Radiofrequency ablation of ventricular tachycardia originating from a lipomatous hamartoma localized in the right ventricle cavity

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Introduction

Ventricular tachycardia (VT) caused by primary cardiac tumors is a rare presentation of this malignant arrhythmia. Management of VT in patients with cardiac hamartomas is especially difficult to control with antiarrhythmic drugs alone, usually requires surgical resection, and imparts a significant risk of sudden cardiac death.[1] In this report, we present a case of VT arising from the recurrence of a ventricular hamartoma 8 years after initial surgical resection, which was successfully treated by radiofrequency ablation (RFA) before placement of an implantable cardioverter-defibrillator (ICD).

Case report

A 55-year-old man with a history of right ventricular lipomatous hamartoma measuring 5.5 cm × 6 cm underwent surgical resection in 2003, following 2 episodes of recurrent, unstable VT. Because the tumor extended into the basal and interventricular septum, only partial tumor resection was undertaken in order to preserve the native conduction system. The patient did well for 8 years, until 2011, when unstable VT was observed in the setting of symptoms of chest tightness, palpitations, and diaphoresis. Urgent synchronized cardioversion was performed after failed attempts at conversion with intravenous amiodarone, but crescendo VT episodes associated with hypotension and syncope recurred, necessitating emergent endotracheal intubation, deep sedation, and mechanical ventilation. After subsequent boluses of intravenous amiodarone and cardioversions, the patient's condition gradually stabilized and sustained sinus rhythm was restored.

Cardiac magnetic resonance imaging demonstrated recurrence of the right ventricular hamartoma (4.7 cm × 4.7 cm × 7 cm) involving the right ventricular septum, but now with extension superiorly to the level of the pulmonary trunk in addition to extension into the right ventricular cavity (Figure 1). Morphologically, the tumor had a wide base and irregular shape. Additionally, mass effect from the tumor resulted in displacement of the interventricular septum (IVS) toward the left ventricular cavity, as well as slight compression of parts of the aortic root, left ventricular outflow tract (LVOT), and pulmonic valve. Echocardiographic imaging revealed evidence of slight right ventricular outflow tract (RVOT) stenosis due to the space-occupying tumor, as well as postoperative changes of the IVS and moderate pulmonic valve regurgitation, which made consideration of surgical reintervention technically challenging.

Despite use for initial stabilization, long-term antiarrhythmic therapy was thought to be ill advised due to concerns about toxicities and likely ultimate ineffectiveness. Additionally, the patient refused surgical reintervention for tumor resection, given the inherent surgical risks and his history of recurrence after prior intervention. Therefore, we decided to offer RFA under 3-dimensional (3-D) guidance before placement of an ICD.

Ventricular tachycardia mapping and ablation

Antiarrhythmic drugs were withdrawn 5 half-lives prior to ablation. The supine patient was prepped and draped in the usual fashion and catheters were placed under local anesthesia through the left subclavian vein into the coronary sinus, and through the femoral veins into the right ventricular apex and His-bundle position. A Navistar ablation catheter was used in conjunction with a CARTO 3-D mapping system (both Biosense Webster, Diamond Bar, CA) for mapping and ablation.

Two morphologies of premature ventricular contractions (PVC) were observed to occur spontaneously: PVC-1 had a left bundle branch block, inferior axis morphology, and

KEY TEACHING POINTS

- Therapy of ventricular tachycardia (VT) induced by primary cardiac tumors has rarely been reported.

- Radiofrequency ablation (RFA) can be an alternative therapy for VT in patients with cardiac hamartomas who are not eligible for, or who choose to forego the risks of, surgical and/or repeat surgical interventions.

- RFA can even help to reduce the tumor size, the so-called "debulking" effect.

12-lead electrocardiogram (ECG) characteristics similar to VT-1, suggesting an RVOT or aortic cusp site of origin (SOO) (Figure 2A). PVC-2 had a right bundle branch block, inferior axis morphology, and 12-lead ECG appearance suggestive of LVOT origin.

