Epicardial radiofrequency catheter ablation of Brugada syndrome with electrical storm during ventricular fibrillation

A case report

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Abstract

Rationale: Brugada syndrome (BrS) is characterized by ST segment elevation at the J point ≥2 mm in the right precordial electrocardiogram (ECG) leads, in the absence of structural heart disease, electrolyte disturbances, or ischemia. It is a well-described cause of sudden death in young patients, especially in the age of between 30 and 40 years old. Here, we reported an unusual case of electrical storm (ES) of ventricular fibrillation (VF) caused by BrS with complete right bundle-branch block (CRBBB) in a 75-year-old male patient.

Patient concerns: A 75-year-old male patient survived sudden cardiac death caused by a ventricular ES. He presented with the cove-shaped ST elevation of 2 mm in lead V1 with typical CRBBB and lacked structural cardiomyopathy and coronary heart disease. The patient suffered ventricular ES again, although the implantable cardioverter defibrillator (ICD) had implanted.

Diagnoses: Brugada syndrome with complete right bundle-branch block.

Interventions: Implantable cardioverter defibrillator (ICD) implantation was performed. But this therapy could not prevent the recurrence of malignant arrhythmia. Finally, the ES was treated successfully using radiofrequency catheter ablation (RFCA) at the area of the free wall of the right ventricular outflow tract (RVOT) epicardium.

Outcomes: During 7 months of follow-up, the patient was asymptomatic and free of arrhythmic events.

Lessons: As far as we know, the patient is the oldest patient reported to have BrS. RFCA offers an alternative therapy for patients with BrS, especially when ICD shocks are encountered.

Abbreviations: BrS = Brugada syndrome, CRBBB = complete right bundle-branch block, ECG = electrocardiogram, ES = electrical storm, ICD = implantable cardioverter defibrillator, RFCA = radiofrequency catheter ablation, RVOT = right ventricular outflow tract, VF = ventricular fibrillation.

Keywords: Brugada syndrome, complete right bundle-branch block, implantable cardioverter defibrillator, radiofrequency catheter ablation, ventricular fibrillation.

1. Introduction

Brugada syndrome (BrS) is an autosomal dominant genetic heart disease with variable penetrance that affects the sodium channel. It is reported to be responsible for 4% of all sudden deaths and 20% of sudden deaths in patients without structural heart disease. The most typical manifestation is syncpe or resuscitated sudden death in the third or fourth decade of life at night or at rest during the day, caused by ventricular tachycardia (VT) or ventricular fibrillation (VF). Thus, BrS is a serious health threat, especially in people aged 30 to 40 years old. In this study, we report a case of ES of VF caused by BrS with complete right bundle-branch block (CRBBB) in a 75-year-old male patient.

2. Consent

The patient signed the necessary documents to consent to the use of his data for teaching and publication.

3. Case report

A 75-year-old male patient was admitted to our hospital at night, who had survived sudden cardiac death caused by a ventricular electrical storm (ES). He had no history of tobacco or drug abuse, and was nondiabetic and normolipidemic, with normal electrolytes and cardiac enzyme levels. His chest x-ray and echocardiography were normal, and a coronary angiogram revealed normal coronary arteries. An initial electrocardiogram (ECG) showed sinus rhythm with CRBBB and J point elevation was observed at 2 mm in lead V1. Unfortunately, ajmaline is not available in China; therefore, we could not perform a provocative
test. Instead, we placed the precordial leads V1, V2, and V3 into the chest in the second and third intercostal space. Before ablation, there was more accentuation in the higher precordial leads (Fig. 1). Questioning the patient revealed that his brother died of sudden death at the age of 30. The clinical manifestation, abnormal ECG, special family background, and the lack of structural cardiomyopathy and coronary heart disease, suggested that the patient suffered from BrS. The patient was implanted with an ICD according to the Position Statement on the Diagnosis and Management of BrS.[1]

A month later, he was readmitted to our hospital after 2 discharges of the ICD caused by VF during the night. That night, the patient suffered ventricular ES again, with 58 consecutive ICD shocks. Subsequently, we decided to perform RFCA to control the malignant arrhythmia. During the electrophysiological study, neither abnormal electrical activity nor low voltage maps were recorded in the endocardium of right ventricle. We suspected epicardial right ventricular outflow tract (RVOT) exit[2]; therefore, epicardium voltage mapping was performed. We recorded abnormal epicardial electrograms characterized by fragmented and late potential, exhibiting low voltage in the area of the free wall of the RVOT epicardium (Fig. 2). We then performed radiofrequency catheter ablation (RFCA) at those regions, which abolished all the abnormal potentials. After 7 months of follow-up, the patient was asymptomatic and interrogation of the ICD showed that he was free of arrhythmic events; the J point was degraded obviously in precardial leads V1 and V2 (Fig. 1).

In addition, a genetic test was conducted. Abnormality of the SCN5A gene was not detected, but mutations in the CAN1, FGF12, and MYL3 genes were revealed (Fig. 3).

4. Discussion

BrS was first described as a clinical entity in 1992. It is characterized by ST segment elevation at the J point ≥2 mm in the right precordial ECG leads, in the absence of structural heart disease, electrolyte disturbances, or ischemia. Recently, Aizawa et al[3] found that BrS could coexist with CRBBB. Furthermore, CRBBB could completely mask BrS on ECG (<2 mm elevation of the J point at the time of CRBBB in the right precordial leads at the fourth intercostal space on ECG).[4] Thus, our patient qualified clinically as having BrS with CRBBB (cove-shaped ST elevation of 2 mm in lead V1 with typical CRBBB in the ECG and more accentuation in the higher precordial leads). The prevalence of CRBBB in patients with BrS is low. A study showed that among 326 patients with BrS, only 25 had CRBBB (7.7%).[4] By contrast, BrS is a well-described cause of sudden death in people aged between 30 and 40 years. Although Wada et al reported that patients with BrS patients and CRBBB were significantly older (54.2 ± 14.5 years) than patients with BrS but without CRBBB[4]; the present patient was 75-year-old. As far as we know, he is the oldest patient reported to have BrS.

