Abstract

A 71-year-old woman was admitted for paralysis on the left side of her body. She developed dyspnea and hypoxemia after admission. Although pulmonary embolism was suspected, hypoxemia and dyspnea occurred repeatedly in spite of anticoagulation therapy. Transesophageal echocardiography revealed a patent foramen ovale (PFO), an atrial septal aneurysm (ASA), and a right-to-left shunt that appeared in an upright position. She was diagnosed with platypnea-orthodeoxia syndrome. Moreover, cardiac catheterization showed congenital anomalies, such as unroofed coronary sinus, partial anomalous pulmonary venous return and persistent left superior vena cava. Simple surgical closure of the ASA and PFO improved all of her symptoms. (Internal Medicine 44: 453–457, 2005)

Key words: atrial septal aneurysm, patent foramen ovale, unroofed coronary sinus, intracardiac shunts, platypnea, transesophageal echocardiography

Introduction

Platypnea is defined by dyspnea induced by an upright position and relieved by recumbency. Orthodeoxia is defined as arterial deoxygenation increased by an upright position and improved by recumbency (1, 2). Platypnea-orthodeoxia (P-O) syndrome is a rare clinical disorder associated with conditions such as pneumonectomy, pulmonary emphysema and liver cirrhosis. Recently, interatrial right-to-left shunting through an atrial septal defect (ASD) or a patent foramen ovale (PFO) was reported to be a common cause of this syndrome (3–6). We present a case of P-O syndrome caused by a PFO.

Case Report

A 71-year-old woman was referred to our hospital for progression of paralysis on the left side of her body. She had a history of hypertension and cerebrovascular disease (cerebral hemorrhage and cerebral infarction). Physical examination revealed no evidence of pedal edema, clubbed finger, cyanosis, or jugular venous distention. Her blood pressure was 150/90 mmHg and pulse rate was 90 beats/min. There was a small systolic ejection murmur on cardiac auscultation. The electrocardiogram showed sinus rhythm, a tall R wave in lead V1, and Q waves in leads III and aVF. Magnetic resonance imaging (MRI) of the brain excluded new lesions due to cerebral infarction.

She did not complain of dyspnea while supine. However, when she was in an upright position, she experienced dyspnea with cyanosis and severe hypoxemia. She did not have any pulmonary diseases that would cause dyspnea and the chest radiograph did not show atelectasis or pleural effusion.

Transthoracic echocardiography showed left ventricle hypertrophy with normal systolic function. The short axis view did not show dilatation of the right ventricle. In addition, color Doppler did not reveal evidence of pulmonary hypertension.

Pulmonary perfusion scintigraphy with 99mTc-macroaggregated albumin and radionuclide venography revealed a small perfusion defect in the left upper lobe. Deep vein thrombosis of the lower limbs was detected through venography. Immediately, we started anticoagulation therapy with heparin and warfarin.

Thoracic MRI and computed tomography revealed elongation of the ascending aorta and compression of the atrial septum, but the relationship between the positional change in
the atrium and alternate shunting was still unclear because these tests could be performed only while the patient was recumbent.

After anticoagulation therapy, pulmonary scintigraphy revealed improvement of the perfusion defect. In the supine position, the arterial pO₂ was 68.8 mmHg and the oxygen saturation was 95.8% in room air. Despite the success of anticoagulation therapy, she still complained of dyspnea with cyanosis; the arterial pO₂ dropped to 40.8 mmHg and the oxygen saturation to 83% in the sitting position.

Transesophageal echocardiography (TEE) revealed a PFO and an atrial septal aneurysm (ASA). ASD and the Eustachian valve were not observed. The R-L shunt through the PFO was not seen when the patient was supine. However, in the upright position, the R-L shunt was visualized by color flow Doppler imaging and bubble contrast studies (Fig. 1), and the arterial oxygen saturation decreased from 96% to 80%. She was diagnosed with P-O syndrome caused by the PFO and ASA.

Cardiac catheterization showed normal pulmonary artery pressures and normal right atrial pressure (Table 1). A Swan-Ganz catheter was easily passed through the PFO into the left atrium. Left atrial pressure was slightly higher than right atrial pressure. The selective right atriotomy showed floppy-mobile ASA (Fig. 2A).

The right and left lower pulmonary veins were intact and they returned oxygen-rich blood to the left atrium. However, the left upper pulmonary vein was hypoplastic, and there was a small persistent left superior vena cava (PLSVC). The left innominate vein was present but there was no connection

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Table 1. (a). Cardiac Catheterization (Room Air, Supine Position)

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<thead>
<tr>
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<tbody>
<tr>
<td>RA</td>
<td>10/9/7 mmHg</td>
<td>Ao</td>
<td>150/94/113 mmHg</td>
</tr>
<tr>
<td>RV</td>
<td>26/4 mmHg</td>
<td>LV</td>
<td>150/19 mmHg</td>
</tr>
<tr>
<td>PA</td>
<td>29/9/18 mmHg</td>
<td>PVRI</td>
<td>224 dynes ⋅ s ⋅ cm⁻² ⋅ m²</td>
</tr>
<tr>
<td>PCWP</td>
<td>77/5 mmHg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>LA</td>
<td>18/12/9 mmHg</td>
<td>BSA</td>
<td>1.53 m²</td>
</tr>
</tbody>
</table>

Ao: aorta, BSA: body surface area, LA: left atrium, LV: left ventricle, PA: pulmonary artery, PCWP: pulmonary capillary wedge pressure, PVRI: pulmonary vascular resistance index, RA: right atrium, RV: right ventricle, SVRI: systemic vessel resistance index. RA, PCW and LA data were expressed as a wave/v wave/mean pressure. RA, PA and PCWP data were expressed as systolic/diastolic/mean pressure.

