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Case Report

A Case Report Looking at an Incidental Finding of a Partial Anomalous Pulmonary Venous Connection Using Magnetic Resonance Angiography

Iqra Qamar,[1] Mohammad Waleed.[2]

Abstract

Background: Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital defect where one or more of the pulmonary veins drain to the right atrium or its tributaries. This can cause a left to right shunt (LTRS) which may need surgical management if symptomatic. **Results:** A 58-year-old female presented in emergency with non-pleuritic, central chest pain. She denied breathlessness and fever. Her past medical history included: non-ST elevated myocardial infarction (2013), anti-phospholipid syndrome, asthma, pulmonary embolism (PE) and deep vein thrombosis many years prior. Differential diagnoses included myocardial infarction, PE and unstable angina. Pulmonary angiography ruled-out PE. Coronary angiography did not show obstructive coronary artery disease. On cardiac MRA an incidental finding of PAPVC was found (left upper pulmonary vein drained into brachiocephalic vein) creating a LTRS which wasn't substantial (Qp/Qs <1.5). Despite being asymptomatic, this patient was followed-up of her incidental finding, and if symptoms developed she would be considered for PAPVC correction surgery. **Conclusion:** This case demonstrated the incidental finding of PAPVC in an otherwise asymptomatic patient. The decision was not for surgery as the patient was not symptomatic from the PAPVC and for follow-up. Previous studies showed PAPVC patients developing symptoms of LTRS are candidates for operation. Physicians should be aware of rare anomalies like PAPVC, their complications if unresolved, and should follow-up these patients closely in case of worsening of the LTRS, in which case surgical correction may be possible. Overall, PAPVC i) can be overlooked ii) diagnosis requires multimodality imaging iii) constant follow up and iv) may be operable.

Key Words: Anomalous Pulmonary Venous Return, Cardiac Magnetic Resonance Angiogram, Pulmonary Vein, Left To Right Shunt, Superior Vena Cava (Source: MeSH-NLM).

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a congenital anomaly in which one or more of the pulmonary veins drain to the right atrium or its tributaries, instead of the left atrium, causing a left to right shunt (LTRS).[1] The prevalence of incidental PAPVC is 0.1--0.2% and connection of the left upper pulmonary vein to the superior vena cava (SVC) is rare.[1,2-4] PAPVC can have associated congenital anomalies like atrial septal defect and has been reported to be syndromic (e.g. Turner syndrome) in up to 25% of patients.[5-6] In adults, PAPVC can be silent or present with its complications like dyspnea secondary to LTRS. If left undiagnosed, complications such as right ventricular hypertrophy can occur secondary to pulmonary hypertension. Ultimately, this can lead to right sided (RS) overload and RS heart failure.[7,8]

The Case

A 58-year-old female presented to the emergency department with non-pleuritic, central chest pain. She denied any breathlessness and did not report feeling febrile. She had a past medical history significant for a non-ST elevated myocardial infarction (non-STEMI) in 2013 with a drug eluting stent to the syndrome (APS), asthma, pulmonary embolism (PE) and deep vein thrombosis (DVT) many years prior. Furthermore, she was on aspirin due to APS, but no current anticoagulation. Therefore, differential diagnoses included myocardial infarction (MI), PE and unstable angina of which pulmonary angiography ruled-out PE.

Key Points:

- PAPVC is a rare, often silent and can be easily missed which clinicians should be aware of.
- Patients should be followed up closely for development of symptoms or worsening of the LTRS.
- It is very important to systematically review all the data obtained from every imaging modality to prevent missing a potential diagnosis.

Investigation findings showed T wave inversion in leads V2-V5 on EKG and an elevated Troponin T (532 ng/L) confirming an NSTEMI. Therefore, coronary angiogram was immediately performed, showing patent LAD artery stent and moderate (50-60%) stenosis in the mid-right coronary artery (RCA). Cardiac Magnetic Resonance (CMR) was performed, showing preserved LV ejection fraction (LVEF=67%), delayed hyper-enhancement of the septum and apex involving more than 50% of the wall thickness which is consistent with myocardial infarction (MI). On the MRA an anomalous pulmonary venous drainage of the left upper pulmonary vein into the brachiocephalic vein was noted as an incidental finding (***Figure 1***). The LTRS was not substantial as Qp/Qs was < 1.5. (Qp/Qs = 1.3). Her case was discussed in a Cardiology-Cardiothoracic Multidisciplinary Team (MDT) meeting and the decision was not for surgery as the patient was not symptomatic from the PAPVC and to follow up the patient in the cardiology clinic.

[1] University of Leeds, Leeds, United Kingdom.
[2] Dr., Leeds General Infirmary, Leeds, United Kingdom.

About the Author: *Iqra Qamar is affiliated with the University of Leeds, Leeds, United Kingdom.*

Correspondence:
Mohammad Waleed
Address: Great George St, Leeds LS1 3EX, UK.
Email: waleed1280@yahoo.com

