Transvenous implantable cardioverter-defibrillator implantation in a patient with arrhythmogenic cardiomyopathy and massive right atrial thrombus

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Introduction

Arrhythmogenic cardiomyopathy (ACM) is an inherited myocardial disease that is characterized by a progressive loss of myocytes with fibrofatty replacement and predisposes to ventricular tachycardia (VT) and sudden cardiac death.[1] Implantable cardioverter-defibrillators (ICDs) are indicated for VT; however, the use of those devices is often challenging because of sensing dysfunction with a low right ventricular sensing amplitude and dilated right atrium (RA). Atrial arrhythmias are common in ACM and are associated with inappropriate ICD shocks and an increased risk of both death and heart failure.[2] In addition, owing to a large hypokinetic right ventricle (RV) and slow blood flow within the RV, ACM may be complicated by thromboembolic events, and screening of thrombi before the procedure is important.[3,4] We present a case of an ACM patient with a massive RA thrombus in whom an ICD implantation was successfully achieved by creating a 3-dimensional echocardiographic image of the thrombus and an electroanatomic voltage map of the RA.

KEY TEACHING POINTS

- Arrhythmogenic cardiomyopathy may be complicated by intracardiac thrombosis; therefore, screening of thrombi before the procedure is necessary.
- When implantation of a transvenous electronic device is required in patients with anticoagulation therapy–refractory intracardiac thrombi, creating a 3-dimensional echocardiographic image of the thrombus may prevent thromboembolic complications during the procedure.
- Evaluating the intracardiac voltage prior to the device implantation is helpful for implanting the leads in the appropriate locations.

Case report

A 53-year-old man diagnosed with ACM and chronic kidney disease was referred to our hospital with palpitations and dyspnea. He had a previous history of catheter ablation of typical atrial flutter and anticoagulation therapy (warfarin) for persistent atrial tachycardia (AT) with prothrombin time–international normalized ratio between 2 and 3. The electrocardiogram upon admission revealed a wide QRS tachycardia with a left bundle branch block morphology and superior axis with an R/S transition in lead V_5 (Figure 1A), which was diagnosed as VT. Transthoracic echocardiography revealed a dilated RA and RV with spontaneous echogenic contrast and severe hypokinesis of the RV. A huge RA along with a massive thrombus extending from the right free wall to the junction of the superior vena cava was documented (Figure 2). Although the VT was terminated and controlled by intravenous nifekalant, the patient was revealed to have a junctional rhythm at 30 beats per minute with retrograde P waves, which caused a low cardiac output syndrome (Figure 1B). As the atrial contraction and atrial-ventricular synchrony are important in patients with heart failure,[5] we decided to implant a dual-chamber ICD system.

Before the ICD implantation, because the manipulation of the atrial lead and ICD shock lead under the presence of a massive RA thrombus might cause a thromboembolism, we established 3-dimensional echocardiographic mapping of the thrombus and RA using a CartoSound module (Biosense Webster, Diamond Bar, CA) to confirm the anatomical distribution of the thrombus within the RA. As the RA was severely dilated, a bipolar voltage map was created using

