IMAGES

Biventricular activation of right bundle branch block pattern ventricular tachycardia originating from the right ventricle in Ebstein's anomaly

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Introduction

Ebstein's anomaly (EA) is a rare congenital heart disease where the attachment points of the septal and posterior leaflets of the tricuspid valve are displaced toward the apex of the right ventricle, which may be complicated by tricuspid regurgitation.[1] Additionally, EA is often complicated by arrhythmias, commonly Wolff-Parkinson-White syndrome and supraventricular tachycardia.[1] Ventricular tachycardia (VT) is a rare occurrence in EA and poses a risk factor for sudden death. Catheter ablation is reportedly useful for the management of VT.[2,3] VT in EA often presents with the left bundle branch block (LBBB) rather than right bundle branch block (RBBB) morphology because the atrialized right ventricle (RV) is an arrhythmogenic substrate.[4] Furthermore, there is no detailed study on the mechanism of VT originating from the RV presenting with an RBBB pattern. Hence, this study aimed to provide a better understanding of VT in EA, its morphological characteristics, its mechanism, and implications for clinical diagnosis and management.

KEY TEACHING POINTS

- In a case of Ebstein's anomaly (EA) with an arrhythmogenic substrate in an atrialized right ventricle (RV), ventricular tachycardia (VT) could potentially present with a right bundle branch block (RBBB) morphology.
- RBBB-pattern VT originating from the RV could propagate slowly across the RV following immediate activation of the entire left ventricle (LV).
- In EA, even when VT exhibits RBBB morphology, it is important to consider the possibility of the presence of an arrhythmogenic substrate in the RV rather than the LV.

Case report

We report a case of a 37-year-old man who was diagnosed with EA during childhood but was asymptomatic. The patient experienced syncope at the age of 29 years, the cause of which was unknown despite thorough examinations, including an electrophysiological study (EPS). Thereafter, there were no recurrences of syncope.

The patient visited the emergency department with a complaint of palpitations and dizziness. The pulse rate was 220 beats per minute, and the patient was in shock. Electrocardiography (ECG) showed wide QRS tachycardia with RBBB morphology, and electrical cardioversion was promptly performed to restore sinus rhythm and facilitate recovery from shock (Figure 1A). ECG in sinus rhythm revealed an intraventricular conduction abnormality of the RBBB, which is commonly observed in EA (Figure 1B). Echocardiography revealed a markedly enlarged right atrium (RA), depressed RV function, and apically displaced tricuspid valve leaflets with regurgitation.

On the following day, EPS and catheter ablation were performed for the VT. Vascular access was obtained from the right jugular vein and right femoral artery/vein, and a 20-pole catheter (BeeAT; Japan Lifeline, Tokyo, Japan) was positioned in the coronary sinus and RA. Catheters were also placed in the bundle of His and RV. A Pentaray catheter (Biosense Webster Inc, Irvine, CA) was used under electroanatomical 3-dimensional (3D) mapping guidance (CARTO system; Biosense Webster Inc). The voltage map in sinus rhythm showed a significantly low-voltage area in the inferior part of the RV, consistent with RV atrialization; however, minimal degeneration was observed in the basal portion of the left ventricle (LV) (Figure 2A).

The stimulus provided by catheter contact in the RV or LV easily induced wide QRS tachycardia (cycle length: 245 ms) with RBBB morphology (positive R waves across the

