CASE REPORT

Ventricular fibrillation via torsade des pointes of cardiac sarcoidosis with preserved left ventricular ejection fraction

Takayuki Sekihara, Eisaku Nakane, Kazutaka Nakasone, Moriaki Inoko

Cardiovascular Center, Tazuke Kofukai Foundation, Medical Research Institute, Kitano Hospital, Osaka, Japan

Correspondence to

Dr Takayuki Sekihara,
t-sekihara@kitano-hp.or.jp

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SUMMARY

Generally, low left ventricular ejection fraction (LVEF) is a risk for ventricular arrhythmia in patients with cardiac sarcoidosis. We present a case of cardiac sarcoidosis with preserved LVEF that evoked ventricular fibrillation (VF). A 73-year-old woman with VF presented to our emergency department. She had a history of ocular sarcoidosis, with gradual thinning of the basal intraventricular septum. LVEF was 62% on the most recent echocardiography. The electrocardiogram after defibrillation showed complete atrioventricular block (CAVB) with QT segment prolongation and frequent ventricular premature beats. VF via torsade des pointes (TdP) was suspected, and temporary intravenous ventricular pacing and magnesium sulfate infusion suppressed her VF. Cardiac sarcoidosis was diagnosed, and an implantable cardioverter defibrillator was implanted. Patients with cardiac sarcoidosis with CAVB are at risk of evoking VF via TdP regardless of LVEF. If cardiac sarcoidosis is suspected, early diagnosis and risk stratification of ventricular arrhythmia are important.

BACKGROUND

Cardiac sarcoidosis is associated with various arrhythmias, including fatal ventricular arrhythmia. In general, patients with ischaemic heart disease or other cardiomyopathy with very low left ventricular ejection fraction (LVEF) have significant risk of

emergence of fatal ventricular arrhythmia.^{1 2} Cardiac sarcoidosis is not an exception.³ Here we present a case of a patient with cardiac sarcoidosis with normal LVEF on the most recent study who experienced ventricular fibrillation (VF) via torsade des pointes (TdP) and emphasise the importance of prevention of sudden cardiac death (SCD) due to ventricular arrhythmia in patients with cardiac sarcoidosis regardless of LVEF.

CASE PRESENTATION

A 73-year-old woman with ocular sarcoidosis was referred to our department for investigation of cardiac involvement in 2011. At the time of referral, none of the findings indicated cardiac sarcoidosis. Her echocardiography showed moderate aortic regurgitation (AR) at initial evaluation (figure 1A, B). During the following 5 years, her basal interventricular septum became subtly thinner (figure 1C, D) and her ECG showed a change to right ventricular bundle branch block (RBBB) with left axis distortion and gradual PR segment prolongation (figure 2A–C). However, no ventricular arrhythmia or complete atrioventricular block (CAVB) had been detected on frequent Holter ECG, and congestive heart failure (CHF) had never occurred. Her LVEF was 62% on echocardiography in September 2015. Although the progression of

Figure 1 Moderate AR demonstrated by echocardiography (A, B). The basal interventricular septum became subtly thinner in September 2015 (D) than in 2011 (C). AR, aortic regurgitation.

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cardiac sarcoidosis had been suspected, further investigations, including cardiac MRI (cMRI) and endomyocardial biopsy, were not performed because she had been completely asymptomatic during these periods.

In April 2016, she had leg oedema and orthopnoea. According to her home doctor, her pulse rate was 52 bpm, which was significantly lower than her usual resting heart rate, which was 81 bpm in November 2015 (figure 2C). After 2 weeks, she had syncope and was referred to the emergency department of our hospital. She did not take any atrioventricular nodal-blocking agents and QT-prolonging drugs. Her initial ECG obtained by the emergency service showed CAVB with ventricular escape beat. Ventricular premature beats (VPBs) were frequently observed. During the ambulance trip, TdP degenerated to VF (figure 3). Although the VF was self-limiting, it recurred in the emergency room and electrical defibrillation was applied. At first, we used intravenous amiodarone to manage the VF storm. After infusing 150 mg of amiodarone, the VF did not recur. Her initial 12-lead ECG showed prominent QT prolongation (figure 4), and frequent VPBs and non-sustaining TdP were still observed. On laboratory test, she showed mild hypokalaemia (3.7 mEq/L) and hypomagnesaemia (1.6 mg/dL). We stopped the amiodarone infusion and started continuous

potassium chloride and magnesium sulfate infusion and temporary intravenous ventricular pacing, which suppressed her VPBs and TdP immediately. She also showed CHF on admission, which was resolved by the aforementioned therapy. Her LVEF was 52% on echocardiography at that point.

OUTCOME AND FOLLOW-UP

The results of the endomyocardial biopsy were concordant with subendocardial granulomatous lesion, and skin biopsy showed epithelioid granulomatous lesion. Late gadolinium enhancement of cMRI showed high-intensity signal in the left ventricular anterior wall (figure 5). LVEF on cMRI was preserved (56%), and right ventricular ejection fraction was decreased to 28%. 18-Fluorodeoxyglucose positron emission tomography (FDG-PET) demonstrated areas of FDG uptake in the interventricular septum, left ventricular free wall and lymph nodes of the mediastinum (figure 6A, B). She received a DDD implantable cardioverter defibrillator (ICD) implantation and was discharged at hospital day 21. No ventricular arrhythmia was detected on ICD records within 1-month after implantation. She is planned to start oral glucocorticoid therapy.

