Platypnea-orthodeoxia Syndrome Caused by a Latent Atrial Septal Defect

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Abstract

A 79-year-old woman presented with unexplained hypoxia that became exacerbated by an upright posture (platypnea-orthodeoxia syndrome). A ⁹⁹mTc-macroaggregated albumin pulmonary perfusion scan revealed a right to left shunt of 25.5% in the supine position and 32.3% in the sitting position. A dynamic CT scan and a transoesophageal echocardiogram confirmed the presence of a shunt across an atrial septal defect (ASD). A percutaneous transcatheter closure of the defect significantly improved the patient’s blood oxygenation levels when she was in the upright position. An ASD should therefore be included in the differential diagnosis of platypnea-orthodeoxia syndrome, regardless of the patient’s age.

Key words: platypnea-orthodeoxia syndrome, atrial septal defect, hypoxia, right to left shunt


Introduction

Platypnea-orthodeoxia, a rare syndrome characterized by hypoxia that is exacerbated by an upright posture and is partially relieved by recumbence, was first described in 1949 by Burchell et al. (1). The causes and pathophysiology of this condition remain uncertain. We herein report the findings of a case of platypnea-orthodeoxia that was successfully treated with a percutaneous transcatheter closure of a pre-existing atrial septal defect (ASD).

Case Report

A 79-year-old woman presented with exertional dyspnoea that had progressively worsened over a 2 month span. She suffered 2 episodes of syncope during this 2-month period, but was sent home from the hospital each time because she recovered completely while being transported by ambulance. She had a history of pulmonary embolisms 2 years earlier and was therefore being treated with warfarin to promote anticoagulation.

During the physical examination, her blood pressure was measured at 136/81 mmHg and her heart rate was 59 bpm. The blood oxygen saturation measurement taken while she was in the supine position was 90% with 3 L/min of oxygen delivered via a nasal cannula, but this reading rapidly decreased to 70% when she was elevated to a sitting position. Her oxygen saturation level while in a sitting position remained unchanged even with increases in the oxygen delivery rates. Auscultation revealed audible fine crackles in the left lung field, but there was no detection of a heart murmur.

The laboratory tests, chest roentgenogram and transthoracic echocardiogram findings were all normal. The electrocardiogram showed an isolated incomplete right bundle branch block. A mild restriction and an airflow obstruction were identified using the following pulmonary function test results: a forced vital capacity (FVC) of 78.2% of the predicted value and a forced expiratory volume in 1 second (FEV₁) of 85.6% of the predicted value, with an FEV₁/FVC ratio of 0.66. Chest computed tomography with contrast was normal, with the exception of faint reticular shadows in both of the lung bases and a minimal loss of volume in the left lower lobe. No thrombus was found in the pulmonary circulation.

The results of the examination indicated that the postural
Figure 1. The ⁹⁹ᵐTc-macroaggregated albumin lung perfusion scan in the supine position (left) and sitting position (right) during Valsalva maneuvers. Note the extrapulmonary accumulation of the radio-labelled albumin in the kidneys and the brain.

Figure 2. A dynamic CT scan of the chest. Blood flow (arrow) was observed between the left atrium (LA) and right atrium (RA). Ao: aorta, PA: pulmonary artery

Figure 3. A transoesophageal echocardiogram that shows a left to right shunt across the ASD (arrow). The eustachian valve (arrow head) is present at the junction of the inferior vena cava and the right atrium. LA: left atrium, RA: right atrium, Ao: aorta, IVC: inferior vena cava

Hyoxia may have been caused by a right to left shunt. Therefore, pulmonary perfusion imaging with ⁹⁹ᵐTc-macroaggregated albumin (MAA) was performed in both the supine and sitting positions. The MAA accumulated in the kidneys and brain, thus suggesting the presence of a right to left shunt (Fig. 1). The estimated shunt ratio was 25.5% in the supine position and 32.3% during a Valsalva maneuver in the sitting position. The differential diagnosis included a pulmonary arterio-venous malformation, hepato-pulmonary syndrome and intracardiac shunts. A dynamic computed tomography scan suggested the presence of an interatrial flow (Fig. 2). The transoesophageal echocardiogram confirmed the presence of a left to right shunt through a 10.7x15.1 mm, ostium secundum type ASD, as well as the presence of the eustachian valve at the junction of the inferior vena cava and the right atrium (Fig. 3). As the oesophagus was separated from the left atrium in the sitting position, we could not detect the redirection of the shunt flow.

