Radiofrequency Catheter Ablation in Familial Paroxysmal Supraventricular Tachycardia Due to Accessory Atrioventricular Pathways

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Radiofrequency catheter ablation (RF-CA) has been widely used to cure paroxysmal supraventricular tachycardia (PSVT). However, its use has never been reported in familial PSVT caused by an accessory atrioventricular pathway (AP), which is known as one of the typical familial cardiovascular diseases. Two cases of using RF-CA for familial PSVT due to APs are presented, in a brother and sister, supporting a potential genetic role in the developmental failure to lose the atrioventricular connection during fetal life. The sister, a 24-year-old woman, had intermittent episodes of palpitation accompanied by chest pain for 2 years. An electrophysiologic study (EPS) confirmed her clinical tachycardia was atrioventricular reentrant tachycardia (AVRT) due to a left lateral concealed AP, which was subsequently successfully ablated with RF-CA. The brother, a 22-year-old man, had a 5-year history of paroxysmal palpitation. A resting electrocardiogram showed a right bundle branch block and left axis deviation with a delta wave. During his EPS, AVRT was reproducibly induced and a manifest AP was localized and then ablated at the left posteroseptal site, resulting in disappearance of the delta wave. PSVT, however, recurred 1 month later and during a repeat EPS the tachycardia was proved to be AVRT due to a right anterior concealed AP. The right anterior AP was successfully ablated with RF-CA. Both patients remained asymptomatic for more than 3 years following the successful ablation procedures. (Jpn Circ J 1998; 62: 883–886)

Key Words: Accessory pathway; Familial supraventricular tachycardia; Radiofrequency catheter ablation

Familial cardiovascular diseases are gaining more interest than before as current molecular studies are successfully demonstrating the genetic basis of many familial diseases. The familial type of Wolff-Parkinson-White (WPW) syndrome, which is caused by an accessory atrioventricular pathway (AP) bridging the atrium and ventricle, has been considered a typical familial cardiovascular disease. An AP conducts impulses in an anterograde, retrograde, or both directions. The classical surface electrocardiogram (ECG) of manifest WPW syndrome, in which the AP has an anterograde conduction property, is characterized by a short PR interval and a delta wave. The diagnosis of a concealed AP can be made when the 12-lead ECG shows no evidence of preexcitation but the electrophysiologic study (EPS) demonstrates an AP capable of only retrograde conduction. However, concealed APs that conduct impulses only retrogradely do not produce an ECG abnormality, thereby rendering it impossible to even suspect their existence in asymptomatic patients.

APs, whether or not their existence is evident on the surface ECG, can serve as an anatomic substrate for atrioventricular reentrant tachycardia (AVRT), which is one of the most common causes of paroxysmal supraventricular tachycardia (PSVT). Although many cases of familial WPW syndrome have been reported, familial PSVT due to concealed APs has been rarely reported probably because AVRT due to a concealed AP is much more difficult to diagnose than WPW syndrome. Almost always requiring an EPS, radiofrequency catheter ablation (RF-CA) of PSVT caused by APs has been shown to be very effective and safe and has been accepted as the first choice of treatment. Numerous studies on using RF-CA for curing PSVT have been reported. However, to our best knowledge, successful use of RF-CA in familial PSVT due to APs has never been reported. Therefore, we report our recent experience of using RF-CA in familial PSVT caused by a concealed AP in a woman and by 2 APs (1 manifest and 1 concealed) in her brother.

