Case Report

Electroanatomical mapping of the atrialized right ventricle: Placement of a transvenous implantable cardioverter-defibrillator in a patient with Ebstein's anomaly

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ARTICLE INFO

ABSTRACT

A 47-year-old woman with Ebstein's anomaly suffered from an out-of-hospital cardiac arrest caused by ventricular fibrillation. Electroanatomical activation mapping showed an atrialized right ventricle. Atrial electrocardiogram, normal atrioventricular node conduction delay, and ventricular electrocardiogram were confirmed in the right atrium. Relatively preserved ventricular amplitude was found in the septal wall. Based on these findings, a transvenous dual-chamber implantable cardioverter-defibrillator was implanted for the prevention of sudden cardiac death. The patient has fared well, without any lead malfunctions, lead dislodgement, or inappropriate shocks. Sufficiently high atrial and ventricular amplitudes were confirmed during 18 months of follow-up.

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1. Case report

A 47-year-old woman (height: 165 cm, weight: 52.8 kg) was transported to the hospital following syncope. According to the automated external defibrillator, she experienced ventricular fibrillation (VF), which was treated with a direct current shock. The patient had a history of cardiac surgery for an atrial septal defect in childhood. Echocardiography and cardiac computed tomography (Fig. 1A) revealed a severely dilated right atrium and right ventricle, and Ebstein's anomaly of the tricuspid valve. On the other hand, the size and contraction of the left ventricle were almost normal (left ventricular ejection fraction, 57%) and her B-type natriuretic peptide level was 63.7 pg/mL. Amiodarone (200 mg/day) and carvedilol (10 mg/day) were administered to prevent lethal ventricular arrhythmia. Initially, electrophysiological testing and cardiac catheterization were performed to identify the substrate of VF and to investigate the appropriate site for the positioning of an implantable cardioverter-defibrillator (ICD) lead. Coronary angiography revealed no significant stenosis in the coronary arteries. Left ventriculography showed normal wall motion in the left ventricle. Consistent with the echocardiographic findings, right atriography showed a significantly dilated right atrium and displacement of the tricuspid valve (Fig. 1B). Ventricular tachyarrhythmia was not induced by a programmed

stimulation with up to three extrastimuli. Electroanatomical activation mapping (CARTO® system, Biosense Webster, Diamond Bar, CA, USA) clearly showed an atrialized right ventricle (Fig. 2A). Atrial electrocardiogram [Fig. 2A-(1)], normal atrioventricular node conduction delay [Fig. 2A-(2)], and ventricular electrocardiogram [Fig. 2A-(3)] were confirmed in the right atrium. Both atrial and ventricular potentials were observed near the atrioventricular groove [Fig. 2A-(2)]. At the border of the anatomical RA and atrialized RV, the ventricular potential was defined as the local activation potential. Even though electroanatomical voltage mapping showed a wide low amplitude area, especially around the atrialized ventricle, the electrograms at the septal site were relatively preserved and were recognized as an optimal lesion for ICD lead implantation (Fig. 2B). Based on these findings, a transvenous dual-chamber ICD (TELIGEN 100 F111, Boston Scientific Inc., St. Paul, MN, USA) was implanted for the prevention of sudden cardiac death. The atrial lead (DEXTRUS 4136/53 cm, Boston Scientific Inc.) and ICD lead (ENDOTAK RELIANCE G 0296/64 cm, Boston Scientific Inc.) were successfully implanted (Fig. 2C). P-wave and R-wave sensing was satisfactory at 3.2 mV and 9.5 mV, respectively. Adequate capture threshold values were measured at the right atrium (pacing threshold: 0.5 V, 0.4 ms) and right ventricle (pacing threshold: 0.5 V, 0.4 ms). Induced ventricular fibrillation was successfully terminated by a 9 J direct current shock.

