A Surgically Treated Case of Both Wolff-Parkinson-White Syndrome and Atrial Septal Defect Complicating Single Coronary Artery and Partial Pericardial Defect

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SUMMARY
A surgically treated case of both Wolff-Parkinson-White syndrome and atrial septal defect complicating a single coronary artery and partial pericardial defect is reported. These complications are very rare. In addition, the accessory pathway of this case had unique decremental conduction characteristics. (Jpn Heart J 36: 675–680, 1995)

Key words: Wolff-Parkinson-White Syndrome Atrial septal defect Single coronary artery Partial pericardial defect Decremental conduction

SINGLE coronary artery is a rare anomaly. It is found in 0.04 to 0.051) percent of patients undergoing coronary arteriography. Wolff-Parkinson-White (WPW) syndrome often complicates Ebstein’s anomaly, ventricular septal defect, atrial septal defect (ASD), patent ductus arteriosus and cardiomyopathy. However, WPW syndrome complicating a single coronary artery is extremely rare. In this paper we report a case of WPW syndrome, whose accessory pathway had unique retrograde decremental properties, complicating a single coronary artery, ASD and partial pericardial defect. The patient underwent successful surgical treatment of the WPW syndrome and ASD.

CASE REPORT
A 25 year-old female was admitted to our hospital for further evaluation of WPW syndrome and ASD. She was known to have had a heart murmur since 10 years of age. She had noted episodes of palpitations lasting for several minutes once or twice per year since she was 15 years old, but she had not received any medication. Recently the episodes had increased in duration and frequency.

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Therefore, she visited our hospital in May 1991.

On physical examination she was 150 centimeters in height and weighed 42 kilograms. A funnel chest was noted, and fixed splitting of the second heart sounds and a systolic ejection murmur were maximally audible at Erb’s area. There were no other abnormal findings.

Chest roentgenogram disclosed a left-sided heart, the funnel chest, dilatation of the hilar pulmonary artery and increased pulmonary vasculature. Electrocardiogram (ECG) (Figure 1-A) revealed a short PR interval (100 msec) and delta waves in leads I, II, aV_{R}, aV_{L}, V_1, V_2, V_3, V_4, V_5, and was classified as type II WPW according to Gallagher’s criteria. During a palpitation episode, paroxysmal supraventricular tachycardia with an RR interval of 370 msec was recorded (Figure 1-B). Echocardiogram revealed left-sided displacement of the heart and paradoxical movement of the interventricular septum, and a central type interatrial defect. The left ventricular apex was shifted downward and posteriorly, suggesting a partial pericardial defect. Doppler examination documented a shunt flow from the left to the right atrium through this defect.

Cardiac catheterization disclosed an oxygen saturation step-up at the mid-right atrium and normal pulmonary pressure. The left-to-right shunt ratio was
49%, pulmonary flow was 10.5 l/min. and systemic flow was 4.1 l/min. A single coronary artery was discovered on coronary arteriography (Figure 2). The coronary artery originated from the left coronary cusp. The left coronary artery ran its usual route; however, the right coronary artery originated from the proximal left anterior descending coronary artery and ran posteroinferiorly to the main pulmonary artery, and then came anteriorly. The single coronary artery in this case was classified as type IIa according to Smith’s modified criteria.2)

In the electrophysiologic study, clinical supraventricular tachycardia was induced. The tachycardia was an orthodromic atrioventricular reentrant tachycardia that conducted through the atrioventricular node in an antegrade fashion and through an accessory pathway retrogradely. At least two types of two different cycle lengths of atrioventricular reentrant tachycardia, of which the difference was caused by differing AH intervals, were recorded (Figure 3-B). This finding suggested the presence of a dual atrioventricular nodal pathway. The earliest atrial excitation site during atrioventricular reentrant tachycardia was located at the anterior site of the right atrium indicated in Figure 3-A. During rapid right ventricular pacing at rates of up to 260 bpm, retrograde Wenckebach conduction (decremental conduction) from the ventricle to the anterior right

