Right Atrial Thrombosis as a Complication of Arrhythmogenic Right Ventricular Cardiomyopathy

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Abstract

A 65-year-old woman was admitted to our hospital due to palpitation. Electrocardiogram (ECG) showed ventricular tachycardia originating from the right ventricle, and transthoracic echocardiography revealed dilatations of the right atrium and ventricle. The diagnosis of arrhythmogenic right ventricular cardiomyopathy was made. Eleven months later, echocardiography revealed a solid thrombus (36×32 mm) attached to the free wall of the right atrium, and it was surgically resected. Four months after the operation, a solid thrombus (48×30 mm) appeared again at the same site despite anticoagulant treatment. The patient died of both left and right heart failure 33 months after the operation.

Key words: arrhythmogenic right ventricular cardiomyopathy (ARVC), right atrial thrombosis, electrocardiogram

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Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC), which was first reported by Fontaine et al in 1977 (1), is a myocardial disease that primarily affects the right ventricle and is characterized by fibrofatty replacement of the myocardium (2-6). In 1994, an international task force proposed a series of diagnostic criteria based on the electrocardiographic and morphologic features to facilitate the clinical diagnosis of ARVC (4). The phenotype of ARVC is variable, including ventricular tachycardia, supraventricular arrhythmia, right heart failure or sudden death. Here, we present a patient with ARVC complicated by right atrial thrombosis which recurred despite surgical and anticoagulant treatments.

Case Report

A 65-year-old female with a history of chronic hepatitis C was admitted to our hospital due to persistent palpitation. Physical examination showed blood pressure of 74/50 mmHg, pulse rate of 165 beats/min and edema on face and lower limbs. Chest X-ray revealed cardiomegaly with a cardiothoracic ratio of 68%. Electrocardiogram (ECG) showed wide QRS tachycardia with atrio-ventricular dissociation (Fig. 1). Because QRS complex mimicked left bundle branch block, the diagnosis of ventricular tachycardia originating from the right ventricle was made. This ventricular tachycardia was spontaneously terminated after the admission. During sinus rhythm, ECG showed low voltage in limb leads, and T wave inversion in V1-2 leads (Fig. 2). Transthoracic echocardiography revealed left ventricular end-diastolic diameter (LVDd) of 49 mm, left ventricular ejection fraction (LVEF) of 50 %, and dilatations of right atrium and ventricle, but did not show pericardial effusion. Signal averaged ECG showed the presence of late potential. Lung perfusion scintigraphy revealed no findings of pulmonary thromboembolism. Cardiac catheterization was performed 7 days after the admission. Pressures in left ventricle, pulmonary artery, right ventricle and right atrium were 72/8 mmHg, 16/6 mmHg, 16/2 mmHg and 6 mmHg, respectively. Coronary angiography revealed no coronary artery disease, but left ventriculography revealed mildly depressed systolic function. Right ventriculography revealed right ventricular dilatation and slow evacuation of contrast medium due to severely depressed systolic function. Endomyocardial biopsy from the interventricular septum showed...
Figure 1. Electrocardiogram (ECG) on admission. ECG showed wide QRS tachycardia with atrio-ventricular dissociation. QRS complex mimicked left bundle branch block, suggesting ventricular tachycardia originating from the right ventricle. P waves are manifest in inferior leads (arrows).

Figure 2. Time course of ECG during the 4 years. ECG showed only T-wave inversion in V1-2 leads at the time of the first admission. However, epsilon wave appeared in V1-2 leads one year later (arrows).

mild interstitial fibrosis, but no fatty infiltration. She had no family history of any heart disease. According to criteria defined by the Task Force of the European Society of Cardiology and International Society and Federation of Cardiology, the diagnosis of ARVC was made. The patient was treated with carvedilol (5 mg/day), furosemide (20 mg/day) and spironolactone (25 mg/day) and flecainide (100 mg/day) and was discharged 9 days after the admission.

