

Pulmonary Arterial Hypertension in An Adult: What Would Be Your Diagnosis?

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Abstract

Case Report

Partial anomalous pulmonary venous return (PAPVR) accounts for approximately 3% of all congenital heart disease. Its diagnosis in adulthood is often tricky as long as clinical symptoms are misleading and sonographic signs are discrete and suggestive of pulmonary hypertension. We report the case of a 65-year-old female patient who was diagnosed with pulmonary arterial hypertension on partial abnormal pulmonary venous return on chest CT.

Keywords: Partial anomalous pulmonary venous return, pulmonary arterial hypertension, chest CT scan.

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INTRODUCTION

Partial anomalous pulmonary venous return (PAPVR) is estimated to occur in approximately 3% of all congenital heart disease. This congenital condition is often associated with an atrial septal defect. Its overall incidence is estimated to be 0.7% in the population [1].

The diagnosis of RVAP is a clinical challenge. The importance of the functional signs is directly correlated to the extent of the drained territory and the obligatory left-right shunt. Often the discovery is made during a cardiac echocardiography prescribed for a heart murmur or for symptoms not specific to this pathology such as dyspnea, atrial arrhythmia or right heart failure.

We report the case of a 65-year-old woman who was found to have RVPAP, and we describe the history of the patient whose symptoms were not suggestive, as well as the contribution of the thoracic CT scan that allowed us to retain this diagnosis.

CASE REPORT

The patient was 65 years old woman, with an unknown pathological history, suffering from exertional dyspnea classified as NYHA stage 2, which had been evolving for more than 1 year and had recently become stage 3.

On the thoracic CT scan in helical acquisition with multy planar reconstruction without and with injection of iodinated contrast, we note: trunk of the pulmonary artery as well as its right and left branches are increased in caliber measuring respectively 45, 35 and 27 mm, opacified after injection of contrast product, dilatation of the right heart chambers, and demonstration of a right superior pulmonary vein flowing into the superior vena cava (SVC) just above its junction with the right atrium (RA).

Note the absence of any other communication between the right and left heart chambers. We therefore conclude that this is a case of pulmonary arterial hypertension due to partial anomalous pulmonary venous return through the presence of a right superior pulmonary vein which terminates in the RA via the SVC.

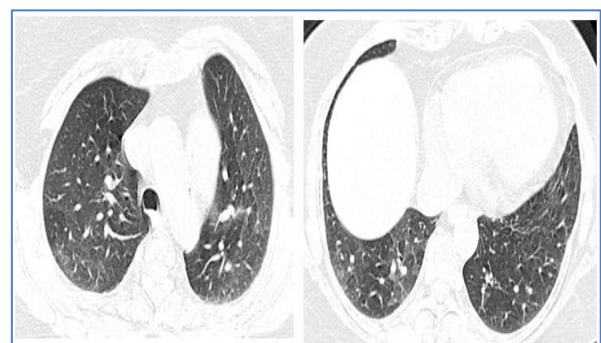


Fig-1: Chest CT axial sections in parenchymal window showing mosaic perfusion

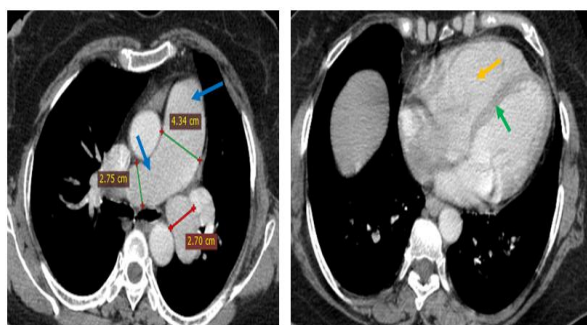


Fig-2: Dilation of the pulmonary artery trunk and the right and left pulmonary arteries (blue arrow). Dilation of the right heart chambers (yellow arrow) with straight interventricular septum (green arrow)

