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Case Report

Partial anomalous pulmonary venous return with secundum atrial septal defect: A case report☆

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ABSTRACT

Partial anomalous pulmonary venous return (PAPVR) is a congenital heart anomaly in which some of the pulmonary veins return to the right atrium or one of its supplying veins instead of normally connecting with the left atrium. Oftentimes it is concurrent with a secundum atrial septal defect. PAPVR is typically asymptomatic, however symptoms of pulmonary hypertension can arise at higher degrees of left-to-right shunting. An 80-year-old male presented with exertional dyspnea and was found to have a secundum atrial septal defect on echocardiogram. A subsequent contrast enhanced computed tomography of the chest revealed a concomitant PAPVR.

Introduction

Partial anomalous pulmonary venous return (PAPVR) is a relatively rare and often asymptomatic congenital heart anomaly in which some of the pulmonary veins return to the right atrium or one of its supplying veins instead of the left atrium. In the adult population, anomalous pulmonary veins most often occur in the left upper lobe, with the right upper lobe being next most common [1]. Left upper lobe PAPVR typically drains into the left brachiocephalic vein via vertical vein and right upper lobe PAPVR drains into the SVC.

Case report

An 80-year-old male with known secundum atrial septal defect (ASD) with right ventricle dilatation and right ventricle volume overload on echocardiogram 3 months ago, presenting to outpatient imaging clinic for computed tomography (CT) of heart to examine ASD. The patient admits to progressive exertional dyspnea over the last few months. Contrast enhanced computed tomography (CECT) Chest redemonstrated the ASD with left to right flow (Fig. 1) and the presence of 2 anomalous right upper lobe pulmonary veins abnormally draining into

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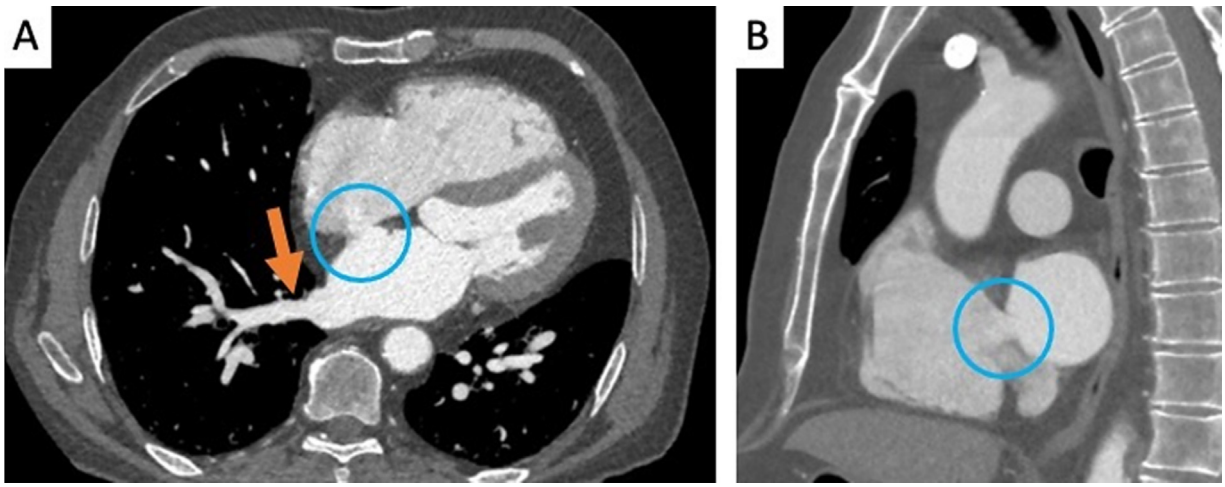

Fig. 1 – Eighty-year-old male with progressive exertional dyspnea. (A) Contrast-enhanced axial CT of the chest showing a secundum ASD (blue circle). Associated features include faint visualization of left to right flow of contrast through ASD, right ventricular dilatation without hypertrophy, and biatrial enlargement. Right inferior pulmonary vein also seen draining normally into the left atrium (orange arrow). (B) Sagittal CECT of the chest again demonstrates the secundum ASD (blue circle).