Figure 2. Coronary arteriogram (CAG) A: left coronary arteriogram, left anterior oblique (LAO) 60° view, B: aortogram (AoG), LAO 60° view, C: AoG, right anterior oblique 30° view, D: Valsalva cusp shot CAG, LAO 60° view.
atrium occurred at 240 bpm (Figure 3-C). The ventriculoatrial conduction time via the accessory pathway increased gradually (decremental conduction) during premature right ventricular stimulation (Figure 3-D). The atroventricular reentrant tachycardia was initiated on a sudden jump-up of ventriculoatrial conduction time (Figure 3-E), suggesting the presence of another accessory pathway. However, the earliest atrial excitation site of this induced atroventricular reentrant tachycardia was not different from that of the other atroventricular reentrant tachycardia. The antegrade refractory period of the accessory pathway was 240 msec (basic cycle length 400 msec), and that of the retrograde pathway was
230 msec (basic cycle length 400 msec). Antegrade one to one conduction from the atrium to the ventricle was 220 bpm, while retrograde conduction was 230 bpm.

The partial pericardial defect was confirmed after median sternotomy. Right atrial mapping was performed using a 6 × 6 cm² mat electrode with an interelectrode interval of 0.8 cm during both supraventricular tachycardia and ventricular pacing. There was only one earliest excitation site, which was located at the anterior right atrium, as suggested by the preoperative electrophysiologic study. After the accessory pathway was incised from the epicardial site, the delta waves disappeared, and supraventricular tachycardia, including atrioventricular nodal reentrant tachycardia, was no longer inducible. However, there was a discontinuous atrioventricular conduction curve (Figure 4), which also proved the presence of the dual atrioventricular nodal pathway. A double accessory pathway was therefore not present. The ASD was then closed directly after starting extracorporeal circulation. The postoperative course was uneventful and supraventricular tachycardia, including atrioventricular nodal reentrant tachycardia, was not induced during the postoperative electrophysiologic study.

**DISCUSSION**

ASD is a surgically correctable disease, and WPW syndrome is also curable by surgery. The ASD of this case was an indication for surgery. Additional
surgical ablation of the accessory pathway at the ASD operation was thought to be ideal in this case. Only surgical ablation of the accessory pathway was performed and no surgical modification of the atrioventricular node was performed. As no atrioventricular nodal reentrant tachycardia was induced by either the preoperative or operative electrophysiologic study after ablation of the accessory pathway, the operative procedure was thought to be successful.

The accessory pathway of this case had unique characteristics. The ventriculoatrial interval during atrioventricular reentrant tachycardia was 70 msec, and therefore, the antegrade PR interval was longer than the retrograde RP' interval. However, decremental retrograde conduction, such as the increment of the ventriculoatrial interval induced by premature ventricular stimulation and Wenckebach conduction by rapid ventricular stimulation, was present. In addition, the sudden jump-up of the ventriculoatrial interval was accompanied by atrioventricular reentrant tachycardia. These decremental characteristics are uncommon in cases of WPW syndrome. Their incidence was reported to be 7.6% in 653 patients with WPW syndrome.4) Furthermore, they are often found in cases with a long retrograde RP' tachycardia.4) The reason why this case did not exhibit a long retrograde RP' sequence could not be clarified. There are two possible reasons for the sudden jump-up of the ventriculoatrial interval. One is that two accessory pathways existed near each other and the second is explained by the character of the accessory pathway showing longitudinal dissociation of the retrograde accessory pathway.4) Although surgical documentation of closely existing accessory pathways may be limited, the surgeon could not find a double accessory pathway. Therefore, the latter reason might explain the sudden jump-up of the ventriculoatrial interval.

In some cases of a single coronary artery, compression of the right coronary artery between the pulmonary artery and the aorta during exercise is reported to occur, leading possibly to sudden death. In this case, however such an episode did not occur, probably due to the route of the coronary artery. Thus, surgical coronary artery intervention was not performed.

REFERENCES