CASE REPORT

Platypnea-orthodeoxia Syndrome Associated with a Thoracic Vertebral Fracture Following a Car Accident

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Abstract

Platypnea-orthodeoxia syndrome (POS) is a rare clinical condition defined by the presence of dyspnea and deoxygenation accompanying changing from a supine to an upright position. We experienced the case of a 75-year-old woman who developed severe acute dyspnea after a car accident. Detailed history taking and a physical examination offered important clues that helped to make an accurate diagnosis of POS. The mechanism of onset is unique and rare; however, it is important for clinical cardiologists to keep this possibility in mind when making a differential diagnosis.

Key words: patent foramen ovale, paradoxical de-oxygenation, contrast computed tomography, tilt transesophageal echocardiography

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Introduction

Platypnea-orthodeoxia syndrome (POS) is a rare clinical condition characterized by dyspnea and hypoxia when the patient changes from a supine to an upright position. It was first described by Burchell in 1949 (1), and more than 100 cases, including 10 Japanese cases, have been previously reported in the literature. We encountered a patient who developed POS after a car accident and was successfully treated by means of a surgical repair.

Case Report

A 75-year-old woman visited our hospital due to palpitations and dyspnea on exertion lasting for 10 months after a car accident. She had a history of paroxysmal atrial fibrillation, for which she had continuously received anticoagulation therapy for 20 years without any known episodes of heart failure or stroke. On a physical examination, the patient’s blood pressure was 134/74 mmHg, and her pulse rate was 60 beats per minute. While the oxygen saturation was 83% on room air, no apparent cyanosis, peripheral edema or clubbing of the extremities were observed. On a chest X-ray, no visible lung congestion was observed, despite the presence of cardiomegaly with a cardiothoracic ratio of 56.1%, while thoracic shortening and aortic distortion due to a thoracic vertebral fracture was noted compared with the findings of a chest X-ray obtained one year earlier (Fig. 1). On an electrocardiogram, left axis deviation and complete right bundle branch block were detected. On a transthoracic echocardiogram (TTE), both atria were enlarged, and an atrial septal aneurysm was detected; however, no interatrial shunt flow was observed. The laboratory data revealed no remarkable abnormalities, and the A-aDo2 level was increased to 38.0 mmHg on an arterial blood gas analysis. Contrast-enhanced chest computed tomography (CT) revealed a deformity of the right atrium caused by an elongated ascending aorta and a left to right atrial shunt flow through a patent foramen ovale in the supine position; however, no lung parenchymal abnormalities, pulmonary emboli or arteriovenous shunts were observed (Fig. 2). A lung perfusion test also revealed no abnormalities. Detailed history taking disclosed that the patient’s symptoms worsened on sitting up...
Figure 1. A chest X-ray obtained on admission showed thoracic shortening and aortic distortion (black arrows) due to a thoracic vertebral fracture (arrowhead) compared to that observed one year earlier (white arrows).

Figure 2. Contrast-enhanced chest computed tomography showed a) a dilated and elongated ascending aorta on three-dimensional reconstruction of the coronary artery and thoracic aorta, b) a deformity of the right atrium caused by the ascending aorta (black arrow) on a coronal view and c) a left to right atrial shunt flow through a patent foramen ovale on a four-chamber long axis plane (white arrow). Ao: ascending aorta, RA: right atrium, LA: left atrium, LV: left ventricle

Table. The Arterial Blood Oxygen Saturation in the Various Postures

<table>
<thead>
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<th>Left lateral</th>
<th>Supine</th>
<th>Right lateral</th>
<th>Sitting</th>
<th>Upright</th>
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<tr>
<td>Saturation</td>
<td>98%</td>
<td>98%</td>
<td>93%</td>
<td>87%</td>
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or being in an upright position and improved in the supine position, while the oxygen saturation improved from 87% on sitting to 98% in the supine position (Table). According to the patient’s history and physical and laboratory examination findings, a diagnosis of POS was suspected. TTE with intravenous agitated saline contrast solution was performed again in both the supine and upright positions, which revealed bubbles that appeared in the left atrium within eight cardiac cycles. This finding suggested either intracardial or intrapulmonary shunting. Transesophageal echocardiography with the administration of intravenous agitated saline contrast solution was therefore performed, and a right to left shunt flow through the PFO was observed. The shunt flow was increased by the Valsalva maneuver and postural changes from a supine to a sitting position (Fig. 3a-c). Cardiac catheterization revealed normal hemodynamics with an influx of blood flow from the inferior vena cava into the left atrium and obvious desaturation in both the left atrium and left inferior pulmonary vein. Based on these findings, the patient was diagnosed with POS, and the cause of hypoxia during the upright position was concluded to be POS. The mechanism of POS in this case was thought to be a deformity of the atrium caused by elongation of the ascending aorta during sitting or being in an upright position. The patient’s symptoms rapidly developed after a car accident; therefore, this phenomenon may have been the result of a vertebral compression fracture caused by the car accident.