The patient has fared well, without any lead malfunctions, lead dislodgement, or inappropriate shocks. Follow-up echocardiography showed no aggravation of tricuspid regurgitation. Sufficiently

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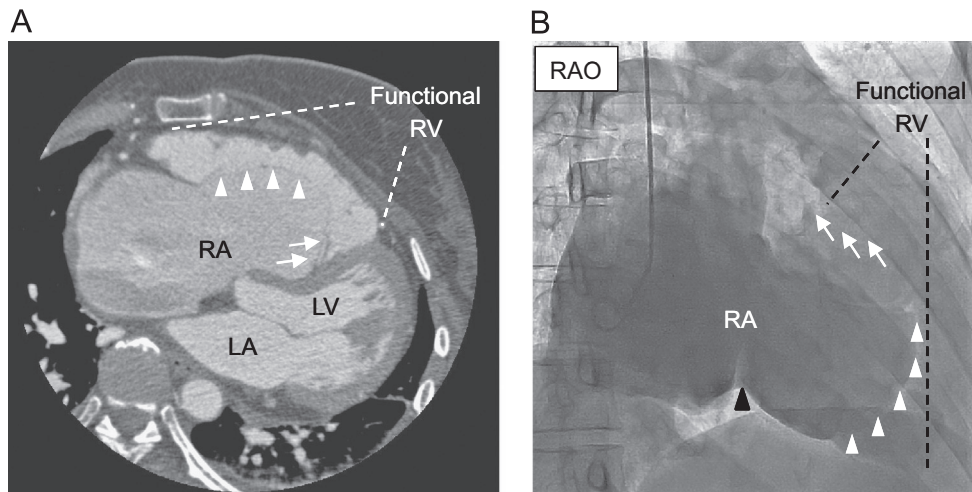E-mail address: t-arimoto@med.id.yamagata-u.ac.jp (T. Arimoto).

Fig. 1. (A) Cardiac computed tomography and (B) right atriography. White arrows and white arrowheads indicate the displaced septal leaflet and enlarged anterior leaflet of the tricuspid valve, respectively. A functional right ventricle (RV) existed between the dotted lines. The black arrowhead indicates the atrioventricular groove. LA, left atrium; LV, left ventricle; RA, right atrium; RAO, right anterior oblique.
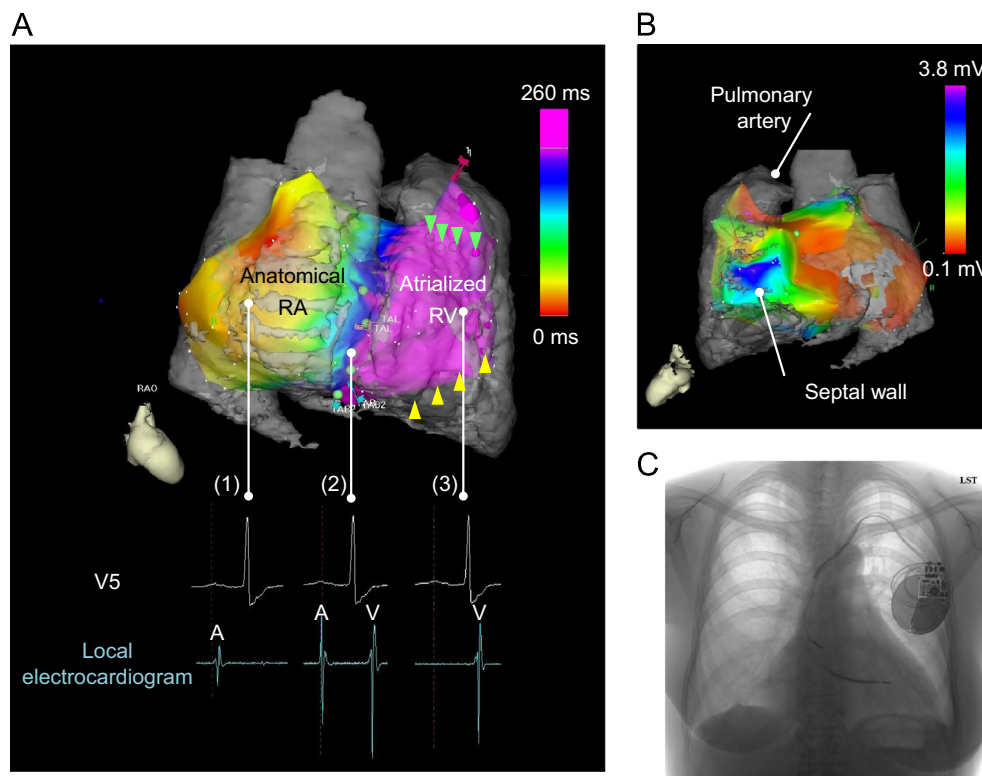

