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Short communication

Pulmonary hypertension due to presence of isolated partial anomalous pulmonary venous connection: A case report

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ABSTRACT

Anomalous pulmonary venous return is an uncommon congenital malformation which can be broadly categorized into partial or total, of which the former is more common. The anomaly is considered to be partial if some of the pulmonary veins drain into the systemic circulation and total if all the pulmonary veins drain into systemic circulation. Isolated partial anomalous pulmonary venous return (PAPVC) is an uncommon finding and is a very uncommon cause of pulmonary arterial hypertension. Whilst many patients with PAPVC remain asymptomatic, some may present at a later age with symptoms related to left-to-right shunt, pulmonary hypertension and right heart failure. We are presenting an interesting case report of an 18 years old patient who presented with exertional dyspnea and fatigue conforming to NYHA class II symptom status. Trans-esophageal echocardiography revealed isolated obstructive PAPVC as the cause for pulmonary hypertension without other demonstrable left-to-right shunts.

1. Introduction

Partial anomalous pulmonary venous connection (PAPVC) is an uncommon congenital abnormality in which some, but not all, of the pulmonary veins connect directly to the right atrium or to one of its venous tributaries.[1] Rarely, PAPVC can present as an isolated cause of pulmonary hypertension.[2,3] We are reporting a case of 18 years old female who presented with effort intolerance. Investigations revealed the presence of obstructive PAPVC being the underlying etiology for pulmonary hypertension in this case.

2. Case report

An 18-year-old female presented with effort intolerance and easy fatigability conforming to New York Heart Association (NYHA) class II symptom status since the last one year. She didn't give any past history of recurrent chest infection or heart failure in infancy which could suggest the presence of a large left to right shunt in infancy. Her family and personal history was non-contributory the diagnosis. Blood investigations revealed that the total and differential leukocyte counts and the other parameters of complete hemogram were normal (hemoglobin – 12.1 g/dl, total leukocyte count – 8500/cu mm, platelet count – 175,000/cu mm) and the liver & thyroid function tests were also normal. Her renal function parameters were deranged (blood urea – 48 and serum creatinine – 2 mg/dl). Ultrasound abdomen revealed the presence of grade I renal parenchymal disease. Urine examination was found to be normal. Electrocardiogram revealed presence of 'P' pulmonale with the presence of right axis deviation. Transthoracic echocardiography (TTE) showed dilated right sided chambers with the presence of mild to moderate tricuspid regurgitation (TR) with a TR jet gradient of 42 mm of Hg (Fig. 1) suggestive of moderate pulmonary hypertension (PHT). Trans-esophageal echocardiography (TEE) was performed to confirm the etiology of PHT in this case which showed the presence of an obstructed PAPVC connecting to the inferior vena cava (IVC) without the presence of any other left to right shunt lesions like atrial septum defect (ASD) or ventricular septal defect (VSD) (Fig. 2; Video 1). Echocardiographic assessment of the Qp:QS was not possible in this case as the pulmonary vein was obstructed and was connected to the inferior vena cava. As the patient had significant renal dysfunction and also because there was no doubt regarding the diagnosis of the patient, only non-contrast computed tomography (CT) scan of chest was performed in order to avoid further worsening of the renal function. The CT did not reveal any pulmonary parenchymal disease or any other etiology which could have contributed to the presence of PHT. She was treated with low dose diuretics and oral sildenafil. She was planned for cardiac MRI or cardiac catheterization after improvement of renal function with a plan to take the patient for surgical correction of the defect, but unfortunately the patient was lost to follow-up.

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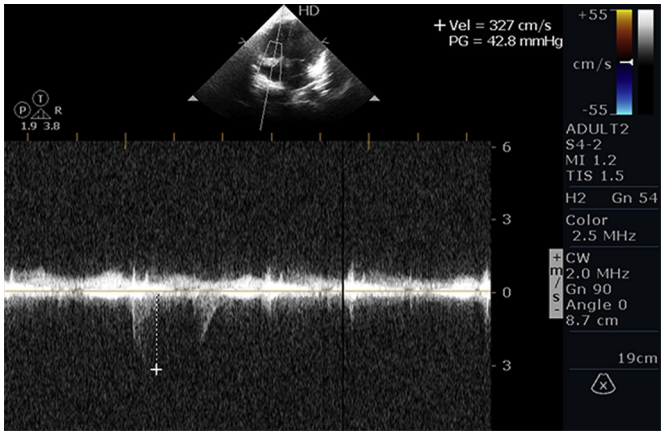

Fig. 1. TTE with CW Doppler showed presence PHT with TR jet of 42 mm of Hg.

