CASE REPORT

Isolated cardiac sarcoidosis masquerading as right ventricular outflow tract ventricular tachycardia

Auras R Atreya, Mitkumar Patel, Senthil K Sivalingam, Mathias L Stoenescu

University of Massachusetts
Medical School-Baystate,
Springfield, Massachusetts, USA

Correspondence to
Dr Auras R Atreya, aurasatreya@gmail.com

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SUMMARY

A 67-year-old man with coronary artery disease (CAD) and left anterior descending artery (LAD) stent presented with symptomatic monomorphic ventricular tachycardia (VT) at a rate of 190 bpm requiring cardioversion. ECG showed left bundle branch block pattern and inferior axis, suggestive of a right ventricular outflow tract (RVOT) focus rather than left ventricular scar due to LAD territory myocardial infarction (MI). Echocardiography showed normal wall motion. Angiography revealed a patent mid-LAD stent. Cardiac MRI with delayed postcontrast sequence revealed several regions of hyperenhancement abnormality within the basal portion of the interventricular septum. Increased metabolic activity on positron emission tomography confirmed active inflammatory sarcoidosis. Although VTs in patients with prior CAD are likely to be related to either scar or ischaemia, alternative diagnoses (eg, infiltrative disorders, RVOT-VT, arrhythmogenic right ventricular cardiomyopathy) should be considered in patients with an apparent right ventricular focus on ECG.

BACKGROUND

Ventricular tachycardias (VTs) occur in patients with and without structural heart disease. Advanced imaging modalities are important to truly differentiate structural abnormalities in the heart. Infiltrative cardiomyopathies may occur in apparently normal hearts and may masquerade as benign VTs, particularly when the morphology of the VT suggests right ventricular outflow tract (RVOT) focus.

CASE PRESENTATION

A 67-year-old man with history of coronary artery disease (CAD) with primary percutaneous coronary intervention of mid-left anterior descending artery (LAD) for non-ST elevation myocardial infarction 3 years prior, atrial fibrillation, posterior circulation stroke and hypertension presented with 7–8 hours of palpitations, presyncopal symptoms, diaphoresis, nausea and diarrhoea. Paramedics obtained a 12-lead ECG that showed a VT at 190 bpm (figure 1A,B). He underwent synchronised cardioversion, initially with 150 J and subsequently with 200 J, as his VT persisted and systolic blood pressure was in the 60 s (mm Hg). Repeat EKG showed non-specific ST-T wave changes. (figure 1B). Intravenous lidocaine was started and the patient was transferred to our hospital. Serum electrolytes were within normal limits and the patient had an acute kidney injury (blood urea nitrogen 31 mg/dL;

creatinine 1.8 mg/dL). Cardiac biomarkers were mildly elevated (creatinine kinase 139 U/L; troponin-T 0.11 ng/mL).

INVESTIGATIONS

The 12-lead ECG showed monomorphic VT with wide QRS complex, left bundle branch block (LBBB) pattern (indicating origin from the right ventricle) and inferior axis (positive QRS complex in leads II, III and aVF) suggestive of an RVOT focus. Transthoracic echocardiogram showed low normal left and right ventricular systolic function with left ventricular ejection fraction of 50%–55% without regional wall motion abnormalities. Cardiac catheterisation revealed a patent stent in the mid-LAD with minimal irregularities in other arteries. Given mild systolic dysfunction, cardiac MRI (CMR) was performed to rule out infiltrative disorders. Delayed postcontrast sequence revealed several regions of hyperenhancement abnormality within the left ventricle, with the overall pattern favouring scar and/or infiltration (figure 2A–C). The lesions were in the basal portion of the interventricular septum. No features suggestive of arrhythmogenic right ventricular cardiomyopathy (ARVC) were noted. Quantitatively, the left ventricular ejection fraction was 36%. Positron emission tomography (PET-CT) demonstrated patchy areas of increased metabolic activity in the region of the interventricular septum consistent with active inflammatory sarcoidosis. Tissue diagnosis was not sought due to high pretest probability of cardiac sarcoidosis based on clinical presentation and low sensitivity of endomyocardial biopsy.

