Unilateral pulmonary vein stenosis

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SUMMARY Unilateral pulmonary vein stenosis is a rare congenital anomaly. A case is described in a girl who first presented at the age of four years with recurring haemoptysis but in whom diagnosis was not established until she was 16 years old. Pulmonary angiography demonstrated a minimally hypoplastic right pulmonary artery, and the laevophase showed normal pulmonary venous return from the left lung, but none from the right.

Surgical treatment was necessary because of life threatening haemoptysis, and pneumonectomy was required in the light of the findings at operation.

Congenital causes of pulmonary venous obstruction have been reviewed elsewhere, and the rarity of pulmonary vein stenosis, whether bilateral or unilateral, has been emphasised.¹⁻⁴ Previous cases have been predominantly reported in infants. We report on a patient who was first seen at the age of four years and in whom diagnosis and successful treatment were not made until the age of 16 years. There may be difficulties in diagnosis, but the changes of unilateral pulmonary venous hypertension on the chest x ray are a useful indicator. Cardiac catheterisation and angiography are necessary, however, to establish the diagnosis.

Case report

A female patient was found to have a cardiac murmur at the age of four years. She had suffered from recurring chest infections in early childhood, and there had been several episodes of minor haemoptysis. The chest x ray showed a normal cardiac contour but increased vascular markings on the right with Kerley B lines at the right base were suggestive of unilateral venous obstruction. At cardiac catheterisation no intracardiac shunt was demonstrated but there was pulmonary hypertension (60/30 mmHg in the main pulmonary artery). Mitral valve disease was suspected at this point, but operation was not indicated. There were two further episodes of haemoptysis several weeks later, but repeat chest x ray and catheterisation were unchanged.

When the patient was 16 she was intensively reinvestigated because of a further major haemoptysis with a fall in haemoglobin concentration to 8-8 g/dl which required blood transfusion. The chest x ray was unchanged. Bronchoscopy immediately after a further minor haemoptysis showed a small blood clot in the right main bronchus. Bronchography showed a normal right bronchial tree. Ventilation of both lungs appeared equal and normal on isotope ventilation perfusion lung scan, but perfusion was absent on the right (Fig. 1). The electrocardiogram was normal as were both M mode and cross sectional echocardiography. The Table shows the relevant findings at repeat cardiac catheterisation. The most important findings were the raised mean pulmonary capillary wedge pressure on the right and the oxygen rise in the right pulmonary artery.

At pulmonary artery angiography contrast delineated only the proximal part of the right pulmonary artery before being redirected retrogradely into the left pulmonary artery. Selective angiography in the right pulmonary artery demonstrated a slightly hypoplastic but normally distributed arterial tree. The laevophase of the pulmonary arteriogram showed normal pulmonary venous drainage from the right lung (Fig. 2). After injection of contrast into the ascending aorta there was late filling of the pulmonary artery indicating a left to right shunt. Selective bronchial arteriography demonstrated two dilated tortuous bronchial arteries which communicated with the terminal branches of the right pulmonary artery through which contrast passed retrogradely to the left pulmonary artery.

The diagnosis was still in doubt, and haemangiomatous malformation of the right pulmonary artery branches or stenosis of the right pulmonary veins were considered as possible diagnoses. Tho
Pulmonary veins outlined right atrium with no pulmonary veins outlined on the right lung.

Fig. 1  Isotope ventilation perfusion scan images. The image on the left shows normal ventilation of each lung. The image on the right shows evenly distributed perfusion of the left lung but none of the right lung.

Table  Haemodynamic data

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mm Hg)</th>
<th>Oxygen saturation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrium (mean)</td>
<td>7/5</td>
<td>72</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>40/5</td>
<td>69</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>40/24</td>
<td>69</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>40/24</td>
<td>77</td>
</tr>
<tr>
<td>Right pulmonary capillary wedge (mean)</td>
<td>24</td>
<td>94</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>96/4</td>
<td>96</td>
</tr>
<tr>
<td>Aorta</td>
<td>96/60</td>
<td>96</td>
</tr>
</tbody>
</table>

Fig. 2  Laevophase of pulmonary angiogram demonstrating pulmonary veins draining from the left lung into the left atrium but a smooth right border of the left atrium with no pulmonary veins outlined on the right.

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Racotemy was performed with the intention of ligating the aberrant collateral vessels. However, the right lung was found to be grossly abnormal with a thickened pleura and diffuse bullous disease. A mass of abnormal collateral systemic vessels entered the lung both in the hilum and across the pleural space from the mediastinum. We therefore decided to remove the entire lung; this was accomplished with some difficulty. The right pulmonary artery was 1 cm to 1.5 cm in diameter and was thickened. Two right pulmonary veins entering the left atrium were identified, but these were reduced in size and had thick dysplastic walls and a grossly reduced luminal calibre. The right pulmonary artery and its branches showed thinning of the vessel walls with loss of elastic tissue and some areas of intimal fibrous proliferation. The bronchi were normal. There was mediastinal thickening and prominent elastic laminae in the bronchial arteries.

