A Case Study on Cardiac Imaging in Patients with Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy—A Comparison between 64-Slice Computed Tomography, Magnetic Resonance Imaging and Electroanatomical Mapping—

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Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is an uncommon type of cardiomyopathy with fibrofatty tissue replacement. Magnetic resonance imaging (MRI) is a sophisticated method for the diagnosis of ARVD/C. Electroanatomical mapping has been reported to rapidly provide accurate data that facilitates catheter ablation of VT in ARVD/C. In addition, multislice CT (MSCT) is fast, easy to perform, and has a more reliable image quality. MSCT is considered a clinically valuable, noninvasive tool for assessment of myocardial pathology. However, the relationship between MSCT, cardiac MRI and electroanatomical mapping imaging in patients with ARVD/C is unknown. We report two cases diagnosed as ARVD/C by endomyocardial biopsy. In one case MRI imaging revealed marked wall thinning and a decrease in wall motion in the inferior wall and basal interventricular septum of the left ventricle. In another case an electroanatomical map of the RV was created. Endocardial voltage mapping in sinus rhythm revealed a large low voltage area and a focal pattern of activation was documented in activation mapping during VT. In both cases MSCT demonstrated low density areas indicative of focal fatty infiltration and morphological and functional abnormalities which were simultaneously assessed with MRI or electroanatomical mapping.

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Key words: ARVC/D, Multislice CT, MRI, Sudden cardiac death, Radiofrequency catheter ablation

Introduction

Arrhythmogenic right ventricular dysplasia/cardio-myopathy (ARVD/C) is an uncommon type of cardio-myopathy with fibrofatty tissue replacement. This disease is one of the causes of sudden cardiac death among young people, particularly in athletes. Magnetic resonance imaging (MRI) is a sophisticated...
method for the diagnosis of ARVD/C. Electro-anatomical mapping has been reported to provide a rapid and accurate mapping that facilitates catheter ablation of VT in ARVD/C. In addition, multislice computed tomography (MSCT) is fast, easy to perform, and has a more reliable image quality. MSCT is considered a clinically valuable, noninvasive tool for assessment of myocardial pathology. However, the relationship between MSCT, cardiac MRI and electroanatomical mapping imaging in patients with ARVD/C is unknown.

Case Report

Case 1

A 69-year-old man with a history of intermittent atrial fibrillation and hypertension was admitted to hospital because of sustained ventricular tachycardia (VT) with right bundle branch block morphology (Figure 1). He had no family history of cardiomyopathy or sudden death. After abolition of VT, an electrocardiogram showed atrial fibrillation, complete right bundle block, and abnormal Q wave in III and ST depression in V4-V6 leads.

Contrast-enhanced MSCT (Aquilion 64, Toshiba Medical Systems Corporation) demonstrated low density areas (−70 to −5 HU) indicative of focal fatty infiltration in the anterior wall of the right ventricle (RV) outflow tract and RV trabeculations along the right ventricular side of the interventricular septum (Figure 1, 2). Significant right ventricular dilatation was not observed. In addition, conspicuous trabeculations with low attenuation (−10 HU) and a wedge-shaped low density area (−50 to −60 HU) in

Figure 1
Panel A: Sustained VT with right bundle branch block morphology (Case 1). Panel B: ECG after abolition of VT showed atrial fibrillation, complete right bundle block, and abnormal Q wave in III and ST depression in V4-V6 leads (Case 1). (Continued)
the left ventricle (LV) myocardium were observed. Cinematic display of basal short axis images reconstructed from the same CT data set revealed marked wall thinning and wall motion abnormalities in the LV inferior wall and interventricular septum. A localized ventricular aneurysm was noted in the inferior wall of the RV. Cardiovascular MRI was performed with a 1.5-T whole body scanner (Symphony, Siemens Medical Systems). Cine MRI imaging using true fast imaging with steady-state precession sequence also revealed marked wall thinning and decrease in wall motion in the inferior wall and basal interventricular septum of the LV as indicated by MSCT imaging (Figure 2). A localized ventricular aneurysm was also noted in the inferior wall of right ventricular base as indicated by MSCT imaging (Figure 2).

An endomyocardial biopsy specimen from the RV disclosed extensive replacement of the myocardium with fibroadipose tissue and the patient was diagnosed with ARVD/C1) (Figure 2).

We also performed ventriculography and coronary angiography. Right ventriculograms revealed an RV ejection fraction of 29.5% and a systolic bulge of reduced RV wall motion and diffuse infero-septal aneurysm. Left ventriculograms revealed an LV ejection fraction of 25.7% and a systolic bulge of the infero-septal LV wall with reduced motion and aneurysm. Angiography revealed normal coronaries, suggesting LV involvement (Figure 1).
The patient did not agree to implantable cardioverter defibrillator (ICD) implantation and radiofrequency (RF) catheter ablation and suffered from sudden cardiac death one year after discharge.

Case 2
A 35-year-old man was admitted to hospital because of sustained VT with left bundle branch block morphology. He had no family history of cardiomyopathy or sudden death. After abolition of VT, an electrocardiogram showed complete right bundle block with $\xi$ wave in V1 and V2.

We also performed ventriculography and coronary angiography. The RV ejection fraction in right ventriculograms was 52.3% and the LV ejection fraction in left ventriculograms was 72.4%. An
Endomyocardial biopsy specimen from the RV disclosed extensive replacement of the myocardium with fibroadipose tissue and the patient was diagnosed with ARVD/C\(^1\) (Figure 3).

Contrast-enhanced MSCT (Aquilion 64, Toshiba Medical Systems Corporation) also demonstrated low density areas (−70 to −5 HU) in the anterior wall of the right ventricular outflow tract and the inferior wall of the right ventricular base (Figure 4).

