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Asymptomatic Total Anomalous Pulmonary Venous Connection with Double Drainage in a Young Adult: A Case Report

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ABSTRACT

Total anomalous pulmonary venous connection is an uncommon congenital anomaly in which all pulmonary venous return drains to the right atrium or one of its tributaries. Survival beyond infancy without surgical palliation is unlikely, so this anomaly is not encountered in the adult population with congenital heart disease. The patient presented here was 22 years old on admission and had no total anomalous pulmonary venous connection–associated symptoms. He underwent transthoracic echocardiographic examination for atypical chest pain. Transthoracic echocardiography along with cardiac catheterization favored the presence of a total anomalous pulmonary venous connection. Surgical correction of pulmonary venous confluence (draining to both the coronary sinus and right atrium) was performed successfully. This is a rare case of total anomalous pulmonary venous connection with no reported symptoms in contrast to the majority of patients who are symptomatic during the first year of life.

CASE REPORT

A 22-year-old male patient was admitted to our center with atypical chest pain. On auscultation, there was a 2/6 systolic murmur on the left upper parasternal area and a fixed, widely split second heart sound. On inspection, there was no abnormality except for mild clubbing. Transthoracic echocardiography (Figure 1) demonstrated the dilated coronary sinus and an echo-free structure above the left atrium (venous confluence). With slight movement of the probe, a large ostium secundum atrial septal defect (Figure 2) was visualized. There was also mild mitral regurgitation due to the prolapsus of the anterior mitral leaflet and moderate tricuspid regurgitation (systolic right ventricular pressure, 45 mmHg). On contrast echocardiography, there was contrast filling of the coronary sinus in the late phase. On cardiac catheterization, pulmonary angiography revealed the opacification of the pulmonary venous confluence (Figure 3) and indistinct opacification of the neighboring right atrium in the late phase of the angiography. Left atrial angiography (Figure 4) through the large atrial septal defect did not demonstrate any opacified pulmonary venous structure around the left atrium. The peak pulmonary systolic pressure was around 50 mmHg. The pulmonary-to-systemic flow ratio (Qp/Qs) was found to be 3.3. The patient underwent surgical correction of the total anomalous pulmonary venous connection (TAPVC) draining to both the coronary sinus and right atrium and recovered uneventfully.

DISCUSSION

TAPVC describes a situation in which all pulmonary veins fail to drain directly to the left atrium, thereby directing all systemic and pulmonary venous return to the right atrium via various routes. Anomalous pulmonary veins generally form a venous confluence around the left atrium and then drain to systemic venous circulation. Survival beyond infancy without

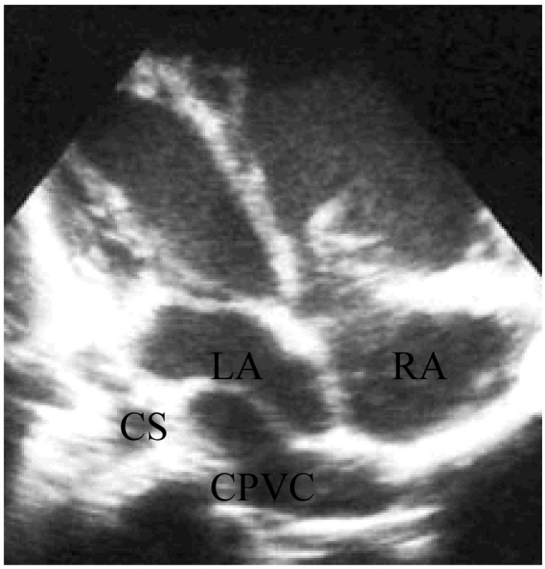

Figure 1. The dilated coronary sinus (CS) and common pulmonary venous confluence (CPVC) above the left atrium (LA) on transthoracic echocardiography (apical 4-chamber view). RA indicates right atrium.

