Radiofrequency catheter ablation for drug-refractory paroxysmal atrial fibrillation in a patient with Ebstein’s anomaly

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Introduction

Ebstein’s anomaly is a rare congenital heart disease in which the tricuspid valve is displaced toward the apex of the right ventricle (RV). It is commonly associated with atrial arrhythmia, especially Wolff-Parkinson-White syndrome. In fact, up to 20% of patients with Ebstein’s anomaly have 1 or more accessory pathways owing to tricuspid valve malformation, most of which are located along these tricuspid valves. In addition, atrial fibrillation (AF) can develop with aging. Although the success rate of catheter ablation of an accessory pathway is lower in patients with Ebstein’s anomaly than in the general population, catheter ablation is the most favorable treatment strategy. In patients with AF, however, concomitant surgical ablation at the time of corrective surgery is preferred. To the best of our knowledge, there has been no report of catheter ablation without corrective surgery for AF in a patient with Ebstein’s anomaly. Here, we present the case of an adult patient with Ebstein’s anomaly who developed drug-refractory and intolerant paroxysmal AF, but did not yet require corrective surgery.

Case report

A 39-year-old man visited the emergency department for recurrent palpitations. He had been diagnosed with Ebstein’s anomaly (Carpentier type B) at 27 years of age. He had a history of receiving radiofrequency catheter ablation (RFCA) for Wolff-Parkinson-White syndrome at 38 years of age at our clinic. A single bidirectional atrioventricular accessory pathway was identified in the right lateral region of the true anatomical tricuspid annulus. This accessory pathway was treated successfully with RFCA using a 3-dimensional (3D) mapping system (EnSite Velocity, St. Jude Medical). Subsequently, the patient had no palpitations, and electrocardiography revealed no sign of ventricular pre-excitation. However, palpitations recurred after 6 months, the characteristics of which were slightly different from the previous palpitations associated with Wolff-Parkinson-White syndrome. Electrocardiography revealed AF without ventricular pre-excitation. We initially prescribed a beta-blocker, sotalol, and amiodarone, but the AF was not controlled during the following 5 months. Because the patient was highly symptomatic (he visited the emergency department once a month, and direct-current cardioversions were performed twice during 5 months), we considered ablation therapy via a catheter or surgical approach.

Echocardiography was performed on admission. The right heart was globally enlarged and the function of the RV was decreased. The insertion of the septal tricuspid valve was markedly displaced toward the apex (the linear distance between this point and the septal insertion of the anterior mitral leaflet was approximately 57 mm). Motion of the anterior tricuspid leaflet was limited because of tethering at multiple points on the free wall of the RV. On color Doppler echocardiography, single tricuspid regurgitation was noted, which was considered to be moderate based on the color jet area method (Figure 1). Chamber size and systolic function of the RV were stationary over 10 years. The patient had no symptoms except for palpitations, and his exercise capacity was tolerable and stationary. Based on these findings, corrective surgery was not yet required, and the patient did not want open heart surgery. Therefore, we decided to perform RFCA for drug-refractory paroxysmal AF.

After we obtained written informed consent, dexmedetomidine and midazolam were administered for deep sedation. The entire procedure was performed via peripheral venous access and transseptal access across the interatrial septum to approach the left atrium (LA). Intravenous heparin was administered to maintain an activated clotting time >250 s after transseptal puncture. A multimode duodopolar catheter was placed in the right atrium (RA) and coronary
sinus via the left femoral vein. A 20-pole circular mapping catheter was used for both mapping and confirmation of pulmonary vein antrum isolation (PVAI). Ablations were performed using an open irrigated-tip catheter, and a 3D mapping system was used to guide the procedure. The size of the RA was huge (anteroposterior diameter, 74 mm), while the LA was slightly enlarged (anteroposterior diameter, 41 mm). We checked the trigger of AF before ablations and found the atrial premature beat from the right inferior pulmonary vein that triggered the AF. Circumferential PVAI was achieved successfully. However, sustained AF was induced by rapid atrial pacing after PVAI. Therefore, linear ablations of the roof, mitral isthmus, and cavotricuspid isthmus (CTI) were performed (Figure 2). Linear ablations of the roof and mitral isthmus were performed according to standard techniques.6 Ablations of CTI were difficult because of the patient’s huge RA and atrialization of part of the RV. Nonetheless, bidirectional block of the CTI was achieved successfully. The patient’s AF was organized and converted to atrial flutter and finally terminated during CTI ablation. We rechecked and confirmed the PVAI and bidirectional block of the roof and CTI line. However, bidirectional block of the mitral isthmus could not be achieved. Neither sustained AF nor atrial flutter (AFL) was induced after ablation. The procedure was completed without any complications. Low-dose sotalol (20 mg twice daily) was prescribed postoperatively because of the persistent arrhythmogenic substrate of the RV and the failed linear ablation of the mitral isthmus. The patient had no further palpitations, and there was no documented AF or AFL during the 15-month follow-up. Postprocedure and 1-, 3-, 6-, and 12-month Holter monitoring did not show any sustained paroxysmal AF or AFL.

