Three-dimensional mapping discovered arrhythmic substrate missed in the initial diagnosis of idiopathic ventricular fibrillation

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Abstract

A 45-year-old man who was diagnosed with idiopathic ventricular fibrillation (IVF) 12 years ago experienced multiple implantable cardioverter defibrillator shocks due to ventricular tachycardia (VT). The recorded electrocardiogram showed VT originating from the right ventricular outflow tract (RVOT). He underwent catheter ablation for VT, and 3-dimensional (3D) mapping revealed a low voltage area in the RVOT. VT was successfully ablated at this site, and his final diagnosis was scar-related VT, not IVF. We suggest that 3D mapping is useful for the reassessment of IVF because an arrhythmic substrate might have been missed in patients who were initially diagnosed with IVF.

Case report

Twelve years ago, a 33-year-old man (now 45 years old) with no medical or familial history except for syncope when playing golf developed sudden cardiac arrest (SCA) after drinking alcohol. His heartbeat was restored by an automated external defibrillator (AED), and polymorphic ventricular tachycardia (PVT) with a short cycle length (CL) was recorded at another hospital (Figure 1). Although various examinations including a pilsicainide provocation test were performed, no underlying heart disease was clear. Therefore, he was diagnosed with idiopathic ventricular fibrillation (IVF). An implantable cardioverter defibrillator (ICD) was not implanted because of his rejection. However, 5 years later, a second SCA occurred due to the same PVT, and he received an ICD implantation after successful resuscitation.

At present, the patient was admitted to our hospital due to frequent ICD shocks. His ICD record showed rapid monomorphic VT (mean cycle length, 185 ms), which was always triggered by the same premature ventricular contraction (PVC) (Figure 2). The morphology of the PVC was a left bundle branch block configuration with an inferior axis, and catheter ablation to the trigger PVC was performed. A three-dimensional (3D) voltage map of the right ventricle was constructed, and a low-voltage area (LVA) on the free wall of the right ventricular outflow tract (RVOT) was detected. A good pacemap was obtained at the border zone of the LVA, and the target PVC was ablated at the site. Shortly after discharge, he experienced the recurrence of ICD shocks and underwent a second ablation session the following month. However, the ablation failed again, and he revisited our hospital a month after the second session due to frequent ICD shocks. Monomorphic VT originating from the RVOT was recorded on an electrocardiogram (Figure 3A), and the third ablation session for recurrent VT was performed. The morphology of the VT was the same as the trigger PVC, which was frequently observed during his sessions. An almost perfect pacemap (score, 96) with PVC was obtained near the LVA, slightly posterior to the prior ablation site. The ventricular potential of the PVC on the ablation catheter showed up earlier than the QRS onset of any leads on the electrocardiogram at this site (Figure 3B–C). Both VT and PVC were successfully eliminated and not inducible after ablation. There were no ICD shocks one year after the last session.

Discussion

In this case, the recorded ventricular tachyarrhythmia on the ICD log at our hospital was monomorphic VT, not VF. On the other hand, the recorded log from the other hospital after SCA was polymorphic VT or VF. This difference could be explained as follows. VF has been reported to be sometimes promoted by VT accompanied with hemodynamic instability and would develop into VF or SCA.[1] Most VTs originating from the RVOT are considered as benign arrhythmias; however, some VTs can be malignant and develop into VF or polymorphic VT, and the characteristics of these malignant VTs are more likely to occur in patients with a history of syncope and short CL.[2] This patient had several events of syncope of unknown cause and monomorphic VT with very short CL (185 ms). Therefore, we estimated that his repetitive SCAs were primarily due to a malignant type of VT originating from the RVOT.

When a patient with SCA is taken to a hospital, there could be an initial lack of clinical history and information, and therefore the initial diagnosis of IVF might be incomplete. In fact, it has been reported that more than one-fifth of patients initially diagnosed with IVF are re-diagnosed with another specific disease; therefore, continuous follow-up and reassessment of patients with IVF has become very important.[3] This patient presented to our hospital with IVF and was uneventfully followed-up in the ICD clinic for several years. When multiple ICD shock events due to monomorphic VT were detected, we suspected that his diagnosis was possibly not IVF. In the subsequent ablation session, 3D mapping revealed the existence of LVA on RVOT, which showed that he had a structural arrhythmic substrate. Moreover, the induced ventricular arrhythmia, which was reproducibly monomorphic VT, showed a regular fixed sequence of ventricular potentials that arose from the LVA. Ablation of the LVA resulted in the successful elimination of the target PVC and VT. Based on these results, the patient was finally diagnosed with a scar-related VT, not IVF.

