Anomalous Unilateral Single Pulmonary Vein Mimicking Pulmonary Varices

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Anomalous unilateral single pulmonary vein (AUSPV) is a rare anomaly of the pulmonary venous system. Six cases have been documented in the literature, and two additional are reported here. Since AUSPV mimics pulmonary varices on pulmonary angiography, careful interpretation of the pulmonary venous anatomy by bi-directional and stereoscopic angiography is mandatory for diagnosing this anomaly.

A nomalous unilateral single pulmonary vein (AUSPV) is a rare anomaly of the pulmonary venous system. Etiologically, it has recently been recognized as an entity distinct from pulmonary varices.1,2 Radiologically, AUSPV may mimic abnormalities such as pulmonary varices and parenchymal nodules. Although AUSPV is regarded as innocuous,3 it must be differentiated from other abnormalities. The AUSPV was first described by Benfield et al4 in 1971. Five additional cases have since been documented.1,4,5 This report describes two more cases of AUSPV mimicking pulmonary varices. The angiographic findings and differential diagnosis are discussed.

CASE REPORTS

CASE 1

This 22-year-old asymptomatic man was referred to Kyushu University Hospital with an abnormal mass detected during chest roentgenography. Chest roentgenography and tomography showed a large vascular structure in his right lower lung field which extended towards the right hilum. Pulmonary arteriography revealed normal right pulmonary arteries. In the venous phase, a large tortuous pulmonary vein drained the right lower lobe, and coursed anteriorly, with a loop configuration. It joined the right superior pulmonary vein, which emptied into the left atrium (Fig 1). The right lower pulmonary vein was absent. In the anteroposterior projection, this anomalous vein mimicked pulmonary varices. However, in the lateral projection (Fig 2) and stereoscopically, there was no evidence of pulmonary varices, such as localized dilatation of the pulmonary vein.

CASE 2

This 57-year-old asymptomatic man had an abnormal mass on chest roentgenography. Bronchography revealed bronchiectasis of the right middle lobe; pulmonary arteriography, normal right pulmonary arteries. In the venous phase, the right pulmonary veins and their tributaries were tortuous, especially near the hilum. It was difficult to distinguish these from pulmonary varices (Fig 3). With pulmonary angiography in the lateral projection (Fig 4), and stereoscopically, there was no localized dilatation of the pulmonary veins. The veins from the posterior basal segment drained into the inferior vena cava.

DISCUSSION

The AUSPV is extremely rare and is one of several congenital anomalies occurring in the pulmonary venous system. It is usually accompanied by tortuous draining veins; one of the six incidences previously reported was diagnosed as pulmonary varices.6 That case, we believe, should be categorized as AUSPV, since it had no localized dilatation of the vein — only venous tortuosity. Embryologically, AUSPV is considered atresia or hypoplasia of one of the major pulmonary veins, with drainage of the ipsilateral pulmonary venous flow via a single pulmonary vein.1 It is believed that pulmonary varices are caused by developmental aberrations during the primitive splanchnic capillary stage.6

Table 1 summarizes the six cases which have so far been reported, and our two cases as well. The age distribution ranged from 21 to 67, with an average of 45 years. None of the cases were symptomatic. The right lungs of five and the left lungs of three patients were involved. Abnormalities associated with AUSPV con-
FIGURE 2. Case 1. Pulmonary angiogram, lateral projection, venous phase. Anomalous course of pulmonary veins are clearly demonstrated without localized dilatation.

FIGURE 3. Case 2. Pulmonary angiogram, anteroposterior projection, venous phase. Markedly tortuous veins drain into left atrium via a single pulmonary vein, and vein of posterior basal segment empties into inferior vena cava. Residual contrast material of previous bronchography is seen.

FIGURE 4. Case 2. Pulmonary angiogram, lateral projection, venous phase. Extremelty tortuous courses of pulmonary veins are seen, draining into left atrium via a single pulmonary vein. Partial anomalous pulmonary venous drainage of posterior basal vein into inferior vena cava is also noted.

Associated partial anomalous pulmonary return (PAPVR) in our case 2 has thus far not been described in the literature. The associated PAPVR indicates that this is not AUSPV in the strict sense. However, we believe that it should be categorized as AUSPV with PAPVR because most of the pulmonary venous flow from the right lung drained into the left atrium via a single pulmonary vein.

