ejection fraction of 32% with thick left and right ventricles, and ECG showed multiple premature ventricular complexes. Cardiac MRI revealed parietal pericardial enhancement with late gadolinium enhancement (LGE) suggestive of infiltrative heart disease (Figure 1B), while computed tomography (CT) of the thorax showed mediastinal lymphadenopathy (Figure 1C). The patient underwent a positron emission tomography (PET) scan, which revealed metabolically active lesions at the cardiac, mediastinal lymph nodes and spleen.

The patient was referred to the pulmonology team, who performed transbronchial needle aspiration (TBNA) of the lymph node at station 7 using Olympus Vizishot-2 (22G Needle) by linear-endobronchial ultrasound (L-EBUS, BF-UC190F-Olympus, Tokyo, Japan). The patient underwent EBUS-TBNA, during which four passes were performed on the selected node, with 10 to 12 revolutions per pass. Histopathology (HPE) of the lymph node showed chronic granulomatous inflammation – the presence of multiple naked granulomas with occasional Schaumann bodies suggestive of sarcoidosis (Figures 1D and 1E). He was treated with a tapering dose of oral prednisolone (20 mg once daily (OD) for 2 weeks, 15 mg OD for 2 weeks and currently on 10 mg OD) as well as antifailure medications. Subsequent follow-up demonstrated resolution of symptoms and improvement in ejection fraction to 50%.

Case 2

A 54-year-old Indian gentleman working as a barber presented with a near-syncopal episode in December 2013 to the cardiology team. ECG showed complete heart block (Figure 2A). Blood investigation showed normal full blood counts, renal profile, electrolytes and thyroid function test.

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Cardiac MRI (Figure 2B) showed thickened myocardium with parietal enhancement and LGE over the lateral, inferoseptal and inferior walls in the midcavity and left ventricle with small pericardial effusion. A coronary angiogram showed normal coronaries, and a dual-chamber pacemaker was inserted in view of his complete heart block. He had not started on any medications then. He was subsequently lost to follow-up for device checks.

In September 2018, he presented again with reduced effort tolerance and bilateral lower limb swelling. ECG showed atrial fibrillation, and ECHO revealed thickened visceral pericardium, pericardial effusion and poor ejection fraction. Investigations for connective tissue disease, tuberculosis, and other infective causes were all negative. Serum angiotensin-converting enzyme (ACE) was also normal. Chest x-ray (CXR) showed bilateral hilar lymphadenopathy (Figure 2C). CT thorax (Figure 2D) demonstrated multiple mediastinal lymphadenopathies and small, calcified granulomas in the right upper lobe. The patient was referred to the pulmonology team, who proceeded with TBNA of the lymph node at station 7 and transbronchial lung biopsy (TBLB) by L-EBUS. Four passes were performed during EBUS-TBNA for the selected node, with 10 to 12 revolutions per pass. HPE of the lymph node and lung parenchyma revealed granulomatous inflammation suggestive of sarcoidosis (Figures 2E and 2F). Ophthalmology assessment revealed bilateral chronic anterior uveitis. He was then diagnosed with sarcoidosis with multiorgan involvement (heart, lung, eye).

The patient was treated with a tapering dose of oral prednisolone (20mg OD for 2 weeks, 15mg OD for 2 weeks, then 10mg OD) and antifailure medications. He succumbed three months later to severe pneumonia, attributed to immunosuppression resulting from prolonged corticosteroid use.

DISCUSSION

Sarcoidosis is an uncommon chronic systemic inflammatory and infiltrative disorder characterised by non-caseating granulomata. A diagnosis of sarcoidosis is based on compatible clinical and radiological findings, histological evidence of non-caseating granulomas and the exclusion of other causes of granulomatous disease. Sarcoidosis is common among adults less than 40 years old and peaks between 20 and 29 years of age, with a modest female predominance.[2] However, both our cases were above the age group, and both were males.