In this case, surgical closure of the patent foramen ovale (PFO) and left atrial appendage excision were performed to
prevent paroxysmal atrial fibrillation. After treatment, the patient’s arterial oxygen saturation recovered up to 98% on room air in the sitting position, and her symptoms disappeared. She was discharged on foot 15 days after undergoing surgery.

**Discussion**

We herein described a typical POS case with a unique onset after a car accident. POS is an uncommon but serious clinical syndrome characterized by dyspnea and deoxygenation accompanying changes from a supine position to a sitting or upright position (2). Some anatomical features and functional components must coexist as causes of POS (3). The former include atrial septal defects (ASDs), a patent foramen ovale, atrial aneurysms forming intracardiac communications and hepatopulmonary syndrome forming an intrapulmonary shunt. Among these, PFO is the most common, with an incidence of 20-25% (4). The latter include cardiac events, such as pericardial effusion and constrictive pericarditis, pulmonary events, such as emphysema, arteriovenous malformations, pneumonectomy and amiodarone toxicity, abdominal events, such as cirrhosis of the liver and ileus, and vascular events, such as aortic aneurysms and elongation (5). Kyphoscoliosis is also associated with a right atrial pressure elevated above the left atrial pressure, which may lead to POS (6). The most common causes are thought to be interatrial right to left shunts through PFOs and atrial septal defects. Some mechanisms of right to left shunting that occur even in the absence of pulmonary hypertension have been considered. Standing upright can stretch an interatrial communication, be it a PFO or ASD, allowing for more streaming of venous blood from the inferior vena cava through the defect, described as the “flow phenomenon” by Zanchetta (7). This redirection of the flow is also caused by anatomical distortions of the right atrium or right septum induced by either aortic aneurysms or lobectomy. In our case, a direct flow from the inferior vena cava to the left atrium through a PFO was observed during catheterization and TEE. The etiology of the distortion of the PFO in our case may be explained as follows: the thoracic vertebral fracture resulted in the shortening of the height in the sitting position. This induced relative aortic elongation and aggravated the aortic enlargement projection toward the right atrium, which was observed as an aortic shadow distortion on chest X-rays. Thereafter, the increased right to left shunt through the PFO resulted in POS. Galiuto et al. (6) also described in their previous report that a similar mechanism of acute onset of dyspnea was observed after a vertebral fracture. Concerning the diagnosis, similar to previous reports, the first step involved paying attention to the patient’s dyspnea pattern, with detailed history taking providing a clue for the differential diagnosis (3). For the second step, as Desouza (2) described, tilt TEE with agitated saline contrast solution is usually the most effective diagnostic tool; however, in our case, TTE with intravenous agitated saline contrast solution performed in both the supine and upright positions was also effective and efficient in distinguishing abnormal shunting from a ventilation-perfusion mismatch, low alveolar ventilation and a diffusion disturbance. Regarding the treatment of POS, the use of open surgery is fundamentally recommended to close the interatrial communication. Recently, transcatheter closure devices have come into use due to their reduced invasiveness and advantages in safety and good long-term outcomes compared to open surgery (8). In this case, we chose to perform surgical repair because we worried that the atrial septal deformity induced by postural changes may cause device erosion or the superior septal rim defect may cause device migration (9). Concerning open surgery, Hojo et al. (10) reported a case of POS with a PFO and an elongated ascending aorta. The PFO was closed and the ascending aorta was shortened with open surgery because relative tricuspid valve stenosis was revealed in the sitting position as a consequence of aortic elongation. In our case, as a result of the absence of relative tricuspid valve stenosis in the sitting position, we decided to perform closure of the PFO using open surgery due to the possibility for erosion and device migration. As described above, we encountered a rare but important case of unique reverse postural desaturation revealed by detailed history taking and

**Figure 3.** Transesophageal echocardiography revealed a) a patent foramen ovale (white arrow) and increased right to left shunt caused by postural changes compared between the b) supine and c) sitting positions using agitated saline. LA: left atrium, PFO: patent foramen ovale

physical findings of interest to general cardiologists. POS is a rare but significant disease with specific findings of paradoxical deoxygenation. Performing detailed history taking and a careful checkup during the physical examination is therefore important for accurately diagnosing this specific disease. This case suggests that it is important for clinical cardiologists to keep this possibility in mind when making a differential diagnosis in patients with dyspnea and deoxygenation in the clinical setting.

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

References