Tortuous draining veins accompanying AUSPV appear as single or multiple masses, usually perihilar, on chest roentgenography. Tomography may reveal them to be vascular.

Angiographically, these tortuous veins mimic pulmonary varices and their differentiation from varices may be difficult in such cases. Bartram and Strickland7 proposed the following angiographic criteria for the diagnosis of pulmonary varices: (1) normal pulmonary arterial trees; (2) opacification of the varices at the same rate as the pulmonary veins; (3) direct drainage of the varices into the left atrium; (4) delayed emptying of the varices compared with other pulmonary veins; and (5) varicose appearance and tortuosity only of the proximal vein with normal ramifications. Localized dilatation of a pulmonary vein normally entering the left atrium is essential for the diagnosis of pulmonary varices.

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ANOMALOUS UNILATERAL SINGLE PULMONARY VEIN
Table 1—Anomalous Unilateral Single Pulmonary Vein (AUSPV): Reported Cases

<table>
<thead>
<tr>
<th>Investigator</th>
<th>Year/Age/Sex</th>
<th>Side</th>
<th>Absent Pulmonary Vein</th>
<th>Reported Diagnosis</th>
<th>Associated Conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benfield et al⁴</td>
<td>1971/67/F</td>
<td>Left</td>
<td>SPV</td>
<td>AUSPV</td>
<td>Unremarkable</td>
</tr>
<tr>
<td>Tretewjay et al⁵</td>
<td>1974/38/M</td>
<td>Left</td>
<td>IPV</td>
<td>AUSPV</td>
<td>None</td>
</tr>
<tr>
<td>Ben-Menachem et al⁶</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Case 1</td>
<td>1975/50/F Right</td>
<td>IPV</td>
<td>AUSPV</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>Case 2</td>
<td>1975/53/F Right</td>
<td>IPV</td>
<td>AUSPV</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>Twersky et al⁷</td>
<td>1976/51/M Left</td>
<td>SPV</td>
<td>AUSPV</td>
<td>Tortuous right SPV</td>
<td></td>
</tr>
<tr>
<td>Moro et al⁸</td>
<td>1978/21/F Right</td>
<td>SPV “Varix”</td>
<td>Hypoplasia of right pulmonary artery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Present Authors</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Case 1</td>
<td>1980/22/M Right</td>
<td>IPV</td>
<td>AUSPV</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>Case 2</td>
<td>1980/57/M Right</td>
<td>IPV</td>
<td>AUSPV</td>
<td>PAPVR Bronchiectasis</td>
<td></td>
</tr>
</tbody>
</table>

*SPV—Superior pulmonary vein; IPV, inferior pulmonary vein; and PAPVR, partial anomalous pulmonary venous return.

Determining whether the tortuous veins are due to pulmonary varices or draining veins of AUSPV is sometimes difficult, but confusion may be avoided by using bidirectional or stereoscopic angiography. In AUSPV, one of two major pulmonary veins is absent, and there are tortuous draining veins without localized dilatation. Careful angiographic interpretation of the pulmonary venous anatomy is essential for diagnosing this anomaly. Three deaths have been reported among varices patients, due to rupture of pulmonary varices⁹¹⁰ and complications such as systemic embolization.¹¹ It is therefore important to determine whether the tortuous veins are pulmonary varices or AUSPV.

In symptomatic patients, pulmonary varices are related to pulmonary venous hypertension.¹² Pulmonary varices may progressively enlarge as the pulmonary venous pressure increases due to deterioration of mitral insufficiency¹³ or the malfunction of a mitral valve prosthesis.² Pulmonary varices in otherwise normal persons are not thought to exhibit such changes in size. The varices of two patients without pulmonary venous hypertension who have been followed for 4 and 15 years have showed no changes in size.¹² Another patient with AUSPV has been followed for 24 years, without change.¹ The natural history of AUSPV is thought to be the same as that of pulmonary varices. We regard AUSPV to be innocuous if there are no associated abnormalities, such as pulmonary venous hypertension.

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REFERENCES