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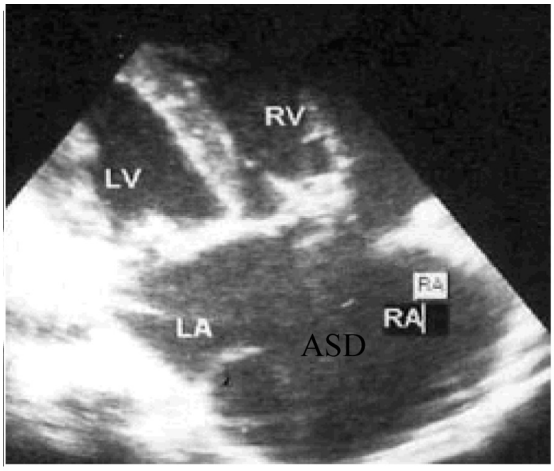

Figure 2. The large atrial septal defect (ASD) on transthoracic echocardiography (apical 4-chamber view). RV indicates right ventricle; LV, left ventricle; LA, left atrium; RA, right atrium.

surgical palliation is unlikely, so this anomaly is not encountered in the adult population with congenital heart disease. The clinical manifestation is closely associated with the size of interatrial communication and the magnitude of pulmonary vascular resistance. In cases of small interatrial communication, there is usually right-sided heart failure with pulmonary hypertension and limited systemic blood flow with mild cyanosis. The anomalous connection may be conventionally classified according to the site of the drainage: supracardiac (innominate vein, superior vena cava, etc), cardiac (right atrium, coronary sinus), infracardiac (portal vein, etc) or mixed type [Darling 1957]. The infracardiac TAPVCs usually have an obstructive component in the venous confluence or in the vessel connecting the venous confluence to the systemic venous circulation. Rarely there may be multiple drainage sites at the same or different levels, particularly the double drainage in which the venous confluence usually drains to the coronary sinus and the

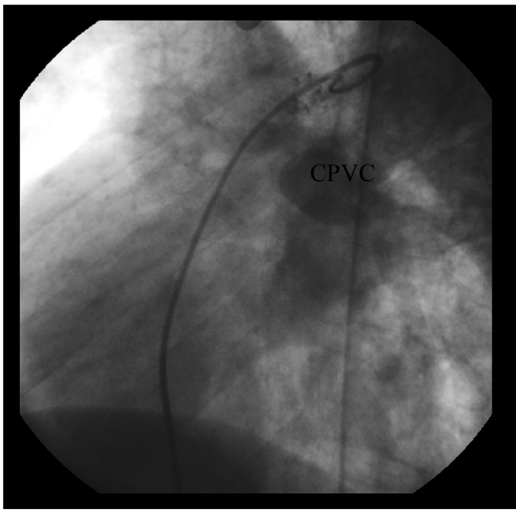

Figure 3. Opacification of common pulmonary venous confluence (CPVC) in the late phase of pulmonary angiography.

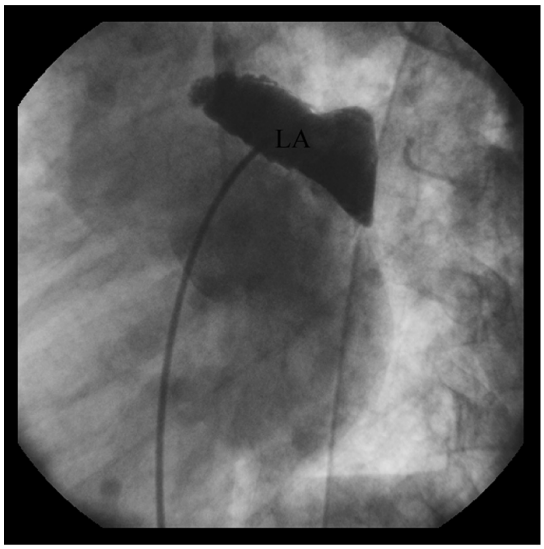

Figure 4. Left atrial angiography demonstrating the absence of any pulmonary vascular structure around the left atrium.

left innominate vein (also the mixed type) [Arciprete 1986; Van de Wal 1993; Lee 1995]. In our patient, the common venous confluence of all pulmonary veins was found to drain to the coronary sinus and the right atrium via a short connecting vessel. Tanabe et al [2000] reported a severely symptomatic newborn with TAPVC draining to systemic venous circulation at both supra and infradiaphragmatic levels, and propounded the persistent connection between the primitive forms of the pulmonary venous plexus, systemic venous channels, and the umbilical line system as the possible underlying factor.

The interesting point in our case other than the drainage pattern was the absence of TAPVC-associated symptoms (cyanosis, right heart failure, etc) until the age of 22. This might be due to the well-balanced amount of right-to-left shunt at the atrial level preventing both severe pulmonary hypertension and severe cyanosis. In conclusion, this is an unusual case of coincidentally diagnosed asymptomatic TAPVC with double drainage.

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