

REPORT OF A RARE ANATOMIC VARIANT: LEFT UPPER LOBE PARTIAL ANOMALOUS PULMONARY VENOUS RETURN

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We report the CT findings in a case of partial anomalous pulmonary venous return (PAPVR) from the left upper lobe in an adult. PAPVR is an anatomic variant in which one to three pulmonary veins drain into the right atrium or its tributaries, rather than into the left atrium. This results in a left-to-right shunt with varying clinical presentation. These can range from asymptomatic patients to advanced cardiac failure.

Key-word: Pulmonary veins, abnormalities.

Case report

A 61-year-old man presented to the emergency department with a 5-week history of dyspnea, cough and transient episodes of fever. These symptoms did not respond to antibiotic treatment. He was known with COPD GOLD III and α 1-antitrypsin deficiency. Initially he had an arterial oxygen saturation of 87%, which could not be increased with O₂-administration.

Clinical findings

Lung auscultation yields bilateral rhonchi and discrete expiratory wheezing. Laboratory findings include elevated C-reactive protein of 225 mg/l, leukocytosis of 10.200/mm³ and increased D-dimer of 1074 ng/ml. Plain chest film showed a Chronic Obstructive Pulmonary Disease (COPD) configuration of the chest, with an opacification in the lower left lobe. To rule out pulmonary embolism a chest CT angiography was performed, using 60cc lobitridol 300mg (Xenetix 300, Guerbet, Belgium). This showed (Fig. 1) predominantly basal centrilobular emphysema with a consolidation in the lower left lobe. There were no signs of pulmonary embolism. By coincidence, we depicted an anomalous drainage of the left upper lobe pulmonary vein via a large (anomalous) vertical vein, into the innominate vein, causing a left-to-right shunt (Fig. 2).

Discussion

Partial anomalous pulmonary venous return (PAPVR) is a rare condition with a prevalence of 0.4-0.7% (1). In PAPVR, up to three pulmonary

veins connect to the right atrium or its tributaries, rather than to the left atrium. The most common presentation of PAPVR is a right upper lobe vein draining into either the right atrium or superior vena cava. The right-sided PAPVR is typically associated with an atrial septal defect (ASD) of the sinus venosus type.

Only 10% of the PAPVR are left-sided, and 3% of the reported cases show drainage from the left lung into the innominate vein, as seen in our case. In contrast to the life-threatening total anomalous pulmonary venous return, the PAPVR is usually detected at a later age or during autopsy. The PAPVR results in a left-to-right shunt, similar to an ASD,

ventricular septal defect or a patent ductus arteriosus (1).

The clinical presentation varies widely from asymptomatic patients to congestive heart failure. Long-standing PAPVR predisposes the patient to right-sided volume overload, tricuspid regurgitation (TR), arrhythmias, pulmonary arterial hypertension, irreversible pulmonary vascular disease, right ventricular dysfunction and right ventricular failure (2). Several pulmonary anomalies may be associated with PAPVR. These malformations include bronchopulmonary sequestration, pulmonary arteriovenous malformation, congenital diaphragmatic hernia and cystic adenomatoid malformation (3). Another rare form of PAPVR, called the Scimitar syndrome or hypogenetic lung syndrome, is characterised by pulmonary venous drainage of a portion or all of the right lung to the inferior vena cava. The anomalous vessel

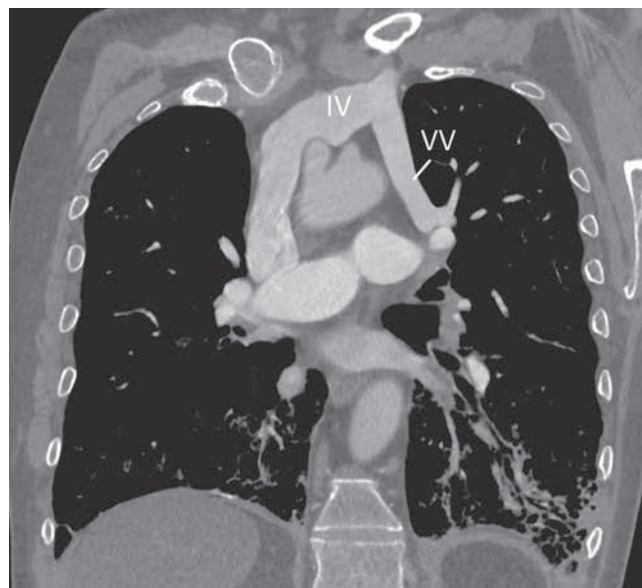


Fig. 1. – Curved coronal CT image of the chest demonstrates a 15 mm-large vertical vein (VV) draining blood from the left upper lobe pulmonary vein into the innominate vein (IV).

