Massive Right Atrial Thrombus Formation Followed by an Atrial Flutter with 1:1 Atrioventricular Conduction in a Patient with Arrhythmogenic Right Ventricular Cardiomyopathy

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Abstract

A 46-year-old man was admitted to our hospital for near syncope and palpitations. An electrocardiogram showed a common type of atrial flutter (AFL) with 1:1 atrioventricular conduction. Transthoracic echocardiography revealed a massive right atrial (RA) thrombus with a huge RA and right ventricle. The patient was diagnosed with arrhythmogenic right ventricular cardiomyopathy. It was difficult to control the heart rate with beta-blockers during AFL, which resulted in the deterioration of right-sided heart failure. The effect of anticoagulation therapy for the RA thrombus was also limited. Restoration to sinus rhythm by catheter ablation effectively improved the right-sided heart failure, and the massive RA thrombus eventually disappeared.

Key words: atrial thrombus, arrhythmogenic right ventricular cardiomyopathy, atrial flutter

Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited myocardial disease that causes dilatation of the right ventricle (RV) and RV wall motion abnormalities. The disease is characterized by a progressive replacement of the RV myocardium with fatty and fibrotic tissue and an episode of ventricular tachyarrhythmias (1). It has recently been reported that the prevalence of atrial tachyarrhythmias such as atrial fibrillation (AF) and atrial flutter (AFL), which are well-known as important causes of atrial thrombus formation, are common and occur in 14% of patients with ARVC (2). However, the incidence of thromboembolic complications in patients with ARVC is very low (3). We herein report a case of a massive right atrial thrombus associated with tachycardiac AFL in a patient with ARVC.

Case Report

A 46-year-old man visited the outpatient clinic of our hospital due to leg edema and an electrocardiogram (ECG) abnormality in November 2012. The ECG showed sinus rhythm with complete right bundle branch block and right axis deviation. Transthoracic echocardiography (TTE) revealed an enlarged right atrial (RA) and RV with thinning of the RV free wall, however, left ventricular (LV) contraction was normal. There was no evidence of pulmonary thromboembolism by enhanced chest CT. Furosemide was initiated for the leg edema. In April 2013, he was admitted to the emergency department of our hospital due to near syncope and palpitations. A physical examination revealed peripheral coldness with hypotension (systolic blood pressure: 70 mmHg) and a regular, rapid pulse (110 bpm). There were no heart murmurs or extra heart sounds. There were no abnormal breath sounds. The ECG on this admission exhibited saw tooth P waves in leads II, III, and aVF, indicating common AFL. The flutter cycle length was 290 msec with 2:1 atrioventricular (AV) conduction (Fig. 1A). He had no family history of sudden death. The serum brain natriuretic peptide (BNP) level was 364.2 pg/mL and D-dimer was elevated up to 2.4 μg/mL on this admission. AV conduction...
Figure 1. A) The 12-lead ECG on admission exhibited saw tooth P waves in the inferior leads with 2:1 atrioventricular conduction. B) Development of 1:1 AV conduction with aberrant conduction after eating.

during AFL converted from 2:1 to 1:1 with eating and exercise (Fig. 1B). The total heart beats evaluated by Holter ECG recordings was 133,162 beats per day with an AFL rhythm throughout the entire day. A chest X-ray showed cardiomegaly without any congestion in the lung field. TTE demonstrated a huge RA along with a massive thrombus extending from the RA free wall to the junction of the superior vena cava, which was not detected during sinus rhythm six months prior to this admission. Ebstein’s anomaly had been ruled out because previous TTE performed in May 2004 showed no evidence of a tricuspid valve malformation. Cardiac magnetic resonance imaging (MRI) also demonstrated a massive RA thrombus along with pronounced thinning of the dilated RV (Fig. 2A). The RV end-diastolic and systolic volumes were 357 mL and 340 mL, respectively. The calculated RV ejection fraction was 5%. There was no evidence of any fatty changes in the RV. However, cardiac computed tomography (CT) showed an inhomogeneous low-density area in the LV apex, suggesting fatty degeneration (Fig. 2B). ARVC was diagnosed according to the presence of epsilon waves in a previously recorded ECG during sinus rhythm (Fig. 2C); regional RV dyskinesia and a ratio of the right ventricular end-diastolic volume (RVEDV) to the body surface area of ≥110 mL/m² estimated by cardiac MRI; and inverted T waves in right precordial leads, non-sustained ventricular tachycardia and over 500 ventricular extrasystoles by Holter ECG. These findings met the 3 major criteria and 1 minor criterion for the diagnosis of ARVC in the proposed modification of the Task Force criteria (4).