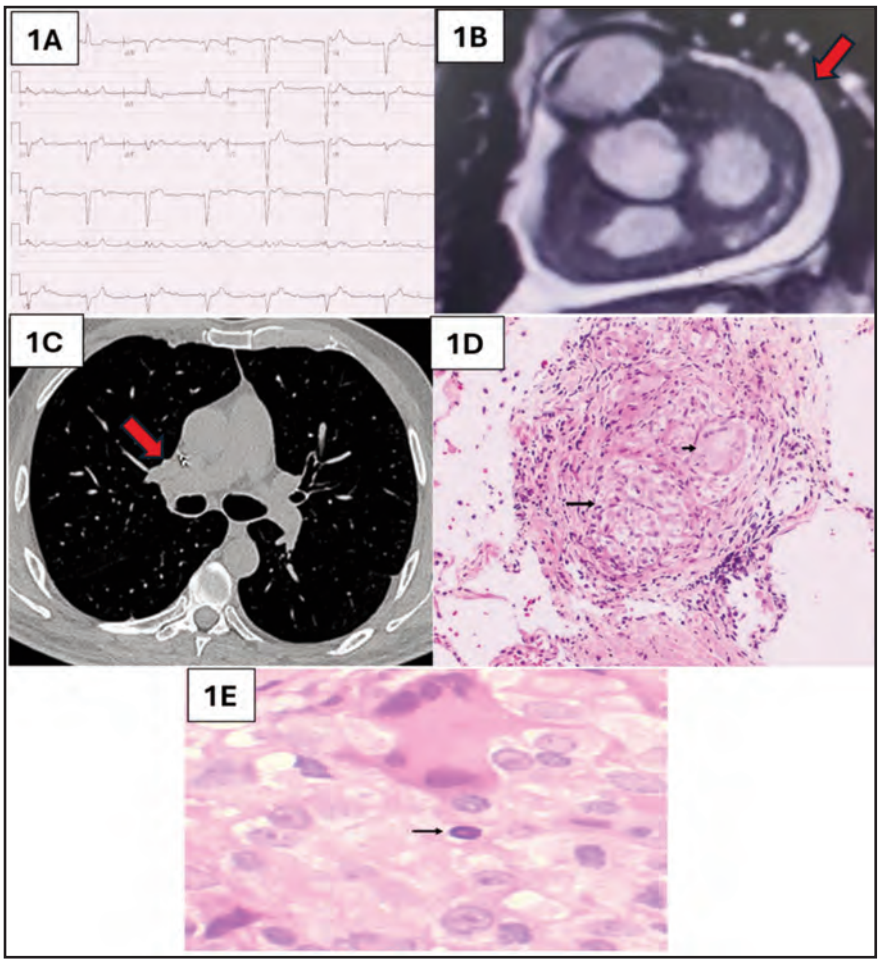

Fig. 1: ECG showing complete heart block (1A), Cardiac MRI showing enhanced parietal pericardium with LGE (1B), CT thorax showing mediastinal lymphadenopathy (1C), HPE lymph node showing naked granuloma with Langhans type multinucleated giant cell (1D) and Schaumann body (1E).