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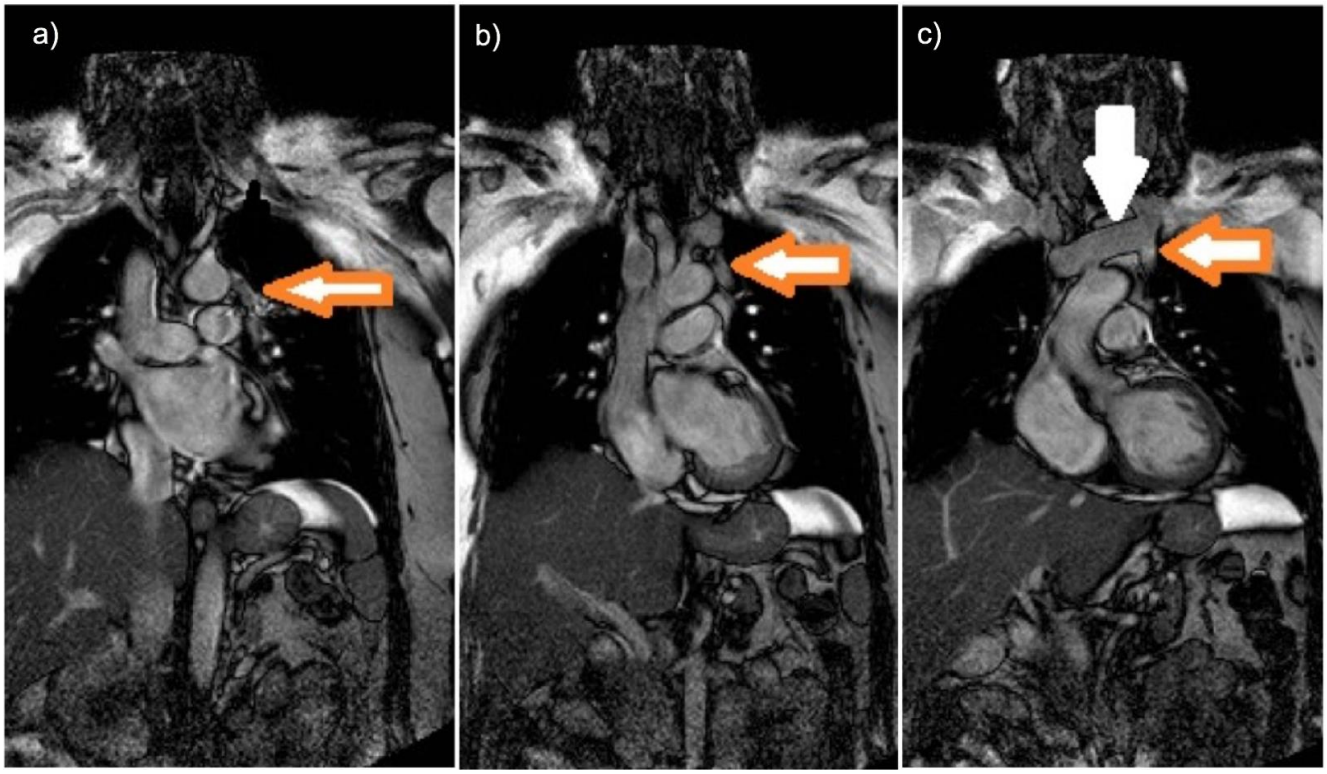
Figure 1. The Patient's Magnetic Resonance Angiogram.



Legend: An MRA Scan Showing (a) An orange arrow points towards the left upper pulmonary vein at the beginning of the anastomosis (b) The orange arrow highlights the middle of the anastomosis between the left upper pulmonary vein and the brachiocephalic vein (c) The arrow is pointing out the left upper pulmonary vein anastomosis with the brachiocephalic vein, marked by the white arrow).

Six months after discharge, the patient was seen in the cardiology clinic and was asymptomatic from the PAPVC, denying any shortness of breath or further chest pain.

Studies have shown asymptomatic PAPVC patients with low post-surgical repair mortality. [13] One study showed that 93% of patients were free from arrhythmia medications at post-surgical follow-up and the remaining 7% had either atrial fibrillation, sinus node dysfunction (SND) or SND ectopic atrial rhythm. [13] Most common surgical risks included getting an arrhythmia, risks of hemorrhage, thrombus, infection, poor tolerance of analgesic methods used and death. However, as these risks are smaller than the risks of untreated PAPVC. Symptomatic patients or those with complications of pulmonary hypertension and right ventricle hypertrophy should be considered for surgical correction to prevent developing a LTRS and eventually RS heart failure.[12-14]

Articles have previously reported diagnosing incidental PAPVC in patients with lung cancer. One patient's PAPVC anatomy mimicked our patient and although asymptomatic, they had increased mean arterial pressure and Qp/Qs ratio > 2.0. [15] Despite a significant co-morbidity of lung cancer, the post-surgical recovery and survival was commendable and supportive of surgery. Despite follow up by cardiologists, some patients were not diagnosed with PAPVC until post CT.[16]

These studies advocate using multimodality imaging to aid diagnosis of PAPVC especially in symptomatic patients with chest symptoms. Ultimately, PAPVC is rare and further research is required showcasing different presentations and ethnic groups.[15-16]

This case highlights incidental PAPVC found on a cardiac MRA performed for assessment of a MI. Our case was discussed in MDT and being asymptomatic from the PAPVC perspective, the decision was not for surgery and to follow the patient in the cardiology outpatient clinic. Symptomatic PAPVC patients can be managed by surgery and repair of the anomalous connection may be performed successfully.

In conclusion, PAPVC is rare, can be fatal and in this case could be easily overlooked as RS heart symptoms were absent. Awareness is crucial as if PAPVC patients become symptomatic, surgical correction may be considered; hence appropriate follow up should be accomplished. Ultimately this case emphasizes the importance of i) advocating relevant multimodality imaging, ii) appropriate patient follow-up iii) and surgical consideration for PAPVC patients who are symptomatic or have developed pulmonary or RS heart complications.

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Author Contributions

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