Mixed Type Total Anomalous Pulmonary Venous Connection in a Newborn

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Abstract

Total anomalous pulmonary venous connection (TAPVC) is referred to a specific condition in which there is no direct connection between any pulmonary veins and the left atrium. In this report, we describe a case of mixed type TAPVC in a newborn. The surgical correction of anomaly was successful. The child had an uneventful post-op recovery and was discharged.

Keywords: Pediatric Cardiology, TAPVC, Pulmonary Vein Anomalous, Cardiac Surgery

1. Introduction

Total anomalous pulmonary venous connection (TAPVC) is referred to a specific condition in which there is no direct connection between any pulmonary veins and the left atrium. All pulmonary veins connect to the right atrium or one of its tributaries. In mixed type TAPVC, there is no common pulmonary venous sinus, and pulmonary veins connect randomly to the heart. TAPVC was first described by James Wilson in 1798 in the autopsy of an infant with ectopia cordis and other congenital malformation (1, 2). Mixed variety of TAPVC is a rare cardiac malformation and includes 5% - 10% of these cases, but wide variations of pulmonary venous anatomy are reported that can make surgery more challenging (1, 3). In this report, we describe an infant who was diagnosed with a mixed type of TAPVC.

2. Case Report

A neonate was referred to pediatric center who was diagnosed with total anomalous pulmonary venous connection (TAPVC) on the second day of life. The baby boy weighing 3.2 kg with tachypnea, central cyanosis, and respiratory distress was admitted for further examinations. A loud P2 murmur on upper left sternal border was heard. Electrocardiogram showed normal sinus rhythm with right axis deviation. Echocardiography revealed the enlargement of both right atrium and right ventricle (Figure 1). Relative shrinkage of left atrium and ventricle was also evident. Base on Doppler echocardiography, the right-sided pulmonary veins with left inferior pulmonary vein formed a confluence and drained via the common chamber to coronary sinus (Figure 2). The left upper pulmonary vein drained via a vertical vein to left innominate vein (Figure 3). There was also an atrial septal defect (ASD) with the diameter of 5 mm and right to left shunt was evident with severe pulmonary artery hypertension. But there was no patent ductus arteriosus (PDA). Computed tomography angiography (CTA) also supported these findings and determined the diagnosis of TAPVC (Figure 4 - 9). So the child was a candidate for surgery and it was performed on the 12th day of birth. Under general anesthesia and sterile conditions, sternotomy was performed. Abnormal pulmonary veins were appeared and separated by vessel tapes. Cardiopulmonary bypass (CPB) was done with aortic and bicaval venous cannulation. Cardiac arrest was induced by antegrade cardioplegia and then mild hypothermia. The roof of left coronary sinus adjacent to left atrium (LA) was excised, then connection between the confluent of pulmonary veins (CPV) and LA was established. The left upper pulmonary vein remained intact. Due to the avoidance of right heart failure, surgeon partially closed the ASD and held open 2 millimeters of the hole. The child was weaned...
off CPB with the support of inotropes. He was then transferred to pediatric intensive care unit with stable hemodynamics. This course was uneventful and post-operative echocardiography was normal. Due to follow-up echocardiography, there was no sign of stenosis in coronary sinus to LA anastomosis (unroofing area). The child was in a good state of health and weighing was acceptable.

3. Discussion

The correct diagnosis and accurate anatomical description of TAPVC is necessary for planning the surgery. Detection is difficult to deal with in the mixed type of anomalous. Echocardiography is sufficient for diagnosis in most cases, but the definitive diagnosis and determination of the anatomical location of the vessels is based on the results of the computed tomography. In TAPVC, the pulmonary venous blood returns to right side of the heart directly or by combination with coronary sinus or systemic veins. It may lead to pulmonary hypertension; so
Figure 5. Confluence of Pulmonary Veins (CPV) or Common Pulmonary Chamber (CPC) to Coronary Sinus

Axial view in CT angiography.

Figure 6. Left upper pulmonary vein connection (LUPV) to left innominate vein (LIV)

Axial view in CT angiography.

Figure 7. Confluence of Pulmonary Veins (CPV) or Common Pulmonary Chamber (CPC) to Coronary Sinus

Frontal view in CT angiography.

Figure 8. Left Upper Pulmonary Vein Connection (LUPV) to Left Innominate Vein (LIV)

Frontal view in CT angiography.

Symptoms of pulmonary congestion and right heart failure are expected (4, 5). Total anomalous pulmonary vein connection is the disease in which structural changes of pulmonary circulation including arterial wall thickness is present in all types. With the development of pulmonary hypertension, septum grows with the right ventricle and their growth together overtakes that of the left (6).

Surgical correction for TAPVC can be beneficial, but complications including severe preoperative cardiopulmonary instability in infants with obstruction, postoperative paroxysmal pulmonary hypertension, and delayed development and progression of pulmonary vein stenosis are still expected (1). Despite the poor condition and severe pulmonary hypertension before the surgery, our patient was discharged in good conditions. Immediate correction of anomaly and hemodynamic status should be considered. The more the surgery is delayed, the child’s condition worsens. By the progression of pulmonary hypertensive crisis, higher mortality rates are expected. In this case,
the surgeon did not completely close the ASD, and let the right ventricular pressure to be adjusted for prevention of right heart failure.

**Footnotes**

**Conflicts of Interest:** The authors declare that they have no conflicts of interests.

**Consent:** Written informed consent was not available because of change of residence but oral agreement from the patient was obtained.

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