A Case of Platypnea Orthodeoxia Syndrome: A Persistent History Taking was the Key to the Diagnosis

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Abstract

A 79-year-old woman who had been suffering from dyspnea on effort for more than 50 years was admitted for further examination and treatment. On the screening respiratory examinations, the A-aDO2 was elevated but none of diffusion disturbance, ventilation-perfusion ratio inequality nor right-to-left shunt was detected. Finally, the fact that the dizziness occurred only in sitting or standing position was revealed by persistent history taking. Transesophageal echocardiography in recumbent and sitting positions revealed the platypnea orthodeoxia syndrome associated with atrial septal defect. This case highlights the necessity of awareness of this syndrome and the occult atrial septal defect.

Key words: hypoxemia, platypnea-orthodeoxia syndrome, atrial septal defect

Introduction

Platypnea orthodeoxia syndrome (P-OS) is a rare syndrome characterized by dyspnea and a decrease in arterial blood oxygen saturation that is most prominent when the patient is sitting or standing, and it is resolved when the patient is supine. Some anatomical causes are known to cause P-OS, such as a patent foramen ovale (PFO), atrial septal defect, and fenestrated aneurism in the interatrial septal region, in which a blood flow shunt from the right to left atrium is formed. Some cases have been reported in the world (1), including several cases in Japan (2-6), since Burchell et al. reported the first case in 1949 (7).

We report a patient with hypoxemia for which the cause had been unclear for more than 50 years, and diagnosed P-OS by transesophageal echocardiography (TEE) using different posture.

Case Report

A 79-year-old woman had been aware of shortness of breath during exercise since she was young. She had consulted about exertional dyspnea when she was admitted for hysterectomy at the age of 33 years old and transient ischemic attack at 66 years old, but no apparent cause had been detected. When she visited a physician for a common cold in January 2009, percutaneous arterial blood oxygen saturation was only in the 70% range, for which the patient was referred to our hospital and admitted for further examination.

She had no particular family medical history. She had a past medical history; hysterectomy, 33 years of age; transient cerebral ischemic attack, 66 years of age; lumbar compression fracture, 76 years of age; surgery for ileus, 78 years of age. Her physical examination findings on admission were as follows: consciousness, clear; body temperature, 35.8°C; blood pressure, 134/84 mmHg; pulse rate, 90 beats/min; resting SpO2, 95% (indoor atmospheric pressure); fine crackles were slightly heard on the dorsal side of the left lower lung region on chest auscultation; and no cardiac murmur was heard.

The findings of laboratory test on admission were as follows (summarized in Table 1). On chest X-ray radiography, the cardiothoracic ratio was 54%, showing mild auxocardia, and the right pulmonary hilum was dilated (Fig. 1a). No apparent abnormality was detected by electrocardiography. On chest contrast CT, linear and reticular shadows were slightly

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observed in the peripheral lung field, and no apparent feature of pulmonary embolism was noted (Fig. 1b).

On arterial blood gas analysis (on supine position under room air): pH, 7.430; PaCO2, 37.3 mmHg; PaO2, 60.7 mmHg; SaO2, 91.9%; and A-aDO2, 42.4 mmHg; showing mild oxygenation failure. On a pulmonary function test: FVC, 1.49 L; %VC, 73.4%; FEV1, 1.17 L; FEV1/FVC, 78.5%; DLCO, 8.78 mL/min/mmHg; %DLCO, 62.3%; DLCO/VA, 3.99 L; and %DLCO/VA, 70.2%; showing no disturbance of ventilation or diffusion. The shunt rate measured with 100% oxygen inhalation was 9%, showing a mild high level. On ventilation-perfusion scintigraphy, no ventilation-perfusion ratio inequality was detected (Fig. 1c). No abnormality was observed on transthoracic echocardiography (TTE).

The pathological condition of hypoxemia was not clarified by imaging procedures or tests at this point, but the gas analysis parameters after walking for 5 minutes were: pH, 7.415; PaCO2, 36.5 mmHg; PaO2, 52.4 mmHg; SaO2, 87.8%; and A-aDO2, 44.6 mmHg; showing that hypoxemia was apparently exacerbated. Detailed physical examination and medical interview were undertaken, in which the patient reported dizziness only in sitting or standing position but not in supine position, and blood pressure reduction upon posture change and simultaneous marked reduction of arterial blood oxygen saturation were found; 155/96, 82 beats/min, SpO2, 95% on supine to 102/77, 103, 85% in sitting position.

Based on these physical findings, P-OS was suspected. The shunt rate was measured with posture changes, and the rate was 9% in recumbence but it markedly increased to 18% in a sitting position. A TEE was performed to identify the cause, through which an atrial septal defect measuring 1.5×0.67 cm was noted, and blood flow in both directions: from the left to right atrium and vice versa, was noted in the supine position (Fig. 2a). After posture change to a sitting position, the right atrium was compressed by the aortic root and blood flow from the right to left atrium became dominant (Fig. 2b). Based on the above findings, it was concluded that the cause of hypoxemia was P-OS with right-to-left interatrial shunt through the atrial septal defect formed on sitting or standing up.