Burst pacing from the right ventricular apex at 150 beats per minute easily induced 2 morphologies of VT: VT-1 shared morphologic similarities with PVC-1 and readily transitioned to VT-2 during nonsustained episodes lasting 10 seconds to 2 minutes in duration (Figure 2B). VT-2 had a right bundle branch block, inferior axis morphology with a wide (>160 ms), notched QRS complex suggestive of an epicardial SOO. VT-2 exhibited subtle beat-beat changes in morphology (Figure S1, available online) and shared 12-lead ECG characteristics similar to the clinically observed sustained VT

(Figure 2C). Activation mapping in the septal RVOT identified local activation time preceding PVC-1 onset by 37 ms and pace mapping identified an 11/12 ECG match. In addition, at an adjacent site, local activation during VT-1 preceded the endocardial QRS onset by 52 ms with a QS complex on unipolar electrograms. Ablation was performed in a linear fashion connecting these 2 SOO at 30 W, 43°C, and 17 cc/min flow for 60–90 seconds per lesion (Figure 3A and B). Immediately following creation of this ablation line, no further spontaneous episodes of PVC-1 or induction of VT-1 were observed. Left ventricular endocardial mapping of VT-2 failed to identify sufficiently early local activation times. Therefore, based on combined anatomic 3-D computed tomographic imaging of the tumor and electrocardiographic activation mapping of the arrhythmias, it was hypothesized that the tumor, infiltrating the high IVS, conducted the critical reentrant wavefront of activation from the RVOT to the right ventricle (VT-1) in one direction, and from the epicardium to the left anterior free wall (VT-2) in another, with continuous, slight changes of endocardial conduction exit. Therefore, local activation time was mapped to the inflexion point of the left ventricular interval on the QRS complex of the surface ECG as a surrogate for endocardial activation timing. The earliest site of local endocardial left ventricular activation was identified in the anterior free wall and preceded the left ventricular interval by 20–30 ms. Detailed mapping was conducted surrounding the target site and a broader area of similar 20–30 ms local activation times and similar QRS morphologies during pace mapping was identified. Ablation lesions were delivered for 60–90 seconds per lesion at 30–35 W, 40°C, and 17–25 cc/min flow in this area, for a total ablation time of 900 seconds
