Platypnea-orthodeoxia syndrome in an elderly woman with a patent foramen ovale

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A case of an 85-year-old woman with gradually progressive but severe dyspnea is reported. A thorough workup disclosed platypnea-orthodeoxia syndrome associated with right-to-left shunting through a patent foramen ovale, in the presence of several other recognized risk factors, despite the absence of significant pulmonary hypertension. Successful percutaneous transcatheter closure of the intracardiac defect resulted in resolution of the syndrome.

Key Words: Dyspnea; Patent foramen ovale; Shunt

CASE PRESENTATION

An 85-year-old woman presented to the hospital with new-onset right leg weakness suggestive of a cerebrovascular event. On admission, the patient also reported gradually progressive dyspnea over the preceding 10 years, which had recently become severe. The patient reported dyspnea with minimal activities such as speaking, and further noted that her dyspnea was worse in an upright posture, and relieved by assuming a supine position. The woman's medical history included hypertension, dyslipidemia, cerebrovascular events, peptic ulcer disease, spinal stenosis and a history of a 'heart murmur' since childhood. Medications included a thienopyridine antiplatelet agent, a statin and an angiotensin-converting enzyme inhibitor.

The physical examination was notable for widely fluctuating and highly positional oxygen saturations, ranging from 97% in the supine position while breathing ambient air, to 75% in the upright position while breathing oxygen at 5 L/min by nasal cannula. Chest examination revealed minimal crepitations at the right lung base. Heart sounds included moderate (grade II/VI), nonradiating systolic murmurs at the cardiac base and apex. The jugular venous pressure appeared normal. The extremities were normal with no edema. The abdominal examination was unremarkable, with no stigmata of chronic liver disease. Neurological findings included mild weakness of the right leg.

Routine initial bloodwork was noncontributory. An echocardiogram showed early Z-axis transition in the precordial leads with an rsr' pattern in lead V₁ and left-axis deviation; no significant dysrhythmias were noted on telemetry. A chest x-ray showed unfolding of the thoracic aorta and thoracolumbar scoliosis only. A computed tomography pulmonary angiogram showed a small amount of scarring or atelectasis at only the right lung base, with no evidence of pulmonary embolism or arteriovenous malformation. A cranial computed tomography scan disclosed evidence of a remote right cerebellar infarct. Doppler ultrasound of the carotid arteries showed less than 50% stenosis of the internal carotid arteries bilaterally. When agitated saline was injected into the venous system, transcranial Doppler ultrasound showed air bubbles in the intracranial arteries, consistent with a right-to-left shunt. A transthoracic echocardiogram (TTE) in the supine position showed a patent foramen ovale (PFO) and a mobile interatrial septum almost meeting the criteria for an interatrial septal aneurysm; agitated saline contrast did not reveal any shunting through the PFO.

A transesophageal echocardiogram (TEE) was then performed, which showed the near-interatrial septal aneurysm along with a

Un syndrome de platypnée-orthodéoxie chez une femme âgée ayant une persistance du foramen ovale

Les auteurs rendent compte du cas d'une femme de 85 ans ayant une dyspnée évolutive, mais grave. Un bilan approfondi a révélé un syndrome de platypnée-orthodéoxie associé à une dérivation droite-gauche par une persistance du foramen ovale, en présence d'autres facteurs de risque reconnus et malgré l'absence d'hypertension pulmonaire. Une fermeture par transcathéter percutané de l'anomalie intracardiaque a permis la résolution du syndrome.

Figure 1 Echocardiogram demonstrating near-interatrial septal aneurysm and a prominent Eustachian valve

prominent Eustachian valve (Figure 1). Mild aortic dilation was also noted. A PFO was again evident (Figure 2). With the administration of agitated saline contrast, copious right-to-left shunting of air bubbles was seen (Figure 3).

After considering her options, and following another cryptogenic cerebrovascular event in the hospital, the patient opted for closure of her PFO with a 35 mm septal occluding device. Of note, cardiac catheterization revealed normal right-side heart pressures, with no evidence of pulmonary hypertension. A subsequent TEE showed a well-positioned device (Figure 4). At clinical follow-up, the patient's dyspnea had improved from New York Heart Association class IV to class II, and oxygen saturation was preserved at 96% to 98% in both the supine and upright positions.

DISCUSSION

Platypnea-orthodeoxia syndrome is described by its cardinal presenting symptom (platypnea: dyspnea in an upright posture, relieved by recumbency) and sign (orthodeoxia: oxygen desaturation in an upright posture). It is seen in association with a small number of clinical conditions, namely PFO and related atrial defects (eg, atrial septal

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Figure 2) Transesophageal echocardiogram without contrast demonstrating a patent foramen ovale

Figure 3) Transesophageal echocardiogram with agitated saline contrast demonstrating significant shunting through the patent foramen ovale, from the right atrium into the left atrium

aneurysm and fenestrated atrial septum), and intrapulmonary shunts (eg, arteriovenous malformation and hepatopulmonary syndrome) (1,2). In the case of PFO – which is present in up to 27% of normal adults (3) – and related interatrial defects, concomitant conditions may be required to precipitate the rare syndrome of platypnea-orthodeoxia. These associated conditions include pulmonary embolism, emphysema, pneumonectomy, pericardial disease, right ventricular remodelling, a prominent Eustachian valve, aortic dilation or elongation, and kyphoscoliosis (1,2). In our patient, platypnea-orthodeoxia syndrome was present by definition. A workup oriented toward conditions associated with this syndrome followed. Although the initial TTE with agitated saline contrast did not demonstrate shunting through the visualized PFO, this test was performed in the supine position; shunting may only become apparent in the upright posture in cases of platypnea-orthodeoxia syndrome. Furthermore, TTE is less sensitive than TEE for the diagnosis of shunting through a PFO (1). Kyphoscoliosis, aortic dilation and a prominent Eustachian valve have each been documented in cases of platypnea-orthodeoxia syndrome associated with a PFO, and were all present in our patient. The gradual

Figure 4) Transesophageal echocardiogram demonstrating good positioning of a septal closure device

natural history of both kyphoscoliosis and aortic dilation may explain the syndrome's insidious development over years in the present case.

The exact mechanistic underpinnings of platypnea-orthodeoxia syndrome are controversial and have not been fully elucidated, especially given the frequent absence of significant pulmonary hypertension at cardiac catheterization – leading some to ask “What causes water to flow uphill?” (4). Whether shunt flow truly occurs in the absence of right-to-left pressure gradients that are too small, dynamic or difficult to measure well, remains debatable (5). A brief physiological reversal in the usual left-to-right atrial pressure gradient has been observed in early diastole, and may be potentiated by concomitant lesions that increase pulmonary vascular resistance and/or decrease right ventricular compliance (eg, pulmonary embolism), to produce shunting in those with interatrial defects. Alternately or concomitantly, mechanical factors that alter the anatomical relationship between inferior vena cava blood flow into the right atrium and an existing interatrial defect, may produce local conditions that favour right-to-left shunt flow in a substream of right atrial blood, despite normal average atrial pressure differentials. According to this model, anatomical changes lead to actual physical stretching of the interatrial septum and a redirection of incoming right atrial blood toward the enlarged interatrial defect (eg, altered mediastinal anatomy following pneumonectomy). Finally, these pressure and/or mechanical flow characteristics may be further altered by physiological manoeuvres, such as sitting up, in such a way that the clinical syndrome is produced (5).

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