Platypnea-Orthodeoxia: A Case of Unexplained Hypoxia

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ABSTRACT

Introduction: Platypnea-orthodeoxia syndrome is a rare clinical syndrome defined by worsening deoxygenation and dyspnea when changing to an upright sitting or standing position. It is seen in 3 different clinical scenarios: intracardiac shunts, pulmonary arteriovenous shunts, and ventilation/perfusion mismatch in the lungs.

Case: An 82-year-old woman with a history of nonischemic cardiomyopathy with reduced ejection fraction was admitted with dyspnea and hypoxemia. She was found to have atrial septal defect with right to left shunting in the setting of normal right atrial pressures.

Discussion: Platypnea-orthodeoxia syndrome is a clinical syndrome where, in the setting of an interatrial communication, a right to left shunt can occur without elevated pulmonary or right atrial pressure, resulting in significant hypoxia.

Conclusion: Platypnea-orthodeoxia syndrome is a clinical condition that is being recognized more frequently due to more accurate diagnosis, and its treatment can alleviate symptomatic hypoxemia.

INTRODUCTION

Platypnea-orthodeoxia is an uncommon clinical phenomenon that usually manifests in older patients and is defined by worsening hypoxemia and dyspnea with the change to an upright sitting or standing position. Patients with platypnea-orthodeoxia syndrome (POS) experience hypoxia from a right to left cardiac shunt without the need for elevated right heart pressure.[1]

The condition was first reported in 1949 in a patient with an intrathoracic arterial venous shunt. Since then it has been described in a small subset of patients and can be seen in 3 different clinical scenarios: intracardiac shunts, pulmonary arteriovenous shunts, and ventilation/perfusion mismatch in the lungs.[1] In POS cases involving intracardiac shunts, 3 different defects have been described: patent foramen ovale (PFO), atrial septal defect (ASD), and a fenestrated atrial septal aneurysm (Box 1). PFO is the most common among these three, but in cases of POS caused by an ASD, the ostium secundum type is most common.[1]

CASE PRESENTATION

An 82-year-old woman with a history of nonischemic cardiomyopathy with reduced ejection fraction was transferred from a referring hospital for new onset of hypoxemia and dyspnea requiring high levels of oxygen. She recently had been hospitalized for a heart failure exacerbation with improvement after standard medical management. She had returned to the referring hospital several days after discharge with new hypoxia and dyspnea without any clinical signs of heart failure. She required 10L/min of oxygen via face mask to maintain oxygen saturation in the low 90s. A transthoracic echocardiography with bubble study led to the diagnosis of an interatrial shunt. Transesophageal echocardiography revealed a secundum ASD measuring up to 8 mm in diameter. The atrial septum demonstrated increased mobility consistent with an atrial septal aneurysm. A right heart catheterization performed while the patient was supine demonstrated hypoxia, which improved with 100% FiO_2 during the procedure, and normal right atrial, right ventricle, and pulmonary capillary wedge pressures.

A pulmonary work-up was performed following the right heart catheterization. Computed tomography scan of the chest showed no evidence of any pulmonary arterial venous malformations (AVM). Ventilation-perfusion scan demonstrated very low probability for acute pulmonary embolism, and pulmonary function testing showed normal spirometry with no significant improvement after bronchodilators.

The patient continued to have high oxygen needs ranging from 4L to 10L on high flow nasal cannula. On further evaluation, she was found to have worsening oxygen needs when changing from a lying to seated position and standing up from a recumbent position. The clinical picture was consistent with platypnea-orthodeoxia

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Box 1. Most Common Types of Intracardiac Shunting in Platypnea-Orthodeoxia Syndrome

1. Patent Foramen Ovale
2. Atrial Septal Defect (Ostium Secundum)
3. Fenestrated Atrial Septal Aneurysm

Box 2. Common Findings in Platypnea-Orthodeoxia Syndrome

Interatrial communication or pulmonary arteriovenous malformation
Right to left shunt without elevated pressure
Worsening oxygenation when sitting/standing
Dyspnea that resolves when lying flat
Resolution with closure of defect

syndrome. Based on this diagnosis, the patient underwent an atrial septum defect closure using a 30 mm Gore Septal Occluder. Her POS promptly resolved, and she was discharged without requiring supplemental oxygen. Follow-up 2 months later showed that she was still oxygenating well on room air and was free of dyspnea.