Anticoagulation therapy with warfarin was initiated for the RA thrombus and AFL. The oral administration of bisoprolol reduced the ventricular response to 4:1 AV conduction. His symptoms were ameliorated along with the control of his heart rate, and the RA thrombus became smaller after the optimal anticoagulant therapy. The serum BNP level also gradually decreased (Fig. 3), and the D-dimer level decreased to 0.5 μg/mL. However, two weeks after discharge, the patient’s heart failure became exacerbated, presumably due to an uncontrolled heart rate on exertion.

During the second admission, his body weight had increased by 10 kg from the last discharge due to systemic edema. His systolic blood pressure was below 80 mmHg, which made it hard to increase the dose of bisoprolol to control his heart rate. According to these findings and the clinical course, we decided to choose a rhythm control therapy using radiofrequency catheter ablation while paying careful attention to the residual RA thrombus.

All catheters for the electrophysiological study and radiofrequency catheter ablation were introduced from the right femoral vein in order to avoid catheter manipulation-induced thromboembolism complications. Activation mapping during AFL revealed a counter-clockwise propagation pattern around the tricuspid annulus with an enlargement of the tricuspid annulus (Fig. 4). The calculated tricuspid annulus orifice area measured by the 3D mapping system (NavX®, St. Jude Medical, St. Paul, USA) was 27.6 cm². Entrainment
Figure 2. The diagnosis of arrhythmogenic right ventricular cardiomyopathy and a massive RA thrombus by cardiac CT and MRI. A) The arrowheads show an extensive enlargement of the RA with a massive thrombus, which was detected by cardiac MRI. B) The arrowheads show a massive RA thrombus and the white arrow shows an inhomogeneous low-density area in the LV apex suggesting fatty degeneration, which were detected by cardiac CT. C) The ECG during sinus rhythm exhibited epsilon waves in the right precordial leads.

We herein reported a case of a massive RA thrombus associated with tachycardiac AFL in a patient with ARVC. ARVC is an inherited myocardial disease affecting the RV or characterized by the biventricular replacement of myocytes with fibrotic tissue and fatty degeneration. The prevalence of ARVC in the general adult population is estimated to be approximately 1 in 2000 to 1 in 5000 (5). The symptoms of patients with ARVC include palpitations, fatigue and syncope. In some cases, cardiac arrest may be the first presentation (6). The most famous clinical presentation is ventricular arrhythmias of an RV origin. A recent study revealed that atrial arrhythmias are common in ARVC and 14% of patients with ARVC develop atrial arrhythmias during a median follow-up of 5.78 years (2). However, despite the high prevalence of atrial tachyarrhythmias in ARVC, thromboembolic complications in ARVC are relatively low (3). It is speculated that most of these patients have a low CHADS2 (7) or CHA2DS2-VASc score (8), which represents the risk of thromboembolism in patients with AF. These ARVC patients are typically younger in age, and therefore, the prevalence of hypertension and diabetes mellitus is less frequent as compared to the general AF population. There have been only a few reports on RA thrombi in ARVC (9-12).

It was speculated that several factors might be involved in the development of the RA thrombus in the present case. AFL, which is well-known to increase the risk of atrial thrombi, as well as AF, is the most important cause of RA thrombus formation. Indeed, there was no detection of a RA thrombus during sinus rhythm in the present case. The rapid ventricular response, especially 1:1 AV conduction during AFL, caused the deterioration of right-sided heart failure in addition to the RV dysfunction due to ARVC.

Enhancement of AV node conduction by catecholamines and anticholinergic agents and/or atrial conduction velocity slowing by sodium channel blockers, such as flecainide, are pacing from the ablation catheter located at the cava tricuspid isthmus (CTI) showed that the post-pacing interval was identical to the tachycardia cycle length, indicating CTI-dependent AFL. CTI liner ablation successfully eliminated AFL and restored sinus rhythm. After catheter ablation, sinus rhythm could be maintained and heart failure was well controlled. Eventually, almost all of the RA thrombus disappeared 5 months after discharge, except for a small amount of organized thrombus (Fig. 5). There have been no recurrences of AFL and no deterioration of the patient’s heart failure for more than 1 and a half years.

