Journal of Rare Cardiovascular Diseases 2013; 1 (2): 75–77
www.jrcd.eu

Partial anomalous pulmonary venous connection (RCD code: II-3C.0)

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Abstract

A 48-year-old woman was admitted to our institution with a year's history of uncharacteristic chest pain episodes and decrease in exercise tolerance. A CT lung scan performed nine months earlier revealed an anomalous vein arising from the right lower pulmonary lobe and draining into the inferior vena cava. A chest radiogram performed 2 months prior to the admission showed a curved shape adjacent to the right heart border. Right heart catheterization revealed: normal systolic, diastolic and mean pulmonary artery pressures, normal pulmonary resistance and insignificant shunt volume. Pulmonary artery angiography demonstrated an anastomotic vessel between the aberrant pulmonary vein and the right upper pulmonary vein. The patient was diagnosed with Partial Anomalous Pulmonary Venous Connection in form of Scimitar syndrome. Given a mild character of symptoms, no signs of the right-sided heart volume overload, insignificant shunt fraction and normal pulmonary circulation parameters, the patient was qualified to a conservative treatment with a close medical observation. JRCD 2013; 1 (2): 75–77

Key words: partial anomalous pulmonary venous connection, scimitar syndrome, left-to-right shunt, pulmonary hypertension

Literature review

Partial Anomalous Pulmonary Venous Connection (PAPVC) is a rare congenital heart defect where some of the pulmonary veins drain into the right atrium either directly or indirectly through its venous tributaries.

In contrast to the Total Anomalous Pulmonary Venous Connection, not all of the pulmonary veins are aberrantly connected. The exact incidence of PAPVC is unknown. In an autopsy series published in 1952 it was estimated to be 0.4% [1]. In a more recent retrospective analysis of a series of CT scans performed for other indications, the condition was found in 0.1–0.2% of examined adult patients [2,3]. PAPVC in a significant proportion of cases is associated with an atrial septal defect (ASD). Anomalously connected right-sided pulmonary veins can drain into the right atrium, the superior vena cava or, less frequently, into the inferior vena cava. The first two variations often accompany the sinus venosus type ASD. Possible connections for the left-sided aberrant pulmonary veins include: the left brachiocephalic vein, the coronary sinus and the hemiazygos vein. PAPVC causes a left-to-right shunt, which if severe enough can cause right ventricle overload and can lead to arterial pulmonary hypertension development. The symptoms depend on the shunt volume and the presence of other concurrent congenital anomalies. Possible clinical manifestations involve fatigue, exercise intolerance, frequent pulmonary infections. Another likely manifestation is palpitations resulting from atrial flutter or atrial fibrillation development, caused by long-standing right-sided heart volume overload. A single, isolated anomalous vein rarely produces any symptoms. A scimitar syndrome is a distinct form of PAPVC in which a pulmonary vein, usually arising from the lower and the middle lobe of the right lung, drain into the inferior vena cava. It has been reported in 3% to 6% of patients with partial anomalous venous connection. The syndrome is frequently associated with other congenital abnormalities like: sequestration of a lung lobe, lung hypoplasia, dextroposition of the heart, an abnormal arterial lung supply from the systemic arteries and an atrial septal defect [5]. The scimitar vein can often be visible on a plain chest X-ray at the right side of the heart silhouette, as a structure resembling in shape a Turkish sword used in the times of the Ottoman Empire – a scimitar.

Conflict of interest: none declared.