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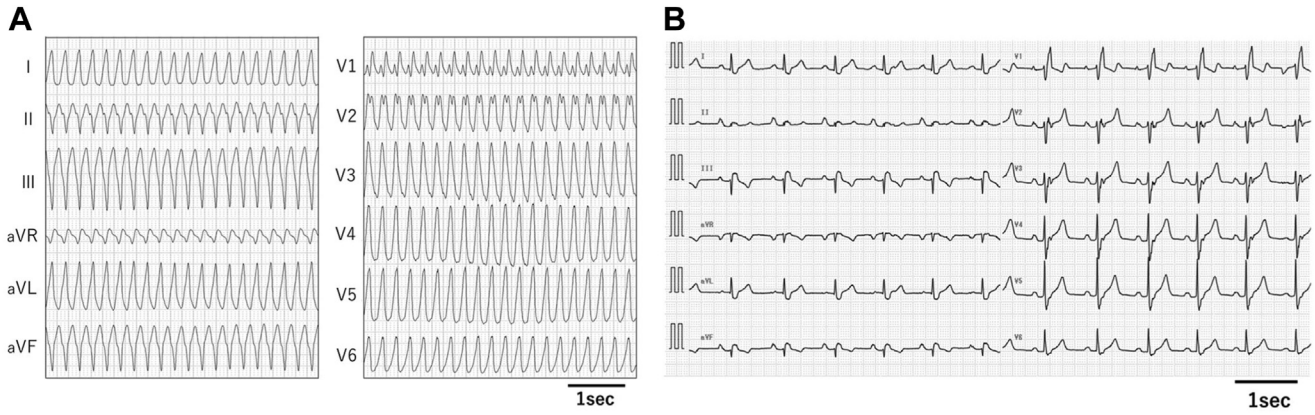

Figure 1 **A:** Surface electrocardiography (ECG) of ventricular tachycardia with right bundle branch block pattern **B:** Surface ECG during sinus rhythm.

precordium) and left axis deviation, akin to clinical VT. Ventriculoatrial dissociation was observed during tachycardia. Burst pacing with different pacing cycles from the RV septum showed constant fusion and progressive fusion, indicating reentrant VT. The activation maps revealed that the posterior portion of the RV was the earliest site during VT, which propagated to both ventricles. The coherence map showed rotating activation around the scar in the inferior part of the RV (Figure 2B). Entrainment mapping from the isthmus between the scar in the inferior RV and the RA showed concealed fusion with an approximate match between the postpacing interval and tachycardia cycle length. The short duration of the pacing stimulus to the QRS implied that the septal aspect of the posterior RV constituted the VT exit site (Figure 2B and 2C). Moreover, the ventricular septum was activated from the right to the left and there was no preceding fascicular potential in the left ventricular septum; therefore, we considered the VT less likely to be bundle branch reentrant VT. These findings were collated and the patient was diagnosed with macroreentrant VT rotating counterclockwise in the inferior RV, with the critical conduction zone being the isthmus between the scar of the inferior RV and RA, and the RV septum as the exit site. Moreover, the ripple map clarified the delay in activation of the lateral RV compared to that of the LV during VT (Supplemental Video 1).

Owing to the unstable hemodynamics during VT, electrical cardioversion was employed for VT termination. Thereafter, a complete block line was created across the critical isthmus in the inferior RV using the ThermoCool SmartTouch irrigated catheter (Biosense Webster Inc) at 35 W for 30–60 seconds during sinus rhythm. Finally, the EPS showed no VT induction.

The patient had no palpitations or syncope for 3 years after ablation, and periodic 24-hour Holter monitoring showed no VT recurrence.

Discussion

Herein, we report a case of RBBB-pattern VT originating from the RV in a patient with EA. Furthermore, we visualized full activation of VT by mapping the RV and LV using a 3D mapping system.

A comprehensive study of VTs in EA reported that VTs often present with LBBB morphology owing to breakout from an atrialized RV, and mapping and ablating were effective in treating this degenerated RV chamber.[2,4]

Although the arrhythmogenic substrate in this case was an atrialized RV, as observed in previous studies, surface ECG of VT showed an RBBB morphology rather than a typical LBBB morphology. VT was diagnosed as a macroreentrant tachycardia rotating counterclockwise in the inferior RV, exiting the RV septum. The exit site of the VT was close to the RV septum, and the entire LV was activated immediately after the breakout. In contrast, owing to degeneration in the atrialized RV, the VT exiting from the septal side of the RV propagated slowly toward the free wall side, right after complete activation of the LV. These observations clarified why VT originating from the RV presented with an RBBB morphology. This electrophysiological feature has also been observed in arrhythmogenic right ventricular cardiomyopathy (ARVC), and most VTs with an RBBB morphology in ARVC originate from the RV rather than the LV.[5] Additionally, this study mentioned that VT from the inferior septal part of a dilated RV produces an RBBB morphology that can be misidentified as VT from the LV in ARVC.

