Anomalous pulmonary venous return: a report of a case diagnosed by CT pulmonary angiogram

Amal Moukhliss¹,², Soukaina Safir¹, Rihab Machtache¹, Hatim Zahidi¹, Abdennaser Drighil¹, Rachida Habbal¹, Kawtar Charef², Fadwa Kossale², Saloua Lazaar², Saloua Elmanjra², Fatiha Essodeigui²

¹Cardiology Department, Ibn Rochd University Hospital Center, Casablanca, Morocco, ²Radiology Department, Ibn Rochd University Hospital Center, Casablanca, Morocco

Corresponding author: Amal Moukhliss, Cardiology Department, Ibn Rochd University Hospital Center, Casablanca, Morocco

Received: 12 Dec 2019 - Accepted: 11 Feb 2020 - Published: 24 Feb 2020

Domain: Cardiology

Keywords: Anomalous pulmonary venous return, pulmonary vein, CT Angiogram

Abstract

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital cardiopathy that can progress to right heart failure in the absence of treatment. It can be associated with an intra-auricular communication. The APVR remains often asymptomatic and its discovery is in the majority of cases fortuitous. The thoracic CT Angiogram is the examination of choice to make the diagnosis by showing the localization, the number and the path APVR drainage. We report the case of a 26-year-old female patient with congenital heart disease type an atrial septal defect; who was diagnosed with a partial bilateral APVR associated with pulmonary arterial hypertension signs (PAH). We describe the history of the patient whose symptomatology was nonspecific and in whom the imaging examinations, in particular the CT angiogram, made it possible to reach the diagnosis.
Introduction

Anomalous pulmonary venous return is an abnormality of connection of the pulmonary veins to the left atrium. It consists in the drainage of one or more pulmonary veins other than in the left atrium. Numerous anatomical variants are encountered, ranging from the partial APVR of incidental discovery to the totally blocked APVR (due to an infradiaphragmatic obstacle) which is considered a neonatal emergency [1].

Patient and observation

A 26-year-old lady, followed for an atrial septal defect at the Cardiothoracic Surgery Department of Ibn Rochd University Hospital of Casablanca, consults for dyspnea and chest pain progressing rapidly without fever. On clinical examination, signs of right heart failure were found; SpO2 was 95%, her blood pressure was at 120/80mmHg, her pulse at 80 beats/min. On auscultation, a pulmonary outflow murmur was detected with some wheezing sounds. Blood tests (blood count, CRP, Troponin and D-Dimers) revealed no abnormalities. The patient underwent an arterial blood gas test. The arterial blood pH was at 7.47, partial pressure of oxygen was at 60mmHg, partial pressure of carbon dioxide was at 37mmHg and oxygen saturation at 95%. The ECG showed a regular sinus rhythm with an incomplete right bundle branch block, without conduction or repolarization disorder or signs of right ventricular hypertrophy. The chest X-ray displayed a cardiomegaly with a cardio-thoracic index equal to 0.6 and a dilatation of the pulmonary trunk without pulmonary opacities or pleural effusion. A CT pulmonary angiogram was then performed, showing abnormal drainage of the left superior pulmonary vein into the innominate vein and right SPV into the superior vena cava. It also reported significant dilatation of the pulmonary trunk, the right and left pulmonary arteries without intraluminal material. The diagnosis was that of partial left and right bilateral APVR with CT signs of pulmonary arterial hypertension, without signs of pulmonary embolism (Figure 1, Figure 2).

Discussion

The incidence of APVR is estimated to be 0.4 to 0.7 in the autopsy series [2]. There is a difference between total abnormal pulmonary venous return PAPVR, which represents 2% of all congenital heart defects, and 30% of pulmonary venous return abnormalities. These are severe malformations in which all pulmonary veins drain into the right atrium or its afferent vessels. This abnormality is responsible for heart failure and cyanosis in the first days after birth. An interatrial communication is obligatorily associated and partial venous return (RVPAP). It accounts for 70% of cases, often discovered by chance. The anomaly concerns either a pulmonary vein or both ipsilateral pulmonary veins [3]. PAPVR is mainly found in adults in a fortuitous way [4,5]; occurring either alone or most often in association with congenital heart disease (interatrial or interventricular septal defects). It is an abnormal connection of the pulmonary vascular plexuses and the primary pulmonary vein [6]. The symptomatology, related to the importance of the left-right shunt (increased by interatrial communication), reflects the elevation of pulmonary arterial pressures.

The PAPVR is most often asymptomatic, as in our observation and fortuitously discovered in adulthood during a CT scan, which represents the best diagnostic tool [7]. Clinical signs are mostly absent but asthenia, exercise dyspnea, revealing dry cough, palpitations and chest pain are reported [4]. Heart failure is also possible. The embryonic development of these malformations is still poorly understood. APVRs are 10 times more frequent on the right than on the left [8]. In our case we noted right and left PAPVR. APVRs are twice as frequent on the
right and are located on the upper lobes. They drain into the superior subazygotic vena cava, into the large azygos vein or directly into the right atrium [5]. Left APVRs are more rarely observed and was analyzed in a recent retrospective series of 29 adult patients [4]. They drain to the left in the left brachiocephalic venous trunk, in the superior vena cava, in the coronary sinus or in the azygous system [4,5]. Chest CT scan with iodinated contrast injection (CT pulmonary angiogram) or injected thoracic MRI (MR angiography), transesophageal echocardiography and cardiac catheterization are, ranked from the least to the most invasive imaging, currently the key diagnostic tests [9]. In asymptomatic patients, therapeutic abstinence is the rule. In case of a significant shunt, the surgical treatment consists of directing the venous return to the left atrium, with a Dacron tube if necessary and to close the possible interatrial communication [6].

**Conclusion**

Abnormal pulmonary venous return (APVR) is a rare condition. The diagnosis in adulthood is often difficult. CT pulmonary angiogram simplifies its diagnosis.

**Competing interests**

The authors declare no competing interests.

**Authors’ contributions**

All the authors have read and agreed to the final manuscript.

**Figures**

**Figure 1**: axial CT angiography with coronal reconstruction and VR showing the entry of the right upper pulmonary vein draining into the SVC

**Figure 2**: axial CT angiography with coronal reconstruction and VR showing the opening of the left superior pulmonary vein which drains towards the innominate venous trunk

**References**


Figure 1: axial CT angiography with coronal reconstruction and VR showing the entry of the right upper pulmonary vein draining into the SVC
Figure 2: axial CT angiography with coronal reconstruction and VR showing the opening of the left superior pulmonary vein which drains towards the innominate venous trunk.