CASE REPORT

Platypnea-Orthodeoxia Syndrome, a Hidden Cause of Dyspnea?

ANÍBAL A. ARIAS[†, 1], PABLO F. OBERTI[MTSAC, 1], MARIANO L. FALCONI[MTSAC, 1], CARLOS ROJAS MATAS[2], DIEGO FUNES[1], ARTURO M. CAGIDE[MTSAC,1]

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Address for reprints:
Dr. Aníbal A. Arias
Servicio de Cardiología
Hospital Italiano de Buenos Aires
Gascón 450
(C1181ACH) Buenos Aires
E-mail: anibal.arias@hospitalitaliano.org.ar

ABSTRACT

Background

A patent foramen ovale (PFO) is an atrial septal defect with or without atrial septal aneurysm, associated with left-to-right shunt. It is also related with embolic stroke and platypnea-orthodeoxia syndrome. This report describes the case of a patient with platypnea-orthodeoxia syndrome, which represented a diagnostic challenge because she was admitted due to pneumonia but then was incidentally diagnosed with pulmonary thromboembolism (PTE). Given the unfavorable progression of her condition, and worsening of dyspnea with hypoxemia upon sitting, a transesophageal echocardiography (TEE) was performed, revealing a PFO with atrial septal aneurysm and marked right-to-left bubble passage occurring spontaneously (in dorsal recumbent position). After completion of antibiotic treatment, percutaneous closure of patent foramen ovale was performed with an Amplatzer device.

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Key words > Echocardiography, transesophageal - Dyspnea – Patent foramen ovale, - Endovascular procedures

Abbreviations >

Stroke (CVA)		**PFO**	Patent foramen ovale
TEE	Transesophageal echocardiography	**PTE**	Pulmonary thromboembolism

BACKGROUND

The presence of patent foramen ovale (PFO) is associated with embolic stroke and platypnea-orthodeoxia syndrome. (1-4) This uncommon syndrome is characterized by dyspnea and atrial hypoxemia induced by orthostatism, whose symptoms and oxygenation are relieved by recumbency. (5, 6) Two conditions are necessary for its development: an atrial septal defect, and a functional cardiac or extracardiac component that produces an elongation of the septum, favoring right-to-left shunting, but pulmonary hypertension is not essential. (5, 6) The diagnosis is obtained with transesophageal echocardiography (TEE). If this syndrome is suspected, some authors suggest TEE with bubble-contrast injection in both recumbent and sitting positions. (7, 8) At present, septal defect closure is recommended. This report describes the case of a patient with platypnea-orthodeoxia syndrome, which represented a true diagnostic challenge.

CASE REPORT

A 79 year-old hypertensive female, with a history of occasional palpitations had been previously hospitalized for ischemic stroke with subsequent recovery of motor function. While in hospital, atrial fibrillation was detected, and a TEE showed patent foramen ovale (PFO) and absence of thrombus. The stroke was considered of cardioembolic origin. Since then, she received enalapril 20 mg/day, carvedilol 25 mg/day, atorvastatin 20 mg/day, and acenocoumarol.
She was hospitalized 8 months later due to febrile respiratory symptoms, and was diagnosed right basal pneumonia. As she developed persistent dyspnea, a chest angiotomography scan was performed revealing filling defects in the segmental arteries of the right upper lobe consistent with pulmonary thromboembolism (PTE) (Figure 1). Clinically, no signs of shock or right ventricular involvement were evident in the transthoracic echocardiography, so she continued with anticoagulation therapy.

Hospital Italiano de Buenos Aires
[MTSAC] Full Member of the Argentine Society of Cardiology
[†] To apply as Full Member of the Argentine Society of Cardiology
[1] Department of Cardiology
[2] Hemodynamics Unit

The patient progressed with dyspnea and hypoxemia, which worsened upon sitting (platypnea and orthodeoxia). The TEE was repeated revealing PFO with atrial septal aneurysm and marked right-to-left bubble passage occurring spontaneously (in dorsal recumbent position). Dilated aortic root (4.30 cm) and Eustachian valve thickening were also observed (Figure 2; see also video on website). A right catheterization evidenced the following hemodynamic parameters: right atrial pressure, 9 mm Hg; pulmonary artery pressure, 29/15/20 mm Hg; pulmonary capillary pressure, 11 mm Hg; cardiac index, 2.7 litres/m2; and systemic vascular resistance, 1,400 dynes/sec/cm-5.

At first, a cautious approach was adopted due to the diverse coexisting causes of dyspnea. However, given the persistent hypoxemia despite resolution of pneumonia, and after 6 days of effective anticoagulation, the chest CT scan was repeated revealing non-infectious infiltrates. A catheterization was performed (Figure 3), and percutaneous closure of PFO with an Amplatzer device was decided. After this procedure, the patient made good progress, saturation was corrected, and she was discharged with follow-up.

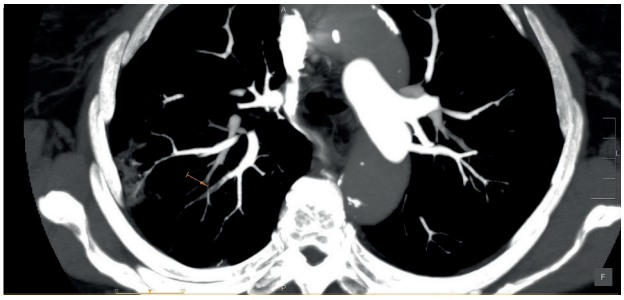

Fig. 1. Filling defects in the segmental arteries of the right upper lobe consistent with pulmonary thromboembolism.