DISCUSSION

Platypnea-orthodeoxia syndrome is an uncommon clinical syndrome. Since it was first reported in 1949 under 200 cases have been reported in the literature.[1] Common findings are listed in Box 2. With prompt recognition of clinical signs—mostly through a thorough history and physical exam—POS can be accurately diagnosed, leading to appropriate intervention and a reduction in patient morbidity. Most intracardiac cases of POS respond to intervention.

In the case of our patient, POS concern was crucial in deciding that intracardiac shunting was the likely cause of hypoxemia. After closure of the ASD she was weaned from supplemental oxygen and returned home to her baseline functional status.

While the exact mechanism of POS is still not known, there have been several theories on its pathophysiology. One possibility is that right to left shunting at the atrial level is caused by transient fluctuations in right atrial pressure, coupled with diminished right atrial compliance when the patient transitions to a sitting or standing position. Decreased right atrial compliance may be related to comorbid conditions, such as right-sided myocardial infarction, right atrial myxoma, or eosinophilic myocardial disease, predisposing some patients to develop right to left shunts.[1] Another potential mechanism is the presence of some distortion in the normal thoracic anatomy leading to more directed flow from the vena cava through the patient's preexisting PFO or ASD.

The most common thoracic abnormality described is dilation or elongation of the aorta. This may affect the architecture of the right atrium, causing horizontalization of the atrial septum.[1] Another possibility is that a persistent eustachian valve or stretching of the ASD with changing body positions may allow blood flow to stream from the inferior vena cava through the anatomical defect and into the left atrium.[2] Other anatomic defects associated with POS include Chiari's network, kyphosis of the spine, previous pneumonectomy, and hemiparalysis of the right diaphragm.[1]

POS also can been seen in noncardiac etiologies, most commonly pulmonary AVM where the upright position increases blood flow to the lung bases, accentuating shunt through the AVM and causing increased hypoxemia.[3] Similar physiology is seen in hepatopulmonary syndrome, where dilated pulmonary capillary vessels can cause V/Q mismatch in an upright position, decreased alveolar-arterial diffusion, and arteriovenous shunting.[3] Even though our knowledge of the pathophysiology of POS has improved in recent years, the exact mechanism that leads to symptomatic shunting in an individual patient may not be clear. In the case of our patient, there was significant kyphosis noted on radiologic imaging.

Proper diagnosis of POS is crucial for appropriate treatment, which can greatly improve the patient's clinical status. In a recent review, the median age at the time of diagnosis was in the seventh decade of life.[1] A retrospective analysis of 78 patients with POS demonstrated that percutaneous closure of the atrial level shunt resulted in significant improvement in New York Heart Association Functional Classification. Successful closure of an atrial defect was shown to produce symptomatic improvement in more than 95% of patients. Given the relative ease of application, percutaneous septal defect closure is the preferred method of treatment of ASD or PFO.[4]

CONCLUSIONS

While POS remains rare, it is being recognized more frequently as a cause of hypoxemia in older patients. Timely diagnosis and treatment are key to reducing morbidity. It is important to report cases of POS to improve understanding about clinical scenarios in which the syndrome may occur. POS appears to be largely a condition of the elderly where chronic structural changes of aging result in misdirection of blood flow, but the condition of right to left shunting via an ASD or PFO occasionally may occur in younger patients as well. While this disease drastically decreases functional status or quality of life, percutaneous closure has been shown to be a reliable and effective treatment.

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