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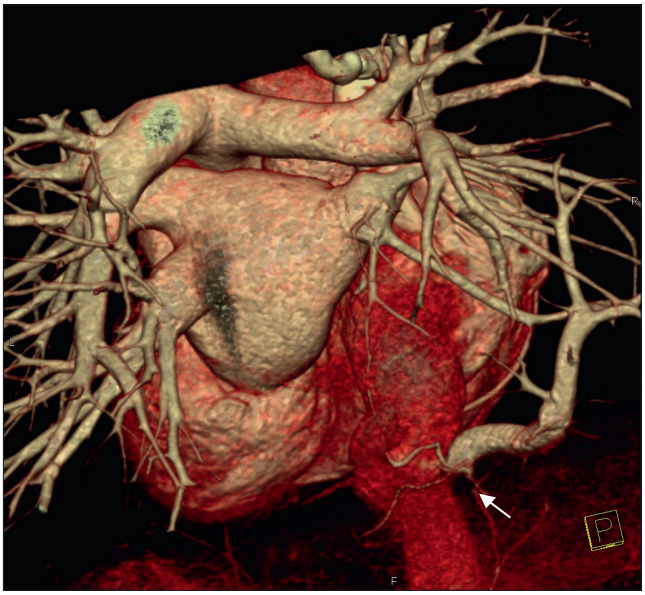

Figure 1. Cardiovascular computed tomography. Reconstruction of heart and great vessels. Anomalous pulmonary vein draining into the inferior vena cava (arrow)

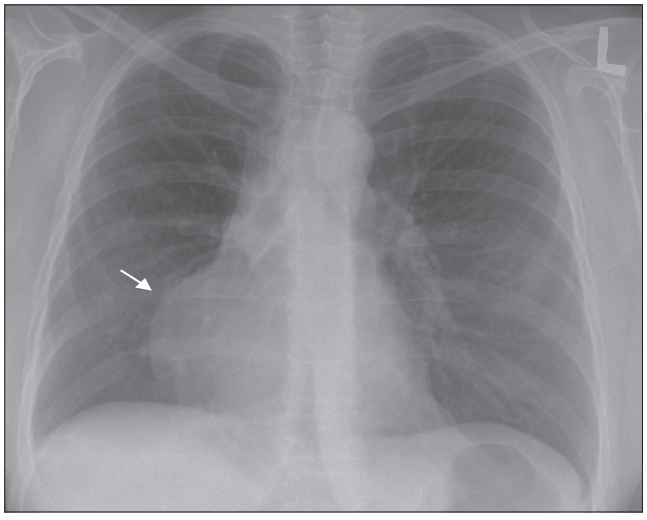

Figure 2. Chest X-ray. Posteroanterior view. Prominent right heart border (arrow)

Case report

We present a case of a 48-year-old woman, with a Partial Anomalous Pulmonary Venous Connection, admitted to our institution with a year's history of uncharacteristic chest pain episodes and decrease in exercise tolerance. A CT lung scan performed nine months earlier, as a part of an initial diagnostic work up of the aforementioned symptoms, revealed an anomalous vein arising from the right lower pulmonary lobe and draining into the inferior vena cava at the level of hepatic veins (Fig. 1). A chest radiogram performed 2 months prior to the admission, showed a curved shape in the lower right pulmonary field adjacent to the heart (Fig. 2).

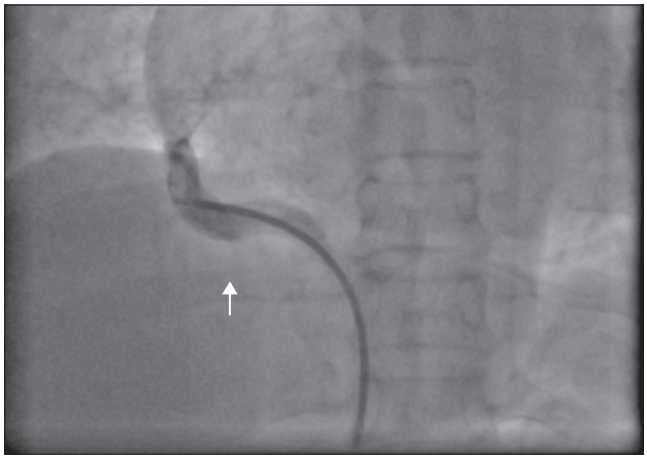

Figure 3. Angiography. Anomalous pulmonary vein (arrow)

