Case Report

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Incidental Diagnosis of Partial Anomalous Pulmonary Venous Return during the Atrial Septal Defect Closure in an Adult Patient: A Case Report

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Partial anomalous pulmonary venous return (PAPVR) is a congenital anomaly that directly connects the pulmonary veins (PV) to a systemic vein or right atrium. Previous studies have reported the association between right-sided PAPVR and sinus venosus-type atrial septal defect (ASD); however, the coexistence of PAPVR and secundum-type ASD is extremely rare. Some patients present with pulmonary hypertension (PHT) and are diagnosed as adults. the clinical manifestation of this typical scenario is due to hemodynamic abnormalities, which develop secondary to the left to right shunt (LTRS) and the possible coexistence of other cardiac abnormalities. Herein, we present a case of PAPVR incidentally diagnosed while closing a secundum-type ASD. Further evaluation using 640 sliced multidetector computed tomography (MDCT) confirmed the diagnosis.

Key words: partial anomalous pulmonary venous return, atrial septal defect, multidetector computed tomography

Introduction

Partial anomalous pulmonary venous return (PAPVR) is a congenital cardiovascular anomaly manifesting as abnormal connection of one or few, but not all, pulmonary vein to systemic veins. The total anomalous venous return (TAVR) is known to be a congenital heart condition which is commonly diagnosed in newborns, whereas, PAPVR is usually asymptomatic and often goes undiagnosed in the majority of the patients. PAPER is a rare condition, occurring in only 0.4% – 0.7% of the population.[1] The adult type is characterized by left superior pulmonary veins (PV) getting connected and draining to the anonymous or right superior PV that is draining directly in the superior vena cava (SVC). PAPVR of the right upper PV can coexist with a sinus venosus atrial septal defect (ASD).[2]

Some patients can be asymptomatic until adulthood. The entity of the left to right shunt (LTRS) determines the clinical manifestation as it is caused by an abnormal pulmonary flow return that presents along with cardiac malfor-

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mations. The sinus venosus ASD is the most common cardiac problem, which may worsen the left-to-right shunt and become symptomatic. In this case report, the patient was incidentally diagnosed with PAPVR diagnosed during ASD closure.

Case Report

A 46-year-old woman presented to our cardiovascular outpatient department with progressive exertional dyspnea. She reported no systematic disease or specific history. She had been completely asymptomatic before this episode. Her electrocardiogram showed sinus tachycardia and delayed borderline atrioventricular conduction. The secundum-type ASD was diagnosed by echocardiography with the left-to-right shunt as shown in Figure 1. Mild right ventricular chamber dilation was also found. The NT-proB-type natriuretic peptide was 20 pg/mL (within the normal limit). Then, further management with ASD closure was arranged as it was symptomatic.

During cardiac catheterization for the closure of secundum-type ASD, pulmonary artery (PA) angiography (4 chambers + lateral view) showed a branch of the right PV connecting to the SVC (Fig. 2). Other pulmonary venous branches showed a normal connection to the left atrium. The calculated Qp/Qs (pulmonary to systemic output) ratio was 3.0, clinically demonstrating a significant left-to-right shunt. The average main PA pressure was 20 mmHg. The bicaval view revealed a 14-mm ASD with the left-to-right shunt on pre-procedure transesophageal echocardiography (TEE). The placement of Amplatzer ASD occluder (9-ASD-015) was conducted smoothly as scheduled under intraoperative TEE assistance. After the occlusion, only the trivial residual shunt was observed on TEE. Instead, of performing further advanced intervention or surgical repair, follow-up cardiac and shunt conditions were closely decided.

Additional evaluation with multidetector computed tomography (MDCT) was arranged to confirm the diagnosis. Computed tomography (CT) angiography performed using 640 sliced MDCT shows right superior pulmonary vein drainage into the SVC (Fig. 3). Other branches of the pulmonary vein drain into the left atrium (LA). Dilated right heart chambers are observed, suggestive of a right heart strain.

A smooth postoperative course was noted in the intensive care unit and general ward.

