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Isolated Partial Anomalous Pulmonary Venous Connection

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BACKGROUND Isolated partial anomalous pulmonary venous connection (PAPVC) is a very rare variant of PAPVC, and involves the anomalous drainage of the right upper pulmonary vein into the superior vena cava (SVC) with no associated inter-atrial septal defect.

CASE We present a case of an isolated PAPVC occurring in 26-year old female patient who presented with palpitations, exertional dyspnea and easy fatigability. Physical examination revealed a dynamic precordium, a right ventricular heave, and a tricuspid regurgitation murmur. Transthoracic echocardiogram showed right atrial, ventricular, and main pulmonary artery dilatation with moderate pulmonary hypertension. Hemodynamic studies revealed an isolated PAPVC with severe pulmonary hypertension. She underwent Warden's procedure (superior vena cava translocation with atriocaval anastomosis), which remarkably improved her symptoms.

CONCLUSION This case report shows a very rare case of isolated PAPVC presenting with heart failure symptoms and right-sided chamber enlargement. The clinical presentation is not specific and may mimic more common diseases, such as acyanotic atrial septal defect and primary pulmonary hypertension. A high index of suspicion is important in diagnosing the disease in patients with right-sided cardiac chamber dilatation.

Isolated partial anomalous pulmonary venous connection (PAPVC) is a very rare variant of PAPVC, and involves the anomalous drainage of the right upper pulmonary vein into the superior vena cava (SVC) with no associated inter-atrial septal defect. It presents with right- sided heart failure, arrhythmias, and pulmonary vascular disease; and may mimic more common congenital anomalies, such as atrial septal defect. This report presents a case of isolated PAPVC in an adult patient who was initially diagnosed to have primary pulmonary hypertension.

CASE REPORT

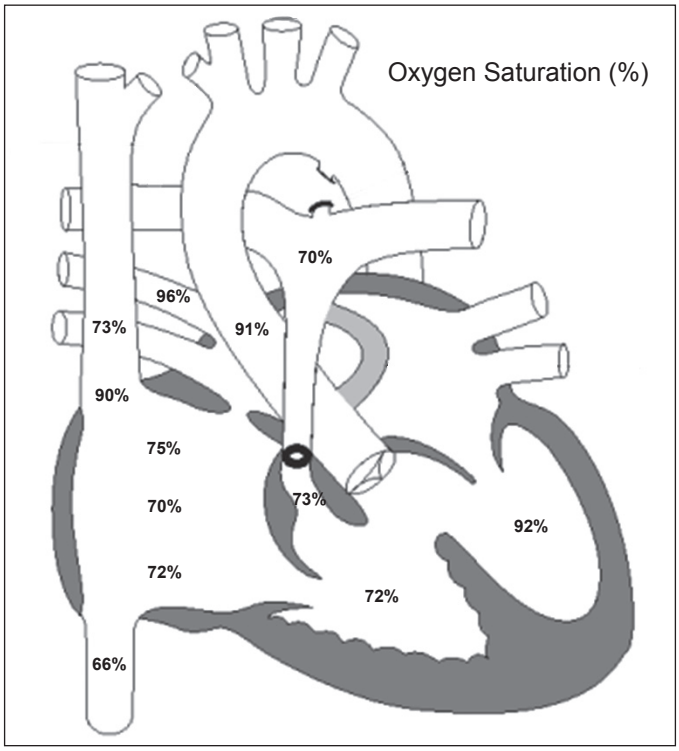
This is the case of GM, a 26-year old female, who was admitted to the Philippine Heart Center for hemodynamic studies. Four months prior to admission, she consulted a cardiologist for palpitations, exertional dyspnea, and easy fatigability. A transthoracic echocardiogram was performed, revealing a dilated right ventricle with good systolic function but with signs of volume and pressure overload. The right atrium and main pulmonary artery (MPA) were also dilated. Moderate tricuspid regurgitation and moderate pulmonary hypertension was also appreciated. The left ventricle had normal dimensions with good contractility and systolic function. No shunt was found. She was diagnosed to have primary pulmonary hypertension and was given appropriate medications. She was then advised for hemodynamic studies, hence the current admission.

On examination, she seemed well. Her blood pressure was both 90/60 mmHg taken at both the upper and lower extremities. Her heart rate was at 68 beats per minute. Her respiratory rate was 18 cycles/minute. Her body temperature was 36.8 ^{o}C. Her body mass index was 20.8 kg/m^2 (weight=52 kg; height=158 cm).

Cardiovascular findings included a dynamic precordium with point of maximal impulse at the 5th mid-clavicular line; right ventricular heave; grade 3/6 systolic murmur on the 4th intercostal space, left parasternal border which accentuates on inspiration (positive Carvallo sign); no thrills; normal rate with regular rhythm; S1 prominent at the apex; and S2 prominent at base (loud P2 component). The rest of the physical examination findings were unremarkable.

Hemodynamic studies revealed a significant step-up of oxygen saturation from the SVC high to SVC low (Figure 1). Levo phase pulmonary angiogram showed drainage of the right superior pulmonary vein into the SVC (Figure 2). No inter-atrial septal defect was noted. Pressure recordings showed an elevated MPA pressure of 100/46 mmHg. These findings were consistent with an isolated PAPVC with severe

Figure 1. Oximetry run during hemodynamic study showing significant step of the O_2 saturation at the level of the superior vena cava (SVC) suggesting an abnormal pulmonary venous drainage to the SVC.



pulmonary hypertension.