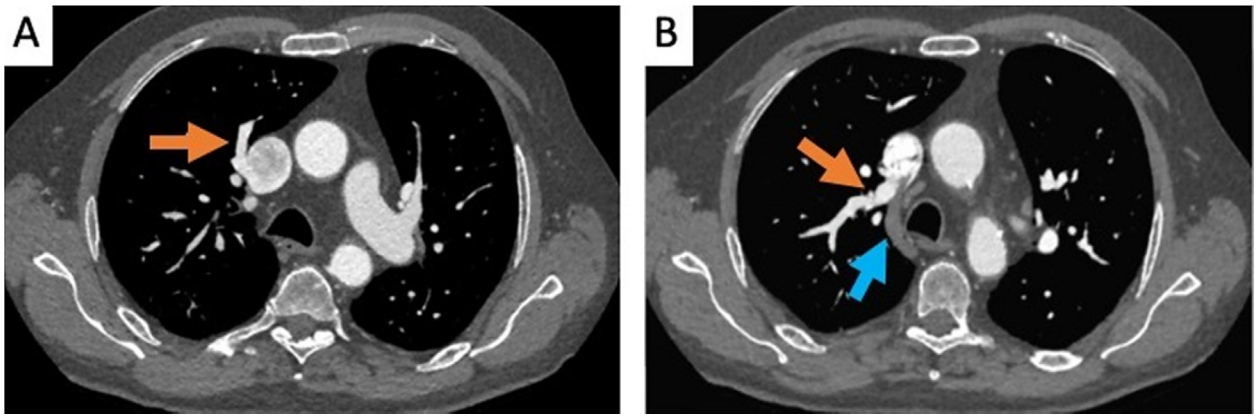

Fig. 2 – Eighty-year-old male with progressive dyspnea. (A) Contrast-enhanced axial CT of the chest showing an anomalous right upper lobe pulmonary vein (orange arrow) draining into the SVC (left-to-right shunt). (B) A second anomalous right upper lobe pulmonary vein (orange arrow) draining into the SVC. Non-opacified blood normally draining into the SVC from the azygos vein is also noted (blue arrow) at this level.

the superior vena cava with the remainder of the pulmonary veins draining normally into the left atrium, confirming PAPVR (Fig. 2). Pulmonary flow: Systemic flow ratio (Qp:Qs) was 1.68:1.

Discussion

PAPVR is relatively rare, only occurring in 0.1%-0.4% of the adult population [2–4]. Oftentimes it is concurrent with a secundum ASD. Although PAPVR is usually clinically asymptomatic, cases with a higher degree of left-to-right shunt may result in pulmonary hypertension and right ventricular volume overload (causing dyspnea, arrhythmias etc.) [5,6]. Shunt reversal or Eisenmenger syndrome can also be a consequence if right heart pressures exceed those on the left [7]. Shunt correction is usually reserved for symptomatic patients, cases with concurrent congenital heart disease or if Qp:Qs >2:1 [5]. The patient in this case had a Qp:Qs of 1.68:1 and was considered for surgical correction given that he was increasingly symptomatic.

The adult presentation of PAPVR most frequently involves anomalous pulmonary veins in the left upper lung lobe with drainage into the left brachiocephalic vein, while the childhood presentation involves the right upper lobe with drainage into the superior vena cava [1,6]. Drainage into the right atrium and inferior vena cava (Scimitar spectrum) is less common. Cardiac magnetic resonance imaging (MRI) or CT can be used to visualize the anomalous veins and frequently associated ASD. Phase-contrast MRI allows shunt quantification (Qp:Qs) [10]. PAPVR can be diagnosed with transthoracic echocardiography, however it is not as reliable as cardiac MR/CT [8].

Surgical repair with ASD patching, anomalous vein anastomosis and other methods may be warranted depending on symptoms and shunt fraction. Patients who are unable to undergo surgery may see improvement with prostaglandins and phosphodiesterase inhibitors [9].

Conclusions

PAPVR is a rare congenital heart defect that can often present with secundum ASD. It is often asymptomatic and discovered incidentally. Symptoms and prognosis depends on the severity of the shunt.

Case report consent statement

Written, informed consent for publication of their case was obtained from the patient. The patient was made aware of the information that would be present in the case report and that identity would not be disclosed. The patient was also informed that although personal information collected or obtained will be kept confidential, there is a limited risk associated with this case report that could result in loss of confidentiality due to the relatively unique case. The patient was made aware that he will not directly benefit from participating in this case report and that the information can be shared within the healthcare community and may improve the care of others in future similar cases. The patient was made aware that this case report involves no cost to him, and he will not receive any compensation. He was also informed that it is not possible to withdraw the case report once it is published.

Patient consent

Written informed consent was obtained from the patient for the publication of patient information in this article.

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