The patient's recovery from operation was uncomplicated and she remains well with no further haemoptysis.

Discussion

Pulmonary venous obstruction can be congenital or acquired. Pulmonary veno-occlusive disease is a rare condition caused by sclerosis or thrombosis and is perhaps the best known acquired cause of the condition, but recurring respiratory infections or more rarely myxoma of left atrium or external compression of the pulmonary veins by neoplasm or mediastinal fibrosis are additional causes. The obstruction is most commonly bilateral in such cases. Congenital pulmonary venous obstruction, however, may be either bilateral or unilateral or may simply affect a single pulmonary vein. Obstruction of the common vein in early development may result in total anomalous pulmonary venous drainage. At a later stage bilateral pulmonary vein stenosis or atresia may occur, presenting with features of bilateral pulmonary venous obstruction. In unilateral pulmonary vein stenosis, however, the situation is modified by the presence of a normal lung through which blood can return to the left atrium. As a result the bronchial arteries on the affected side become enlarged and blood passes through bronchial-pulmonary vascular anastomosis into the ipsilateral pulmonary artery and then retrogradely into the contralateral pulmonary artery. This backward flow may at times be insufficient to drain all the blood from the affected lung, and this leads to pulmonary congestion, recurring respiratory infections, and haemoptysis.
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A comprehensive review of the published reports of congenital pulmonary venous obstruction has yielded 38 patients to date, of whom 20 were affected bilaterally and 18 either unilaterally or in whom only a single vein was affected. Twenty five patients had accompanying congenital cardiac defects, but in 13 pulmonary vein stenosis was the only anomaly. The lethal nature of the condition is highlighted by the fact that 24 of the 38 patients died in infancy or early childhood.

Most of the 38 previously reported cases had important pulmonary arterial hypertension, the cause of which may have been multifactorial since 25 had accompanying congenital cardiac anomalies. The effects of pulmonary venous hypertension resulting from obstruction may be offset by an increase in pulmonary lymphatic flow, the development of collateral communications with bronchial venous system, alteration of the pulmonary capillary permeability, or decrease in pulmonary blood flow to areas with venous obstruction. In our patient pulmonary artery pressure was considerably raised in early childhood (60/30 mm Hg) at the first catheterisation. At repeat catheterisation, however, at the age of 16 years, there was only slight pulmonary hypertension (40/20 mm Hg). This is difficult to explain in the context of the four mechanisms referred to above, and in particular there was no evidence of fresh bronchial venous drainage having developed on the ipsilateral lung. All the blood entering the right lung did in fact drain retrogradely to the contralateral lung. It is conceivable that in our patient the progression of pulmonary hypertension was prevented by the opening up of the pulmonary capillary bed on the contralateral lung which helped in off-loading blood from the right lung.

Twenty four of the 38 patients presented in infancy with congestive cardiac failure; some of them also had haemoptyisis. Most of the remaining 14 (aged 2–15) had either dyspnoea or haemoptyisis, but in a few who were symptom free the chance finding of a cardiac murmur led to investigation and diagnosis of the anomaly. A cardiac murmur was first heard in our patient when she was four years old, although there had been previous chest infections and several minor haemoptyisis. Other possible causes of hae- moptyisis such as mitral valve disease, pulmonary thromboembolism, and pulmonary arteriovenous fistula were excluded by appropriate investigation.

The diagnosis was established rather late, after careful study of the data obtained at the third catheterisation, which were supplemented by the findings at operation. Unilateral pulmonary venous obstruction had been observed in the initial chest x ray when she was four years old, but its relevance was not appreciated at that time. The diagnostic features at the final catheterisation were the rise in oxygen saturation in the right pulmonary artery, a raised pulmonary capillary wedge pressure on the ipsilateral side, and the laevophase of the pulmonary artery angiogram which showed that contrast entered the left atrium from the left pulmonary veins but not from the right. Important features diagnostic of pulmonary venous obstruction are the finding of pulmonary hypertension, a raised pulmonary capillary wedge pressure, and normal left atrial pressure. A pulmonary capillary wedge injection may visualise an obstructed pulmonary vein, or the stenosis may be demonstrated by a selective pulmonary vein injection.

If the local anatomy is favourable operation is the treatment of choice. Transvenous balloon dilatation appears attractive, particularly when the stenosis is localised, but in practice the results have been disappointing. When the stenosis is of the diaphragmatic type excision of the obstructing membrane may be possible, where necessary combined with patch angioplasty. Direct anastomosis of the vein to the left atrium may be possible if the stenosis is more extensive or the vein is atretic. Whether any of these procedures would have been feasible in our patient at an earlier date is debatable given the findings at operation. At thoracotomy the widespread nature of the parenchymal damage in the right lung together with the pulmonary venous drainage which ruled out any reconstructive surgery left no option other than pneumonectomy. This we feel was fully justified because of the recent history of life-threatening haemoptyisis.

References