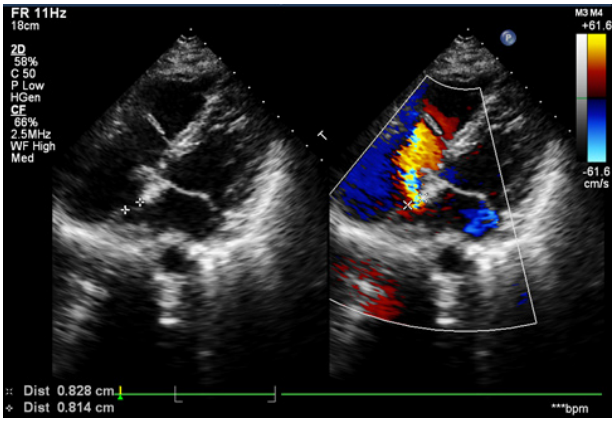

Fig. 1 Transthoracic echocardiography shows a 0.8-cm defect between the right atrium (RA) and left atrium (LA), representing ASD. Color duplex ultrasound shows a left-to-right shunt.

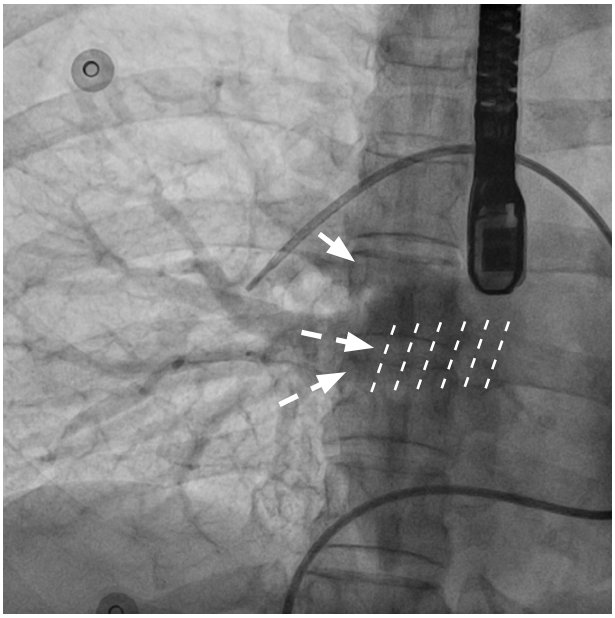

Fig. 2 Pulmonary artery angiography shows a branch of the right pulmonary vein drains into the SVC (arrow) and other branches drain into the LA (dotted arrows).

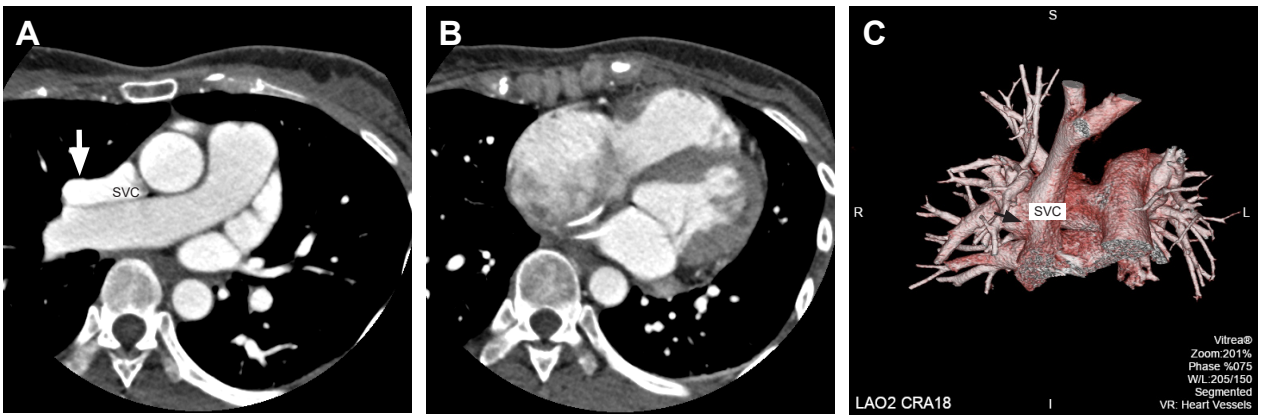

Fig. 3 MDCT angiography reveals (A) a single right superior pulmonary vein (white arrow) connected with SVC and (B) dilated right heart chambers. An ASD occlusion is placed at the proper position. (C) 3D reformatted images show PAPVR from the right upper lobe to the SVC.

Postoperative electrocardiogram, echocardiogram, and chest X-ray are unremarkable. On follow-up after a few weeks, the patient's symptoms improved. The estimated Qp/Qs ratio decreased to < 1.5 on the follow-up cardiac color duplex.

