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A Case Report of Total Anomalous Pulmonary Vein Connection in an Adult through Application of Multi-Modality Imaging

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ABSTRACT

Total Anomalous Pulmonary Vein Connection (TAPVC) is often reported in neonates and children. This report demonstrates an unusual adult TAPVC that the patient is survived in the fourth decade of life. It was diagnosed via application of echocardiography paired with multi detector-row computed tomography angiography. A 39-year-old man was referred for evaluation of dyspnea and cyanosis at rest worsening with exertion. On physical examination, decreased oxygen saturation, clubbing, and central and peripheral cyanosis were detected. In primary evaluation, large Atrial Septal Defect (ASD) was discovered. Cardiac catheterization showed a large ASD and benign variant of Persistent Left Superior Vena Cava (PLSVC). On Trans-Thoracic Echocardiography (TTE), no pulmonary hypertension was defined in direct contradiction of Eisenmenger syndrome. Due to suspicion of a complex shunt, he underwent Trans-Esophageal Echocardiography (TEE) followed by CT-angiography. Both of these image modalities clearly demonstrated the presence of TAPVC in addition to ostium secundum ASD. Due to the change in the main diagnosis, he underwent surgical correction with immediate and complete relief of both cyanosis and dyspnea after operation. Although some complex congenital heart diseases are usually seen in newborns and infants, their presence in adulthood is not impossible. Thus, some unusual findings similar to presence of cyanosis in the absence of pulmonary hypertension should be questioning. In such cases, alternative imaging studies can provide additional guidance in diagnosis. It is not an infrequent occurrence, as this case underscores, where more complete imaging overturns an initial diagnosis leading to completely different management.

1. Introduction

A part of complete anomalous drainage of pulmonary veins into systemic venous circulation is due to abnormal development of pulmonary veins. Partial anomalous pulmonary vein connection is usually a non-cyanotic disease. Total Anomalous Pulmonary Vein Connection (TAPVC) is associated with cyanosis due to mixing of pulmonary and systemic blood flow through Patent Foramen Ovale (PFO) or inter Atrial Septal Defect (ASD). Anomalous return could be cardiac, supra cardiac, infra cardiac, or a combination of them. This anomaly has been reported in 6.8 per 100,000 live births and 1.5% of all cardiovascular malformations (1). In the previous literature, most cases of TAPVC were reported in neonates and children and survival until adulthood has been rarely reported (2).

Knowing the characteristics of the disease is essential for therapeutic strategies. Presence of abnormal pulmonary vein communication may be discovered by echocardiography through showing small left atrium, enlarged right ventricle, or abnormal pulmonary venous connection. However, definitive depiction of pulmonary veins problems is one of the limitations of echocardiography due to lack of acoustic window.

For a long time, catheterization was known as a diagnostic method for TAPVC cases. Recently, however, Multi Detector-row Computed Tomography (MDCT) angiography and Magnetic Resonance Imaging (MRI) have been introduced as powerful non-invasive diagnostic tools

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for evaluation of TAPVC (3).

Hereby, we report an adult patient who was nominated for surgical repair of ASD based on heart catheterization. However, thanks to Trans-Esophageal Echocardiography (TEE) in addition to MDCT angiography, final diagnosis and management completely changed to TAPVC and ASD.