This is the first report of a patient who was diagnosed with scar-related VT via 3D mapping, even though he was initially diagnosed with IVF. In most cases, 3D mapping has been rarely performed as one of the initial diagnostic tests for IVF.[4] Therefore, we consider that a 3D mapping system might be useful in finding a missing substrate and to clarify another specific arrhythmic disease like in the present case.

Conclusion

We diagnosed a scar-related VT based on 3D mapping results in a patient who was initially diagnosed with IVF 12 years ago. When managing patients with IVF, continuous careful follow-up is very important, and 3D mapping could be useful for the reassessment of IVF because an arrhythmic substrate might have been missed.

References

1. Koplan BA, Stevenson WG. Ventricular tachycardia and sudden cardiac death. Mayo Clin Proc 2009;84:289-297.
2. Noda T, Shimizu W, Taguchi A, et al. Malignant entity of idiopathic ventricular fibrillation and polymorphic ventricular tachycardia initiated by premature extrasystoles originating from the right ventricular outflow tract. J Am Coll Cardiol 2005;46:1288-1294.
3. Visser M, van der Heijden JF, van der Smagt JJ, et al. Long-term outcome of patients initially diagnosed with idiopathic ventricular fibrillation: a descriptive study. Circ Arrhythm Electrophysiol 2016;9:e004258.
4. Waldmann V, Bougouin W, Karam N, et al. Characteristics and clinical assessment of unexplained sudden cardiac arrest in the real-world setting: focus on idiopathic ventricular fibrillation. Eur Heart J 2018;39:1981-87.

