

CARDIAC SARCOIDOSIS: MULTIPLES ASPECTS IN ONE HEART CASE REPORTSasbou Lamyae^{*2}, Boussima Hanane², El Gueddar F. Z.², Barsoum Mikhail Paul¹, Maroni Jean Pierre¹¹Medical Cardiology Department, Hospital Center Robert Ballanger, Hopitaux Seine-Saint-Denis, Aulnay Sous Bois.²Medical Cardiology Department, IBN SINA Teaching Hospital, Mohamed V University, Rabat.***Corresponding Author: Dr. Sasbou Lamyae**

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ABSTRACT

Cardiac sarcoidosis is a manifestation of sarcoidosis that is challenging to diagnose due to its clinical silence, but its identification is vitally important, as its repercussions are potentially devastating. These repercussions include conduction abnormalities, arrhythmias, cardiomyopathy, congestive heart failure and sudden cardiac death. This case describes a 40-year-old Indian female with a atrio ventricular block and a dilated cardiomyopathy who had a delayed diagnosis of sarcoidosis with cardiac involvement. It was later deemed to be cardiac sarcoidosis after the onset of PET scan, cardiac MRI and biological elements. Treatment focuses on optimising heart failure therapy, placement of a biventricular implantable cardioverter defibrillator for primary prevention of sudden cardiac death and glucocorticoid. Although some studies suggest a potential benefit from glucocorticoid therapy, conclusive evidence revealing the true value of this treatment regimen has yet to be determined.

KEYWORDS: Cardiac sarcoidosis, Conduction abnormalities, Dilated cardiomyopathy, Cardiac MRI, Non-caseating granuloma, Corticosteroids.

INTRODUCTION

The clinical presentation of cardiac sarcoidosis (CS) ranges from an incidentally discovered condition to heart failure and sudden death.

The diagnosis of CS is difficult to establish, and as a result, CS is often under-recognized in clinical practice.^[1] CS most often occurs as a manifestation of systemic sarcoidosis, although isolated CS can occur in patients who do not have evidence of sarcoidosis in other organs.^[2]

Given nonspecific symptoms and baseline tests, noninvasive imaging with echocardiography, cardiac magnetic resonance imaging (CMRI) and positron emission tomography (PET)-computed tomography (CT) performed with fluorine-18-fluoro-deoxy-glucose (FDG-PET-CT) is instrumental.^[3]

While endomyocardial biopsy provides a high specificity for diagnosing CS, this invasive test has a limited sensitivity. Furthermore, there is limited understanding of disease progression and a lack of consensus on the optimal methods for disease detection.

This report describes a case of cardiac sarcoidosis focusing on workup that led to the diagnosis.

CASE REPORT

A 40-year-old female Indian patient with a history of gestational diabetes is admitted for shortness of breath and epigastric pain. There was no history of hypertension, dyslipidemia, coronary artery disease (CAD), and no family history of premature CAD or stroke. On physical examination, the patient appeared physically fit in no apparent distress. The vital signs were normal. There was no jugular venous distention and no carotid bruits. Lungs were crackling to auscultation. The heart rate was slow with an occasional premature beat. The first and second heart sounds were normal, and no murmurs, rubs, or gallops were appreciated. The abdomen was soft, nontender, non distended, with no hepatosplenomegaly. There was no peripheral clubbing, cyanosis, or edema. No skin lesions were noted. The 12-lead electrocardiogram (ECG) demonstrated an atrio ventricular block with junctional escape and Q wave of necrosis in DII, DIII and AVF (**Figure 1**). The echocardiography was notable for mild-moderate dilation of the left ventricle, with overall severe reduction of LV systolic function with an ejection fraction (EF) of 25 %. The infer posterior and the inferior walls were akinetic, and the rest of the cardiac muscle was hypo kinetic. The LV wall thickness was normal and the right ventricular (RV) size and function, atria, pericardium, and pleura were each normal.

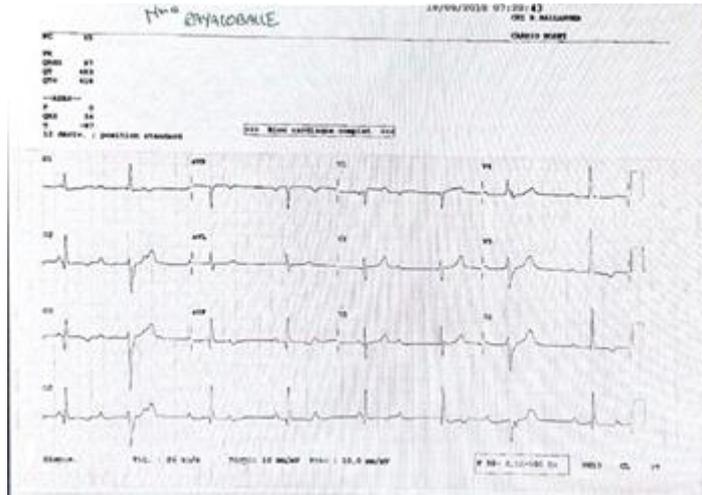


Figure 1: Initial electrocardiogram tracing notable for an atrio ventricular block with junctional escape and Q wave of necrosis in DII, DIII and AVF.