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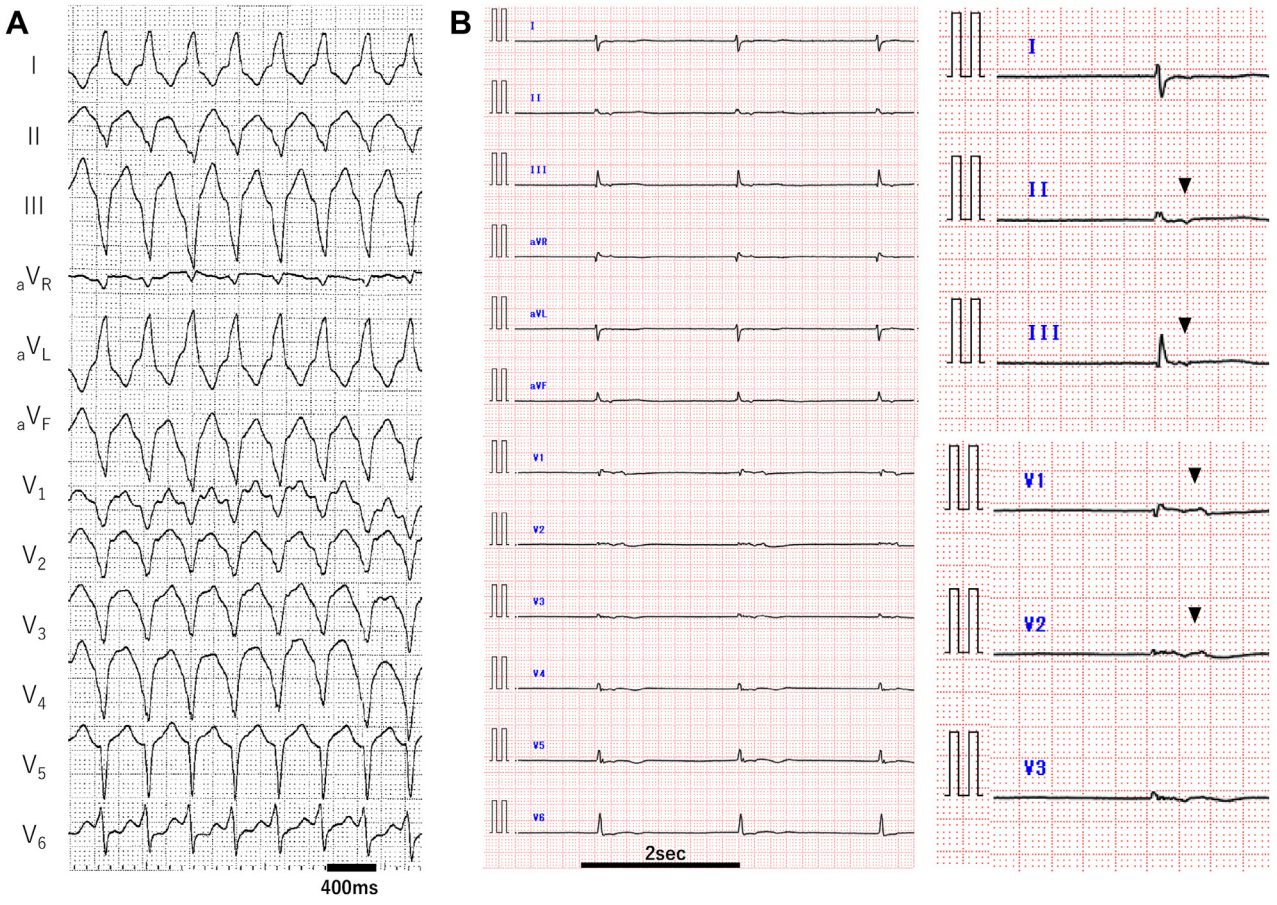

Figure 1 **A:** A 12-lead electrocardiogram of the clinical ventricular tachycardia (VT). **B:** After the termination of VT, the patient revealed a junctional rhythm at 30 beats per minute with retrograde P waves (*black arrows*).

the CARTO system (Biosense Webster) in order to identify the suitable lead position sites. Multiple endocardial sites were sampled with a 3.5-mm-tip catheter (ThermoCool NaviStar; Biosense Webster) advanced via the right femoral vein during junctional rhythm. The 3-dimensional echocardiographic imaging revealed a massive thrombus in the RA appendage that extended to the superior vena cava and dilatation of the RA (Figure 3A and B). The low voltage zone (LVZ) was defined by contiguous areas of bipolar voltages of $\leq$0.05 mV.[6] The electroanatomical map revealed an extensive LVZ in the RA excluding the RA septum. The voltage recorded in the RV ranged from 1.3 to 5 mV. We carefully manipulated the leads while referring to the thrombus mapping in order to avoid the thrombus area. A RA bipolar screw-in lead (INGEVITY MRI; Boston Scientific, Marlborough, MA) was inserted via the left subclavian vein and positioned on the RA septum, where the voltage map showed a relatively high voltage with the best sensed P wave of 1.5 mV and a pacing threshold of 1 V at a pulse width of 0.4 ms. An RV bipolar screw-in lead (RELIANCE 4-FRONT; Boston Scientific, Marlborough, MA) was positioned in the RV apex with a sensed V wave of 7.8 mV and pacing

