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A patient with atrioventricular block and ventricular tachycardia: think sarcoid!

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Abstract

Cardiac involvement in sarcoidosis is often difficult to diagnose, and most alarmingly can lead to sudden cardiac arrest as its first manifestation. We report the case of a 45-year-old Indian woman with an implanted permanent pacemaker for atrioventricular block, who presented with haemodynamically stable ventricular tachycardia and was found to have impaired left ventricular function. Subsequent investigations established the diagnosis of cardiac sarcoidosis. The patient was treated with prednisolone initially at 40 mg a day for 3 months. Left ventricular function improved over 3 months of treatment and there was no further recurrence of ventricular tachycardia. Screening for cardiac sarcoidosis should be considered in a patient with unexplained atrioventricular block and ventricular tachycardia, particularly if young, even in the absence of clinical findings of extracardiac sarcoidosis. Treatment of the cardiac sarcoidosis could control ventricular tachycardia and improve left ventricular function.

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Introduction

Sarcoidosis is a multisystem disease characterised by non-caseating granulomas. Most commonly it affects the lungs, but may affect any organs such as skin, heart, liver or the nervous system. Cardiac involvement occurs in 25% of cases of systemic sarcoidosis. The aetiology of sarcoidosis is uncertain, but believed to be due to an immune reaction to unidentified antigens. Cardiac involvement occurs in three forms: conduction system disease, ventricular tachyarrhythmia (VT) and left ventricular (LV) dysfunction. Most alarmingly, cardiac sarcoidosis can cause sudden cardiac arrest which may even be the first manifestation.[1]

We describe a case of a young female with unexplained atrioventricular (AV) block, VT and LV dysfunction. Cardiac sarcoidosis was diagnosed on further testing. Following treatment with corticosteroids, the LV function improved and there was no further recurrence of VT.

Case presentation

A 45-year-old woman presented with an episode of palpitation lasting for one hour; her pulse rate was 180 beats/minute and blood pressure was 110/60 mmHg. Irregular cannon waves were seen in the jugular venous pulsation. The chest was clear on auscultation and the general examination was normal except for pallor. She had a dual-chamber permanent pacemaker implanted two years before for episodes of syncope with intermittent 2:1 AV block in presence of baseline trifascicular block. At the time of implantation, echocardiography showed preserved LV systolic function and the coronary angiogram was normal. She did not have hypertension or diabetes, nor did she give any other significant medical history except long-standing mild anaemia, for which she was taking iron supplements.

Electrocardiogram (ECG) in the emergency room showed monomorphic VT of left-bundle branch morphology, the transition at V4, discordance in leads II/III indicating an origin from the right ventricular septum. Echocardiography revealed impaired LV function with 35% ejection fraction (EF) with global hypokinesis. As she was haemodynamically stable, intravenous infusion of amiodarone was started, and sinus rhythm was reverted within an hour. Pacemaker interrogation showed multiple episodes of non-sustained ventricular high rate events. A 24-hour ambulatory ECG done after a week confirmed significant ventricular ectopic load including couplets, triplets, and non-sustained VT.

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Figure 1 Cardiac PET scan. Heterogenous FDG uptake pattern in LV myocardium involving the lateral wall, anterior wall and septum, but most marked in the lateral wall.