Table 1. (b). Oxymetry (Room Air, Supine Position)

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<thead>
<tr>
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<tbody>
<tr>
<td>VO₂</td>
<td>186 ml/min</td>
<td>CS</td>
<td>88.7%</td>
</tr>
<tr>
<td>Hemoglobin</td>
<td>12.0 g/dl</td>
<td>LV</td>
<td>93.3%</td>
</tr>
<tr>
<td>IVC</td>
<td>74.7%</td>
<td>Ao</td>
<td>93.3%</td>
</tr>
<tr>
<td>SVC</td>
<td>75.7%</td>
<td>Qs</td>
<td>6.5 l/min</td>
</tr>
<tr>
<td>RA</td>
<td>81.6%</td>
<td>Qp</td>
<td>7.1 l/min</td>
</tr>
<tr>
<td>RV</td>
<td>79.3%</td>
<td>Qe</td>
<td>5.8 l/min</td>
</tr>
<tr>
<td>PA</td>
<td>79.0%</td>
<td>Qp/Qs</td>
<td>1.1</td>
</tr>
<tr>
<td>LA</td>
<td>94.8%</td>
<td>L-R shunt</td>
<td>18.3%</td>
</tr>
<tr>
<td>PV</td>
<td>95.3%</td>
<td>R-L shunt</td>
<td>10.8%</td>
</tr>
</tbody>
</table>

with the PLSVC between the left innominate vein and the left atrium. The left pulmonary vein partially drained into the left innominate vein resulting in partial anomalous pulmonary venous return (PAPVR) (Fig. 2B).

Left ventriculography showed rotation of the heart in a counterclockwise direction and enlargement of the ascending aorta (Fig. 3). Coronary angiography showed normal coronary arteries with normal branching patterns. The cardiac veins opened into the left atrium as well as the right atrium. The R-L shunt occurred through the aperture of the coronary sinus. We could not measure the aperture size precisely, however it appeared to be less than 10 mm in diameter. We were able to insert a 5-F Judkins right coronary catheter easily into the coronary sinus. The aperture was present in the mid-portion of the wall between the coronary sinus and left atrium, the so-called “partially unroofed” mid-portion of the coronary sinus (Fig. 4). The PLSVC was too small (approximately 2 mm in diameter) to have an influence on the hypoxemia. Thus, pressure in the PLSVC was not measured.

After the patient gave informed consent, we surgically closed the PFO and the unroofed coronary sinus. During the operation, the PFO was found to be approximately 10 mm in diameter. Since the relationship between the coronary sinus aperture and the pulmonary veins could not be verified, we performed only surgical excision and patch closure of the PFO and ASA. Fortunately, dyspnea and hypoxemia in the upright position was almost completely resolved. Anticoagulation therapy was continued after the operation.

Discussion

Approximately 25 to 30% of the American population has a defect in the atrial septum known as PFO (7). The presence of PFO and ASA are currently thought to be significant predictors of an increased risk of recurrent stroke (8). In addition, the present patient had partially unroofed mid-portion of the coronary sinus, PLSVC and PAPVR. Unroofed coronary sinus syndrome is also a rare cardiac anomaly in which a communication occurs between the coronary sinus and left atrium as a result of the absence of the roof of the coronary sinus. Unroofed coronary sinus and PLSVC cause R-L shunts, and PAPVR causes an L-R shunt.

In the present patient, the dyspnea and hypoxemia in the upright position were not apparent until this admission. These findings suggested that there was little influence of the PLSVC, unroofed coronary sinus and PAPVR on the patient’s previous status. Sudden onset of the pulmonary embolism might cause elevation of the right atrial pressure and increased shunt flow through the PFO. However, in spite of the success of anticoagulation therapy, the increased R-L shunt flow in the upright position led to more severe hypoxemia.

Recently, it has been reported that a PFO could be the cause of the P-O syndrome (4, 5). In our patient, the enlargement of the ascending aorta rotated the heart in a counterclockwise direction. In the upright position, the patient’s ectatic aorta compressed the right atrium. This anatomical distortion of the atrial septum directs the atrial venous inflow through the PFO, and the R-L shunt flow is exacerbated (9).
This event was more marked when she was in the kyphotic position. During hospitalization, she tended to lean forward in sitting position owing to the loss of muscular strength. Interestingly, holding her in the correct sitting position with a physiological lordosis improved the oxygen saturation. These findings may be able to support our consideration.

The optimal treatment for PFO is controversial and varies from anticoagulation or aspirin to surgical closure (10). In the present case, we selected surgical patch closure of the PFO for the treatment of P-O syndrome. Recently, percutaneous PFO closure was proven to be a safe and effective technique for the prevention of recurrent thromboembolism in patients with PFO (10, 11).

Unroofed coronary sinus could be the cause of cerebral infarction, infectious diseases, and heart failure (12). However, ligation of the PLSVC associated with unroofed coronary sinus is still controversial (13).

In conclusion, we present a patient of P-O syndrome owing to PFO. The P-O syndrome is thought to be a rare cause of dyspnea, and many patients may not be diagnosed correctly from routine examinations. It is important to note the pattern of occurrence of dyspnea, and if it occurs in the
upright position, the most sensitive modality for noninvasive diagnosis is peripheral contrast TEE (on the tilt-table) to establish the presence of an intracardiac R-L shunt.

References