Discussion

PAPVR is often clinically silent. Autopsy results clearly shows that at least > 1% of the human suffer with PAPVR.[1] It rarely manifest any symptoms even when > 50% of the pulmonary blood flows directly to the right heart. Based on the extent of the LTRS, the clinical manifestation widely varies. There are asymptomatic patients as well as history of severe heart failure characterized by right-sided volume overload. Clinical presentation ranges from breathlessness, general fatigue, reduced exercise capacity, palpitations, syncope to more severe features like atrial arrhythmias, right heart failure, and pulmonary hypertension (PHT) is possible.

PAPVR aggravates when there is a failure of primitive lung drainage recovery. This happens if pulmonary vascular bed is seen connected to the common PV from the LA from the embryonic stage. Based on the location of this abnormality, PAPVRs can be classified as three predominant types namely supra-cardiac (e.g., linkage to the SVC), cardiac (e.g., linkage to the right atrium [RA]), infra-cardiac or infra-diaphragmatic (e.g., linkage to the inferior vena cava), and finally a mixed types. Based on the number of PV that are involved in the malformation, the condition can be further divided into various branch types (BT) namely single BT, unilateral two BT, and bilateral single BT.

PAPVR sometimes presents bilaterally, however in majority of the patients (80% – 90%) it drains from right side into the SVC.[3] The right-sided PAPVR can drain into the RA or inferior vena cava (IVC). The right-sided PV drains into IVC and presents with concomitant right lung and right PA hypoplasia along with heart dextroposition, this scenario is called scimitar syndrome. In the contrary, left-sided PAPVR, results in the anomalous vein draining into the left innominate vein (LIV) via an anomalous vertical vein. in case of a bilateral PAPVR, the interatrial septum is seen to be normal and intact if the drainage happens into the LIV and the SVC.

The persistent systemic venous connectivity presents similar as LTRS, where a part of the right ventricle (RV) output is constantly recirculated and the reoxygenated blood reaches back the right heart avoiding the systemic circulation. Progressive remodeling happens with an increased pulmonary vascular resistance

when there is an excess pulmonary blood flow which leads to PHT and the right heart goes for a negative remodeling due to overactivity or demand on the right heart. ASD can result from Right-sided anomalous connections, which is the comments scenario. ASD in turn leads to the LTRS and forms the primary contributing factor. Hence, Right-sided PAPVRs along with ASD can potentially develop symptomatic PHT because of a clinically significant LTRS.

PAPVR usually presents with other cardiac defects and it is documented in few congenital syndromes, like Turner's syndrome (monosomy X). These abnormal veins are often related to ASD, with anomalous PV connection reported in 10% – 15% of patients on average and present with ostium secundum ASD and among 85% of the patients with sinus venosus ASD.[5]

Symptomatic patients with PAPVR require primary reconstruction which is a life-saving repair. Among patients with large LTRSs, when presenting with multiple PV, significant RV overload and PHT are seen. Through surgical correction the anomaly can be reversed and heart failure can be averted. At present the best available surgical options are shunt fraction, Qp/Qs of ≥ 1.5 presenting with or without clinical symptoms.[6] Patients undergoing Qp/Qs of < 1.5; when they are symptomatic should undergo complete evaluation and work-up for finding the risks and benefits of performing the correction.

PAPVR is better diagnosed using transthoracic echocardiography (TEE) and catheter-based angiography. TEE provides only limited information due to its limited acoustic window. Right heart catheterization performed with pulmonary angiography is considered as an operator-dependent and invasive procedure and it is hard to adequately depict, especially because of the small accessory and anomalous blood vessels. ASDs and PAPVR can be precisely defined using electrocardiography (ECG)-gated MDCT scans which is a noninvasive offering rapid image acquisition along with high spatial and temporal quality. Through MDCT, contrast bolus timing can be optimized and a wide anatomic coverage can be attained. MDCT angiography can give a reliable view of the presence, course, quantification anomalous veins, and concomitant cardiovascular defects.

Conclusion

PAPVR, a rare congenital abnormality in the adult population is due to LTRS which when not corrected PV remodeling results, and that leads to PHT. When PAPVR is associated with ASD, there is high chances for severe PHT. If PAPVR is detected, ASD should be carefully considered. The sinus venosus ASD is the most common; however, the secundum-type ASD can occur in 10% – 15% of patients with ASD.

The utilization of contrast-enhanced examinations with MDCT technology enables both detection and characterization of these anomalies.

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Conflicts of Interest

The authors declare no conflict of interest.

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