Idiopathic ventricular fibrillation triggered by premature ventricular complexes originating from the false tendon of the left ventricle

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Introduction

Idiopathic ventricular fibrillation (VF) is a diagnosis of exclusion when no evidence of a structural or metabolic cause is found. It is a rare cause of sudden cardiac death reported in 6.8% of patients who survive an out-of-hospital cardiac arrest and is more frequently seen in young adults.[1] Premature ventricular contractions (PVCs) originating from the Purkinje network can induce polymorphic ventricular tachycardia (PMVT) or VF in rare cases. The electrophysiologic mechanisms, although not completely clear, have been thought to be related to abnormal automaticity and triggered activity.[2] Besides the Purkinje network, other potential sources of PVCs include the right ventricular outflow tract, papillary muscles, the moderator band, and, rarely, the left ventricular false tendon (LVFT).[3]

Drug therapy with antiarrhythmic agents rarely prevents recurrent VF episodes, and catheter ablation of the PVCs triggering VF is potentially lifesaving.[4,5] Very few studies have looked at outcomes after radiofrequency (RF) catheter ablation in PVCs originating from the LVFT.[6,7] Our case is a rare presentation of PVCs originating from the LVFT triggering VF in a patient without structural heart disease.

Case report

A 59-year-old male patient with no prior medical or family history presented with 3 episodes of syncope preceded by palpitations for 2 days. Initial electrocardiogram showed sinus rhythm with a ventricular rate of 81 beats per minute, normal axis, positive Sokolow-Lyon criteria for left ventricular hypertrophy, and T-wave inversions in leads III and aVF (Figure 1A). The QT interval was normal with no pre-excitation or epsilon wave. After inpatient admission, he had recurrent syncopal episodes correlating with pulseless PMVT on telemetry requiring resuscitation and

KEY TEACHING POINTS

- Left ventricular false tendons (LVFTs) are endocavitary structures in the ventricle that comprise fibrous tissue, myocardial fibers, blood vessels, and Purkinje fibers. Premature ventricular complexes (PVCs) originating from LVFTs may be due to the increased automaticity of Purkinje cells within muscle fibers or increased excitability owing to mechanical traction at the false tendon attachment site.
- PVC triggering ventricular fibrillation (VF) storm is a true electrophysiological emergency and prompts careful evaluation. Treatment of PVC-triggered VF in patients without structural heart disease should first be aimed at studying and promptly removing any reversible proarrhythmic cause, like myocardial ischemia, electrolyte imbalances, QT-prolonging drugs, and inflammation.
- Radiofrequency ablation with an intracardiac echocardiography–guided electroanatomical approach should be considered in patients with PVCs originating from LVFTs to improve the efficacy and safety by direct visualization of these dynamic endocavitary structures.
- An implantable cardioverter-defibrillator is recommended for secondary prevention of sudden cardiac death in patients with idiopathic ventricular tachycardia/VF in the absence of reversible causes.