Eleven months later, transthoracic echocardiography revealed LVDd of 48 mm, LVEF of 48% and dilatations of right atrium and ventricle. The interventricular septum shifted from the right to left heart system during the diastolic period. Tricuspid valve remained open even during the systolic period. A solid mass (36×32 mm) attached to the free wall of the right atrium and spontaneous echo contrast in the right atrium were seen (Fig. 3). ECG showed epsilon wave in V1-2 leads (Fig. 2). Atrial fibrillation or flutter had not been detected by the monitoring. Protein C decreased (41%; normal: 55-140%), but protein S (104%; normal: 65-135%) and anti-beta-2-glycoprotein I antibody (<1.2 U/ml; normal: <3.5 U/ml) were normal. The solid mass (42×34×32 mm) was attached to the free wall of the right atrium (6×4 mm), and it was surgically resected. Tricuspid annuloplasty was also performed. Microscopic examination revealed old mural thrombus, and right atrial wall with myocardial hypertrophy, intramural hemorrhage and slight chronic inflammation. Anticoagulant treatment with warfarin was initiated and the thrombo test was meticulously kept between 5% and 20%. The patient was discharged 26 days after the operation.

Four months after the operation, transthoracic echocardiography revealed LVd of 51 mm and LVEF of 28%. Both left and right ventricular systolic functions were depressed. Although the thrombo test was kept within 20%, a solid thrombus (48×30 mm) appeared again at the same site in the right atrium. Repeat surgical treatment was not performed, and the thrombus did not increase in size under anticoagulant treatment during follow-up.

Thirty-two months after the operation, the patient was admitted again due to appetite loss and edema. ECG showed an enlarged epsilon wave, and lung perfusion scintigraphy revealed no findings of pulmonary thromboembolism. She died of both left and right heart failure 33 months after the operation.

Discussion

We present a patient with ARVC complicated by right atrial thrombosis which recurred in spite of surgical and anticoagulant treatments. Endomyocardial biopsy has a limited role for the diagnosis of ARVC, because the interventricular septum is rarely involved and biopsy of the thinned free wall is potentially dangerous. As shown in the current report, the diagnosis of ARVC relies on the clinical demonstration of electrocardiographic, structural and functional abnormalities resulting from the underlying histologic changes.

The occurrence of right heart thrombosis in ARVC is very rare, and there have been only a few reports demonstrating this complication (7-10). In the current patient, atrial fibrillation or flutter had not been detected, but spontaneous echo contrast was seen in the enlarged right atrium even during sinus rhythm. Thus, slow evacuation of blood in the enlarged right atrium appeared to cause right atrial thrombosis. As shown by Marcus et al, right atrial dilatation is not a rare anatomical feature of ARVC (2), and it may be a risk
Figure 3. Follow-up echocardiography. Echocardiography revealed dilatation of right atrium and ventricle (A). Interventricular septum shifted from right to left heart system during the diastolic period (B). A solid mass (36 × 32 mm) attached to the free wall of the right atrium was seen (C).

factor of right atrial thrombosis. On the other hand, protein C, protein S, antithrombin III and plasminogen are produced by the liver, and thrombotic risk factors are frequently detected in patients with chronic viral hepatitis (11). The current patient had chronic hepatitis C, and protein C decreased to 40%. Thus, decreased protein C also might affect right atrial thrombosis. In the current patient, right atrial thrombosis recurred, although anticoagulant treatment with warfarin was kept in the standard therapeutic range after the operation. It remained unclear whether more intensive anticoagulant treatment could prevent the recurrence of right atrial thrombosis. In the current patient, lung perfusion scintigraphy revealed no findings of pulmonary thromboembolism. However, we should take care of right atrial thrombosis in patients with ARVC during follow-up, because it can suddenly cause fatal pulmonary thromboembolism.

In the current patient, left ventricular function also became worse during the 4 years. In fact, several studies have reported that there is also involvement of the left ventricle with subepicardial posterior wall fibrosis and fibro-adipose infiltration (12, 13). In view of the biventricular involvement of this disease, Gallo et al suggested that it should be simply termed “arrhythmogenic cardiomyopathy” (12).

We also demonstrated the time course of ECG during the 4 years. ECG showed only T-wave inversion in V1-2 leads at the time of the first admission. However, the epsilon wave appeared in V1-2 leads one year later, and enlarged 4 years later. Thus, the presence of ECG features appears to be dependent on the stage of this disease. In conclusion, we should recognize right atrial thrombosis as a complication of ARVC.

References