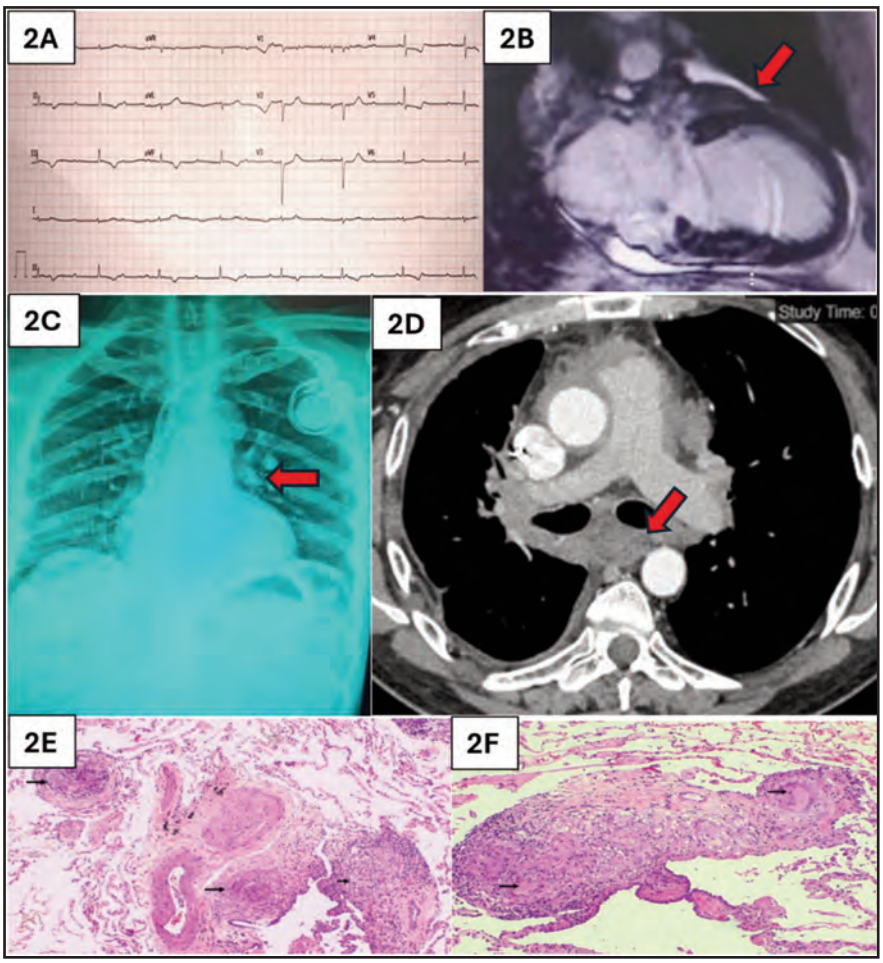

Fig. 2: ECG showing complete heart block (2A), CMRI showing enhanced parietal pericardium with LGE (2B), CXR showing bilateral hilar lymphadenopathy (2C), CT Thorax showing mediastinal lymphadenopathy (2D), EBUS-TBNA HPE showing small tight granulomas within the lung interstitium (2E) and naked granulomas with Langhans type multinucleated giant cells (2F).

A subset of cases involves the heart, which is calculated to be around 5% of patients with systemic disease. Cardiac manifestations vary from conduction abnormalities, ventricular arrhythmias, atypical chest pain, pericarditis, ventricular aneurysms, and congestive cardiac failure to sudden cardiac arrest.[3] The most common sites of involvement in the heart are the interventricular septum base (causing conduction abnormality and heart block), the left free wall (causing aneurysms, heart failure and arrhythmias) and the papillary muscles (causing heart failure).[3] Both of our patients presented with recurrent syncope due to complete heart block and heart failure symptoms.

Cardiac MRI is a striking diagnostic non-invasive method due to its ability to identify small regions of myocardial damage, even with preserved left ventricular systolic function.[1] Presence of LGE on cardiac MRI is the best independent predictor of fatal events, though bigger studies are needed to determine the sensitivity and specificity of screening processes for uncovering asymptomatic cardiac involvement. There is no specific pattern of LGE that is pathognomonic for cardiac sarcoidosis. Hence, images must be interpreted in the context of the patient's history and other relevant investigations under the expertise of a trained cardiologist and/or radiologist.

ACE is produced by epithelioid cells of activated macrophages. Serum ACE levels are often elevated, which may correlate with the total granuloma burden and disease severity but are not specific for sarcoidosis. However, a normal serum ACE level should not exclude the diagnosis, as seen in Case 2. Serum ACE can also be elevated in leprosy, histoplasmosis, hyperthyroidism and lymphoma. ECHO may reveal some abnormalities which may provide a clue to the diagnosis, but it is not specific either. Some of the positive ECHO findings are basal thinning of the ventricular septum, ventricular aneurysm and reduced left ejection fraction.