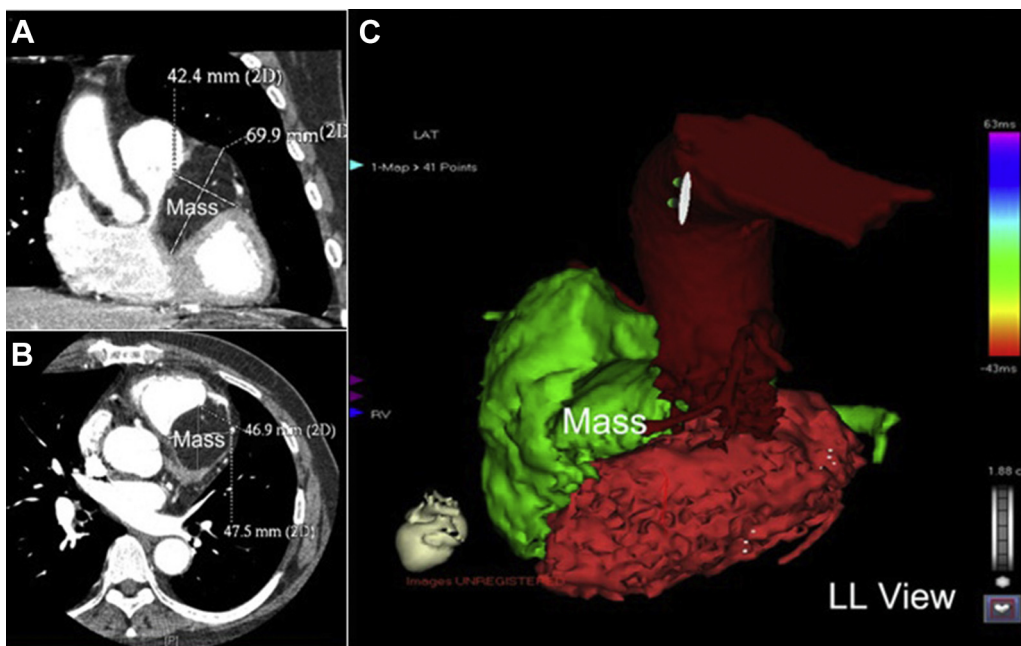
Figure 1 Images of right ventricular hamartomas. **A:** Coronal image of cardiac spiral 2-dimensional computed tomography. **B:** Axial image of cardiac spiral 2-dimensional computed tomography. **C:** Three-dimensional CARTO composite image. The right ventricular hamartoma in the upper side of the ventricular septum (4.7 cm × 4.7 cm × 7 cm) extended upward to the level of the pulmonary trunk and partially grew into the right ventricle cavity. The left ventricular cavity, parts of the aorta and outflow tract, and valve region of the pulmonary artery were slightly compressed.
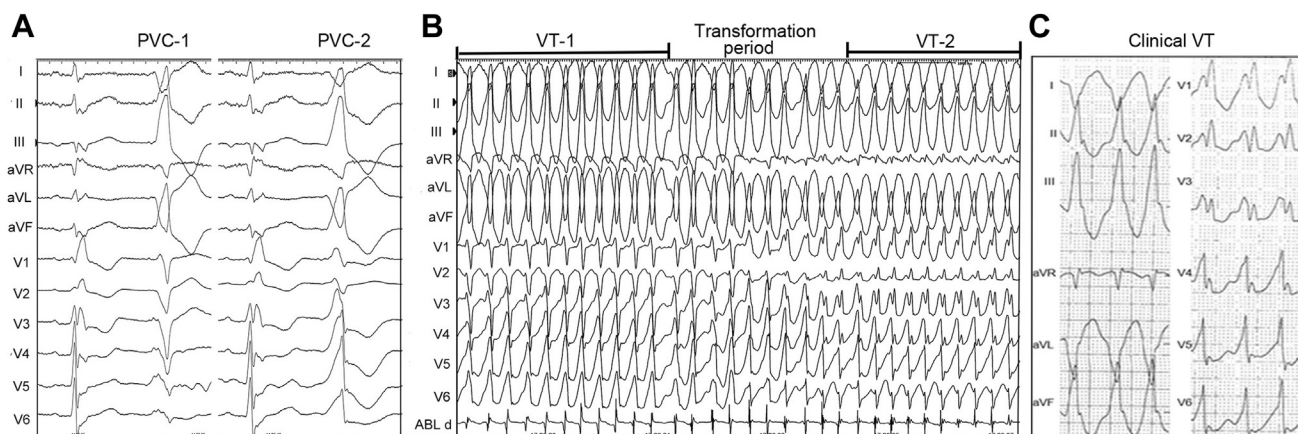
Figure 2   Twelve-lead electrocardiogram of (**A**) premature ventricular contraction (PVC)-1 and PVC-2; (**B**) induced ventricular tachycardia (VT)-1 transforming to VT-2; and (**C**) recorded clinical VT. PVC-1, PVC-2, and clinical VT occurred spontaneously; VT-1 and VT-2 could be induced and transformed between each other. The morphology of PVC-1 and VT-1 is similar, with a left bundle branch block, inferior axis morphology consistent with a site of origin (SOO) in the right ventricular outflow tract. The morphology of PVC-2 and VT-2 is consistent with SOO from the left ventricular outflow tract or left ventricular free wall and the >160 ms width of QRS suggests an epicardial circuit. VT-2 is similar in morphology to the clinical VT.

and a T-shaped, linear length of about 2 cm (Figure 3C and D). No further VT could be induced following ablation, despite utilization of a more extensive programmed stimulation protocol (Figure S2, available online). The patient received a secondary-prevention ICD and treatment with combined metoprolol (23.75 mg daily) and amiodarone (200 mg daily) therapy prior to discharge. Interrogation of the ICD during 5 years of follow-up demonstrated no discharges and no recorded VT episodes. The patient denied symptoms of palpitations, chest tightness, or diaphoresis. A postablation echocardiogram performed 1 year after ablation showed the cardiac tumor to be smaller in size (3 cm × 4 cm), with improvement of the pulmonic stenosis.