KEYWORDS Idiopathic VF; Short-coupled PVCs; Left ventricular false tendon; Radiofrequency ablation; Intracardiac echocardiography
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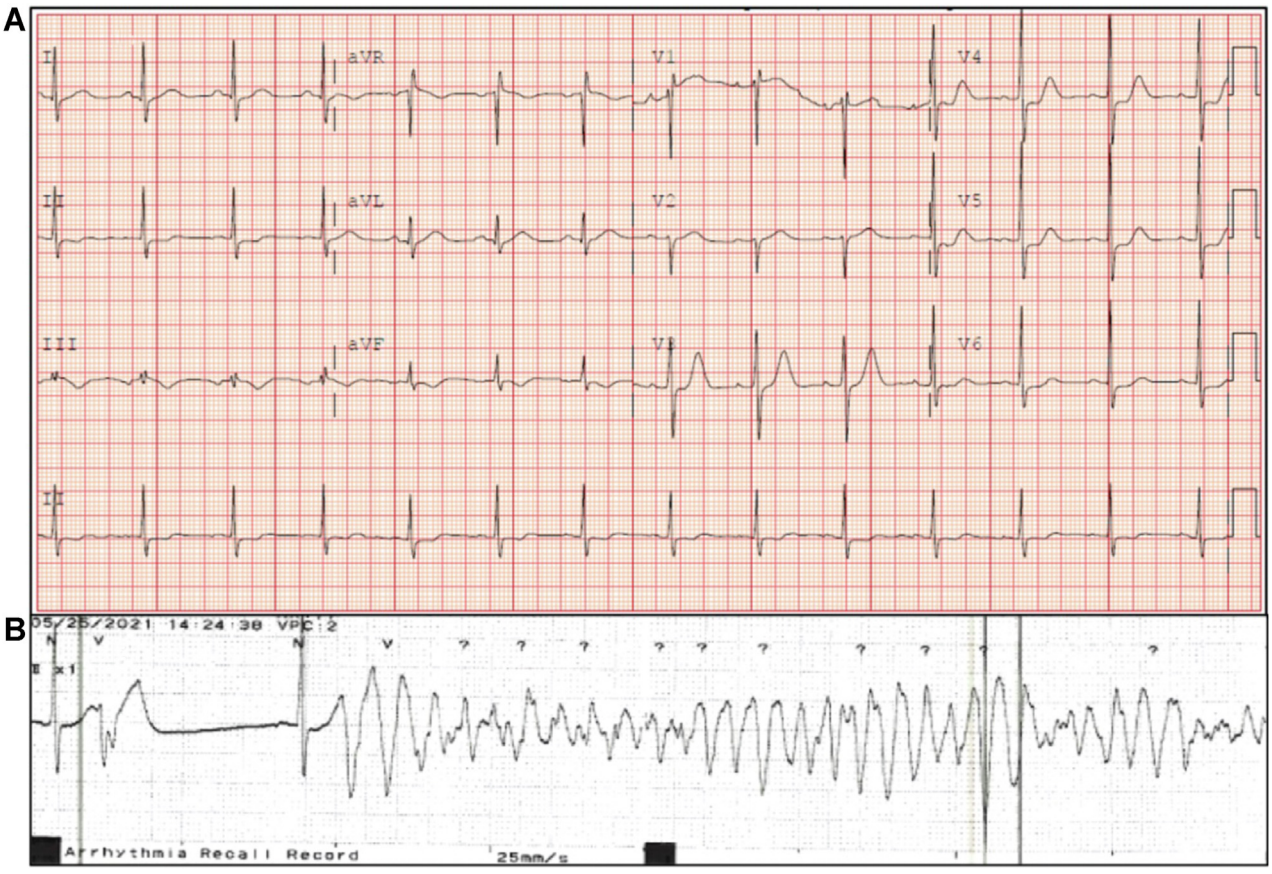

Figure 1 **A:** Initial electrocardiogram with sinus rhythm, normal axis, and left ventricular hypertrophy. **B:** Rhythm strip showing short-coupled premature ventricular contractions triggering ventricular fibrillation.

defibrillation (Figure 1B). The PMVT episodes appeared to be triggered by PVCs with a rapid initial deflection with rSR′ in V_1 and a short coupling interval (380 ms) to initiate VF each time. Echocardiogram was reported as mild left ventricular (LV) dysfunction with an ejection fraction of 40%–45% with normal right ventricular size and function. However, subsequent cardiac magnetic resonance imaging revealed normal LV size and thickness, normal ejection fraction (66%), and no evidence of LV noncompaction or infiltrative or inflammatory cardiomyopathy. Coronary angiography revealed no significant epicardial coronary artery disease.

At this point, a decision was made to pursue an endocardial-only catheter ablation to eliminate the PVCs initiating VF. The patient provided informed consent and was brought to the electrophysiology lab. The procedure was performed under general anesthesia. The patient presented to the electrophysiology lab in sinus bradycardia. He had a spontaneous episode of a PVC triggering VF requiring defibrillation with 360 J. PVC morphology triggering VF was right bundle, superior axis, with precordial transition in V_2 or V_3 and some variation in lead V_2 (Figure 2A). The patient subsequently had multiple spontaneous episodes of PVCs triggering VF requiring defibrillation 11 times. Intracardiac echocardiography (ICE) showed a trabeculated LV apex and a prominent false tendon. Endocardial definition of the LV and the papillary muscles was created using the CartoSound module for image integration. Electroanatomical mapping and RF ablation was performed using the Biosense Webster CARTO 3D electroanatomical mapping system (CARTO; Biosense Webster, Diamond Bar, CA). Transseptal access was obtained and pace mapping and activation mapping of the PVCs were performed in the LV using a contact force–sensing, irrigated ablation catheter (ThermoCool SmartTouch Surround Flow; Biosense Webster). Initial pace mapping demonstrated best pace-map correlation of only 80% over the apical septum. RF ablation was performed over the apex and distal LV inferoseptum but the patient continued to have salvos of PVCs with slightly varying morphologies in V_1 and V_2 triggering VF and requiring defibrillation. The earliest electrogram during PVCs preceding the salvos of PMVT was noted over a false tendon connecting the anterolateral papillary muscle and septum and was preceded by a very early high-frequency Purkinje potential (101 ms early) (Figure 2B). RF ablation was performed over the LV false tendon using high power of 45 watts, as the contact force was variable (2–6 g) over this dynamic, thin fibromuscular structure. Catheter stability was ensured by use of a deflectable sheath (Agilis Nxt; Abbott, Chicago, IL) and intracardiac echocardiography (Supplemental Video 1).