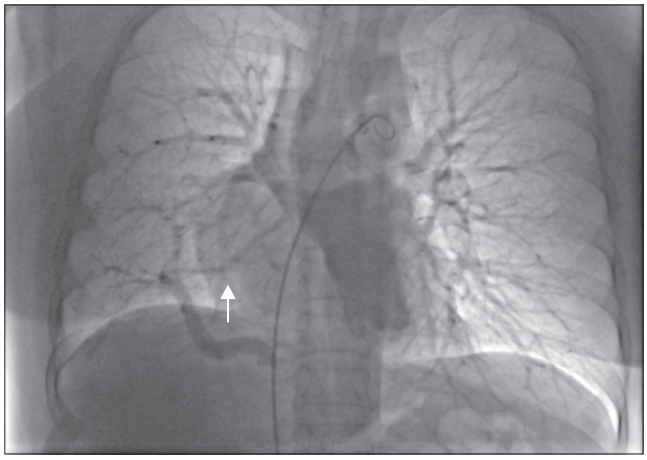

Figure 4. Pulmonary artery angiography. Anastomotic vessel (arrow) connecting an anomalous pulmonary vein with the right upper pulmonary vein

At the time of the admission the ECG showed the evidence of the right ventricular hypertrophy and overload.

A transthoracic echocardiogram did not confirm right ventricular dilation, however it did reveal an additional blood vessel with a turbulent flow draining into the inferior vena cava 2.5 cm from the right atrium. Both left ventricular function and size were normal and there were none significant valvular dysfunctions. Laboratory studies including complete blood count, electrolytes, liver and kidney function tests were within normal limits. Nevertheless, there were signs of hyperlipidemia. An exercise ECG test with ventilatory gas analysis demonstrated no signs of myocardial ischemia, and a normal peak oxygen uptake (Peak VO2: 21,0 ml/kg/min).

A transesophageal echocardiogram revealed no pathological connection between the atria, although it did demonstrate only three pulmonary vein ostia in the left atrium. Coronary angiography showed no evidence of coronary artery disease. Right heart catheterization was performed revealing: normal systolic, diastolic and mean pulmonary artery pressures (consecutively 19, 5, 10 mmHg) and normal pulmonary resistance (1.6 Wood Units). Selective angiography demonstrated an anomalous pulmonary vein draining

into the inferior vena cava (Fig. 3). A direct catheter measurement revealed a saturation of 98% in the anomalous vein.

The shunt volume however, was insignificant and the Qp:Qs ratio estimated with the use of the modified Fick equation was 1:1. A performed pulmonary artery angiography demonstrated that the aberrant pulmonary vein had an additional connection with the right upper pulmonary vein through an anastomotic vessel (Fig. 4). The patient was diagnosed with Partial Anomalous Pulmonary Venous Connection in form of Scimitar syndrome. No associated congenital anomalies were identified.

Discussion

Definitive treatment of PAPVC consists of a surgical redirection of an aberrant pulmonary venous drain into the left atrium [4]. There are no widely accepted indications for surgical repair. Currant European Society of Cardiology Guidelines for the management of grown-up congenital heart disease do not specifically address PAPVC. Indications for surgical repair are similar to those of ASD [5]. Management depends in great deal on the magnitude and the effects of the left-to-right shunt. Patients with a significant shunt resulting with the right ventricular volume overload and the right sided heart dilation are candidates for surgical repair. On the other hand, in many asymptomatic patients with a low shunt fraction, no signs of the right heart overload or pulmonary artery hypertension surgery can be avoided. However, given the fact that the age-related changes, mainly the fall in the left ventricle compliance, can increase the shunt magnitude later in life, these patients should undergo regular evaluation in a center specialized in the management of grown-up congenital heart diseases.

Given a mild character of symptoms, no echocardiographic signs of the right-sided heart volume overload, insignificant shunt fraction and normal pulmonary circulation parameters, the patient was qualified to a conservative treatment with a close medical observation in our outpatient department.

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