A 16-year odyssey of cardiac sarcoid masquerading as idiopathic premature ventricular contractions and then arrhythmogenic cardiomyopathy

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Introduction

Cardiomyopathies can initially present with ventricular ectopy, which can be difficult to differentiate from idiopathic premature ventricular contractions (PVCs). Distinguishing cardiac sarcoidosis from arrhythmogenic right ventricular cardiomyopathy (ARVC) also can be challenging. We report the case of a patient who presented with benign PVCs that progressed to multiple recurrent ventricular arrhythmias over 16 years, was diagnosed as having ARVC, and eventually was found to have sarcoidosis.

Case report

A 44-year-old man with a diagnosis of ARVC was admitted for management of repetitive monomorphic ventricular tachycardia (VT) terminated by antitachycardia pacing from his implantable cardioverter-defibrillator. The patient's past history was remarkable for a diagnosis of idiopathic PVCs 16 years ago (Figure 1). Over the years, his PVCs had been highly symptomatic. Because the patient was intolerant of multiple medications, including beta-blockers, he underwent repeat electrophysiological studies (EPS) with ablation. Each was followed by symptomatic improvement but subsequent recurrent arrhythmias. Notably, he had no family history of cardiomyopathy or sudden death. The patient exercised routinely, including running, but he was not a competitive athlete. At his third EPS performed 4 years after initial presentation, the right ventricular (RV) voltage map was normal (no areas of electrograms $<$1.5-mV bipolar amplitude), and no sustained VT was inducible with programmed stimulation. Ablation targeted 3 different RV PVCs. Cardiac magnetic resonance imaging showed possible thinning of the anterior RV, but no late gadolinium enhancement was observed. Six years later at the fourth EP study, again performed to ablate symptomatic PVCs, inducible sustained monomorphic VT was found, and a small low-voltage ($<$1.5 mV) area at the RV outflow region was noted. No ablation was performed at this time. Cardiac magnetic resonance imaging showed RV enlargement and severe hypokinesis. Late gadolinium enhancement was observed at the RV base to mid-free wall and mid-inferior wall but not in the LV. An implantable cardioverter-defibrillator was inserted. A positron emission tomography (PET) scan for sarcoid, RV endomyocardial biopsy, and genetic testing for ARVC were unrevealing. The fifth EPS performed 1 year later because of recurrent PVCs and VT revealed no inducible VT. Endocardial and epicardial mapping showed low-voltage areas (Figure 2, bottom), and substrate-guided ablation of these areas was performed. Symptoms improved, but frequent PVCs led to a sixth procedure 1 year later, targeting multiple morphologies of PVCs. The low-voltage endocardial scar was noted to have extended (Figure 2, middle). Symptoms again improved, but occasional symptomatic

KEY TEACHING POINTS

- In its early phase, cardiac sarcoidosis can present with isolated premature ventricular contractions, which makes the differentiation from benign premature ventricular contraction challenging.
- The clinical course of cardiac sarcoidosis can resemble that of arrhythmogenic right ventricular cardiomyopathy (ARVC), evolving over years.
- Myocardial scar in cardiac sarcoidosis can evolve with epicardial predominance, which also resembles ARVC.

KEYWORDS Arrhythmogenic right ventricular cardiomyopathy; Cardiac sarcoidosis; Epicardial mapping; Premature ventricular contraction; Ventricular tachycardia

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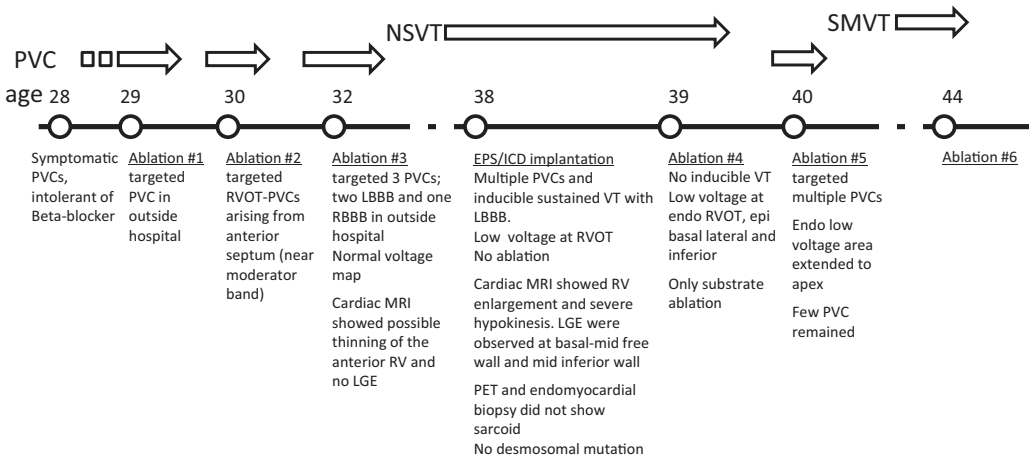

Figure 1 Time line of the disease. The patient had a history of 6 ablation procedures over 16 years with progression of arrhythmia over time. EPS = electrophysiological study; ICD = implantable cardioverter-defibrillator; LBBB = left bundle branch block; LGE = late gadolinium enhancement; MRI = magnetic resonance imaging; NSVT = nonsustained ventricular tachycardia; PET = positron emission tomography; PVC = premature ventricular contraction; RBBB = right bundle branch block; RV = right ventricle; RVOT = right ventricular outflow tract; SMVT = sustained monomorphic ventricular tachycardia; VT = ventricular tachycardia.