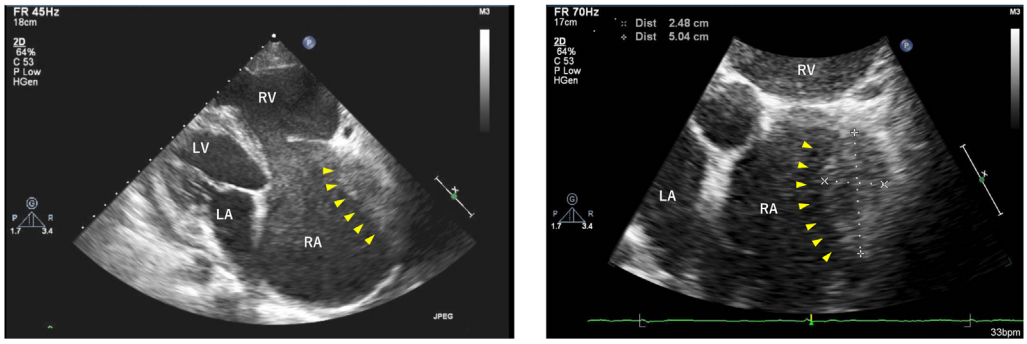

Figure 2 Echocardiographic image showing an enlargement of the right atrium (RA) and right ventricle (RV). A massive thrombus (*yellow arrows*) extending from the right free wall to the junction of the superior vena cava is documented. LA = left atrium; LV = left ventricle.

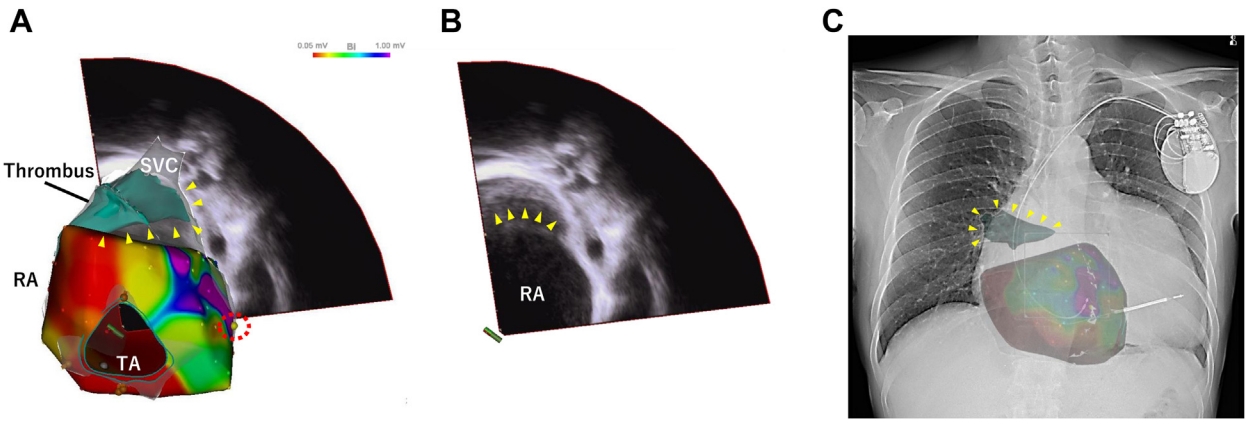

Figure 3 **A:** Bipolar voltage map of right atrium (RA). Large areas of low voltage were observed except for on the RA septum. The dotted red circle had the best amplitude and the pacing threshold was 1.5 mV and 1 V/0.4 ms. The 3-dimensional echocardiographic image (**A**) and intracardiac echocardiogram (**B**) show the thrombus (*yellow arrows*) located in the RA appendage and extending to the superior vena cava (SVC). **C:** The chest radiograph after the implantable cardioverter-defibrillator implantation. The voltage map of the RA is merged with the radiograph, which reveals that the RA lead is implanted on the RA septum with a preserved RA amplitude. The yellow arrows indicate the RA thrombus. TA = tricuspid annulus.