Case Reports

Case 1
A 24-year-old woman, the third of 4 children (3 boys and 1 girl), presented with a 2-year history of intermittent sudden palpitation. She stated that her palpitation started and stopped abruptly. It usually lasted about 20–24 h and did not evoke any other discomfort other than occasional chest pain. She had been observed for a hemodynamically insignificant small ventricular septal defect (Qp/Qs=1.2) for 10 years. However, 2–3 months prior to admission, the episodes of sudden palpitation became more frequent and did not stop spontaneously, requiring pharmacologic intervention.
On physical examination, a pansystolic murmur was heard at the cardiac apex and the left lower sternal border. Blood pressure was 120/70 mmHg and the pulse was 96 beats/min. A 12-lead ECG revealed a normal sinus rhythm at a rate of 88 beats/min, incomplete right bundle branch block (RBBB), and left ventricular hypertrophy (Fig 1A, upper panel). During an episode of paroxysmal palpitation, a narrow QRS-complex regular tachycardia at a rate of 195 beats/min was recorded on the surface ECG and retrograde P waves were clearly seen 120 ms after the onset of the preceding QRS complexes. This suggested AVRT (Fig 1A, lower panel). A 2-dimensional echocardiography revealed a small membranous ventricular septal defect.

Cardiac EPS was performed. Two 6F quadripolar electrode catheters (DAIG Co, MN, USA) were inserted via the right and left femoral veins and positioned at the His bundle region and the right ventricular apex, respectively. A 7F deflectable quadripolar ablation catheter with a 4-mm tip (Webster Inc, CA, USA) was inserted via the right femoral vein and positioned at the right atrial appendage for the diagnostic EPS. During the diagnostic EPS, a narrow QRS tachycardia was reproducibly induced with atrial premature depolarization (APD), ventricular premature depolarization (VPD), and ventricular burst pacing. The earliest atrial activity was localized at the lateral coronary sinus, indicating that her clinical tachycardia was AVRT using a left lateral concealed AP. The AP was then successfully ablated in 1.3 s after delivering 30 W of RF energy using the retrograde aortic approach (Fig 2, upper panels). During delivery of the RF, the retrograde atrial activation sequence changed substantially as the earliest atrial activation shifted from the ablation site to the His bundle recording site (Fig 2, lower panels). After ablation of the AP, tachycardia was not able to be induced. She has remained asymptomatic for 3.5 years.

Case 2

A 22-year-old man, the younger brother of case 1, had suffered from intermittent paroxysmal palpitation for 5 years. His palpitation developed suddenly without specific cause and usually lasted from 20 min to 1 h. His past medical history was unremarkable. Physical examination failed to reveal any abnormalities. Blood pressure was 110/70 mmHg and pulse was 80 beats/min. A resting 12-lead ECG showed sinus rhythm at a rate of 83 beats/min, incomplete RBBB, and delta waves that were positive in leads I, II and V2–V6, isoelectric in leads III and aVF, negative in aVR, and QRS transition between V1 and V2 (Fig 1B, upper panel). Unfortunately, an ECG had never previously been recorded during a palpitation attack. An EPS was performed under the preliminary diagnosis of WPW syndrome, based on the delta waves on the surface ECG and the recurrent attacks of paroxysmal palpitation. Two 6F quadripolar electrode catheters (DAIG Co) were inserted via the right and left femoral veins and positioned at the His bundle region and the right ventricular apex, respectively. A 7F deflectable quadripolar ablation catheter with a 4-mm tip (Webster Inc) was inserted via the right
The catheter was advanced through the femoral vein and positioned at the right atrial appendage. A narrow QRS tachycardia at a cycle length of 320 ms was reproducibly induced. During the tachycardia, the earliest retrograde atrial activation was observed at the left posteroseptal area. A left posteroseptal AP was then successfully ablated at the ventricular insertion site 1.2 s after applying 30 W of RF energy (Fig 3, left panels). As a result, the delta waves were eliminated with normalization of the PR interval, and the QRS transitional zone was shifted to a position between V3 and V4. Also, the retrograde atrial activation sequence was clearly changed, showing retrograde atrial activation at the His bundle electrode earlier than that at the ablation site of the left posteroseptum (Fig 3, lower left panel). After ablation of the left posteroseptal AP, no tachycardia was inducible in the baseline state, but a rapid supraventricular tachycardia with RBBB at a rate of 240 beats/min was induced with double APDs during the infusion of isoproterenol at a rate of 2 μg/min. We suspected at the time that the tachycardia was nonclinical atrial tachycardia, in that the tachycardia was very fast and was induced only during isoproterenol infusion. We therefore recommended him to watch for recurrence of his previous symptoms. He was discharged without medication.