Kamakura et al[5] found patients with BrS aged ≥70 years might belong to a group of patients at lower risk of VF compared with younger patients, and the avoidance of ICD implantation or replacement should be considered. However, in this case, the patient survived cardiac arrest at 75 years old. ICD implantation was performed; however, fortunately, he was rescued from the appropriate ICD shock. This event indicated that ICD remains a cornerstone for the treatment of high-risk BrS, regardless of the age of the patient.

In our case, we could not ignore another obvious problem: Despite ICD being able to terminate VF, it cannot prevent the recurrence of malignant arrhythmia. Frequent ICD shocks can result in depression, posttraumatic stress, or other factors that would affect the patient’s quality of life seriously. Up to 48% of such high-risk patients experience frequent ICD shocks.[6] Although quinidine and isoproterenol are effective in suppressing VF,[1] they are generally proarrhythmic. In addition, patients find it difficult to tolerate them over a lifetime because of their side effects.[7] Finally, we chose RFCA to control the malignant arrhythmia.
arrhythmia. During 7 months of follow-up, the patient was asymptomatic and free of arrhythmic events. Nademanee et al showed that during a mean of almost 2 years of follow-up, there were no recurrent VT/VF episodes requiring ICD discharges in any of the study patients. These results indicated that catheter ablation offers an alternative therapy for patients with BrS, especially when ICD shocks are encountered. In addition, Talib et al were the first to demonstrate the feasibility of “Ablation and implantation” as an alternative approach to suppress ES, especially when the origin of the culprit premature ventricular contraction is easily accessible.

The role of the arrhythmogenic substrates in BrS has been studied extensively. Although the evidence suggests abnormal activation of the RVOT, which is coincident with the ST segment elevation of the Brugada ECG, the target site for catheter ablation varies. In our case, we found that the low-voltage zones with fractionated and delayed potentials were located at the epicardial, rather than the endocardial, RVOT, and the electrocardiographic features and substrate patterns were similar as those found by Nademanee. Nevertheless, the epicardial RFCA of abnormal electrograms localized in the area of the free wall of the RVOT were different and larger than those reported by Nademanee. In a recent study, Josep Brugada found that after flecainide testing, the low-voltage areas expanded by about 2 times in almost all patients, accompanied by subsequent elimination of arrhythmogenic substrates, which in many patients extended beyond the anterior aspect of the RVOT. They pointed out that the epicardial areas with variable extension and distribution were located mainly in the right ventricular free wall and RVOT, as revealed by flecainide testing. This phenomenon indicates that the area of arrhythmogenic substrates maybe wide. Besides, as far as I know, this is the first report of radiofrequency ablation of BrS.

Figure 2. The map A shows the voltage map of the endocardial right ventricle. The map B shows the voltage map and electrograms of right ventricular outflow tract (RVOT) epicardium. Both the voltage maps show low voltage (≤1 mV) in red and high voltage in purple. Red dots represent ablations. The map A express no signs of low-voltage zones. By contrast, low voltage zones are located in the area of the free wall of the RVOT epicardium shown in B. The right side of the map in B displays the abnormal electrograms recorded from the pentaray catheter at the site of posterior-lateral aspect of the RVOT epicardium. Abnormal electrograms are defined as electrograms that have low voltage (≤1.0mV); split electrograms; wide duration (>80 ms); or late potentials.
combined with CRBBB, and whether CRBBB affects its ablation area remains to be studied.

BrS is an autosomal dominant channelopathy. Mutations in the SCN5A gene, which encodes the Nav1.5 protein forming the α subunit of the sodium channel, are common in BrS, accounting for approximately 20% to 25% of cases.[1] In our case, no abnormality in the SCN5A gene was detected. Nevertheless, we also identified the mutation in CACNA2D1. In 2007, Anzelevitch et al. identified loss of function mutations involving the L-type calcium channel alpha1, beta2, and alpha2delta subunits encoded by the CACNA1C, CACNB2B, and CACNA2D1 genes in probands with BrS. Furthermore, the 3 mutations may cause 10% to 15% of BrS, respectively.[10] Of the known BrS susceptibility genes, the loss-of-function mutations in SCN5A or CACNA1C and their auxiliary subunits are most common.

Besides, we identified heterozygous splicing mutation in FGF12. Based on the present research, the fibroblast growth factor (FGF) homologous factors regulate cardiac Na+ and Ca2+ channel currents. Hennessy JA et al.[11] suggest that FGF12 is a BrS locus, and Q7R-FGF12 that affects Na+ channel trafficking and kinetics with minimal effects on Ca2+ channel function is a disease-associated BrS mutation using a native cardiomyocyte system.

Furthermore, we found a novel missense mutation in the MYL3 gene, with a nucleotide change at 42G>T and an amino acid change comprising Lys14Asn (P.Lys14Asn). Mutations in the MYL3 gene, which encodes a component of myofilaments, are usually relevant to hypertrophic cardiomyopathy. Thus, the genetic mutations associated with BrS require further research.

In conclusion, according to what we are informed, the patient is the oldest patient reported to have BrS. RFCA offers an alternative therapy for patients with BrS, especially when ICD shocks are encountered.

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