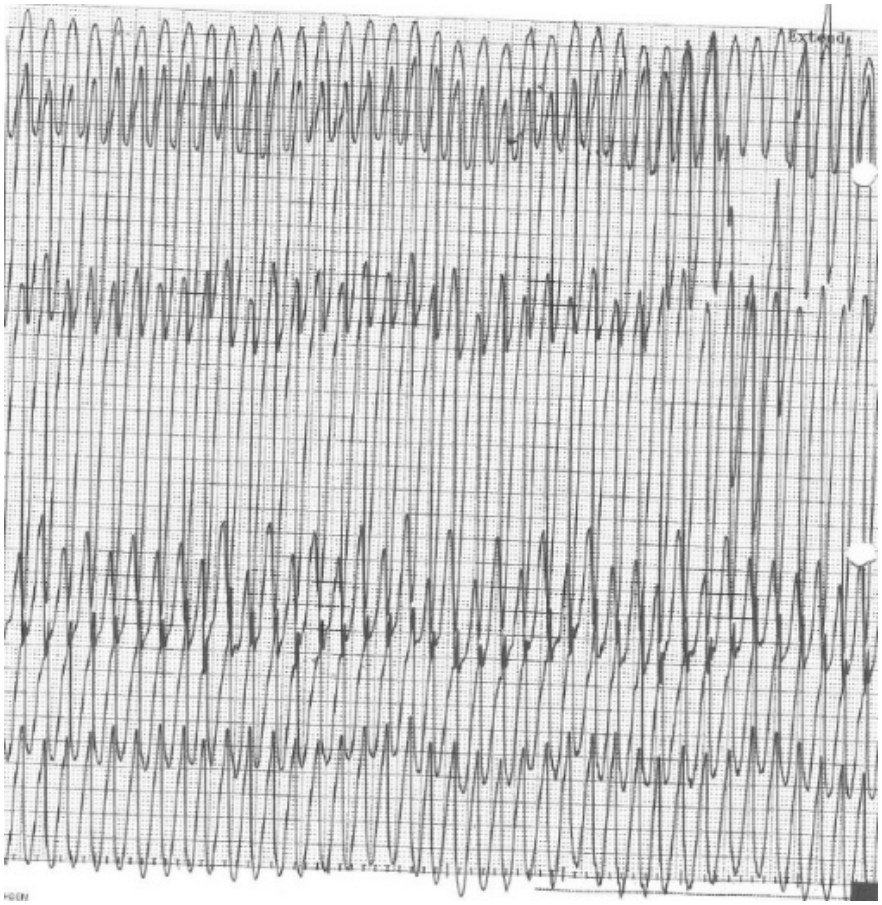
Figure Legends

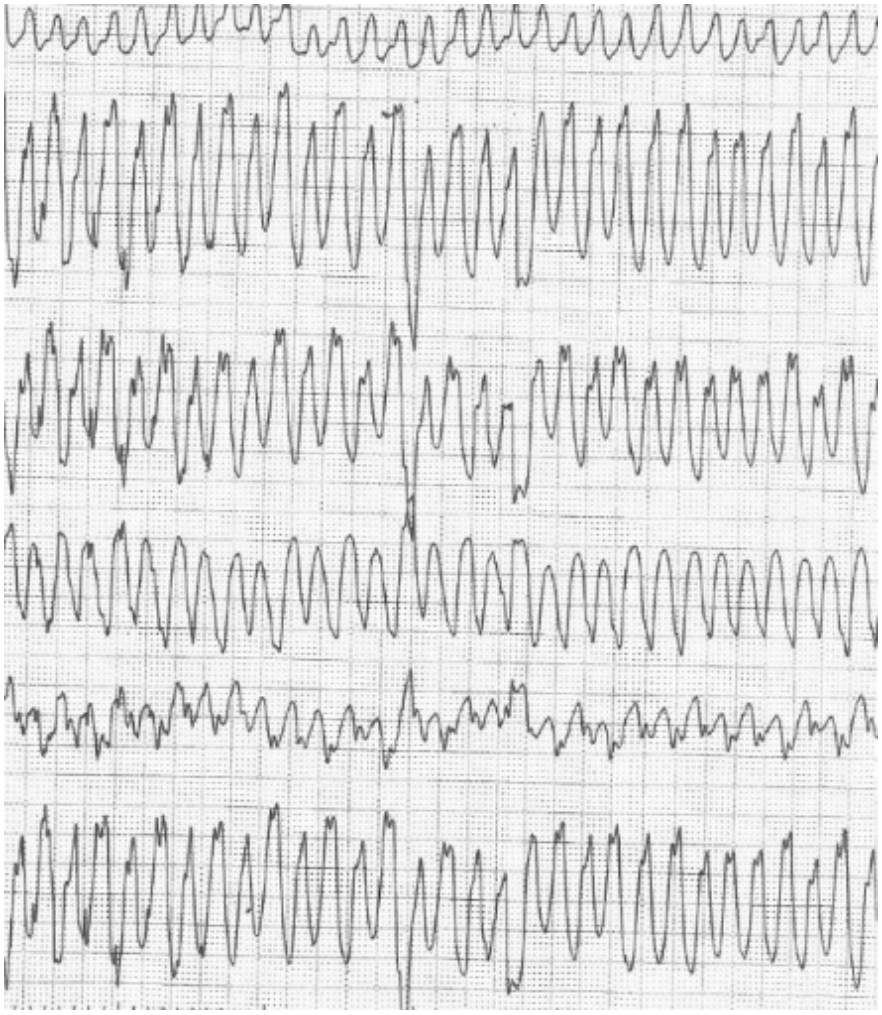
Figure 1. Electrocardiogram on his first admission at 33 years old. Polymorphic wide QRS complex was recorded. Heart rate was 330 bpm.

Figure 2. Intracardiac ICD record of the monomorphic VT. Monomorphic VT (mean cycle length, 185 ms) was recorded and consistently triggered by the same PVC. ICD: implantable cardioverter defibrillator; VT: ventricular tachycardia; PVC: premature ventricular contraction. **Figure 3-A. Electrocardiogram on admission when VT storm before 3rd ablation therapy.** The morphology of VT was a left bundle branch block configuration with an inferior axis. Its origin was estimated to be located in the right ventricular outflow tract. VT: ventricular tachycardia. **3-B. 3D voltage map during sinus rhythm.** Anterior to posterior view of 3D mapping of RV in the 3rd session. Blue tag indicates the successful ablation site for PVC, where good pacemap and a score of 96, was obtained. Brown tags indicate the ablation sites. Yellow tags indicate the pacemap site. 3D: three-dimensional; RV: right ventricle; PVC: premature ventricular contraction: PV: pulmonary valve; TV: tricuspid valve.

3-C. Intracardiac electrogram in the success site of 3rd ablation therapy. Distal part of ablation catheter (ABL 1U and ABL 1-2) detected depolarization 34 milliseconds earlier than any other leads of electrocardiogram. ABL: ablation; HBE: his bundle electrogram; RVA: right ventricular apex.