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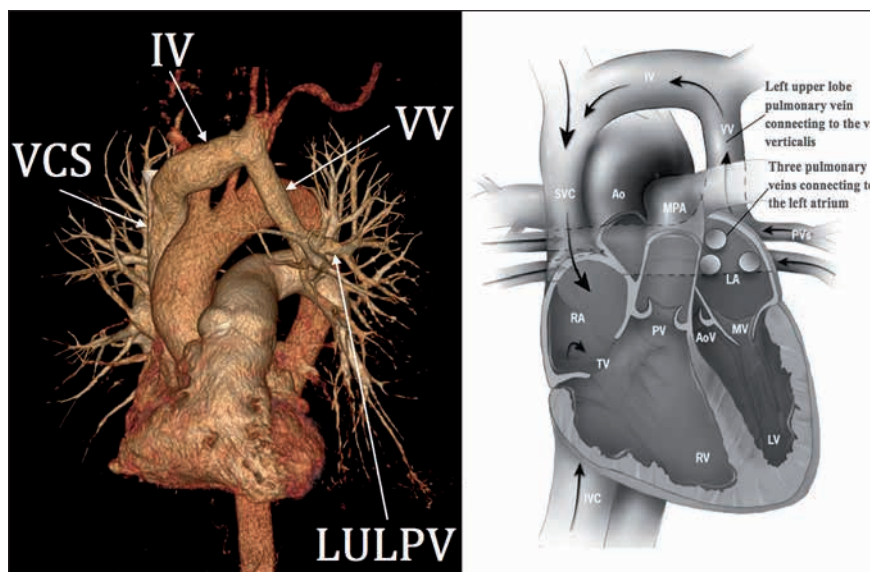


Fig. 2. – A. 3D CT of the heart and shows the early opacification of the left upper lobe pulmonary vein (LULPV) draining into the vena cava superior (VCS) via the anomalous vertical vein (VV) and the innominate vein (IV). B. Illustration of PAPVR of the left upper lobe.

has a characteristic scimitar appearance on chest radiographs as it runs from the middle of the right lung to the cardiophrenic angle (1).

The embryonic pathway of development of the pulmonary veins helps to understand the cause of the anomalous character of the pulmonary venous return. Initially, blood from both embryonic lungs drains into the splanchnic plexus, in turn communicating with a systemic cardinal vein (CV) on each side. Later the right CV becomes the VCS and the left CV involutes or persists as a left VCS or vertical vein. Primitive common pulmonary veins (PCPV) arise as outpouching structures at the dorsal wall of the left atrium, and with time communicate to the splanchnic plexus and CV. In normal situation, the venous connection between the PCPV and the CV involutes, giving rise to four separate pulmonary veins draining to the left atrium. In our case the venous connection between the PCPV and the CV of the left upper lobe failed, re-

sulting in a persistent vertical vein, which drains into the innominate vein (4).

Chest radiograph is often normal, however secondary signs of left-to-right shunt can be seen. These signs include cardiomegaly and pulmonary vascular prominence. Anomalous tip positioning of a left jugular/subclavian central venous catheter at the left mediastinum, should raise suspicion of PAPVR or persistent left-sided VCS. If the vertical vein is large, it can be depicted on X-ray as a small knob at the left side of the aortic arch ("aortic nipple").

CT angiography and 3D CT clearly shows the common upper pulmonary vein connecting to the anomalous vertical vein, which in turn drains into the innominate vein. In a well-performed CT angiography of the pulmonary arteries, the pulmonary veins of the upper lobe may show early and intense opacification, due to retrograde flow of the contrast. This will be more obvious when the contrast media is being

administered via the left arm or left jugular vein.

Echocardiography may also directly show the PAPVR, by identifying the vertical vein draining into the innominate vein. Since some authors believe that PAPVR becomes clinically significant when 50% or more of the pulmonary blood flow returns anomalously, velocity-encoded MRI can play an important role in measuring the ratio of pulmonary to systemic blood flow ($Q_p:Q_s$) (5).

Surgical or transcatheter endovascular treatment is indicated in symptomatic PAPVR and asymptomatic patients with $Q_p:Q_s > 1.5$, right ventricular dilation, mild-to-moderate tricuspid regurgitation (TR), or early stages of hypertensive pulmonary vascular disease. Treatment can prevent the development and progression of right ventricular failure and irreversible pulmonary vascular disease. Watchful waiting is recommended in asymptomatic patients without evidence of right ventricular dilation or TR (6).

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