The patient underwent Warden's procedure (SVC translocation with atriocaval anastomosis). The post-operative course was unremarkable. She was discharged with markedly improved symptoms.

DISCUSSION

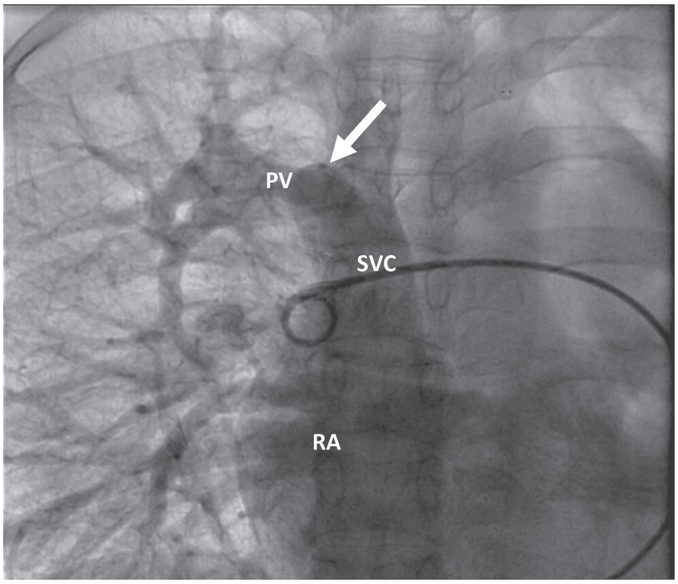
PAPVC is a rare congenital heart disease present in only 0.7% of the general population. It is usually characterized by terminal insertion of a pulmonary vein into the right atrium or superior vena cava in conjunction with a sinus venosus atrial septal defect (ASD).[1] PAPVC with ASD (usually sinus venosus type) is the most common type, which occurs in 80% to 90% of PAPVC cases. PAPVC with intact atrial septum (isolated PAPVC) is a very rare finding, and mostly involves the anomalous drainage of the right upper pulmonary vein into the SVC.[2] This condition results from the failure of the complete separation of the splanchnic plexus-derived pulmonary vasculature from the right common cardinal vein, the embryologic precursor of the SVC, during fetal development.[3] Patients with isolated PAPVC rarely present with symptoms until the third decade of life. The incidence among females is higher than in males.[1]

Our patient was a 26-year old female who presented with palpitations, exertional dyspnea and easy fatigability. Her symptoms can be attributed to a right-to-left shunt resulting from severe pulmonary arterial hypertension from PAPVC-dependent increased pulmonary blood flow.[4] Palpitations may reflect cardiac arrhythmias, which are almost always supraventricular in origin, and may be due to right atrial dilatation. Exertional dyspnea and easy fatigability may occur secondary to right-sided volume overload or pulmonary vascular obstructive disease.[1] These symptoms usually occur at an older age because they are directly related to progressive right chamber enlargement resulting from chronic shunting of blood from the pulmonary vein.

Transthoracic echocardiography can be used to diagnose PAPVC and obviate the need for an invasive cardiac catheterization. However, the lesion can be easily missed if routine echocardiography does not incorporate definition of the entire pulmonary venous return. The sonographer must identify all four pulmonary veins and visualize their connections to the heart. This is complicated by the fact that not all the pulmonary veins may be identified, especially in adults.

The atrial septum also needs to be evaluated for defects.[1] In our patient, the diagnosis of PAPVC was not demonstrated by the initial echocardiogram, but a right-sided chamber

Figure 2. Frontal view (RAO 0° Caudal 0°) showing the right superior pulmonary vein (PV) draining into the superior vena cava (SVC) (white arrow) during levo phase pulmonary angiogram.



RAO=right anterior oblique; PV=pulmonary vein; SVC – superior vena cava; RA=right atrium.

dilation was noted. This may be the first observation that indicates the presence of abnormal venous drainage. Hence, a high index of suspicion for the presence of the lesion is helpful to properly diagnose the condition.

Magnetic resonance imaging (MRI) is rapidly becoming the procedure of choice for further investigation of PAPVC.[5] PAPVC may present a characteristic sign termed the "broken ring sign on MRI.[6]

Cardiac catheterization may be a more preferable diagnostic tool in infants with complex congenital heart conditions in whom PAPVC is one component.[1] In our patient, cardiac catheterization was performed for precise anatomic diagnosis and hemodynamic evaluation. Oxygen saturation in the SVC that is higher than that found in the right atrium strongly indicates PAPVC to the SVC. The abnormal pulmonary venous drainage from the right lung was confirmed on the levo phase of the right pulmonary artery angiogram (Figure 2), which showed that the venous return from the right lung was partly directed via one anomalous vein to the SVC. The left pulmonary veins connected and drained normally.

CONCLUSION

This case report shows a very rare case of isolated partial anomalous venous connection. The clinical presentation may not be specific for particular cardiac disease entity and may simulate more common diseases like acyanotic ASD and primary pulmonary hypertension. It is important to report such a case, because this can highlight the importance of high index of suspicion in considering isolated PAPVC in patients with right sided cardiac chamber dilatation.

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