Annals of Case Reports
Baig M, et al. Ann Case Rep: 9: 101644
www.doi.org/10.29011/2574-7754.101644
www.gavinpublishers.com

Case Report

Unusual Doppler in Pulmonary Veins Leads to the Diagnosis of Unusual Partial Anomalous Pulmonary Venous Return with Dual Connections

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Citation: Baig M, Nasir A, Ou J (2024) S Unusual Doppler in Pulmonary Veins Leads to the Diagnosis of Unusual Partial Anomalous Pulmonary Venous Return with Dual Connections. Ann Case Report 09: 1644. DOI: 10.29011/2574-7754.101644.

Received Date: 06 February 2024; **Accepted Date:** 12 February 2024; **Published Date:** 14 February 2024

Abstract

In this paper, we report an unusual case of Partial Anomalous Pulmonary Venous Return (PAPVR) in a patient with new onset, decompensated heart failure and right sided chamber enlargement.

Keywords: PAPVR; Right Heart Enlargement

Introduction

Partial anomalous Pulmonary Venous Return (PAPVR) can include a variety of congenital abnormalities that are caused by the abnormal return of one or more pulmonary veins to the right side of the heart. Clinical presentation often varies as it is related to the degree of shunting and presence of other abnormalities including pulmonary and cardiac [1]. It can often be difficult to identify and remains underdiagnosed. Imaging is essential to diagnosis and required to obtain accurate anatomy. This includes echocardiography, cardiac CT, cardiac MRI and often, cardiac catheterization, to verify the degree of shunting. Color flow mapping and Doppler methods are all helpful tools when evaluating potential anomalous connections. PAPVR should be considered in situations where SVC, innominate vein or IVC appear dilated, as well as isolated right sided enlargement [2]. In this paper, we report an unusual case of PAPVR in a patient with new onset, decompensated heart failure and right sided enlargement.

Case Presentation

A 68 year old male with a past medical history of hypertension, hyperlipidemia and chronic kidney disease presented for progressive dyspnea on exertion and bilateral lower extremity edema over three weeks. He was found to be in decompensated heart failure and new onset Atrial Flutter with rapid ventricular rate. Transthoracic echocardiography revealed severely impaired Left Ventricular Systolic function with Ejection Fraction (LVEF) <20%, enlarged right ventricle with moderate right ventricular systolic dysfunction, biatrial enlargement, mild pulmonary hypertension. He was started on guideline directed medical therapy and referred for Transesophageal Echocardiogram (TEE) with cardioversion for new onset Atrial Flutter and possible tachycardia induced cardiomyopathy. During TEE, markedly dilated Superior Vena Cava (SVC) (3.6 cm) was seen and routine Pulse wave Doppler of Right Upper Pulmonary Vein (RUPV) showed unusual predominant outflow with brief mid late systolic inflow (Figure A,B). Careful color Doppler interrogation of RUPV revealed normal inflow of two branches of pulmonary veins into

RUPV while significant reversal flow in distal RUPV was seen. These findings suggest Partial Anomalous Pulmonary Venous Return (PAPVR) with dual connection to left atrium and systemic veins. All three other pulmonary veins appeared normal during careful TEE exam. He was also found to have positive bubble study likely related to PAPVR since no Patent foramen ovale or other form of atrial septal defect was identified. Subsequent right heart catheterization revealed left to right shunt with Qp/Qs 1.6, oxygen saturation step up noted between SVC and right atrium. Subsequent CT Angiography (CTA) of heart revealed partial anomalous right upper pulmonary vein had dual connection to both SVC via window like communication and left atrium (Figure C), no sinus venosus defect or atrial septal defect was seen. Patient underwent successful cardioversion and was treated with guideline direct medical therapy for heart failure. His repeat echocardiogram 6 month after cardioversion showed recovered LVEF to 45-50% and moderately enlarged right ventricular with normal systolic function. Patient is referred to cardiac surgery for surgical correction of PAPVR.