Supplementary video related to this article can be found online at http://dx.doi.org/10.1016/j.jcdr.2014.01.002.

3. Discussion

PAPVC occurs when some of the pulmonary veins connect to directly to the right atrium or one of its venous tributaries rather than the left atrium and is commonly associated with an ASD, found in 80% of patients in one pediatric series.[4] Most anomalous pulmonary veins arise mainly from the right lung; connecting primarily to the superior vena cava (SVC), less commonly to the right atrium (RA) or the inferior vena cava (IVC). Only 3–8% of the anomalous pulmonary veins have been reported to originate from the left lung, connecting to the left brachiocephalic vein in all cases.[5] PAPVC is often clinically silent and recently in a retrospective review of computed tomography (CT) series in adults who had undergone imaging for other indications, have identified prevalence rates of 0.1–0.2% in the adult population.[6] Persistent anomalous pulmonary venous connection acts similar to a left-to-right shunt and over time (which may span between few years to decades), the increase in pulmonary blood flow can lead to progressive remodeling of the pulmonary vasculature which may cause increased pulmonary vascular resistance. If the remodeling and vasoconstriction of the pulmonary vasculature is severe enough, pulmonary arterial hypertension (PAH) and right ventricular pressure overload occur which may subsequently lead to RV failure.[7]

The patients can present anytime from infancy to the seventh decade, depending on the size of the shunt, as well as upon the presence of other cardiac anomalies. The presenting manifestations may also vary from being nearly asymptomatic to patients presenting with frank hypoxemia and signs of RV failure. Though among the non-invasive imaging CT scan is an extremely effective diagnostic modality, especially when iodinated contrast is used to delineate the pulmonary vasculature,[8] but in our case due to renal dysfunction we were not able to perform it.

TTE, which is often obtained in patients being evaluated for cardiac symptoms, cannot reliably delineate pulmonary venous anatomy due to technical limitations. In pediatric patients, approximately one third of cases of PAPVR are missed by TTE and the proportion is likely higher in adults.[9] TEE is quite sensitive and specific in the hands of experienced operators[10] and the flame shaped image are characteristic for obstructed PAPVC as seen in our case.

Shunt fraction calculation in our case was not possible as the patient could not be subjected to cardiac catheterization or MRI due to renal dysfunction at presentation and the patient having subsequently been lost to follow up. Echocardiographic assessment of the shunt fraction was not possible in this case as the anomalous pulmonary vein was obstructed and was connecting to the IVC.[11,12]

The mechanism behind the presence of pulmonary hypertension in our case could be pulmonary arterial hypertension (PAH) caused by the obstructed PAPVC can increase pressure over pulmonary artery and results in reactive PHT as is seen in cases with mitral stenosis.

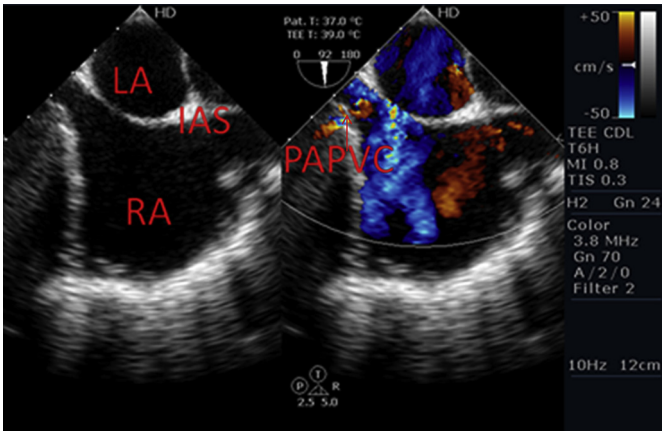

Fig. 2. TEE with color compare image showed presence of isolated right inferior pulmonary vein draining to IVC with intact intra atrial septum (IAS).

4. Conclusion

In conclusion, PAPVR is a rare congenital condition which is usually recognized in the pediatric population but may also be diagnosed late during adulthood in patients who develop PAH. Physicians who diagnose and treat adult patients with PAH should consider PAPVC as a possible etiology, particularly in cases with volume or pressure overloaded right heart chambers and in cases not having any other obvious explanation for the same like the presence of left to right shunt lesions like ASD, VSD or PDA.

Conflicts of interest

All authors have none to declare.

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