One month after the initial catheter ablation, the palpitation recurred. The ECG recorded during the attack showed a regular wide QRS tachycardia with a RBBB pattern and a rate of 230 beats/min (Fig 1B, lower panel). Retrograde P waves were visible 120 ms after the onset of the QRS complexes. A repeat EPS revealed that the tachycardia was AVRT due to a right anterior concealed AP, which was then successfully ablated at the atrial insertion site via the right femoral vein (Fig 3, right panels). After ablation of the second bypass tract, retrograde ventriculoatrial conduction disappeared (Fig 3, lower right panel). He has remained asymptomatic with no recurrence of the delta waves for 3 years.

**Discussion**

Accessory atrioventricular pathways are known to result from the unpredictable developmental failure to lose the atrioventricular connections during fetal cardiogenesis. Since Öhnell reported familial WPW syndrome in 2 kindreds in 1944,1 several other studies of the familial occurrence of WPW syndrome have been reported.2–7 These studies suggested that WPW syndrome may be caused by genetic defects, based on the observations that WPW syndrome developed in infancy or childhood and in multiple generations in certain families.

In 1987, Vidaillet et al examined 2,343 first-degree relatives of 383 patients with APs proven by EPS and compared them with 427,639 subjects undergoing EPS in 14 medical centers in order to evaluate the genetic basis for the pathogenesis of APs.8 The results showed that the prevalence of manifest AP in first-degree relatives was significantly higher than that of the general population (0.55% vs 0.15%). Also, they examined the pedigrees of patients with
first-degree relatives with AP and proposed autosomal dominance as a mode of transmission of familial WPW syndrome. Compared with sporadic WPW syndrome, familial WPW syndrome is characterized by a high incidence of multiple pathways, lack of male dominance, high risk of sudden death, and low association of structural heart disease. Concealed APs that conduct impulse only in the retrograde direction can also be an anatomic substrate for AVRT. However, the existence of a concealed AP cannot be suspected until the EPS-confirmed PSVT develops. Therefore, the prevalence of concealed AP is subject to underestimation and the familial occurrence of concealed AP has rarely been reported.

In our study, AVRT developed in 2 siblings: an elder sister with a left lateral concealed AP and a younger brother with 2 APs, including a left posteroseptal manifest and a right anterior concealed AP. They were 2 of 4 children and different sexes, supporting autosomal dominant transmission. The remaining 2 children were said to have no symptoms suggestive of PSVT and had normal ECG. The mother confessed to having intermittent palpitation, but the symptom was not typical of PSVT and she had no ventricular preexcitation. Thus, she was not studied further.

RF-CA was safely and effectively used to cure PSVT in our 2 cases. Of note, in the case of the brother with 2 APs, a mistake was made in interpreting the tachycardia induced after ablating the left posteroseptal manifest AP without a meticulous mapping study. This necessitated a repeat RF-CA to eliminate the remaining AP. RF-CA was as effective as in sporadic cases of AVRT; however, it should be noted that multiple pathways are more prevalent in the familial type of AVRT than in the sporadic type.

We have presented 2 cases with familial occurrence of AVRT, which were cured using RF-CA, and which provide evidence for a potential genetic role in the failure to lose the atrioventricular connection during fetal development.

References

Fig 3. Catheter ablation of the left posteroseptal and right anterior accessory pathways (AP) in case 2. A left posteroseptal manifest AP was ablated first. Note the delta wave disappeared in 1.2 s after applying 30 W of RF energy (lower left panel). A right anterior concealed AP was successfully ablated at the atrial insertion site via the right femoral vein. After ablation of the APs, no retrograde ventriculoatrial conduction was demonstrated (lower right panel). ABL, ablation catheter; HRA, high right atrium; HIS, His bundle; RVA, right ventricular apex.