The coronary arteriography revealed a right dominant circulation with stenosis of 50% to 70% left main coronary artery that was dilated with an actif stent, left

anterior descending artery, and left circumflex artery with no significant lesions (**Figure 2**).

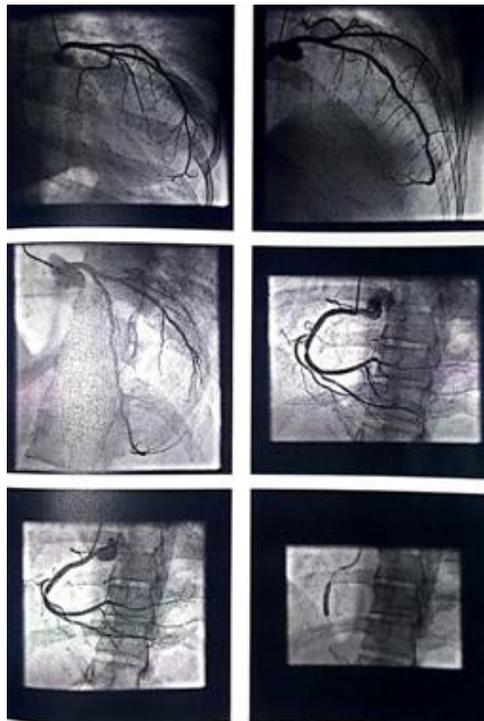


Figure 2: coronary arteriography revealed a right dominant circulation with stenosis of 50% to 70% left main coronary artery.

A cardiac MRI was performed and documented a late enhancement related to a dense fibrosis in epicardial muscle (**Figure 3**).

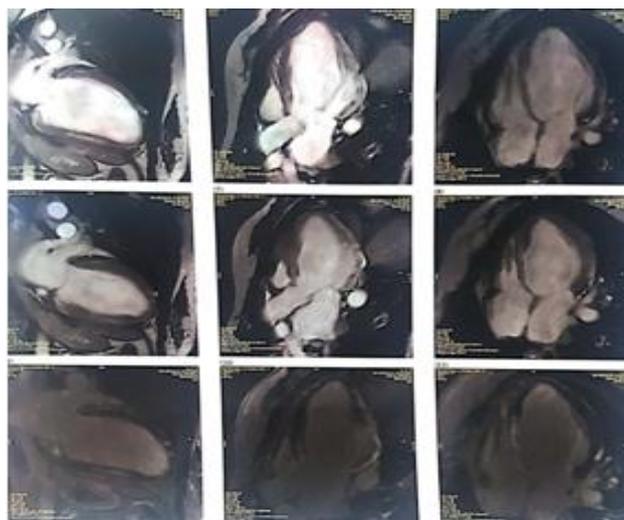


Figure 3: Cardiac MRI documented a late enhancement related to a dense fibrosis in epicardial muscle.

A PET scan showed significant anterior mediastinal and bilateral hilar lymphadenopathy with FNAC of hilar

nodes showing features of non-caseating granuloma type SARCOIDOSIS (Figure 4).

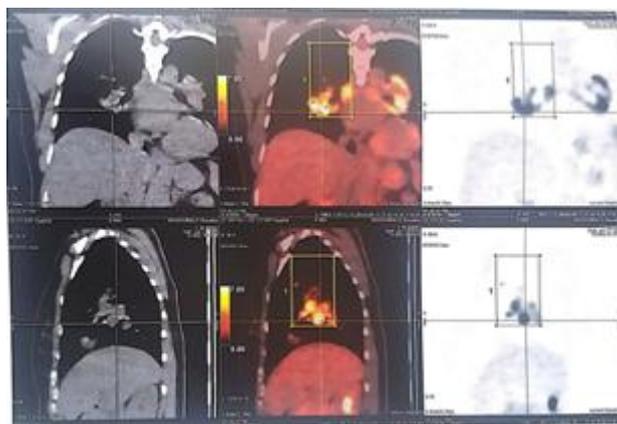


Figure 4: fluorodeoxyglucose positron emission tomography computed tomography (FDG-PET CT) demonstrating significant anterior mediastinal and bilateral hilar lymphadenopathy with FNAC of hilar nodes showing features of non-caseating granuloma type SARCOIDOSIS.

