Isolated pulmonary vein stenosis in complex congenital heart disease, simulating cor triatriatum by cardiac catheterization and transoesophageal echocardiography

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A case of localized stenosis of the lower left pulmonary vein, associated with perimembranous ventricular septal defect (VSD) and secundum atrial defect (ASD), a borderline sized mitral valve apparatus and left ventricle, is described. This final diagnosis was made at surgery, as the localized stenosis of the left pulmonary vein was misinterpreted as partial cor triatriatum by cardiac catheterization, cineangiocardiography and extensive echocardiographic studies. Surgical correction of all the defects was successful, but pulmonary vascular resistance was still increased 2 years after surgery.

Introduction

Stenosis of one or more of the pulmonary veins in the absence of an anomalous pulmonary venous connection is rare. Localized stenosis of a single pulmonary vein may be an isolated phenomenon or may be associated with minor or major cardiac abnormalities1-3. One or more of the pulmonary veins may stenose at the junction with the left atrium. The length of the stenosis varies and may extend for a considerable distance in extra- and intra-pulmonary portions. Most patients present in the first years of life with respiratory symptoms, ultimately progressing to right heart failure. However, clinical presentation may be determined by the associated lesions. Differential diagnosis in patients with clinical findings of pulmonary hypertension include left heart obstructive lesions and idiopathic pulmonary hypertension. Cardiac catheterization and cineangiocardiography are usually diagnostic, although the clues may be subtle.

Case report

The patient presented at the age of 8 weeks with congestive heart failure with severe right ventricular hypertrophy on the electrocardiogram. Chest X-ray showed a very large heart with increased vascular markings. Because of the enlarged heart, no difference could be seen between left- and right-sided vascular markings. The diagnosis of a large secundum ASD and a perimembranous VSD accompanied by pulmonary hypertension was made after a complete precordial echocardiographic study. Furthermore, in the left atrium on the left lateral side, a membrane-like structure was seen with a high velocity turbulence jet through its centre. On measuring the dimensions of the aorta, left ventricle and mitral annulus their valves were shown to be on the 5th percentile. Cardiac catheterization confirmed the diagnosis.

The haemodynamic data are summarized in Table 1. Withdrawal pressure curves from the lower left pulmonary vein towards the left atrium showed a pressure drop of 4 mmHg at the level of the membrane. The membrane-like structure, as visualized by echocardiography, was also seen on cineangiocardiography. To define the left atrial anatomy, transoesophageal echocardiography was performed. This showed normal drainage of the right pulmonary veins into the left atrium. Both left pulmonary veins seemed to drain into an accessory chamber with obstructed outflow to the left atrium. Surgery was planned following a diagnosis of partial cor triatriatum, perimembranous VSD and secundum ASD.

Table 1 Cardiac catheterization data

<table>
<thead>
<tr>
<th></th>
<th>Preop.</th>
<th>Postop.</th>
<th>Oxygen saturations (%)</th>
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</thead>
<tbody>
<tr>
<td><strong>RV</strong></td>
<td>72/10-9</td>
<td>36/5-0</td>
<td>85</td>
</tr>
<tr>
<td><strong>LV</strong></td>
<td>66/1-4</td>
<td>100/0-8</td>
<td>95</td>
</tr>
<tr>
<td><strong>PA</strong></td>
<td>50/23</td>
<td>38/18</td>
<td>96</td>
</tr>
<tr>
<td><strong>AO</strong></td>
<td>76/48</td>
<td>70/50</td>
<td>95</td>
</tr>
<tr>
<td><strong>PCW</strong></td>
<td>12</td>
<td>12</td>
<td>—</td>
</tr>
<tr>
<td><strong>Q/Q</strong></td>
<td>5:1</td>
<td>1:1</td>
<td>—</td>
</tr>
<tr>
<td><strong>SVR</strong></td>
<td>6.5 U. m²</td>
<td>17.5 U. m²</td>
<td>12</td>
</tr>
<tr>
<td><strong>PVR</strong></td>
<td>6.3 U. m³</td>
<td>3.8 U. m³</td>
<td>12</td>
</tr>
</tbody>
</table>