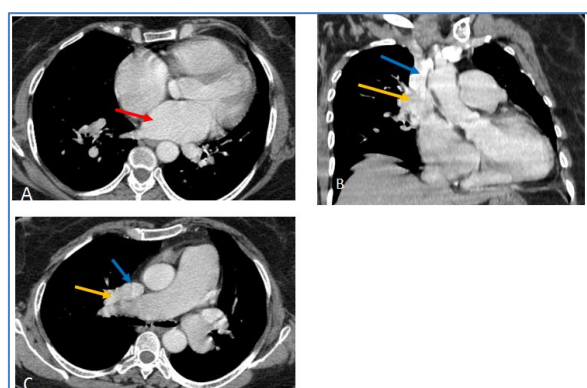


Fig- 3: A: Absence of individualisation of the right superior pulmonary vein (red arrow) B, C: Right superior pulmonary vein (yellow arrow) discharging into the SVC (blue arrow)

DISCUSSION

Partial anomalous pulmonary venous return, a milder form of total anomalous pulmonary venous return, occurs when one, most often, or more pulmonary veins drain into the venous or right side of the heart instead of the left atrium. In the most common form of partial anomalous pulmonary venous return, drainage occurs into the superior vena cava. Less commonly, it may drain into the inferior vena cava, right atrium or coronary sinus. These abnormal communications cause a left-to-right shunt of oxygenated blood to the atrium, which results in dilation of the right heart [1].

In 80-90% of cases, abnormal partial upper lobar pulmonary venous returns are associated with a sinus venosus type atrial septal defect but other anomalies may be observed such as mitral or pulmonary stenosis, a ductus arteriosus. These anomalies are less frequent in partial anomalous pulmonary venous returns of the lower lobes [1].

PAPVR are congenital anomalies. In adult subjects, the average age of discovery varies in the literature from 19 to 83 years. The predominance of women is the rule [2].

Frequency estimates in the pediatric population range from 0.3 per 1,000 (clinical series) to 0.6 per 1,000 (autopsy series). Half of them would therefore not be recognized clinically [3].

Only a few patients with a large increase in pulmonary blood flow have symptoms (e.g., dyspnea on exertion, asthenia). Severe symptoms such as cyanosis and congestive heart failure are relatively rare. In addition to right ventricular failure, patients also have right atrial dilatation and a high number of dysrhythmias [4].

The severity of symptoms correlates with the number of segments involved, the sites of connection, and the presence of other pulmonary vascular anomalies or associated cardiac defects [3].

Most single pulmonary vein anomalies are asymptomatic in childhood and youth. This is the case in our patient. The left-to-right shunt is normally well tolerated and the diagnosis may remain unrecognized until adulthood. It is only when the pulmonary hypervascularization secondary to the left-right shunt causes significant pulmonary hypertension that patients develop symptoms (left-right shunt is greater than 50%) [4, 5].

With age, the increase in left filling pressures promotes the increase of the shunt, because of the low pressures in the right atrium. The shunt is responsible for radiological abnormalities that are long interpreted as being of bronchopulmonary origin.

Sometimes, an abnormal vein draining into the IVC is visible on the chest X-ray as a crescent-shaped shadow of vascular density along the right edge of the cardiac silhouette (scimitar syndrome) [6].

Visualization of the pulmonary veins and their drainage can be difficult with transoesophageal echocardiography (TEE), but Cardiac catheterisation and transthoracic echocardiography (TTE) or thoracic CT angiography is often used to make the diagnosis. Cardiac catheterization can demonstrate elevated filling pressures as well as oxygen saturations of the cardiac chambers and large vessels [7].

Thoracic CT angiography has a primordial role in the different stages; diagnosis and management via millimetric slices and multy planar reconstruction, and so allows precise characterization of the cardiac anatomy including pulmonary venous drainage, as in our patient's case [8].

MRI allows a global evaluation of patients with PAPVR, overcoming most of the limitations of echocardiography, and can be considered a suitable alternative to catheterization [7, 8]. The prognosis of

partial anomalous pulmonary venous return is excellent [5].

Surgical repair is indicated when there is a large left-to-right shunt. Similarly, when an AIC is associated, it should be closed so that pulmonary venous return is directed to the left atrium. A single abnormal pulmonary vein without an atrial septal defect may be difficult to redirect to the left atrium; if the shunt is small and the right ventricle is not dilated, it may be left unoperated [8].

CONCLUSION

A very large number of partial abnormal pulmonary venous returns are misdiagnosed as primary pulmonary hypertension. Our case highlights the role of CT scanning in establishing both the positive and causal diagnosis.

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