Discussion

Ebstein’s anomaly is a rare congenital heart disease that is frequently associated with supraventricular tachycardia. Although accessory pathway-related arrhythmia is most common—occurring in 6%–30% of patients with Ebstein’s anomaly—AFL or AF also may develop. AFL is usually observed after surgery involving atriotomy or other scar tissue. Catheter ablation is the treatment of choice for AFL, and has a short-term success rate of approximately 80%–90% since the introduction of 3D mapping and irrigated-tip ablation catheter.7,8 AF also may be observed in Ebstein’s anomaly, especially in older patients, the mechanism of which may be explained by secondary change of RA myocardium from previous cardiac surgery or chronic hemodynamic stress toward the LA.8,9 Antiarrhythmic drugs may be prescribed, but their use is restricted by structural heart disease. Furthermore, the long-term prognosis of medical treatment is poor. According to American College of Cardiology/American Heart Association guidelines,5 a biatrial Maze procedure is recommended for patients with Ebstein’s anomaly and AF who require corrective surgery because of hemodynamic problems. To the best of our knowledge, there has been no report of RFCA for AF in a patient with Ebstein’s anomaly. This may be explained by the rare incidence of Ebstein’s anomaly (1 in 20,000 live births). In addition, most patients with AF require corrective surgery for hemodynamic reasons. In contrast, our patient developed AF when corrective surgery was not yet required. Therefore, we believe there is a group of patients with

Figure 1  Two-dimensional (A) and color Doppler (B) echocardiographic images. A: Right atrium and right ventricle were globally enlarged. Insertion of septal leaflet (white arrow) was markedly displaced toward the apex. B: Moderate tricuspid regurgitation (central jet area 7.84 cm²). aRV = atrialized right ventricle; fRV = functional right ventricle; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.
Ebstein’s anomaly and AF who do not yet require corrective surgery. In the present case, we performed RFCA successfully without any procedure-related complications, and there was no recurrence of AF or AFL during the 15-month follow-up. Our RFCA strategy focused on the LA, similar to patients with AF without structural heart disease. It seems that the main cause of AF in patients with Ebstein’s anomaly is a huge RA; thus, RFCA focusing on the LA was not effective. However, our results of RFCA were satisfactory, suggesting that the pulmonary vein trigger and LA substrate are still main causes of AF in patients with congenital heart disease, such as Ebstein’s anomaly. Although it is difficult to modify the substrate of a huge RA, modification of the pulmonary vein trigger and LA substrate with congenital heart disease is achievable. Some may doubt the efficacy of RFCA because of the patient’s concomitant use of an antiarrhythmic drug (sotalol). However, sotalol was not effective before RFCA. Thus, at the very least, RFCA successfully reduced the burden of AF until it responded to the antiarrhythmic drug.

Conclusion
Based on this case, RFCA could be an alternative treatment for drug-refractory paroxysmal AF in patients with Ebstein’s anomaly who do not yet require corrective surgery. In addition, our case highlights the importance of the pulmonary vein trigger and LA substrate in the pathogenesis of AF even in patients with apparent right-side heart diseases such as Ebstein’s anomaly.

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