2. Case Presentation

A 39-year-old man was referred for precise cardiac evaluating by his cardio surgeon. He was suffering from dyspnea and cyanosis at rest, worsening with exertion. On physical examination, decreased oxygen saturation, clubbing, and central and peripheral cyanosis were detected. In cardiac auscultation, a brief diamond shaped early systolic murmur followed by another brief diamond shaped early systolic murmur was audible. O2 saturation at rest was 79%, which reached 90% by oxygen treatment. The 6-minute walk test was made by the patient to about 330 minutes. First cardiac evaluation with complementary cardiac catheterization indicated an isolated large ostium secundum ASD with Persistent Left Superior Vena Cava (PLSVC). Therefore, he was nominated for surgical repair. However, because of the obvious cyanosis, his surgeon came to the conclusion that he needed more evaluation in order to rule out Eisenmenger syndrome. Thus, the patient was referred to our institution for further evaluation. Electrocardiography showed fibro-flutter rhythm with tall R wave in V1 lead. Indeed, CXR showed typical snowman appearance due to special configuration of heart and superior mediastinal borders seen in TAPVC. On routine Trans-Thoracic Echocardiography (TTE), no pulmonary hypertension was detected and in the first step, Eisenmenger were ruled out. Large ostium secundum ASD was present. Nonetheless, Color Doppler study unexpectedly depicted that cardiac shunt was completely right to left. Contrast study also showed a similar shunt. This finding forced us to further evaluate systemic and pulmonary veins. TTE showed normal left ventricular size and systolic function, normal diastolic function, severe right ventricular enlargement with mild dysfunction, moderate pulmonary artery hypertension (systolic pulmonary artery pressure = 55 mmHg), and large ostium secundum ASD (4.4×3.8cm). TEE confirmed large ASD with fully right to left shunt, and the main input of the Left Atrium (LA) was through ASD. Pulmonary vein return to LA was totally anomalous with a consequent formed vertical vein by Left Pulmonary Veins (LPVs), which passed under the left pulmonary artery and then connected to right pulmonary veins. The vertical vein finally drained to the innominate vein towards SVC (Figure 1). After all, MDCT angiography confirmed not only the large ASD but also the absence of normal connection of pulmonary veins to the left atrium. This modality demonstrated pulmonary veins that terminated at the supra diaphragmatic level. These veins converged to form a large confluent vein; i.e., the vertical vein. MDCT angiography clearly showed this large vertical vein, which redirected pulmonary veins blood flow to SVC (Figure 2). To evaluate pulmonary vascular resistance, injection of the right ventricle and pulmonary artery was done, which showed aneurismal dilation of the pulmonary artery with anomalous venous return to the right atrium. Pulmonary artery pressure was 60/28 mmHg (mean = 40 mmHg) before oxygenation and 47/17 mmHg (mean = 30mmHg) after that, with acceptable response of pulmonary artery pressure to oxygen treatment.

TAPVC is usually recognized as a cyanotic congenital defect in newborns in which all pulmonary and systemic venous blood enters the right atrium and nothing drains into the left atrium. A right to left shunt like PFO or less commonly ASD is necessary for survival. This anomaly results in embryological failure of pulmonary venous development. In this disease, there is persistent patency

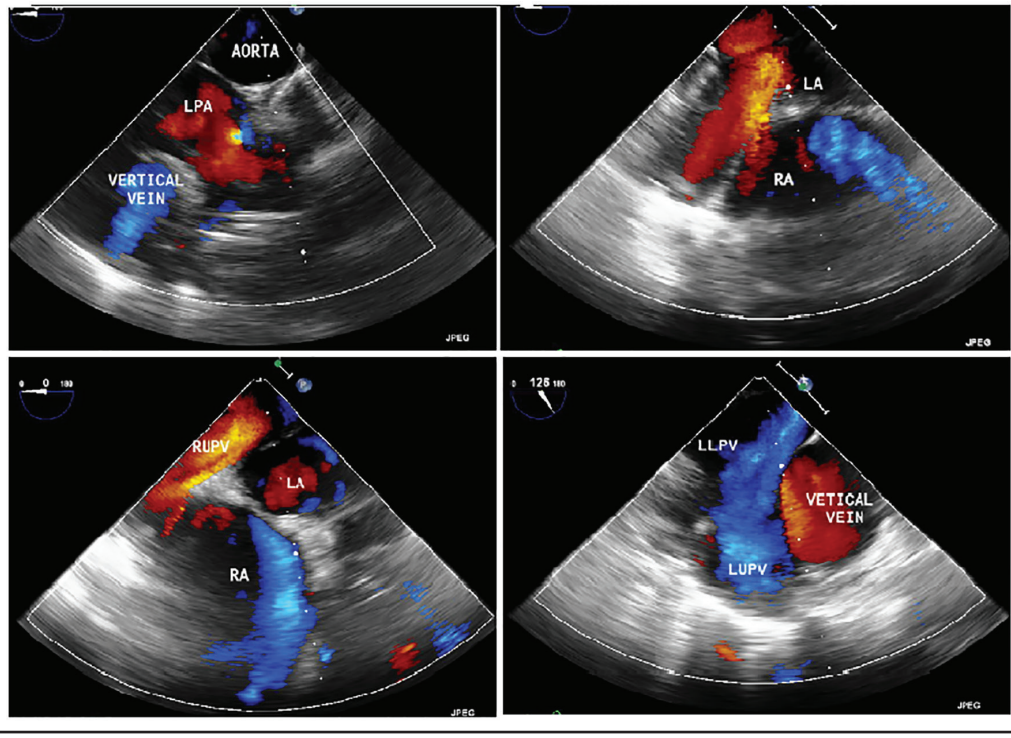
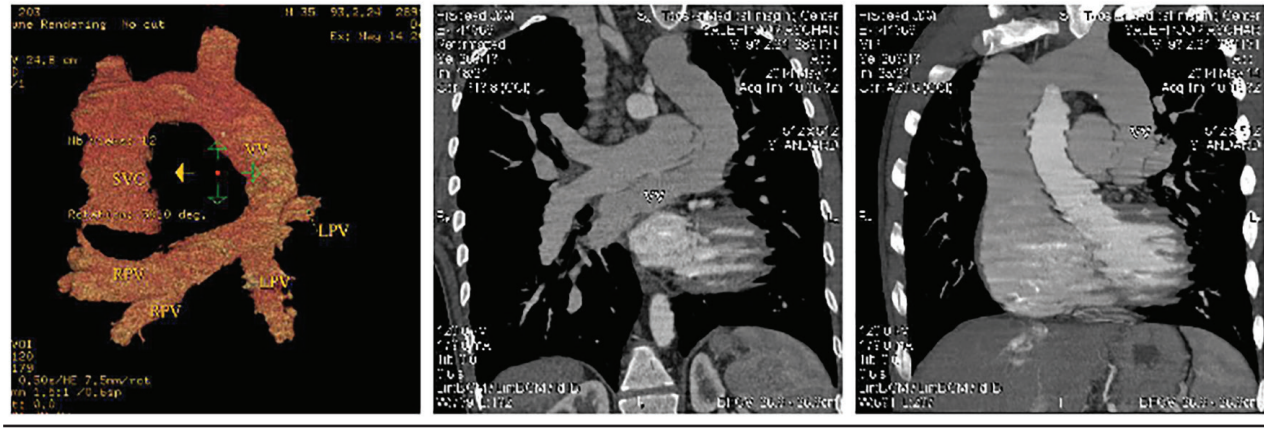