Endomyocardial biopsy specimen from the RV disclosed extensive replacement of the myocardium with fibroadipose tissue and the patient was diagnosed with ARVD/C\(^1\) (Figure 3).

Contrast-enhanced MSCT (Aquilion 64, Toshiba Medical Systems Corporation) also demonstrated low density areas (−70 to −5 HU) in the anterior wall of the right ventricular outflow tract and the inferior wall of the right ventricular base (Figure 4).

Significant right ventricular dilatation and LV involvement was not observed. Because he suffered from incessant VT, we performed RF catheter ablation. Three-dimensional endocardial activation maps of the RV were created with the CARTO system and NAVI-STAR catheter (Biosense-Webster, Diamond Bar, CA, USA). Endocardial voltage mapping in sinus rhythm revealed a large low voltage area in the base of the RV (Figure 4). A
Figure 4
Panels A through D: Series of representative axial CT images from right anterior oblique view illustrated low density areas (−120 to −10 HU), suggesting adipose tissues in the anterior wall (white arrows) and along the base of the right ventricle (white circles).

Three-dimensional endocardial activation maps of right ventricle was created with the CARTO system and NAVI-STAR catheter (Biosense-Webster, Diamond Bar, CA, USA). Endocardial voltage mapping of three-dimensional endocardial activation maps of the RV in sinus rhythm revealed a large low voltage area in the base of the right ventricle (panel E). A focal pattern of activation which showed a wavefront radially spreading from an earliest activation site from the base of the RV was documented in activation mapping during ventricular tachycardia (panel F).
focal pattern of activation which showed a wave front radially spreading from an earliest activation site at the base of the RV was documented in activation mapping during VT. The range of activation times was considerably less than the tachycardia cycle length (Figure 4). After RF catheter ablation the patient underwent ICD implantation.

Discussion

We report two cases which were diagnosed ARVD/C by endomyocardial biopsy with RV aneurysm (Case 1) or wave of electrocardiogram (Case 2).1

In Case 1 MRI revealed marked wall thinning and decrease in wall motion in the inferior wall and the basal interventricular septum of the left ventricle. Contrast-enhanced MSCT demonstrated low density areas indicative of focal fatty infiltration not only in the anterior wall of the right ventricular outflow tract and along the right ventricular side of interventricular septum but also in the inferior wall and the basal interventricular septum of the LV. In this case the patient suffered from VT with right bundle branch block and superior axis morphology, which indicated the origin of VT was the inferior wall of the LV.

Because RV dilatation and LV involvement was associated with the risk of SCD in patients with ARVC/D, morphological evaluation is highly useful in risk stratification and prevention of SCD, and MRI was demonstrated to be useful for noninvasive diagnostic imaging.5-14 Compared with MRI, it was reported that MSCT had demonstrated excellent spatial and temporal resolution which could provide anatomical and functional change in ARVC/D.15 Bomma et al. emphasized 3 main attributes of MSCT in the evaluation of patients with ARVD/C.16 First, MSCT detected many characteristic abnormalities of the right ventricle. Second, MSCT provided quantitative assessment of RV size. Third, cine evaluation images could provide visual assessment of contractile function. They also demonstrated in quantitative analysis of ventricular function the data set of MSCT were similar to values obtained from cardiac MRI. In this case MSCT demonstrates more detailed spatial and temporal resolution than MRI, which revealed many characteristic abnormalities and cine evaluation images, and assisted the diagnosis of ARCV/D and the estimation of the risk of SCD.

In Case 2 an electroanatomical map of the RV was created. Endocardial voltage mapping in sinus rhythm revealed a large low voltage area in the base of RV and a focal pattern of activation from the base of RV also was documented in activation mapping during VT. Contrast-enhanced MSCT also demonstrated low density areas in the inferior wall of the right ventricular base, which involved the epicardial myocardium. In this case the patient suffered from VT with left bundle branch block and superior axis morphology, which indicated the origin of VT existed in the inferior wall of the right ventricular base. It may be inferred that the origin of VT exited in epicardial myocardium of RV because MSCT demonstrated low density areas in epicardial myocardium of the base of RV and epicardial mapping may be needed for further examination.17,18

RF ablation has been performed in selected patients for VT in medically refractory patients.19 It was reported electroanatomical mapping was valuable in identifying the VT mechanism, and in guiding RF ablation in patients with ARVD/C.4 However, the correlation of MSCT and electroanatomical mapping was not clear. In this case the low density area which indicates focal fatty infiltration was visualized along the base of the RV in MSCT, which was the same location as the low voltage area in SR and the earliest activation site of VT in electroanatomical mapping. Merging MSCT and electroanatomical mapping was reported to be useful for substrate-based mapping of scar-related complex ventricular arrhythmias.18

Compared with MRI, MSCT shows excellent spatial and temporal resolution, which could provide anatomical and functional change in ARVC/D. Moreover MSCT can be used even after ICD implantation. Collaboration and merging MSCT and electroanatomical mapping may be useful in RF treatment of VT in patients with ARVC/D. Further study may be needed because there are few reports demonstrating the relation and comparison between MSCT, cardiac MRI and electroanatomical mapping in patients with ARVD/C.

Abbreviations

ARVD/C = arrhythmogenic right ventricular dysplasia/cardiomyopathy, MRI = magnetic resonance imaging, MSCT = multislice computed tomography, VT = ventricular tachycardia, RV = right ventricle, LV = left ventricle, ICD = implantable cardioverter defibrillator, RF = radiofrequency

References


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