Discussion

Cardiac hamartomas are exceedingly rare, with a reported incidence of 0.001%–0.03%.[1] These tumors occur as aberrations in normal cardiac tissue development and can vary histologically as fibromatous,[2] mature myocardial,[3] vascular,[4] lipomatous,[5] or mesenchymal tissue types.[6] Lipomatous hamartomas are unusual and extremely rare lesions (less than 0.0001% among the population). Described by Inoue and colleagues,[7] they define a lesion composed of irregularly distributed mature fat and muscle cells throughout the cardiac valves or the interatrial septum. Information concerning these lesions has accumulated largely from single-patient case reports, most of which relate nonspecific clinical presentation or, in some cases, no symptoms at all. Thus, clinical diagnosis is difficult and most are detected incidentally at autopsy or during echocardiographic examination in evaluation of a cardiac murmur or a nonspecific symptom. However, among symptomatic patients, PVCs, presyncope, and dyspnea were the most frequent presenting complaints. The mitral and tricuspid valves were the most commonly encountered sites of origin for clinically relevant tumors, such that surgical

resection is the recommended treatment, especially given its good short-term prognosis.[8] However, longer-term follow-up is rarely reported.

In this case, we reported long-term recurrence of a cardiac hamartoma that had been incompletely surgically resected 8 years prior to clinical presentation of tumor recurrence. Resection was limited by tumor invasion into the right IVS in an effort to preserve intrinsic cardiac conduction. Although the correlation between postoperative scar and VT cannot be completely ruled out, we suggest that tumor recurrence was the substrate for the clinical VT, not scar, since the VT was not observed clinically during shorter-term follow-up.

This case is also unique in illustrating the utility of RFA in treating VT in patients with cardiac hamartomas who are not eligible for, or who choose to forego the risks of, surgical and/or repeat surgical interventions. Additionally, ICD therapy, though strongly advised, may not be acceptable to some patients, or might be fraught with undesirable discharges. Further, antiarrhythmic therapy alone may not be completely effective in preventing VT. In these instances, as demonstrated by this report, RFA might also be considered a therapeutic option.