The most reliable method of diagnosis is via a tissue biopsy of any obvious lesions either from endomyocardium, lymph nodes or other organs. However, because of the scattered nature of the lesions, the diagnostic rate achieved with biopsy in cardiac sarcoidosis is low. Thus, a negative biopsy does not rule out cardiac sarcoidosis. Since the procedure is invasive and not without significant risk, it is done only in selected cases.[3] As illustrated by both cases above, both patients had lesions in the lungs which can be biopsied more safely

Table I: Diagnostic Guidelines for Cardiac Sarcoidosis (JCS 2016 guideline)

Clinical findings defining cardiac involvement

Cardiac findings should be assessed based on the major criteria and the minor criteria. Clinical findings that satisfy the following 1) or 2) strongly suggest the presence of cardiac involvement.

1. Two or more of the five major criteria (a) to (e) are satisfied.
2. One in the five major criteria (a) to (e) and two or more of the three minor criteria (f) to (h) are satisfied

Criteria for cardiac involvement

1. Major criteria

a. High-grade atrioventricular block (including complete atrioventricular block) or fatal ventricular arrhythmia (e.g., sustained ventricular tachycardia, and ventricular fibrillation)
b. Basal thinning of the ventricular septum or abnormal ventricular wall anatomy (ventricular aneurysm, thinning of the middle or upper ventricular septum, regional ventricular wall thickening)
c. Left ventricular contractile dysfunction (left ventricular ejection fraction less than 50%) or focal ventricular wall asynergy
d. 67Ga citrate scintigraphy or 18F-FDG PET reveals abnormally high tracer accumulation in the heart
e. Gadolinium-enhanced MRI reveals delayed contrast enhancement of the myocardium

2. Minor criteria

f. Abnormal ECG findings: Ventricular arrhythmias (non-sustained ventricular tachycardia, multifocal or frequent premature ventricular contractions), bundle branch block, axis deviation, or abnormal Q waves
g. Perfusion defects on myocardial perfusion scintigraphy (SPECT)
h. Endomyocardial biopsy: Monocyte infiltration and moderate or severe myocardial interstitial fibrosis

Diagnostic guidelines for cardiac sarcoidosis

Histological diagnosis group (those with positive myocardial biopsy findings)

1. Cardiac sarcoidosis is diagnosed histologically when endomyocardial biopsy or surgical specimens demonstrate non-caseating epithelioid granulomas.
2. Clinical diagnosis group (those with negative myocardial biopsy findings or those not undergoing myocardial biopsy) The patient is clinically diagnosed as cardiac sarcoidosis (1) when epithelioid granulomas are found in organs other than the heart, and clinical findings strongly suggestive of the above-mentioned cardiac involvement are present; or (2) when the patient shows clinical findings strongly suggestive of pulmonary or ophthalmic sarcoidosis; at least 2 of the five characteristic labouratory findings of sarcoidosis; and clinical findings strongly suggest the above-mentioned cardiac involvement.

Characteristic labouratory findings of sarcoidosis

1. Bilateral hilar (or mediastinal) lymphadenopathy
2. High serum angiotensin-converting enzyme (ACE) activity or elevated serum lysozyme levels
3. High serum soluble interleukin-2 receptor (sIL-2R) levels
4. Significant tracer accumulation in 67Ga citrate scintigraphy or 18F-FDG PET
5. A high percentage of lymphocytes with a CD4/CD8 ratio of > 3.5 in BAL fluid

Diagnostic guidelines for isolated cardiac sarcoidosis

Prerequisite

1. No clinical findings characteristics of sarcoidosis are observed in any organs other than the heart (The patient should be examined in detail for respiratory, ophthalmic, and skin involvements of sarcoidosis. When the patient is symptomatic, other aetiologies that can affect the corresponding organs must be ruled out.).
2. 67Ga scintigraphy or 18F-FDG PET reveals no abnormal tracer accumulation in any organs other than the heart.
3. A chest CT scan reveals no shadow along the lymphatic tracts in the lung or hilar and mediastinal lymphadenopathy (minor axis >10mm).