Under the light of the echocardiographic data, the persistence of the atrio ventricular block in the electrocardiogram, and mediastinal lymphadenopathy, our team establishes a diagnosis of sarcoidosis with heart damages. Although endomyocardial tissue biopsy was not performed, a presumptive diagnosis of cardiac sarcoidosis was made given the results of PET scan, the persistent conduction disorder, the enzymatic assay of ACE and the reduced left ventricular function. An implantable cardioverter defibrillator (ICD) was placed and a corticosteroids therapy was established. The patient was discharged home. At 3 months follow up, and since implantation of BiV-ICD, the patient's heart failure symptoms have remained stable. His baseline New York Heart Association (NYHA) classification is Class II. The patient demonstrated incremental improvement in his symptoms with his medical therapy consisting of furosemide, bisoprolol, spironolactone and glucocorticoid.

DISCUSSION

Cardiac sarcoidosis is a known but uncommon manifestation of sarcoidosis. It commonly presents as a disorder of cardiac rhythm which may be life threatening requiring urgent life saving measures or it may be discovered coincidentally while evaluating a patient with conduction defects^[4] CS may present with a variety of symptoms; most frequent presentations are severe conduction abnormalities in young patients, complex ventricular arrhythmias and dilated cardiomyopathy, as in the case here presented.^[5] Revised Japanese Ministry of Health Welfare (JMHW) criteria and biopsy results were used to confirm the diagnosis of CS. Systemic sarcoidosis is a clinical diagnosis supported by laboratory, radiologic and nuclear imaging studies and histopathologic or cytologic examinations. Furthermore, Heart Rhythm Society expert consensus recommendations on criteria for the diagnosis of CS provides 2 pathways to a diagnosis of CS: First, histological diagnosis from myocardial tissue and

second, clinical non-invasive and invasive imaging studies for establishing the diagnosis of CS^[6] When CS is suspected, several diagnostic modalities have been recommended. Assessments by TTE, ambulatory ECG monitoring, myocardial perfusion imaging, cardiac MRI or 18F-FDG PET-CT are recommended.

Echocardiography is readily available and provides valuable information of myocardial, valvular, pericardial, and congenital heart defects associated cardiac sarcoidosis. LV dysfunction, WMAs and abnormal septal thickness are most frequently reported. Also recently, Joyce *et al.*, have shown that RV dysfunction detected using right ventricular global longitudinal peak systolic strain (RVGLS) is commonly found in sarcoidosis in the absence of manifest cardiac involvement or pulmonary hypertension.^[7] Coronary angiography is commonly performed as part of the diagnostic evaluation, to rule out coronary vessel anomalies or malformations. In addition to echocardiography, CMRI can characterize tissue revealing inflammation and fibrosis. Diagnostic MRI findings in cardiac sarcoidosis are midwall rather than subendocardial or transmural LGE not corresponding to any particular coronary artery distribution. 13 LGE has a high sensitivity (100%) and moderate specificity (78%) for diagnosing cardiac sarcoidosis suggesting that myocardial scars indicated by LGE were an independent predictor of sudden cardiac death in patients with suspected CS. In our current case, MRI revealed several features of cardiac involvement such as delayed enhancement in RV and LV, edema and fibrosis of LV^[8] FDG PET can detect sarcoid lesions of the whole body and might be more sensitive as a diagnostic tool in detecting CS.

The most common histological features of sarcoidosis are non-caseating sarcoid granulomas with limited lymphocyte infiltration and patchy fibrosis. Endomyocardial biopsy (EMB) is not indicated as long as systemic sarcoidosis has been confirmed by lymph node biopsy or extracardiac biopsy. EMB has several limitations (invasive procedure and insensitivity due to focal involvement of the myocardium).^[9] CS is associated with high rate of mortality due to heart failure, complex ventricular arrhythmias, or even sudden cardiac death. The cornerstone of therapy for sarcoidosis is immune-suppression (corticosteroids) either alone or combined with other immune modulators (methotrexate, azathioprine). Corticosteroids improve survival, and pacemaker implantation for advanced heart block and ICD treatment should be considered to decrease the incidence of sudden cardiac death. ICD implantation is a class IIA indication in patients with cardiac sarcoidosis for primary prevention and class I for secondary prevention.^[10]

CONCLUSION

The diagnosis of CS can often be established with non-invasive modalities when consistent clinical and/or histological/cytological findings are consequent with

systemic sarcoidosis. Identifying early cardiac dysfunction with non-invasive modalities is a critical step in providing opportunities for risk stratification and early intervention to decrease the associated risk of long term cardiovascular disease.

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