Figure 1. Three-Dimensional Trans-Esophageal Echocardiography Showing Large Ostium Secundum Atrial Septal Defect

Figure 2. Multi Detector-Row Computed Tomography Angiography Showing All Pulmonary Veins Draining to the Vertical Vein



Abbreviations: RPV, right pulmonary vein; LPV, left pulmonary vein; VV, vertical vein; SVC, superior vena cava

of primitive veins. Our patient underwent total surgical correction with immediate and complete relief of both cyanosis and dyspnea after operation.

3. Discussion

Diagnosis of TAPVC is a real challenge for cardiologists. Indeed, adult TAPVC cases are rarely reported with few surviving until adulthood. In a long-term follow-up, satisfactory results and excellent long-term survival of adult TAPVC cases within the age range of 18-38 years have been reported (4). Without a high degree of suspicion, this kind of abnormality can be quite difficult to diagnose via TTE and/or on non-ECG-gated CT angiography. It should be noted that anatomically, the vertical vein is similar to PLSVC (5). In this regard, MDCT angiography has been reported to be superior to echocardiography whether it would be TTE or TEE for pre- and post-operative evaluation of TAPVC (6). Novel expansions of CT technology provide a fast, accurate, and non-invasive diagnostic modality with high spatial and temporal resolution. Besides, detection rate of anomalous pulmonary veins by MDCT angiography is close to 100%. Additionally, this technique needs lower radiation exposure than cardiac catheterization, which makes MDCT angiography a superior technique (7). MRI and gadolinium-enhanced MR Angiography (MRA) have high spatial resolution with wide field of view and lack of ionizing radiation. The advantage of CT over MRA in diagnosis of TAPVC is its faster image acquisition and compatibility with metallic implants (8). Anyway, assuming a correct diagnosis, the surgical treatment of TAPVC is accompanied with low operative risk and quite satisfactory immediate and intermediate results (9). On the other hand, 'correction' of an ASD without further imaging in this case would have been catastrophic. Indeed, closure of ASD which was the only receiving orifice of blood flow to the left atrium would have ended up the patient in death.

3.1. Conclusion

The main message of this case report is highlighting the importance of performance of complementary imaging modalities rather than sole reliance on catheterization findings to dictate congenital heart disease diagnosis and the subsequent therapeutics. The utilization of alternative imaging with inherent imaging attributes can often provide additional guidance in patient care. It is not an infrequent occurrence, as this case, where more complete imaging and more precise evaluation changes an initial management to correct therapeutic implications.

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Authors' Contribution

Mirdamadi developed the original idea and the protocol, abstracted and analyzed data; Nejati contributed in development of the protocol, analysis of data and drafting of the manuscript; Behjati developed the protocol and revised the manuscript and supervised study.

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