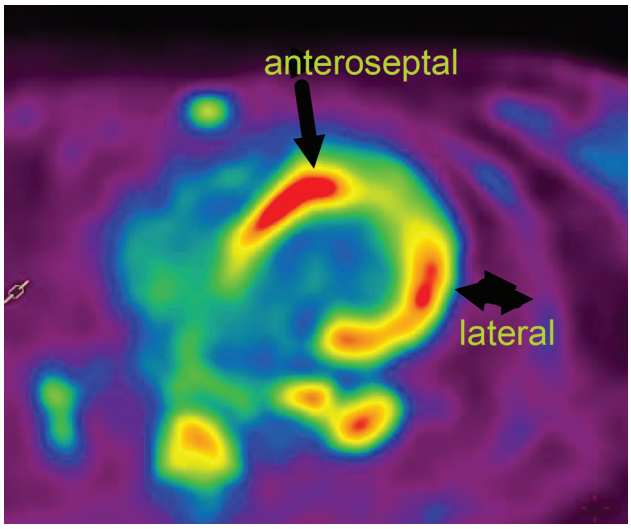
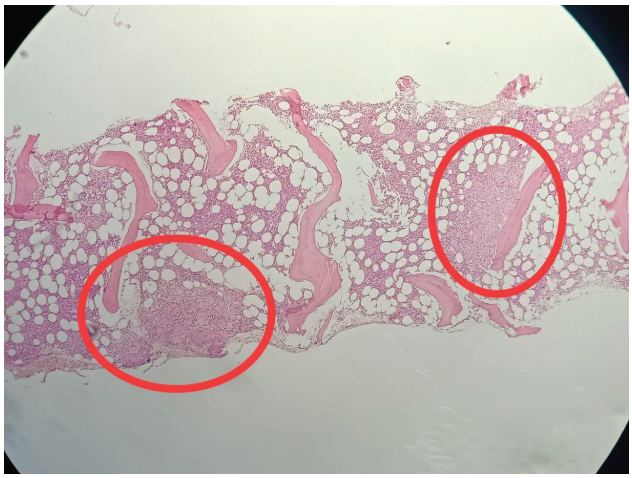


Figure 2 Low power (x10) view of sarcoid granuloma in the bone marrow biopsy section. Focal non-caseating epithelioid cell granuloma (circled) is seen.



For this triad of AV conduction abnormalities, VT and new-onset LV dysfunction, a differential diagnosis of infiltrative cardiomyopathies (sarcoidosis and tuberculosis), tachycardiomyopathy and pacing-induced LV dysfunction were initially considered. Clinical examination did not reveal any palpable lymph nodes or skin lesions. Cardiac MRI was not performed as the patient had an MRI non-conditional pacemaker. A PET scan revealed a heterogenous fluorodeoxyglucose (FDG) uptake pattern in LV myocardium involving the lateral wall, anterior wall and septum (Figure 1). Multiple FDG avid lesions were seen in cervical, thoracic and para-aortic lymph nodes, liver, spleen and bone marrow. These findings were suggestive of inflammatory pathology (sarcoidosis or tuberculosis). A Mantoux test and TB Quantiferon Gold test were performed and found negative. Bone marrow biopsy (Figure 2) showed focal non-caseating epithelioid cell granuloma with Langhans type giant cells supporting the diagnosis of sarcoid infiltration of the bone marrow. Acid-fast bacilli were not found in the stain and culture of the bone marrow biopsy samples. A diagnosis of systemic sarcoidosis with cardiac involvement was made, and the patient was treated with prednisolone initially at 40 mg a day for 3 months. She was advised to have implantable cardioverter defibrillator (ICD) implanted in view of the potential for further ventricular arrhythmic events.

On follow-up, left ventricular ejection fraction (LVEF) improved to 50% over the next 3 months. She had no further sustained VT confirmed on pacemaker interrogation and ambulatory ECG recording.

Discussion

We reported a case of a female in her forties with a permanent pacemaker implanted for AV block, who presented with haemodynamically stable VT and was found to have impaired LV function. Although no other clinical features suggested sarcoidosis, further investigations confirmed cardiac sarcoidosis. The condition was successfully treated with corticosteroids that led to an improvement in LV function and the abolition of recurring sustained VT. This case highlights the need for screening for sarcoidosis in such patients even in the absence of obvious extracardiac manifestation of sarcoidosis.