DIFFERENTIAL DIAGNOSIS

In this patient with a syncopal episode in the past, with known CAD, the first consideration was for myocardial scar related re-entrant VT or ischaemic re-entrant VT in the setting of a new coronary artery stenosis. Since the morphology of the VT suggested an RVOT pattern that could not be explained by his echocardiographic and angiographic findings, idiopathic RVOT-VT, unrelated to scar/CAD, was also on the differential diagnosis. Ultimately, CMR findings were crucial for diagnosis of infiltrative cardiomyopathy. CMR with postcontrast imaging is considered gold standard for detecting scar tissue as it retains gadolinium longer than the normal myocardium. The patterns of delayed hyperenhancement (DHE) are specific for disease processes; DHE in sarcoidosis and myocarditis tends to be

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Figure 1 (A) 12-lead ECG obtained prior to cardioversion shows monomorphic ventricular tachycardia at a rate of around 190 bpm with wide QRS complex, left bundle branch block pattern and inferior axis, suggestive of a right ventricular outflow tract focus. (B) 12-lead ECG obtained in normal sinus rhythm (after cardioversion) shows non-specific ST-T wave abnormalities in the inferolateral leads.

Figure 2 (A) Midventricular short axis view of cardiac MRI, delayed hyperenhancement sequence showing a focal 7×9×20 mm abnormality involving the midwall and outer portion of the mid-left ventricle at the junction of the posterior wall of the right ventricle. (B) Basal short axis view of cardiac MRI, delayed hyperenhancement sequence showing a focal, basal, midseptal defect (arrow) involving the subendocardium and midwall, measuring 3–5 mm in thickness, and 2.7 cm circumferentially at midwall. (C) F-18 FDG-PET and CT scan, axial view. PET imaging reveals patchy increased metabolic activity demonstrated in the region of the interventricular septum (circle). CT, computed tomography; FDG-PET, fluorodeoxyglucose-positron emission tomography; MRI, magnetic resonance imaging; PET, positron emission tomography.

localised, while in amyloidosis it is often diffuse and circumferential. CMR findings excluded other conditions such as ARVC (characterised by fatty infiltration in the ventricles along with dyskinesia of ventricles) and RVOT-VT (usually completely normal CMR or fibrosis/scar in RVOT).

Imaging findings were also consistent with ECG findings. Since the infiltrative focus was within the basal interventricular septum, it generated VT with LBBB morphology (indicating right-sided origin).

TREATMENT

After the clinical diagnosis of cardiac sarcoidosis was established based on mildly depressed left ventricular ejection fraction, VT and CMR findings, treatment was initiated with oral steroid therapy for isolated cardiac sarcoidosis (no other organ system involvement was detected at the time) and an implantable cardioverter-defibrillator (ICD) was implanted for secondary prevention of haemodynamically unstable VT. He was also started on sotalol therapy for suppression of VT focus.

OUTCOME AND FOLLOW-UP

Repeat PET-CT 1 year after initial presentation continued to show increased uptake in the myocardium despite steroid therapy. Additionally, hilar lymphadenopathy with mild uptake was noted suggesting a new diagnosis of pulmonary sarcoidosis. He was continued on low-dose steroid therapy with no repeat ventricular arrhythmia episodes. The patient is doing well nearly 5 years after the initial diagnosis of isolated cardiac sarcoidosis (which subsequently has shown extracardiac involvement).

DISCUSSION

The RVOT is the most common source of VT in patients without structural heart disease, accounting for nearly 70% of cases of idiopathic VT.^{1,2} cAMP-mediated triggered activity and intracellular calcium overload is thought to be the primary mechanism of RVOT-VT.³ On the other hand, infiltrative disorders such as sarcoidosis, amyloidosis, haemochromatosis and ARVC cause VT by facilitating re-entry around the infiltrative focus.