This disease is usually treated by closure of the interatrial shunt. Thoracotomy was considered too stressful because of the advanced age of the present patient. Thus, a percutane-

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### Table 1. Laboratory Data on Admission

<table>
<thead>
<tr>
<th>Hematology</th>
<th>Normal Range</th>
<th>Biochemistry</th>
<th>Normal Range</th>
<th>Serology</th>
<th>Normal Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>WB</td>
<td>4840 /µL</td>
<td>3500-9500</td>
<td>TP</td>
<td>6.9 g/dL</td>
<td>6.5-8.0</td>
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<tr>
<td>Neu</td>
<td>63.5 %</td>
<td>52-80</td>
<td>Glu</td>
<td>117 mg/dL</td>
<td>70-110</td>
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<tr>
<td>Eos</td>
<td>2.5 %</td>
<td>1-5</td>
<td>T-bil</td>
<td>0.3 mg/dL</td>
<td>0.3-1.2</td>
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<tr>
<td>Bas</td>
<td>0.4 %</td>
<td>0-1</td>
<td>ALP</td>
<td>307 IU/L</td>
<td>110-360</td>
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<tr>
<td>Mon</td>
<td>4.1 %</td>
<td>1-5</td>
<td>γ-GTP</td>
<td>12 IU/L</td>
<td>5-40</td>
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<tr>
<td>Lyn</td>
<td>29.5 %</td>
<td>20-40</td>
<td>LDH</td>
<td>231 IU/L</td>
<td>120-240</td>
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<td>Alb</td>
<td>3.8 g/dL</td>
<td>3.8-4.9</td>
<td>SP-D</td>
<td>60.5 ng/mL</td>
<td>&lt;110</td>
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<tr>
<td>RBC</td>
<td>4.17×10⁶ /µL</td>
<td>360-480</td>
<td>Glib</td>
<td>3.1 g/dL</td>
<td>2.3-3.5</td>
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<tr>
<td>Hb</td>
<td>12.9 g/dL</td>
<td>11.5-14.5</td>
<td>ChE</td>
<td>211 IU/L</td>
<td>222-248</td>
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<tr>
<td>Ht</td>
<td>39.8 %</td>
<td>34-44</td>
<td>ALT</td>
<td>10 IU/L</td>
<td>7-42</td>
</tr>
<tr>
<td>Plt</td>
<td>18.4×10⁶ /µL</td>
<td>15-35</td>
<td>AST</td>
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<td>10-35</td>
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<td>CRN</td>
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<td>UN</td>
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<td>UA</td>
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<tr>
<td>Na</td>
<td>140 mEq/L</td>
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<tr>
<td>K</td>
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<td>3.6-5.0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cl</td>
<td>106 mEq/L</td>
<td>101-110</td>
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</table>
Discussion

Although a PFO/ASD is present in 25-30% of adults (8), P-OS does not typically develop even if these causes are
present because the left atrial pressure is 5-8 mmHg higher than the right atrial pressure (2). Right-to-left shunt may occur when the right atrial pressure rises due to kyphoscoliosis, pulmonary hypertension, surgical pneumonec- tomy, etc (9-11). Various P-OS cases have recently been reported, such as those associated with hypothyroidism-induced pericardial effusion collection, pulmonary hypertension in obstructive sleep apnea syndrome (12), constrictive pericardi- tis (13), and pneumocystis pneumonia (14, 15). In the present patient, the aorta projected toward the right atrium in a sitting position, which may have mechanically induced tri- cuspid stenosis, elevating the right atrial pressure and inducing right-to-left shunt.

To treat hypoxemia-inducing diseases, differential diagnosis is performed in consideration of a low alveolar ventilation, diffusion disturbance, ventilation-perfusion ratio inequality, and right-to-left shunt. For the chief complaint of dizziness on standing up in this patient, the presence of a P-OS was suspected by paying attention to changes in oxygenation failure, in addition to hypotension on standing up, leading to the diagnosis.

Briefly summarizing the previous reports, quite a few of such cases were suspected by the exacerbation of the dyspnea on effort or the decrease of artery oxygen saturation at the position change (2). There is no gender difference on the developing frequency of this syndrome and several cases have been diagnosed at the age of 70 years or more as in the presented case (5, 7, 12). Most of the patients improved almost completely by thoracotomy or transcatheter closure of the interatrial shunt (3, 5, 7, 9, 13).

The present patient had been suffering from activity limitation for decades, which was finally resolved at an advanced age of 79 years. Although P-OS is a rare disease, it is important to be careful to pay attention to the dyspnea pattern with detailed history taking. When P-OS is suspected as the etiology, even if a TTE is not able to detect the intracardiac shunt, TEEs not only in the supine position but also in the upright position should be carried out.

The authors state that they have no Conflict of Interest (COI).

This case was reported at the 44th Meeting of the Japanese Respiratory Society Chugoku-Shikoku Area.

References