CASE REPORT

Arrhythmogenic Right Ventricular Cardiomyopathy with Multiple Thrombi and Ventricular Tachycardia of Atypical Left Branch Bundle Block Morphology
Shenzhen Gong,1 MD, Xin Wei,1 MD, Guyue Liu,1 MD, Feng Wu,2 MD and Xiaoping Chen,1 MD

Summary
A 61-year-old male patient was admitted to our hospital with recurrent palpitations and syncope. Electrocardiography, echocardiography, and contrast-enhanced computed tomography were performed. The patient was diagnosed with arrhythmogenic right ventricular cardiomyopathy (ARVC) complicated by multiple thrombi, and ventricular tachycardia (VT) without typical left bundle branch block (LBBB) morphology. This case suggests that VT is not always the sole contributor to syncope and death in patients with ARVC, and pulmonary embolism should be considered. Furthermore, VT with typical LBBB morphology is not an absolute necessity as a major criterion for the diagnosis of ARVC when the right heart is extremely enlarged.

Key words: Ventricular arrhythmia

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a rare inheritable myocardial disorder pathologically characterized by myocardial atrophy in the right ventricle (RV), with fibrofatty replacement and age-dependent left ventricle (LV) involvement. The diagnosis of ARVC relies on the major and minor criteria in the 2010 European Society of Cardiology revised Task Force Criteria1 including RV imaging findings, myocardial histology, arrhythmias diagnosis, family history, and abnormalities of repolarization and depolarization on ECG. ARVC with excessively dilated right heart and severely diminished systolic function, sometimes mistaken for dilated cardiomyopathy, may lead to refractory congestive heart failure and fatal arrhythmia. The major item of arrhythmia diagnosis is non-sustained or sustained ventricular tachycardia (VT) of left bundle-branch morphology (LBBB) with superior axis.2 We herein report a case of ARVC complicated by multiple thrombi, and VT without typical LBBB or right bundle branch block (RBBB) morphology.

Case Report
A 61-year-old male patient was admitted to our hospital on an emergency basis with acute chest pain, deterioration dyspnea, recurrent palpitations, and syncope. Physical examination revealed blood pressure of 109/79 mmHg, a regular heart rate of 56 bpm, a beating jugular, enlarged heart border bilaterally, and a systolic grade 3/6 murmur at the third and fourth left parasternal border most obviously. Twelve-lead electrocardiography (ECG) (Figure 1B) in sinus rhythm showed pathological Q waves on inferior wall leads, poor R-wave progression on chest leads, epsilon waves in V3, first degree atrioventricular block, and intraventricular block. The cardiac troponin T (cTnT) level was elevated moderately at 475.8 ng/L. The levels of fibrinogen degradation products (FDP) and D-dimer were markedly increased (20.8 mg/L, 14.57 mg/L respectively). Therefore, a diagnosis of myocardial infarction was favored. However, emergency coronary angiography excluded coronary heart disease.

Episodes of symptomatic sustained VT were recorded with the morphology of down-direction end-waves from V1 to V6 leads, originating from RV (Figure 1A and 1C). Furthermore, transthoracic echocardiography showed severe hypokinesis and extreme dilatation of the right atrium (RA) at 81 mm, RV at 53 mm and right ventricular outflow tract at 31 mm, extreme thinning and excessive trabeculations of the RV free wall, fractional area change (FAC) of 6% in the RV, and a normal-sized left heart. Additionally, massive thrombi at the RV posterior wall (40 × 32 mm) and right atrial appendage (RAA) (22 × 17 mm) were detected. Suspecting probable subsequent pulmonary embolism, we performed contrast-enhanced computed tomography (Figure 2) and confirmed multiple emboli in the right upper and inferior pulmonary arteries, and their branches, and thrombus formation in the RAA. Based on the clinical picture, the patient presented here was diagnosed with ARVC associated with thrombi and syncope, after differential diagnoses were made morphologically in-
In the heart J. May 2018 653

ARVC WITH MULTIPLE THROMBI AND VT OF ATYPICAL LBBB

Figure 1. 12-lead electrocardiogram. A: Ventricular tachycardia with negative-direction end-wave of leads V1 and V6, rather than typical left bundle branch block morphology, seeming to originate from right ventricular outflow tract. B: Sinus rhythm with pathological Q waves in inferior wall leads, poor R-wave progression on chest leads, Epsilon waves in V3 (red arrows), first degree atrioventricular block, and intraventricular block with prolonged QRS complex. C: Ventricular tachycardia with negative-direction end-wave from V1 to V6, seeming to originate from right ventricular apex.

Discussion

It is frequently reported that VT of LBBB morphology in ARVC reflects origins from the RV, and sporadic VT of RBBB morphology represents left ventricle involvement. 1) Typical LBBB morphology manifests negative-direction end-waves on the V1 lead and positive-

including Ebstein’s anomaly, atrial septal defect, pulmonary disease, and tetralogy of Fallot in echocardiography, though the patient had a negative family history and refused cardiac magnetic resonance or endomyocardial biopsy after sufficient anticoagulant therapy and disappearance of the thrombi.
Figure 2. Contrast-enhanced computed tomography scan of pulmonary artery with 3-dimensional reconstruction. A and B: Multiple embolism in both right upper and inferior pulmonary arteries, even in branches (white arrows), and thrombus formation in RAA (red arrow). C: Extremely enlarged right heart, irregular clockwise rotation, and extended trabeculations (white arrow). Furthermore, right heart occupies anterior and lateral chest wall, and left heart is being pressed upon.

direction end-waves on the V6 lead. However, VT without LBBB or RBBB morphology could also be identified at standard lead sites, because an extremely enlarged right heart leads to irregular clockwise rotation and displaced left heart, so that leads V1-V6 record electrical activity over RV (Figure 2C), like this case. Negative-direction end-waves on the V1 lead reflect that VT originates from the RV, which can be called atypical LBBB morphology. To the best of our knowledge, this is the first reported case of VT with atypical LBBB morphology in ARVC, and it suggests that VT with typical LBBB morphology is not an absolute necessity as a major criterion for the diagnosis of ARVC. Furthermore, only 2-4% of patients are diagnosed with intracardiac thrombosis\(^5\) and literature data concerning ARVC associated with massive and multichamber intracardiac thrombosis is very rare. Thrombolytic and anticoagulation therapy, therapy for heart failure, and implantable cardioverter defibrillator are important after disappearance of the thrombi. Furthermore, ultrasound accelerated thrombolysis\(^5\) or veno-arterial extracorporeal membrane oxygenation\(^6\) can be effective treatments for submassive or massive pulmonary embolism. The present case demonstrates that VT is not always the sole contributor to syncope and death in patients with ARVC, and pulmonary embolism should be considered. At the end stage of ARVC, an extremely enlarged right heart could lead to irregular clockwise rotation and subsequent VT with atypical LBBB morphology.

Disclosures

Conflicts of interest: The authors declare no conflicts of interest.

References