threshold of 0.4 V at a pulse width of 0.4 ms and was connected to the ICD (RESONATE; Boston Scientific) (Figure 3C). There were no perioperative thrombotic events and no pacing or sensing failure occurred. The increase in the heart rate during RA pacing improved the low cardiac output syndrome. As the therapeutic range of warfarin is narrow and the bleeding risk of the patient was high (HAS-BLED score 4), the warfarin therapy was changed to direct oral anticoagulant therapy (apixaban 5 mg twice daily). The patient was discharged without any adverse events. It has been 6 months since the ICD implantation and the patient is still in sinus rhythm, with no recurrence of AT or VT. Oral amiodarone (100 mg daily) had been administered since the ICD implantation and owing to the sinus bradycardia, the device counters showed 87% RA pacing. There was no ICD shock, either appropriate or inappropriate, during the follow-up.

Discussion

The incidence of thromboembolic complications in ACM is reported to be 0.5 in 100 patients,[3] which is not common. Most of the thrombi associated with ACM are thrombi in the RV and only a few case reports have shown ACM complicated with RA thrombi.[7–9] In the present case, the patient was under warfarin therapy with adequate prothrombin time–international normalized ratio level (2–3). The mechanism of the anticoagulation-refractory thrombus formation was speculated to be the blood stasis caused by right ventricular systolic dysfunction, observed as a RA spontaneous echo contrast, and underlying persistent AT. In addition, the presence of heart failure and concomitant chronic kidney disease might have played a role as a predisposing factor for the thrombus formation. In the present case, as the risk of perioperative embolic complications during the procedure was high, we performed the ICD implantation without interruption of warfarin anticoagulation, and intensive monitoring of vital signs was also performed during the operation to detect thrombotic events immediately. After the operation, as the patient was speculated to be a high risk for lead-associated thrombus formation, a lifelong optimal anticoagulation therapy and echographic follow-up of the leads and RA thrombus size is essential.

The present case involved sinus bradycardia and hemodynamically intolerable VT, and therefore a dual-chamber ICD was essential to prevent heart failure and sudden cardiac death despite the presence of an RA thrombus. There has been only 1 case report of an ACM patient with an RA thrombus that successfully underwent an epicardial pacemaker implantation.[8] Fortunately, the present case could avoid any thromboembolism complications associated with the ICD implantation, and a pacemaker with an epicardial lead normally would be chosen in the presence of optimal anticoagulation therapy–refractory RA/RV thrombi. However, implanting an epicardial device system usually involves a surgically invasive procedure, which might be intolerable for a critically ill patient.

A wide area of the LVZ in the RA was observed in the present case. Some studies have suggested that ACM-related fibrofatty lesions may involve atrial chambers.[10,11] Takemura and colleagues[12] reported an ACM patient undergoing electroanatomical mapping of the RA, which showed extensive scarring with no recordable electrical potentials. It is assumed that the atrial remodeling and LVZ was caused by desmosomal dysfunction within the atrium and/or RV dysfunction. Evaluating the intracardiac voltage prior to the device implantation in ACM associated with a dilated atrium may be helpful for implanting the leads in the appropriate locations.

Conclusions

We reported the case of a 53-year-old man with ACM with a massive RA thrombus who underwent a transvenous ICD implantation. Creating a 3-dimensional echocardiographic image of the thrombus and an electroanatomic voltage map are attractive therapeutic options that might be considered

when thromboembolic complications and sensing difficulties are of concern.

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