Preop. = pre-operatively; postop. = postoperatively; RV = right ventricle; LV = left ventricle; PA = pulmonary artery; AO = aorta; PCW = pulmonary capillary wedge; SVR = systemic vascular resistance; PVR = pulmonary vascular resistance.
Surgery

At operation, the left atrium was inspected through the ASD. The orifices of both the right and the upper left pulmonary veins were normal. The left atrial appendage was inverted. On the cranial side of the left upper pulmonary vein a ridge was seen, which appeared to be a fold in the atrial wall. The lower left pulmonary vein drained normally into the left atrium, but was obstructed in the ostium by a ring of fibrous tissue. The left atrial appendage was ligated at its base. The ridge in the atrial wall was resected and the ostium of the lower left pulmonary vein dilated by removing the fibrous tissue. Inspection of the mitral valve showed a normal, although small, mitral annulus with normal valve leaflets. The perimembranous VSD with a diameter of approximately 6 mm was closed through the tricuspid valve with a Gore-Tex patch. The secundum ASD was primarily closed.

The postoperative course was prolonged by pulmonary hypertensive crises with right heart failure, for which oxygen, diuretics, cardiotoxic and vasodilative drugs were administered. Over a period of time the baby's condition improved; he started to grow and developed well. At the age of 2 years a cardiac catheterization was performed. No residual shunt was present, but a peak systolic pressure gradient of 30 mmHg across the aortic valve was found. (For the haemodynamic data see Table 1). Right and left ventricular angiography showed fast transit of the blood through the lungs without evidence of obstruction. The aortic valve was abnormal, but contractility of both the right and left ventricles was normal.

Discussion

Congenital stenosis of a pulmonary vein is a rare malformation that may be difficult to assess. Since pulmonary vein stenosis has been associated with progressive pulmonary hypertension and pulmonary vascular obstructive disease, the recognition of this malformation is of crucial clinical relevance. Unilateral banding of a pulmonary vein in rats has been reported to produce pulmonary vascular obstructive disease that mimicked the key aspect of human disorder. There is still controversy whether the morphological changes due to pulmonary venous obstruction are reversible. In our patient the pulmonary hypertension and the increased resistance were initially explained by the presence of a non-restrictive VSD. The presence of an obstructive membrane-like structure in the left atrium, giving rise to unilateral pulmonary venous obstruction, was also regarded as an extra clue for the presence of pulmonary hypertension. However, at surgery the membrane, as such was not present. Cardiac catheterization, 2 years after surgery excluded pulmonary vein stenosis.

In the presence of an unrestrictive VSD, the pre-operative elevation of the pulmonary vascular resistance cannot be related to the isolated pulmonary vein stenosis only. However, the potential effects of a relatively long-standing isolated pulmonary vein obstruction could be an additional explanation for this feature. This observation is supported by others. Fortunately, in our patient re-catheterization revealed a decrease of pulmonary vascular resistance to lower levels although they were still above normal.

Although transoesophageal echocardiography has been reported to give more accurate information about the pulmonary venous anatomy and flow patterns, the disturbed high velocity jet proved to be due to a stenosis of a single pulmonary vein and not to a stenosis at the side of a suspected membrane. The inverted left atrial appendage and the ridge in the left atrial wall mimicked the image of a membrane, which emphasises the imperfection of both diagnostic methods. Management of pulmonary vein stenosis can be difficult. Both balloon dilation and surgical treatment have been discussed. Balloon dilation proved to be unsuccessful in nearly all cases, while surgical management is successful, especially when a localized stenosis is present. However, in all reports the unexpected finding of pulmonary vein stenosis in patients dying from pulmonary hypertension with right heart failure has been stressed. Theoretically, the final diagnosis could have been confirmed if selective contrast angiography into the lower left pulmonary artery and a lung scan had been performed. As the diagnosis was not suspected, this was not done. The presence of small left heart compartments before surgery probably reflects the low flow through the left heart due to the pathological situation. Both echocardiography and cineangiocardiography demonstrated that the left heart dimensions had also increased with time.

We conclude that single pulmonary vein stenosis, whether associated or not with other cardiac lesions, may be difficult to assess and may potentially give rise to pulmonary obstructive disease. In cases of associated cardiac lesions, single pulmonary vein obstruction must be eliminated, to avoid persisting pulmonary hypertension after management of the cardiac lesion itself.

References