Figures





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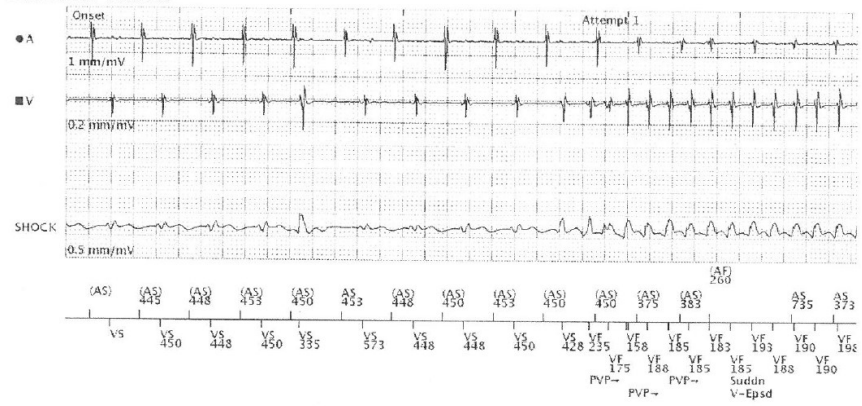

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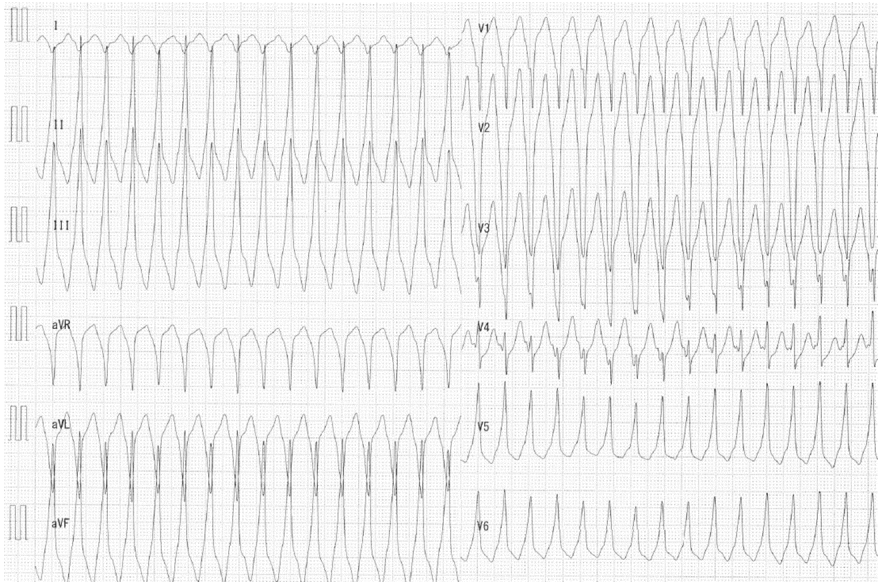
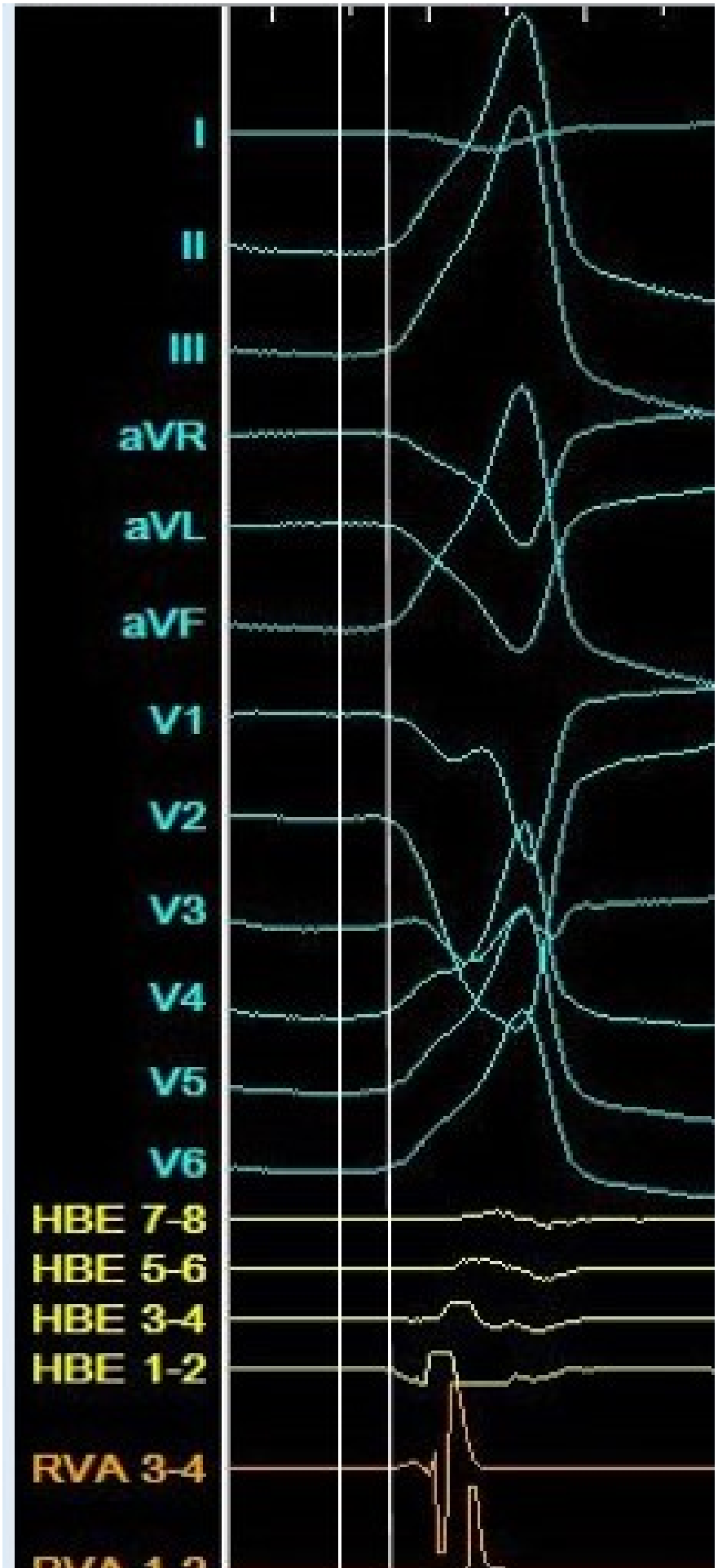
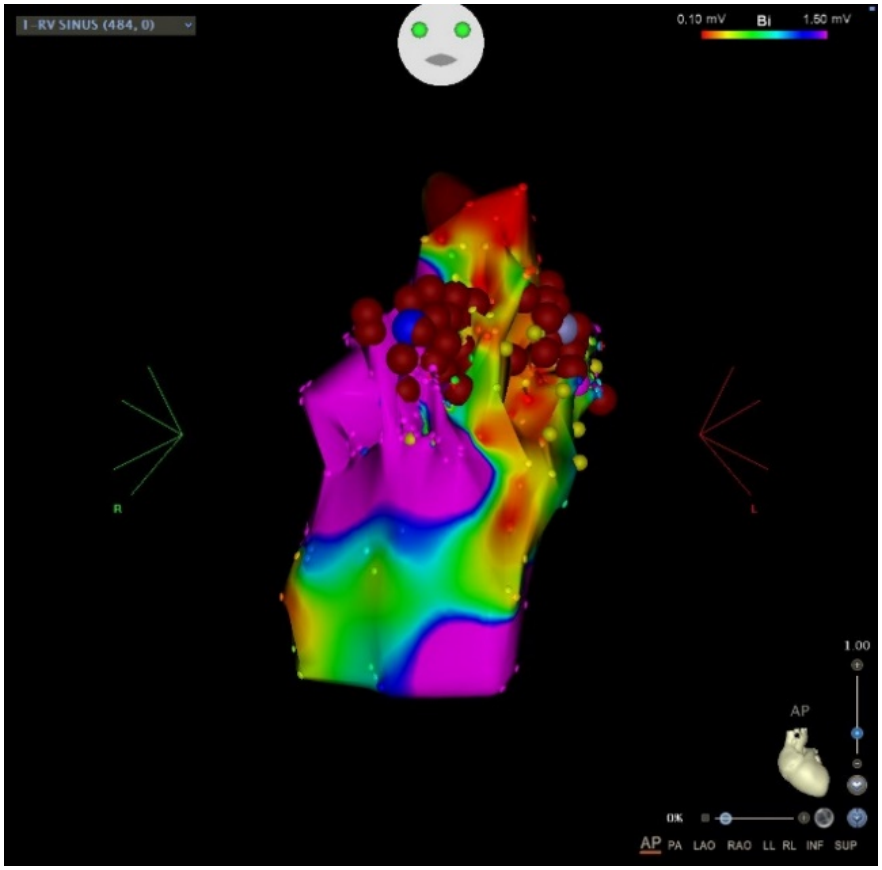




3-A 3-C



3-B