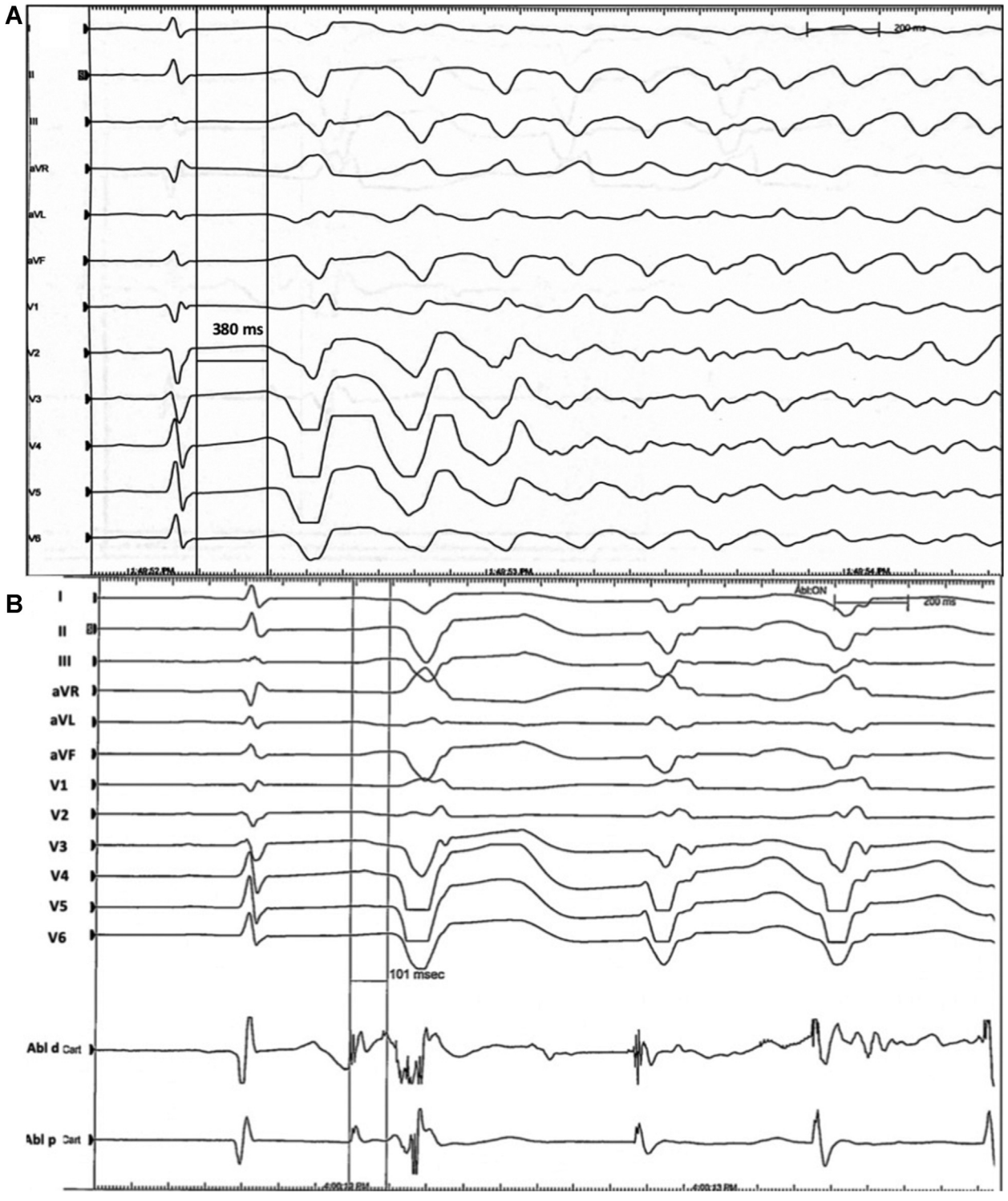

Figure 2 **A:** Ventricular fibrillation triggered by short-coupled premature ventricular contractions (PVCs) of right bundle morphology, superior axis, and negative precordial concordance. **B:** Signals recorded from the ablation catheter positioned over the false tendon. High-frequency, early Purkinje potential (101 ms pre-QRS) is noted on the second beat preceding the PVC.

