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CASE REPORT

Clinical and diagnostic features of partially anomalous pulmonary venous connection in an adult female patient: a case report and review of the literature

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Abstract A 40-year-old woman presented with dyspnoea, chest pain and fatigue. Her medical history was unremarkable. An early systolic ejection murmur was heard in the 3D left inter-costal space. Chest X-ray revealed normal cardiothoracic ratio with an anomalous vessel adjacent to the left pulmonary hilum. Echocardiography and exercise tolerance test were normal. Right heart catheterisation revealed normal pulmonary pressures with normal cardiac output. CT scan and MRI of the thorax were diagnostic for an aberrant pulmonary venous connection between the left lower lobe pulmonary vein and the left brachiocephalic vein without atrial septal defect. She was treated conservatively and remained well.

Keywords Congenital anomaly · Partially anomalous pulmonary venous connection · Conservative strategy

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is an uncommon congenital anomaly [1]. It is considered extremely uncommon in the absence of atrial septal defect (ASD) [2].

From an embryological view point, the lungs are derived from the foregut. The pulmonary and systemic venous circulations are in direct communication early in the development of the foetus. Persistence of the venous drainage of the lung into the systemic cardinal and umbilical vitelline systems apparently results in a variety of anomalies of pulmonary venous return [3–6]. The main physiological disturbance of PAPVC is similar to that of ASD.

The symptoms and complications of PAPVC depend on the shunt magnitude and number of pulmonary veins draining into the right side of the heart. Presenting symptoms can be dyspnoea and palpitations secondary to arrhythmias which are almost always supraventricular in origin; haemoptysis is a rare symptom reflecting either chest infection or the development of pulmonary vascular disease. Chest pain may be evidence of right heart ischaemia but does not occur in childhood. More commonly, chest pain may be a manifestation of recurrent bronchitis. Peripheral oedema can occur in adults with cardiac failure [1]. The purpose of this paper is to describe the clinical and diagnostic features of an adult case of isolated PAPVC.

Case report

A 40-year-old woman presented with dyspnoea, chest pain and fatigue. She had previously been diagnosed with gastric complaints and pregnancy-related temporary anaemia which responded well to medical substitution with iron preparation.

Her blood pressure was 120/80 mmHg, pulse regular at 60 beats/min with normal central venous pressure and an

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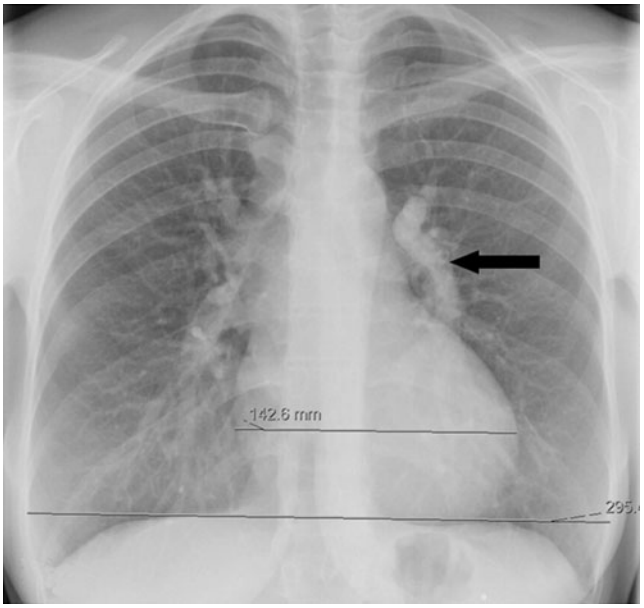

Fig. 1 Posteroanterior chest X-ray. *Arrow* indicates the abnormal left pulmonary venous connection

ejection systolic murmur which could be most clearly heard in the third left inter-costal space without clinical evidence of congestive heart failure. An electrocardiogram depicted sinus rhythm with a normal pattern. During maximal exercise tolerance testing, the patient reached 97% of expected heart rate without signs of coronary insufficiency. The reason for termination was dyspnoea and fatigue. Transoesophageal echocardiography demonstrated normal-sized and normokinetic biventricular function, intact interatrial septum and normal biventricular wall thickness. There was a mild tricuspid regurgitation with estimated right ventricular systolic pressure of 28 mmHg.

Chest X-ray demonstrated an abnormal vessel adjacent to the lower pole of the left hilum without pulmonary venous congestion (Fig. 1). Left-to-right shunt was measured by the nuclear method and showed a shunt of at least of >50%.

Results of right cardiac catheterisation demonstrated (in millimetres of mercury) pulmonary artery pressure of 20:15:18 systolic/diastolic/mean without tall V waves, right ventricle of 33:5 and mean right atrium pressure of 10. Chest CT scan showed an aberrant pulmonary vein (Fig. 2a–c) draining into the left brachiocephalic vein and small liver haemangioma. Cardiovascular magnetic resonance imaging was performed for detection of anatomy and physiology of the anomaly. This depicted a vessel running to the left of the aortic arch giving a left-to-right shunt of 1:1.6. The patient was treated conservatively. She remained well after a follow-up period for more than 4 years.