Lastly, beyond the observed antiarrhythmic benefits in terms of elimination of VT, a reduction in tumor size was observed following RFA. It was hypothesized that this "debulking" was due to scarring of the matrix of the tumor from the saline-cooled ablation lesions, as has been described for other, noncardiac tumors, such as metastatic tumor to the liver, the hypertrophic pulmonary osteoarthropathies associated with pulmonary carcinoma, etc.[9,10] Similarly, RFA has been shown to reduce interventricular septal wall thickness, significantly reduce LVOT gradients, improve symptoms, and increase efficacy rates in patients with hypertrophic obstructive cardiomyopathy.[11]
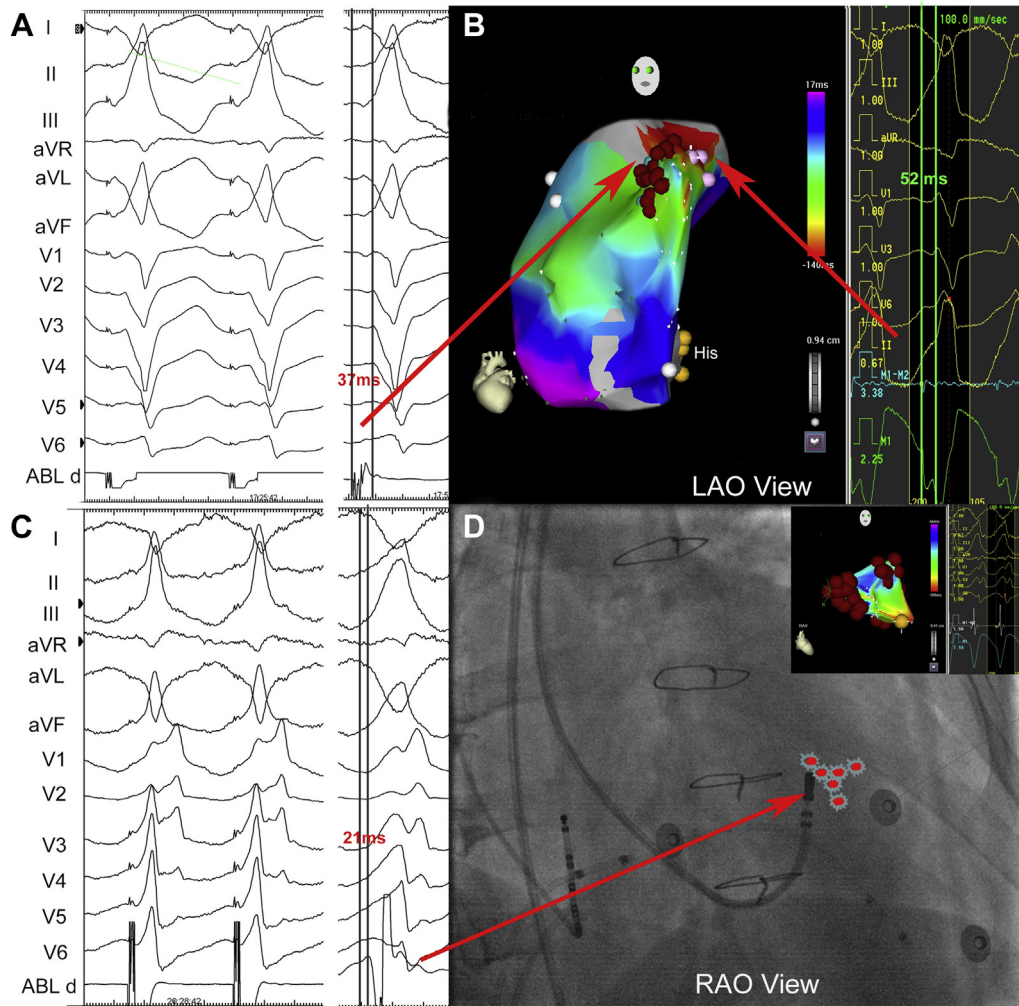
Figure 3   **A:** Pace and activation mapping of premature ventricular contraction (PVC)-1. **B:** CARTO image of PVC-1 and ventricular tachycardia (VT)-1 (with activation map in right panel); PVC-1 (brown tags) and VT-1 (pink tags) shared adjacent site of origin (SOO) in the anteroseptal right ventricular outflow tract. PVC-1 had -37 ms local activation time (LAT) at SOO with 11/12 electrocardiogram pace-map match. VT-1 had -52 ms LAT with QS unipolar morphology at SOO. A linear ablation line of 1 cm was created between the SOO for PVC-1 and VT-1. **C:** Pace and activation mapping of VT-2. **D:** CARTO and fluoroscopic images of PVC-2 and VT-2. LAT was only -20 to 30 ms over a diffuse area in the left ventricular anterior free wall, but pace mapping revealed a reasonable enough similarity to VT-2 (C) that a 2-cm T-shaped linear ablation line (*arrow*) was created at what was suspected of being an area of endocardial breakthrough for VT-2.

Conclusion

VT induced by primary cardiac tumors is very rare. Patients with inducible VT have a higher risk of sudden cardiac death. In this case, RFA proved to be an effective alternative therapy for a patient who suffered from unstable VT associated with a cardiac hamartoma.

Appendix
Supplementary data

Supplementary data associated with this article can be found in the online version at http://dx.doi.org/10.1016/j.hrcr.2017.04.001.

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