Clinical presentations of cardiac sarcoidosis are wide ranging, from silent disease to sudden cardiac death, and primarily manifest as rhythm disorders or heart failure.^{4,5} Conduction abnormalities due to involvement of basal interventricular septum are perhaps the most common arrhythmias and include first, second and third-degree atrioventricular (AV) block as well as bundle branch blocks.⁵ Sustained VT is the second most common rhythm disorder in cardiac sarcoidosis and has been reported in 23% of patients with cardiac sarcoidosis.⁵ Although monomorphic VT is usually seen, polymorphic VT has also been reported.⁶

Electrophysiological studies in patients with cardiac sarcoidosis have demonstrated that close to 70% of VT episodes are due to re-entry, as in the case of other infiltrative diseases.⁷ Automaticity in the area of granuloma may also contribute to VT initiation.⁷ Either left or right ventricle may be involved, generating right bundle branch block or LBBB pattern, respectively.⁸ Occasionally, cardiac sarcoidosis has been reported to mimic ARVC due to LBBB morphology.⁹

The diagnosis of cardiac sarcoidosis is based on clinical diagnostic criteria.¹⁰ The 2006 Japanese Ministry of Health and Welfare Criteria for Diagnosis of Cardiac Sarcoidosis suggests a diagnosis of cardiac sarcoidosis if the patient has extracardiac sarcoidosis and meets two major criteria (advanced AV block, basal thinning of the interventricular septum, positive gallium-67 uptake in the heart, depressed left ventricular ejection

fraction <50%) or one major criterion plus two minor criteria (abnormal ECG findings, abnormal echocardiographic findings, perfusion defects on nuclear imaging, delayed gadolinium enhancement on CMR, cardiac biopsy findings).¹⁰ On the other hand, the 2014 Heart Rhythm Society Consensus statement places a greater emphasis on histological diagnosis of either cardiac or non-cardiac sarcoidosis to truly make a diagnosis of cardiac sarcoidosis.¹⁰ More recently, the combined use of CMR and PET in the evaluation of suspected isolated cardiac sarcoidosis has been emphasised as the concomitant demonstration of fibrosis and inflammation is attractive and possibly provides the highest diagnostic accuracy.¹¹

Treatment options for cardiac sarcoidosis include antiarrhythmic drug therapy, corticosteroids/immunosuppressive therapy, device therapy radiofrequency ablation, and in few cases cardiac transplantation for refractory VT. Corticosteroid use is universally advocated, but the evidence for corticosteroid use is based on modest data from small retrospective observational studies.⁵ ICD placement is standard practice for patients with cardiac sarcoidosis with sustained VT, while in asymptomatic cardiac sarcoidosis prophylactic ICD implantation is a class IIa indication.¹² Radiofrequency ablation is considered for patients with recurrent VT who fail medical therapy with steroids and antiarrhythmic therapy.

Learning points

- ▶ Ventricular tachycardia (VT) occurring in patients with history of coronary artery disease is usually due to healed scar or ischaemia; however, other diagnoses should be investigated, such as concomitant infiltrative diseases, inflammatory conditions, electrolyte disorders and drug effects.
- ▶ Left bundle branch block pattern QRS complex morphology indicates origination of VT from the right ventricle and is most often indicative of idiopathic right ventricular outflow tract (RVOT)-VT or arrhythmogenic right ventricular cardiomyopathy (ARVC).
- ▶ Cardiac sarcoidosis has been known to present with a variety of arrhythmias, most notably atrioventricular block or sustained VT. If the infiltrate is in the right ventricle or the interventricular septum, it may mimic RVOT-VT or ARVC.
- ▶ Implantable cardioverter-defibrillator (ICD) placement is the routine practice for patients with cardiac sarcoidosis with sustained VT; whereas prophylactic ICD implantation is a class IIa indication for asymptomatic cardiac sarcoidosis

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