DISCUSSION

Here we presented a case of cardiac sarcoidosis with preserved LVEF that evoked VF. This case clearly demonstrates the risk of SCD in patients with cardiac sarcoidosis with preserved LVEF and the importance of early investigation for potential patients with cardiac sarcoidosis.

In general, low LVEF is a risk factor of fatal ventricular arrhythmia. ICD implantation is recommended as the primary preventive measure in patients with structural heart diseases with LVEF lower than 35%.^{1 2} Patients with cardiac sarcoidosis are no exception, and ICD is recommended for such patients with LVEF lower than 35% or a history of ventricular arrhythmia.⁴ In addition, especially for patients with cardiac sarcoidosis, ICD implantation can be considered for those with LVEF between 35% and 49%.⁴

However, we encountered VF in a patient with cardiac sarcoidosis with an almost normal LVEF on the most recent examination. Our hypothesis regarding the aetiology of her VF is as follows. The crucial factor is the emergence of CAVB. According to her medical history, changes in ECG and echocardiography findings, we suspect her cardiac sarcoidosis had been silently progressing and finally developed into acute sarcoid myocarditis. As a result, CAVB might have occurred several weeks before the VF. Along with the bradycardia caused by CAVB, pre-existing hypertension and moderate AR evoked CHF. In addition, severe QT prolongation with notched T wave (figure 2), which was reported as a risk factor of TdP,⁵ was also observed. These conditions contributed to the induction of frequent VPBs and finally resulted in TdP probably due to the R-on-T phenomenon.

If the final condition was TdP, her symptom might have been milder because TdP is usually a self-limiting arrhythmia. However, her TdP degenerated to VF. Some risk factors that accelerate TdP to VF have been reported,⁶ such as existence of structural heart diseases, low LVEF and length of QT interval. In this case, cardiac sarcoidosis itself and prominent QT prolongation were thought to be the factors that accelerated her TdP to VF. As described above, some pre-existing conditions and the progression of cardiac sarcoidosis can cause VF via TdP even if LVEF is preserved. Therefore, when cardiac sarcoidosis is suspected especially for those with extracardiac sarcoidosis, early investigation for cardiac

Figure 2 ECG obtained in 2011 (A), 2014 (B) and November 2015 (C), showing a gradual change to RBBB with left axis distortion and gradual prolongation of the PR segment. RBBB, right ventricular bundle branch block.

Figure 3 Monitoring electrocardiogram obtained in the ambulance. The TdP degenerated to ventricular fibrillation but was self-limited. Multifocal VPBs were frequently observed. TdP, torsade des pointes; VPBs, ventricular premature beats.

involvement and close follow-up are important even if the patients are totally asymptomatic. According to expert consensus, cMRI or FDG-PET should have been performed at the time of the emergence of RBBB in this case.⁴

To prevent SCD of cardiac sarcoidosis with CAVB, a correction of bradycardia by permanent pacemaker is not enough for patients with cardiac sarcoidosis. Expert consensus of the Heart Rhythm Society recommended that ICD implantation can

be considered for patients with cardiac sarcoidosis who have indication for permanent pacemaker as primary prevention of SCD,³ because patients with active cardiac sarcoidosis have potential risk of ventricular arrhythmia independent of bradycardia-induced TdP when active cardiac sarcoidosis progresses. Our patient already had late gadolinium enhancement lesion on cMRI, which is predictive of ventricular arrhythmia in patients with preserved LVEF.⁷

Figure 4 Twelve-lead electrocardiogram after defibrillation. CAVB with marked QT segment prolongation and notched T wave was observed. CAVB, complete atrioventricular block.

Figure 5 Short-axis section of late gadolinium enhancement on cMRI. A spotty high-intensity signal was observed in the left ventricular anterior wall (yellow arrows). cMRI, cardiac MRI.

Figure 6 FDG-PET demonstrates areas of FDG uptake in the interventricular septum and left ventricular free wall (yellow arrows) (A) and lymph nodes of the mediastinum (B). FDG-PET, 18-fluorodeoxyglucose positron emission tomography.

Herein, we presented a case of VF storm of cardiac sarcoidosis with preserved LVEF. Even with preserved LVEF, severe bradycardia secondary to atrioventricular block can immediately cause VF via TdP. Close observation and early risk stratification for SCD are essential in patients with potential cardiac sarcoidosis. Once CAVB occurs, ICD implantation, not a permanent pacemaker, can be considered for patients with high risk of ventricular arrhythmia.

Learning points

- Patients with cardiac sarcoidosis with normal or near-normal ejection fraction can be at risk for ventricular tachycardia/ventricular fibrillation.
- Patients with extracardiac sarcoidosis should be systematically screened for cardiac involvement.
- Implantable cardioverter defibrillator implantation can be considered for patients with cardiac sarcoidosis with complete atrioventricular block as the primary measure for those with high risk of ventricular arrhythmia.

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