Screening for cardiac sarcoidosis is recommended in the presence of specific cardiac manifestations even in the absence of clinical evidence of extracardiac sarcoidosis. In a study with 49 patients aged 18–60 years presenting with unexplained Mobitz II- or third-degree AV block with no history of sarcoidosis, 15 patients (32%) were diagnosed with cardiac sarcoidosis.[2] Patients under 60 years of age presenting with unexplained high degree AV block should be screened with high resolution CT scan chest and advanced imaging (cardiac MRI or whole-body fluorodeoxyglucose [FDG]-positron emission tomography [PET]). If imaging is abnormal, an extracardiac biopsy should be performed from the sites suggested by the scan. Alternatively, an image or voltage-guided endomyocardial biopsy should be performed to confirm the diagnosis of cardiac sarcoid. Screening for sarcoid in patients with idiopathic VT (except typical fascicular VT and outflow tract VT) may also be fruitful.[3] For patients with biopsy-proven extracardiac sarcoidosis seen in subspecialty clinics, such as pulmonary or dermatology clinics, it is recommended to screen for cardiac symptoms, and have ECG and echocardiography done.[4] If these are normal, there may not be any need to investigate further. However, if any of these are abnormal, FDG-PET or cardiac MRI should be done to look for cardiac involvement.

This case emphasises the need for screening for sarcoid in a young patient with an unexplained AV block. Also, in patients who develop LV dysfunction after pacing, it is prudent to rule out sarcoidosis as a correctable cause before diagnosing pacing-induced LV dysfunction. Pacemakers also provide a unique opportunity to detect any VT from the stored events. In patients with a pacemaker implanted for unexplained AV block, where stored events show VT, screening for cardiac

sarcoidosis may be important.The Heart Rhythm Society (HRS) Expert Consensus Statement diagnostic criteria include two pathways: a histological diagnosis from myocardial tissue or a clinical diagnosis from invasive and non-invasive studies.[4] In our case, unexplained 2:1 AV block, sustained VT, reduced LVEF, patchy LV myocardial FDG-uptake on PET scan, non-caseating granuloma in the bone marrow biopsy sample, and response to corticosteroids established the diagnosis of cardiac sarcoidosis through the clinical pathway. The involvement of other sites contributed towards the diagnosis of the cardiac sarcoidosis in this case, otherwise, an endomyocardial biopsy would have been necessary. Cardiac tuberculosis may mimic cardiac sarcoidosis, but the former was excluded by the negative results of a Mantoux test and interferon-gamma release assay.

Ventricular arrhythmias in cardiac sarcoid are commonly due to macro re-entry around the granuloma. Treatment of VT involves a stepwise escalation of therapy starting with immunosuppression (activity assessed by FDG-PET), followed by anti-arrhythmic drug therapy, and finally catheter ablation if immunosuppression and anti-arrhythmic therapy have been unsuccessful. In one study of patients with sarcoidosis related VT,[3] the majority of VT were mapped to the right or left ventricular septal wall, or the peritricuspid or perimitral valve area. Diagnosis of cardiac sarcoidosis gives an opportunity to treat the underlying cause of AV block and ventricular arrhythmias. Heart block of any degree can occur in cardiac sarcoidosis due to the predilection of granulomas for the interventricular septum where the conduction system resides. Immunosuppression can be useful in cardiac sarcoid patients with Mobitz II or third-degree heart block. In a recent systematic review,[5] 27 of 57 patients with cardiac sarcoid (47.4%) treated with steroids had improved AV conduction. However, such AV block which reverses on immunosuppression may recur, hence permanent pacemaker implantation is advisable.

The indication of ICD is dictated by risk stratification among patients with cardiac sarcoid.[4] ICD implantation may be considered in patients with cardiac sarcoid and an indication for permanent pacemaker implantation. This is because such patients with enough granulomatous burden to cause high-degree AV block necessitating permanent pacing are also likely to have the substrate for re-entrant VT and therefore should have a device that treats both bradyarrhythmias and tachyarrhythmias. Also, response to cardiac resynchronisation therapy in sarcoidosis may be poorer than in other non-ischaemic cardiomyopathies.[6]

Conclusion

This case report emphasises the need to suspect and screen for cardiac sarcoidosis especially in young patients with unexplained AV block and VT. Subsequent treatment of cardiac sarcoidosis may reverse AV block, control ventricular arrhythmia and improve LV function in such patients. It is important for the physician not to miss the diagnosis of cardiac sarcoid, especially in light of the potential for reversibility of the cardiac abnormalities with appropriate treatment.

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