This case report is the first to provide a detailed description of biventricular activation of RBBB-pattern VT originating from the RV in a patient with EA. Furthermore, even when VT presents with a morphology resembling RBBB, it is important to consider the possibility that VT originates from the RV rather than the LV. However, further accumulation of cases and studies is required to gain a better understanding of this phenomenon.

Conclusion

This case report describes biventricular activation of RBBB-pattern VT originating from the RV in a patient

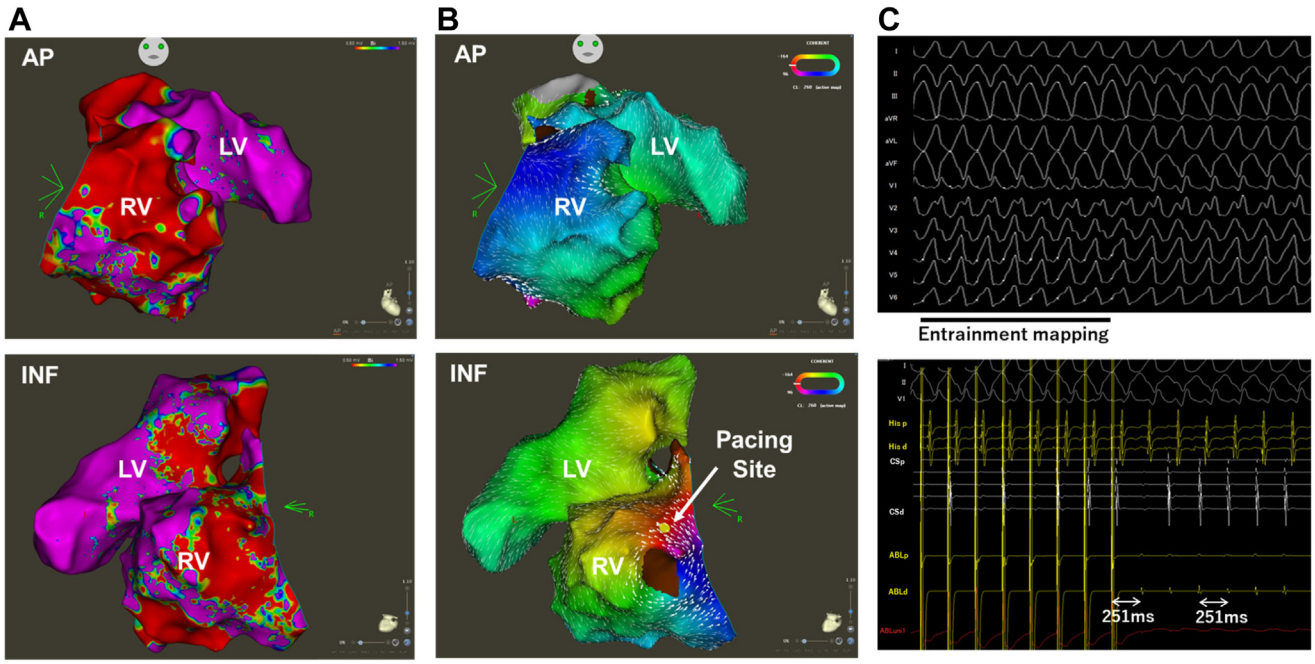

Figure 2 **A:** Voltage maps of the right ventricle (RV) and left ventricle (LV) in sinus rhythm. The following standard scar settings were used: scar <0.5 mV; border zone, 0.5–1.5 mV; and healthy myocardium >1.5 mV. **B:** Coherent maps of RV and LV in ventricular tachycardia (VT). The inferior (INF) view (lower panel) showed VT rotating around the scar in a counterclockwise manner. **C:** Surface electrocardiography (upper panel) and intracardiac electrography (lower panel) during entrainment mapping of VT from the pacing site (yellow tag in panel B). ABL = ablation catheter; AP = anteroposterior; CS = coronary sinus catheter.

with EA. These findings highlight the need to consider RV origin even when VT presents with RBBB morphology in patients with EA. Further studies are required to elucidate the mechanism and clinical implications of this distinct VT pattern in EA.

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Appendix

Supplementary Data

Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2023.10.023.

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