The left heart function and the pulmonary artery pressures were normal during a cardiac catheterization (Table 1). The leftatriogram showed a left to right, but not a right to left, atrial blood flow. An upright position caused a worsening of the arterial oxygenation level (SpO₂ of 58.8%), followed by transient bradycardia (40 bpm) and hypotension (systolic blood pressure of 60 mmHg). A balloon closure of the ASD increased the ascending aortic oxygen saturation to 88.2% in the upright position (Table 2), thus suggesting that the hypoxia was caused by a right to left shunt through the ASD. The patient underwent a percutaneous transcatheter closure of the defect with an Amplatzer® device, which markedly alleviated her symptoms and increased the arterial saturation on room air in the upright position to 93%.

Discussion

Platypnea-orthodeoxia is a syndrome that can be caused by various arteriovenous malformations (2), pulmonary parenchymal diseases (3), or the presence of intracardiac shunts. The latter is predominantly due to a patent foramen ovale (PFO) and, less frequently, due to an ASD (4). In the 33 reported cases of platypnea-orthodeoxia syndrome that were caused by intracardiac shunts in the last 5 years, the mean age of the patient was 67 years. Of these 33 cases, 24 (73%) were due to PFO and 6 (18%) were caused by ASD. Platypnea-orthodeoxia is caused by an increase in right atrial pressure that can be induced by postural changes, inspiration, cough, or Valsalva maneuvers (5); although, in the absence of pulmonary hypertension, the mean left atrial
patients suffering from platypnea-orthodeoxia syndrome due to an intracardiac shunt is associated with an enlarged ascending aorta (6). It has been hypothesized that the ascending aorta causes an anterior-posterior displacement of the aortic root when the patient is in the upright position, which leads to compression of the right atrium and an enlargement of the PFO aperture (7). The bulging of the aortic root above the tricuspid orifice may behave functionally like an acquired cor triatriatum dexter (6). A previous report has described a large aneurysm of the ascending aorta that caused a significant lateral displacement of the superior vena cava (SVC) and considerable compression of the right atrium, thus leading to the development of a direct shunt between the vena cava and the left atrium across a PFO (8). Furthermore, the enlargement of the ascending aorta rotates the heart counterclockwise, thus distorting the position of the atrial septum relative to the caval inflow, and directing the right atrial venous inflow toward the PFO (9).

In the present case, however, the size of the ascending aorta was normal and the heart was rotated clockwise due to loss of volume in the left lower lung. The eustachian valve, an embryonic remnant that was present in this patient, may have directed the blood flow from the inferior vena cava to the left atrium across the ASD. The development of manifestations of a congenital abnormality in later life may be explained by the loss of right heart compliance due to aging (10). In addition, the MAA that was injected from the arm was distributed outside the lungs, thus suggesting that the blood flow from the SVC may have contributed to the development of the right to left shunt. However, since we could not visualise the shunt flow with an echocardiogram or by cardiac catheterization, the precise mechanism remains uncertain.

Our patient’s symptoms were immediately alleviated and her quality of life was rapidly improved following a percutaneous transcatheter closure of the ASD. Previous reports have described the safe and successful treatment of elderly patients suffering from platypnea-orthodeoxia syndrome due to an ASD or PFO using similar procedures (11, 12). Therefore, it is essential to suspect this syndrome in patients who present with unexplained hypoxia, and to order the appropriate diagnostic tests.

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

**References**