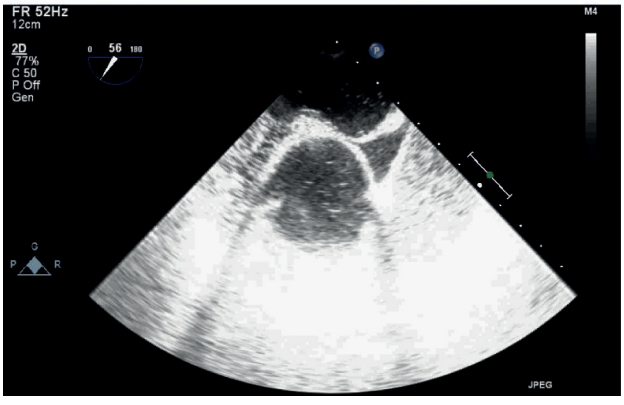

Fig. 2. Transesophageal echocardiography in a short-axis view at 56 degrees. Opacification of the right atrium and spontaneous right-to-left bubble passage (in dorsal recumbent position) are observed.

DISCUSSION

Platypnea-orthodeoxia is an uncommon syndrome characterized by dyspnea and atrial hypoxemia induced by orthostatism, whose symptoms and oxygenation are relieved by recumbency. Since Burchell et al described this syndrome over half a century ago, no more than 50 cases have been reported in the literature. (5, 6) Two conditions must coexist to cause this syndrome: on the one hand, the presence of an anatomical component in the form of an atrial septal defect (interatrial communication, patent foramen ovale, or fenestrated atrial septal aneurysm); on the other hand, the presence of a functional component that produces a deformity in the atrial septum and results in a right-to-left redirection of shunt flow following the upright position, mainly from the inferior vena cava flow. The functional component may be cardiac (pericardial effusion or constrictive pericarditis), pulmonary (emphysema, arteriovenous malformation, pneumonectomy, or amiodarone toxicity), abdominal (cirrhosis of the liver or ileus), or vascular (aortic aneurysm or elongation). (5-9)

What calls the attention of this case is that this syndrome appeared after a pneumonia, and perhaps also after PTE. Moreover, in addition to the patent foramen ovale with atrial septal aneurysm, two predisposing factors were identified to cause right-to-left shunting in the absence of pulmonary hypertension: one is the persistent Eustachian valve, and the other is the dilation and elongation of the thoracic aorta.

If this syndrome is suspected, some authors suggest TEE with bubble-contrast injection in both recumbent and sitting positions. (7, 8)

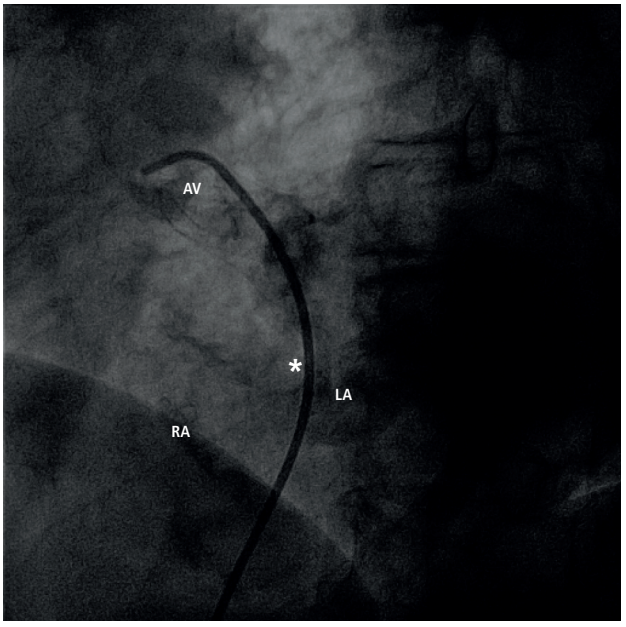

Fig. 3. Contrast passage through the patent foramen ovale (*). LA: Left atrium. RA: Right atrium. AV: Arterial vein.

Regarding treatment, the ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease state that the septal defect closure, either percutaneously or surgically, is reasonable in the presence of platypnea-orthodeoxia syndrome (Class IIa, level of evidence B). (10) While most authors are in favor of percutaneous closure, (5, 11) surgical closure is often preferred when there is an additional indication, such as aortic dilation. (7)

RESUMEN

Síndrome de platipnea-ortodesoxia, ¿una causa oculta de disnea?

El foramen oval permeable (FOP) es un defecto del septum interauricular que puede coexistir con aneurisma de éste y se asocia con pasaje de flujo de izquierda a derecha. Su presencia se ha vinculado con accidente cerebrovascular (ACV) embólico y con el síndrome de platipnea-ortodesoxia. En esta presentación se describe el caso de una paciente con síndrome de platipnea-ortodesoxia que representó un verdadero desafío diagnóstico, ya que ingresó a nuestra institución por neumonía y luego incidentalmente se diagnosticó una tromboembolia de pulmón (TEP). Dada la tórpida evolución y al evidenciarse disnea con desaturación que se acentuaba al sentarse, se realizó un ecocardiograma transesofágico (ETE) que mostró un FOP con aneurisma del septum interauricular y pasaje marcado de burbujas de derecha a izquierda en forma espontánea (en decúbito dorsal). Luego de completar tratamiento antibiótico, se procedió al cierre percutáneo del FOP con la colocación de un Amplatzer.

Palabras clave > Ecocardiografía transesofágica - Disnea - Foramen oval permeable - Procedimientos endovasculares

ConflictS of interest
None declared

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