At the end of the procedure, no PVCs triggering PMVT/VF were observed. The patient underwent an intracardiac dual-chamber implantable cardioverter-defibrillator placement for secondary prevention. Amiodarone was stopped at 1 month follow-up. Genetic testing was negative for arrhythmogenic right ventricular dysplasia, catecholaminergic PMVT, long QT syndrome, Brugada syndrome, and LV noncompaction. The patient had no recurrent ventricular tachycardia (VT)/VF and PVC burden was $<1\%$ on device interrogation at 6 months follow-up.

Discussion

The natural history of idiopathic VF is unknown and accounts for 8% of individuals with sudden cardiac death.[8] While this arrhythmia may be an early manifestation of a developing cardiomyopathy, idiopathic VF is diagnosed when the heart is structurally normal on imaging and no identifiable genetic cause or accessory pathway is found on testing. Traditionally idiopathic VF is seen in younger patients between ages 35 and 50 years. However, there are reports of idiopathic VF occurring at a later age. Almahameed and colleagues[9] describe a case of idiopathic VF in a 56-year-old female patient. They also outline baseline characteristics of idiopathic VF patients in published reports in which the age range is anywhere between 21 and 60 years of age.[9]

Most idiopathic ventricular arrhythmias are due to abnormal automaticity or a focal mechanism of triggered activity. The clinical manifestations of idiopathic ventricular arrhythmia are highly variable and range from benign, asymptomatic PVCs to sustained VT or, as seen in our case, VF.[3]

Anatomical considerations of the LVFT

LVFTs are single or multiple, thin or fibromuscular endocavitary structures in the ventricle that extend between the ventricular septum and the papillary muscles, LV free wall, or apex. They consist of fibrous tissue, myocardial fibers, blood vessels, and Purkinje fibers.[10] LVFTs are found more often on autopsy than on echocardiography, with improved detection rates since the advent of harmonic imaging. However, traditional imaging planes are unsuitable for detection and off-axis imaging is often required.[11] Clinical studies have suggested a significant correlation between LVFTs and PVCs.[12]

Zhang and colleagues[7] have described 10 patients with PVCs originating from the LVFT and successful RF ablation using an ICE-guided electroanatomical approach. The origin of PVCs was noted to be the sites of attachment of the false tendon to the LV septum, papillary muscle, or LV apex. High-frequency Purkinje potentials were observed in 7 of 10 patients at the target sites of ablation over the LVFT with the earliest timing pre-PVC being 36 ms.[7] Interestingly, in our patient, we noted a very early high-frequency Purkinje potential (101 ms pre-PVC), as shown in Figure 2B. Usually such early Purkinje/conduction system potentials are observed in bundle branch VT or fascicular VT. Supporting this theory, there are other studies that have reported that LVFTs may play important roles in forms of VT, such as idiopathic fascicular ventricular tachycardia.[13] Another notable finding is that Zhang and colleagues noted a pre-QRS Purkinje potential on the sinus beats in 7 of 10 patients, which we did not note in our patient.

Our case is unique in its clinical presentation of PVCs from the LVFT, which triggered VF storm and a malignant involvement of the Purkinje network in this location. The mechanisms of PVCs originating from LVFTs may be owing to the increased automaticity of Purkinje cells within muscle fibers or increased excitability owing to mechanical traction at the false tendon attachment site.[7]

PVC triggering VF storm is a true electrophysiological emergency and prompts careful evaluation. Treatment of PVC-triggered VF in patients without structural heart disease should first be aimed at studying and promptly removing any reversible proarrhythmic cause, such as myocardial ischemia, electrolyte imbalances, QT-prolonging drugs, and inflammation. There is paucity of data regarding effectiveness of antiarrhythmic drugs in this population. Quinidine has been shown to be effective, with a ventricular arrhythmia burden reduction in some short-coupled VF and idiopathic VF studies.[14,15] Implantable cardioverter-defibrillator implant is recommended for secondary prevention for patients who experience sudden cardiac death caused by VT/VF or sustained or hemodynamically unstable VT in the absence of reversible causes.[3]

Conclusion

LVFTs can be associated with PVCs that can trigger VF storm in structurally normal hearts. RF ablation with an ICE-guided electroanatomical approach should be considered to improve the efficacy and safety of this procedure by direct visualization of these dynamic endocavitary structures.

Appendix

Supplementary data

Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2022.05.006.

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