Patent foramen ovale and ascending aortic aneurysm causing the platypnea-orthodeoxia syndrome

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ABSTRACT

Patent foramen ovale (PFO) is present in about 25% of the population. Platypnea-orthodeoxia syndrome (POS), which is dyspnea and hypoxemia in the upright position that is relieved when supine, is a rare manifestation of PFO. We describe a case of a 74-year-old woman who presented with new-onset hypoxia. A PFO and a dilated aorta causing POS were found on workup. Symptoms were resolved after undergoing percutaneous PFO closure. This case highlights the mechanism by which an ascending aortic aneurysm can alter hemodynamics through an existing PFO, leading to symptoms of a previously clinically insignificant PFO, and emphasizes how early recognition of POS allows for appropriate intervention.

KEYWORDS Ascending aortic aneurysm; patent foramen ovale; platypnea-orthodeoxia syndrome

Patent foramen ovale (PFO) is usually an incidental finding with no clinical repercussions, but it harbors the potential for catastrophic diseases.^{1,2} PFO has been implicated in ischemic stroke, migraine, and decompression illness among high-volume divers.¹ A rare manifestation of PFO is hypoxemia from platypnea-orthodeoxia syndrome (POS). Platypnea is the subjective sensation of dyspnea in the upright position relieved in the supine position, and orthodeoxia is the objective hypoxemia while in the sitting or standing position.³ Most individuals with PFO almost never develop POS due to the higher left atrial pressure.⁴ In certain situations, right-to-left shunting may occur because of a transient elevation in the right atrial pressure and/or through a flow phenomenon.⁴

CASE DESCRIPTION

A 74-year-old woman with known ascending aortic aneurysm measuring 4.3 cm (*Figure 1a*), hypertension, obstructive sleep apnea, degenerative joint disease, and a body mass index of 34.27 kg/m² presented for an elective right shoulder surgery. In the procedure room, she was noted to be hypoxic with oxygen saturation at 76% on room air, prompting emergency department evaluation. On further

history, she had been having progressive shortness of breath for 3 months. Physical examination was only significant for tachypnea at 28 breaths per minute with accessory muscle use. Cardiac and pulmonary exam were unremarkable. Initial workup demonstrated a normal chest x-ray. Chest computed tomography showed mild atelectasis without evidence of pulmonary embolism, and a ventilation-perfusion scan showed radiotracer activity in both brain and kidneys concerning for possible intracardiac shunt. She then had a right heart catheterization, which revealed normal right heart pressures, including a normal pulmonary artery pressure. Shunt run did not show any evidence of left-to-right shunt, and a coronary angiogram only showed mild coronary artery disease. Laboratory tests including hemoglobin, platelets, creatinine, and brain natriuretic peptide were all normal. Arterial blood gas was consistent with respiratory alkalosis. Pulmonary function test demonstrated normal lung volumes and normal diffusion capacity. Her symptoms improved with oxygen supplementation at 6 L. Finally, a transthoracic echocardiogram with bubble study showed an atrial septal aneurysm and a right-to-left shunt at the level of the atrial septum at baseline and with provocation suggestive of a possible PFO (*Figure 1b*). She was discharged with home oxygen at 2 to 4 L.

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Figure 1. Transthoracic echocardiograms (a) demonstrating a dilated aortic root measuring 4.3 cm in diameter and (b) confirming a right-to-left shunt during a bubble study. The arrow points to the agitated saline, which has crossed the shunt.

Figure 2. Fluoroscopic images of Gore Cardioform device in interatrial septum: (a) anteroposterior view with arrows pointing to the left and right atrial disc; (b) en face view of a 30-mm Gore Cardioform device deployed in the interatrial septum with the arrow pointing to the pigtail catheter in the aorta.

She continued to have significant limitation in physical activity and could only ambulate <1 block and climb <1 flight of stairs. She was diagnosed with POS, and closure of her PFO was subsequently recommended. During the procedure, intracardiac echocardiogram redemonstrated the ascending aortic aneurysm. Intracardiac echogram also confirmed the PFO with a large right-to-left shunt by color Doppler and bubble study. The patient underwent a successful implant of a 30-mm Gore Cardioform Septal Occluder (Figure 2). Postprocedure transthoracic echocardiogram showed no residual shunt. Immediately after the procedure, the patient's arterial blood saturation increased to 92% (supine), an improvement from 86% (supine) preprocedurally. She had complete resolution of symptoms after the procedure and was discharged without any home oxygen requirements. She was seen in the outpatient clinic 1 month later with no symptoms, with pulse oximetry showing 99% on room air.

DISCUSSION

POS requires two factors to be symptomatic: the anatomical factor, which pertains to an interatrial communication via an atrial septal defect, PFO, or fenestrated atrial septal aneurysm, and the functional factor, which may be cardiac, pulmonary, abdominal, or vascular.⁵ Any of these functional factors leads to a deformity of the atrial septum that could drive hemodynamic changes that lead to symptoms from

otherwise small defects.^{2,3,5} Diagnosis of intracardiac shunt and intrapulmonary vascular dilation is confirmed via transthoracic or transesophageal echocardiogram done in both supine and upright positions with use of contrast medium and/or Doppler studies.⁴

In the absence of pulmonary hypertension, right-to-left shunting across a PFO is thought to occur either through a transient reversal of the pressure gradient between the right and left atria during each cardiac cycle due to the right atrium being depolarized slightly earlier from the localization of the sinoatrial node or through the displacement of the septum primum during respiration.⁶ Respiratory changes can cause the septum primum to deflect either to the left of the limbus, causing marked right-to-left shunting, or against the limbus of the fossa ovalis, preventing right-to-left shunting.⁶

Among 25% of patients with PFO who present with POS, there has been an identified coexistence of aortic abnormalities.^{4,7,8} The enlarged aortic root reduces the distance between the aorta and the atrial posterior wall, causing right atrial distortion and decreased atrial septal tautness leading to sustained caval inflow against a PFO. This distortion also results in the flap valve of the fossa ovalis moving freely, leading to a spinnaker effect, with the venous flow moving to the left, maintaining the foramen ovale wide open and allowing for persistent right-to-left shunting.^{9–12} In the upright position, there is an increased PFO aperture size and enhanced streaming of blood flow from the inferior vena cava, causing greater shunting due to the effects of gravity.^{9,13} Some patients with PFO only become symptomatic in adulthood because aortic dilation, which is a significant component in POS, is often due to longstanding hypertension and age.¹⁰

Definitive management of POS with identified interatrial communication is surgical or percutaneous closure of the shunt.³ Closure often leads to immediate improvement in aortic oxygen saturation and systemic blood saturation.¹¹ Occasionally, patients may have persistent symptoms despite PFO closure. In these circumstances, an alternative underlying cause for dyspnea, likely pulmonary, is hypothesized.¹² If symptomatic patients are left untreated, they suffer from worsening dyspnea and hypoxemia with significant effects on functionality.³

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