Discussion

We herein reported a case of a massive RA thrombus associated with tachycardiac AFL in a patient with ARVC. ARVC is an inherited myocardial disease affecting the RV or characterized by the biventricular replacement of myocytes with fibrotic tissue and fatty degeneration. The prevalence of ARVC in the general adult population is estimated to be approximately 1 in 2000 to 1 in 5000 (5). The symptoms of patients with ARVC include palpitations, fatigue and syncope. In some cases, cardiac arrest may be the first presentation (6). The most famous clinical presentation is ventricular arrhythmias of an RV origin. A recent study revealed that atrial arrhythmias are common in ARVC and 14% of patients with ARVC develop atrial arrhythmias during a median follow-up of 5.78 years (2). However, despite the high prevalence of atrial tachyarrhythmias in ARVC, thromboembolic complications in ARVC are relatively low (3). It is speculated that most of these patients have a low CHADS2 (7) or CHA2DS2-VASc score (8), which represents the risk of thromboembolism in patients with AF. These ARVC patients are typically younger in age, and therefore, the prevalence of hypertension and diabetes mellitus is less frequent as compared to the general AF population. There have been only a few reports on RA thrombi in ARVC (9-12).

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Figure 3. The clinical course of the treatment of heart failure and RA thrombus. It was hard to control the heart rate, despite the administration of an optimal dose of beta-blockers during atrial flutter (AFL). The RA thrombus had not completely disappeared following anticoagulation therapy during AFL. Restoration to sinus rhythm by catheter ablation was effective in treating the heart failure and RA thrombus.

Figure 4. Activation mapping during AFL with a 3D mapping system revealed a counter-clockwise propagation pattern around the tricuspid annulus with an enlargement of the tricuspid annulus.

known to be risk factors for 1:1 AV conduction during AFL (13, 14). However, in the present case, neither catecholamine/anticholinergic agents nor anti-arrhythmic agents, including sodium channel blockers, were used.

In the present case, an enlargement of the tricuspid annulus with a low voltage area in the RA free wall was confirmed by 3D electroanatomical mapping. RA structural remodeling, such as fibrotic changes, and a tricuspid annulus
enlargement caused by ARVC might have led to slowing of the conduction velocity, and a longer reentrant circuit might have facilitated 1:1 AV conduction, especially concomitant with an enhancement of the sympathetic nervous activity. Both the loss of the atrial kick and hemostasis in the RA due to right-sided heart failure were associated with the RA thrombus formation.

In the present case, an atrial thrombus was detected in the RA and not in the LA. The incidence of left atrial thrombi in patients with AF/AFL has been widely investigated. However, little focus has so far been placed on RA thrombi in those patients. Several reports have indicated that RA appendage thrombi are detected in 0.7% to 2.4% of patients with AF/AFL, however, it is less frequent than for the LA appendage (15). In patients with ARVC complicated by atrial tachyarhythmias, the risk of thrombus formation might be more increased in the RA than the LA.

The present case showed that the resistance to heart rate control therapy with bisoprolol resulted in persistent tachycardiac AFL. It was difficult to control the heart failure, and the residual RA thrombus was thereby refractory to anticoagulation therapy. Therefore, radiofrequency catheter ablation was performed for AFL with careful attention being paid to the residual thrombus. The restoration to sinus rhythm by catheter ablation effectively improved the heart failure, implying that the maintenance of sinus rhythm was essential for the treatment of heart failure as well as the thrombus prevention in this case.

RA hemostasis associated with a loss of the atrial kick and right-sided heart failure due to AFL with 1:1 AV conduction promoted a massive RA thrombus formation. A thrombus formation in the atrium might cause a fatal thromboembolic complication. Therefore, if an atrial tachyarrhythmia occurs in patients with ARVC, then anticoagulation therapy and a prompt restoration to sinus rhythm by catheter ablation or electrical cardioversion should be considered to prevent any thrombus formation and a deterioration of heart failure in such cases.

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References