PVCs still occurred and preceded the development of increasingly frequent sustained VT.

At the current presentation, the electrocardiogram (ECG) during sinus rhythm showed slight prolongation of the PQ interval of 210 ms, T-wave inversion in the inferior and precordial leads, and low-voltage QRS complexes (Supplemental Figure 1). The ECG of the current VT is shown in Figure 3. Echocardiography revealed significant dilation and wall-motion abnormalities of the RV, but left ventricular size and function were normal.

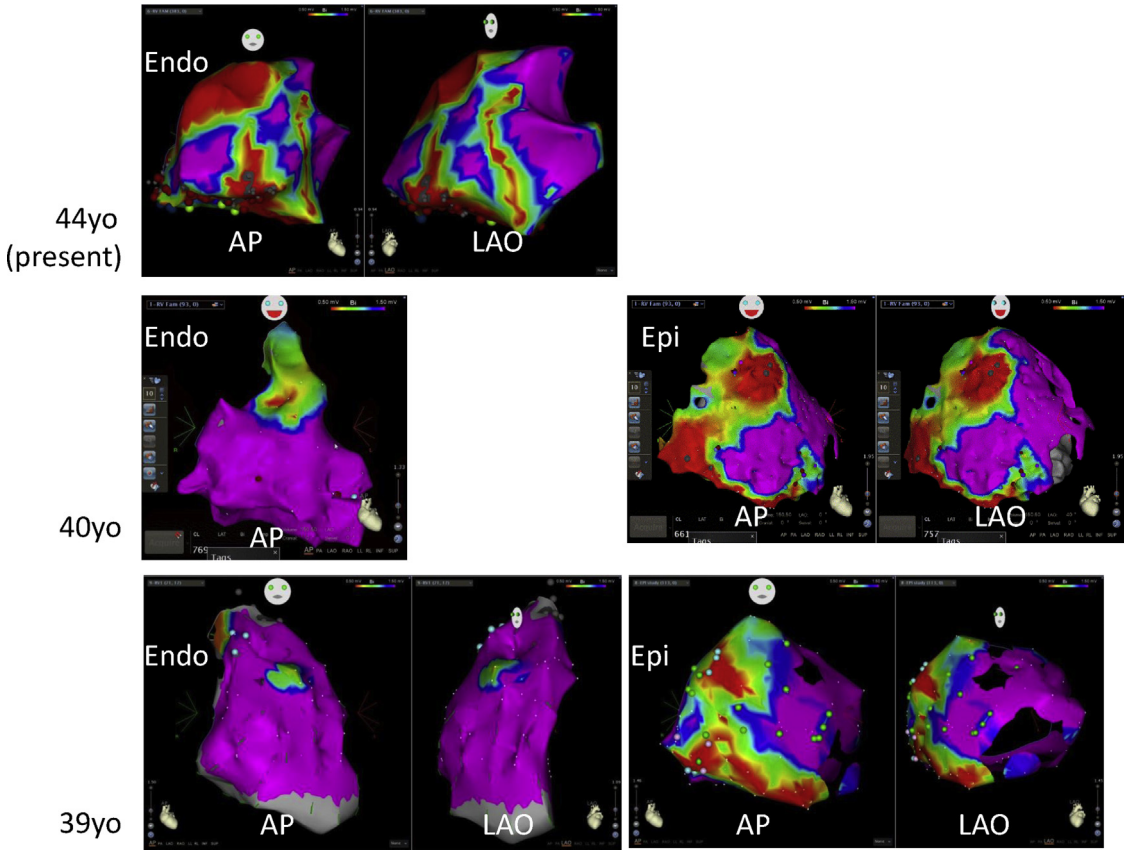

Figure 2 Electroanatomic sinus rhythm voltage maps in anteroposterior (AP) and left anterior oblique (LAO) projections. Normal voltage myocardium (>1.5 mV) is depicted as *purple*, and very low voltage (<0.5 mV) is depicted as *red*. Ablation sites are marked by *red circular tags*. **Top:** Low-voltage scar areas extend from the inferior to anterior free wall and inferobasal aspect of septum superiorly. Those areas showed normal voltage in previous endocardial mapping, whereas epicardial mapping showed low voltage in previous sessions (**middle, bottom**).