Fig. 2. (A) Electroanatomical activation mapping. The green arrowheads indicate the displaced septal leaflet of the tricuspid valve, confirmed by transthoracic echocardiography. Electroanatomical activation mapping demonstrated the atrialized right ventricle. Atrial electrocardiogram (1), normal atrioventricular node conduction delay (2), and ventricular electrocardiogram (3) were confirmed in the right atrium. Both atrial and ventricular potentials were observed near the atrioventricular groove (2). An enlarged anterior leaflet of the tricuspid valve (yellow arrowheads) existed in this patient, and therefore, it was impossible to advance a catheter and shock lead near the apex. A, atrial potential; RA, right atrium; RV, right ventricle; V, ventricular potential. (B) Electroanatomical voltage mapping. Relatively preserved ventricular amplitude was found in the septal wall. (C) Posterior-anterior chest X-ray after transvenous implantation of an implantable cardioverter-defibrillator.

high atrial and ventricular amplitudes (P-wave: 3.5 mV and R-wave: 9.9 mV) were confirmed during 18 months of follow-up.

2. Discussion

Because of significant progress in pediatric cardiac care, a growing number of patients with congenital heart disease survive well into adulthood [1,2]. The management of arrhythmia in adult patients with congenital heart disease represents an important issue for cardiologists [3–5]. Ebstein's anomaly is characterized by apical displacement of the septal and posterior leaflets of the tricuspid valve. Approximately 25% of these patients have associated accessory pathways and are at risk of supraventricular tachycardia. Unless atrial fibrillation degenerates into ventricular fibrillation in patients with manifest accessory pathways, ventricular arrhythmia is a rare complication of Ebstein's anomaly [6]. In a multicenter cohort study, ventricular arrhythmia occurred in only 1 of 77 young patients with Ebstein's anomaly who received an ICD [7]. In our case, electroanatomical voltage mapping found a

wide low amplitude area around the atrialized ventricle. Although ventricular tachyarrhythmia was not induced by programmed stimulation, this low voltage area may be associated with an arrhythmogenic region in this patient. Lacking a histological investigation, we could not specify the pathological mechanism underlying these electrophysiological changes.

Implantation of the device in this patient was technically challenging because of the unusual orientation of the right ventricle. Electroanatomical mapping can be used to determine the exact intracardiac anatomy and confirm the amplitude of the cardiac chambers, especially in patients with complex cardiac anatomy [8,9]. In the present case, prior electroanatomical information was helpful in planning the transvenous procedure strategy. Therefore, we believe that electroanatomical mapping is a useful, minimally invasive strategy to be performed prior to the implantation of devices in patients with a complex cardiac anatomy. We selected the thinnest ICD device (TELIGEN 100 F111, Boston Scientific Inc.) because the size of the patient's body was quite small, and her precordial subcutaneous fatty layer was assumed to be relatively thinner. The Guidant ICD lead (a shock lead) is an integrated bipolar system, and consequently, we should pay close attention to avoiding atrial oversensing, especially when deep insertion of the shock lead is impossible. Although we could not advance the lead into the apical site, probably because of an enlarged anterior leaflet of the tricuspid valve, atrial double sensing was never observed during the implantation procedures.

We successfully visualized the atrialized right ventricle and implanted a transvenous ICD, without any complications, in a patient who had not undergone surgery for Ebstein's anomaly.

Conflict of interest

No author has a real or perceived conflict of interest.

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