Discussion

Persistence of the venous drainage of the lung into the systemic cardinal and umbilical vitelline systems apparently results in a variety of anomalies of pulmonary venous return [3–6]. The main physiological disturbance of PAPVC resembles that of ASD. This is in the form of increased pulmonary blood flow as a consequence of recirculation of oxygenated blood through the lungs. PAPVC has high physiological significance when the atrial septum is intact because only one lung is draining into the left atrium and a large left–right shunt is present leading to high pulmonary pressure and development of pulmonary hypertension [7].

Partial anomalous pulmonary venous connection is defined as some, but not all, of pulmonary veins are aberrantly connected [8]. The term connection refers to anatomical relation while drainage refers to physiological effect [9].

Drainage of the right upper lobe pulmonary vein directly into the superior vena cava (SVC) is the most common type of PAPVC. Our patient presented with left lower lobe pulmonary vein drainage to the brachiocephalic vein.

The most frequent presenting signs and symptoms were a heart murmur and dyspnoea on exertion [10]. The ECG may be normal but it may demonstrate signs of right ventricular dilatation or hypertrophy. In our case, ECG

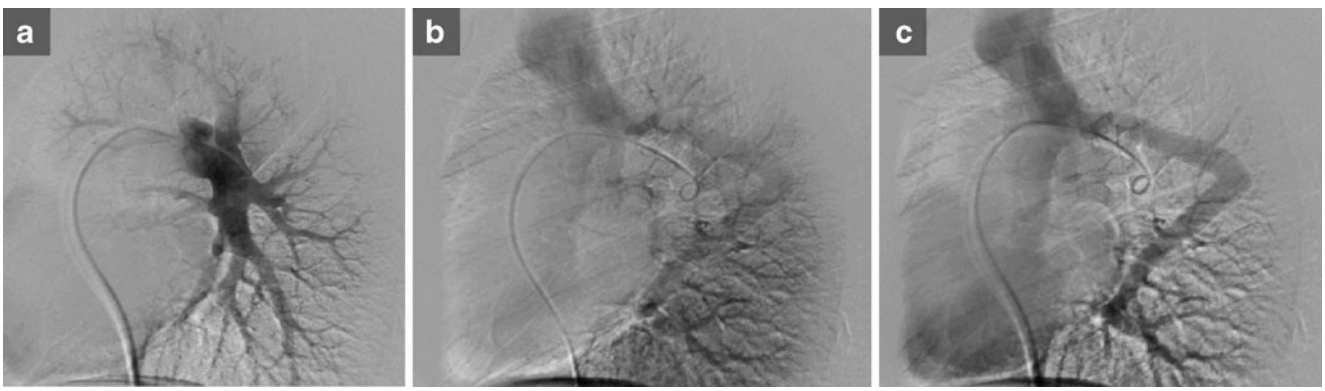

Fig. 2 CT scan thorax: demonstrating **a** arterial, **b** capillary and **c** venous phases showing the abnormal vein

depicted a normal pattern. The diagnosis of PAPVC may be suggested by echocardiography and further localised by CT scan, cardiac catheterisation, radionuclide studies and cardiovascular MRI.

It is estimated that one of every 140 autopsies had PAPVC and that most patients were asymptomatic [10]. In 1985, Weiman et al. described their good surgical experience with 24 patients with PAPVC; eight had left-sided drainage to the innominate vein. The goal of surgical treatment is to divert the pulmonary venous flow in the anomalous pulmonary vein to the left atrium in an unobstructed fashion.

The position of the anomalous pulmonary vein determines the site of cannulation in the SVC. Typically, a glutaraldehyde-treated pericardial patch can be used to create a baffle where pulmonary venous blood from an anomalous right upper pulmonary vein flows beneath the baffle and through an ASD into the left atrium.

Lobectomy is described as a treatment modality for selected cases of PAPVC, particularly Scimitar syndrome, in which there is aplasia of one or more lobes of the right lung. In Scimitar syndrome, the anomalous venous return drains into the inferior vena cava which means that correction of the venous flow is not possible.

In our case, the surgical team had advised lobectomy as a treatment modality. Considering the absence of hypoplasia of the affected lobe and the lack of significant clinical symptoms, we decided to choose conservative strategy with close regular follow-up.

Conclusion

PAPVC does not always indicate surgical intervention, particularly in the absence of ASD and pulmonary hypoplasia. In the current case, the symptoms were well tolerated by the patient, and considering the morbidity associated with lobectomy, we chose a conservative strategy.

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