1. Histological diagnosis group
   Isolated cardiac sarcoidosis is diagnosed histologically when endomyocardial biopsy or surgical specimens demonstrate non-caseating epithelioid granulomas.
2. Clinical diagnosis group
   Isolated cardiac sarcoidosis is diagnosed clinically when the criterion (d) and at least three other criteria (a) to (e) are satisfied

compared to an invasive endomyocardial biopsy. Overall, EBUS-TBNA has sensitivity up to 89% with good diagnostic yield (86%) and negative predictive value (79%). 4EBUS-TBNA is also generally safe for adults, even those above 70 years old. EBUS-TBNA has also shown superiority compared to mediastinoscopy, as it is safer, less invasive and able to sample stations that are difficult to reach by mediastinoscopy, such as hilar and posterior carinal nodes. EBUS-TBNA was preferred for both cases in view of poor ejection fraction, which was deemed high risk for mediastinoscopy under general anaesthesia. Transbronchial cryobiopsy of mediastinal lymph nodes can be utilised in cases of suspected sarcoidosis to yield bigger samples. Endoscopic ultrasound (EUS) fine needle aspiration (FNA) and EBUS FNA also provide high diagnostic yield in sarcoidosis.

On microscopic examination, the granulomas are composed of epithelioid cells and multinucleated giant cells which are characteristically non-caseating. Commonly there are small numbers of surrounding lymphocytes and plasma cells (tight or naked granuloma). The giant cells may contain asteroid bodies, Schaumann bodies or other microcalcifications which are classically seen but not pathognomonic for sarcoidosis.[3] This was clearly demonstrated in both cases.

Sarcoidosis is mainly diagnosed via biopsy (heart or other organs) with other supportive findings such as bilateral hilar lymphadenopathy, high serum ACE level or a CD4/CD8 ratio of >3.5 in bronchoalveolar lavage fluid.[5] Cardiac sarcoidosis is diagnosed based on the Japanese (JCS 2016) guideline for the diagnosis of cardiac sarcoidosis (Table I)5. The guideline defines cardiac sarcoidosis when two or more major criteria are fulfilled or one major and two or more minor criteria.[5] Both our cases fulfilled the criteria of cardiac sarcoidosis based on the guideline. As illustrated by both cases, the diagnosis of cardiac sarcoidosis was challenging at initial presentation, as it requires a high index of suspicion and can only be confirmed via biopsy, which is difficult to obtain, especially in a case of isolated cardiac sarcoidosis.

Immunosuppression with corticosteroids remains the standard therapy for the acute inflammatory phase of cardiac sarcoidosis, but there is an evolving role for steroid-sparing agents. Systemic corticosteroids remain the first-line therapy owing to their efficacy in achieving an excellent clinical response within a shorter timeframe. Non-steroidal agents such as methotrexate, azathioprine, mycophenolate mofetil, leflunomide, cyclosporine or cyclophosphamide may be considered if treatment fails or if patients develop adverse reactions to systemic corticosteroids.[1]

Since cardiac arrhythmias in cardiac sarcoidosis may be fatal, permanent pacemakers (PPM) or implantable cardiac defibrillators (ICD) play an important role in management. Current guidelines recommend ICD implantation in cardiac sarcoidosis patients with sustained ventricular tachycardia, post cardiac arrest, and LVEF of 35% or less, while PPM is suggested for those patients with high atrioventricular blocks. Both our patients had dual-chamber pacemakers inserted, as both initially presented with symptomatic complete heart block.

This case series highlights that cardiac sarcoidosis should be considered in any patient presenting with conduction abnormalities, especially complete heart block or non-ischaemic cardiomyopathy. A multidisciplinary team approach is invaluable for establishing a definitive diagnosis. EBUS-FNA provides a high diagnostic yield in cases of sarcoidosis.

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