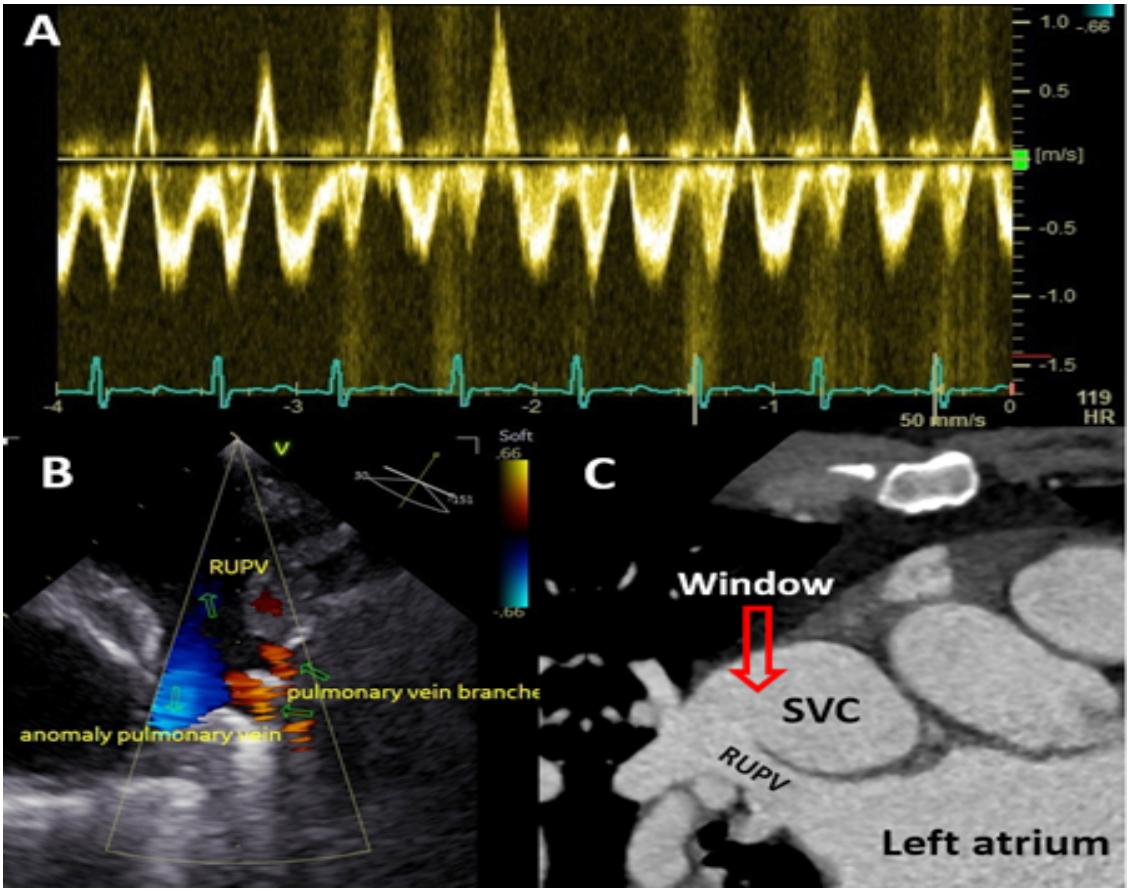

Figure A-C: A. Pulse wave Doppler of right upper pulmonary vein (RUPV) during transesophageal echocardiogram (TEE) exam. B. Color Doppler of RUPV during TEE exam; C. 3-D reconstruction of CT angiography of heart. Red arrow showed window like communication between SVC and RUPV.

Discussion

PAPVR is a rare congenital abnormality, consisting of abnormal blood return from one or more pulmonary veins to the right atrium. It often occurs with other cardiac abnormalities [2]. Clinical presentation often varies depending on the degree of shunting and other associated cardiac or pulmonary abnormalities. Patients with isolated PAPVR frequently remain asymptomatic [3]. Patients with an associated ASD may present with significant left to right shunting, right sided chamber enlargement and subsequent pulmonary hypertension [3]. This case illustrates an unusual example of PAPVR with bidirectional flow, as well as the importance of thorough evaluation even on routine transesophageal echocardiograms especially for patients with right heart enlargement.

Conclusion

Patients with significant right heart enlargement warrant careful evaluation of left to right shunt with multimodality imaging approach.

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