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Acute coronary syndrome and platypnoea-orthodeoxia with thoracic and interauricular septal aneurysms

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Abstract. – OBJECTIVE: Platypnea-orthodeoxia is a rare syndrome characterized by dyspnea induced by the upright position and relieved by supine position and an arterial deoxygenation increased by the upright position which improves during recumbency. Several anatomical factors that can alter the atrial anatomy and facilitate shunting through an interatrial defect have been related to this syndrome. In many cases, this syndrome has been associated with patent foramen ovale (PFO) and right-to-left shunt. Rarely platypnea-orthodeoxia syndrome has been described associated with an aortic and with an interauricular septal aneurysm too.

CASE PRESENTATION: We present a case of platypnea-orthodeoxia syndrome in a 85-year-old woman with patent foramen ovale, interauricular septal aneurysm and ascending aortic aneurysms who was admitted for an acute coronary syndrome which could be of embolic origin and was responsible for ventricular fibrillation during the transfer to the hospital.

PFO closure was performed by percutaneous device and right coronary artery obstruction was treated by transluminal angioplasty and stenting.

Key Words:

Ascending aortic aneurysm, Patent foramen ovale, Platypnea-orthodeoxia syndrome, Coronary, Interauricular aneurysm.

Case Presentation

A 85-year-old wo man was admitted to the Emergency Department, Brugmann University Hospital, Université Libre de Bruxelle for chest pain with ventricular fibrillation during the transfer to the hospital which needed electric defibrillation by the medical team in the ambulance. Blood pressure was 130/50 mmHg with heart rate 72 b/min; no abnormal cardiac murmur was audible in all areas. Respiratory rate was normal; while breathing room air oxygen saturation was 88%. Chest radiography revealed mild vascular congestion and ascending aortic ectasia. ECG showed sinus rhythm of 85 b/min with signs of acute inferior coronary syndrome. Coronarography revealed a distal obstruction of Right Coronary Artery (RCA): transluminal angioplasty with stenting of the RCA was immediately performed with a good angiographic and clinical result (Figure 1). Transthoracic echocardiography on admission showed a dilated and hypertrophic left ventricle. An ejection fraction of 50% was measured, no pericardial effusion was observed. Right heart cavities weren't dilated and a moderate mitral and tricuspid regurgitation and a mild aortic incompetence were present. No pulmonary hypertension or intracardiac shunt were initially found. A thoracic CT scan confirmed a voluminous ascending aortic aneurysm (60 mm) (Figure 1).

After the ICU admission patient referred important and persistent episodes of dyspnea with cyanosis, chest oppression and acute desaturation in the upright position. Arterial blood taken from the patient in the upright position showed a severe hypoxemia: pH 7.48, PaO_2 44 mm Hg, $PaCO_2$ 41 mm Hg and blood saturation of 87%; after giving 10 L/min of oxygen there was no particular improvement. A thoracic CT scan with contrast excluded acute aortic dissection, pulmonary embolism, lung arteriovenous fistula and anomalous venous return.

In the supine position, blood saturation suddenly increased to 99% and cyanosis disappeared: patient did not prefer an upright position. Another transthoracic echocardiography was inconclusive.

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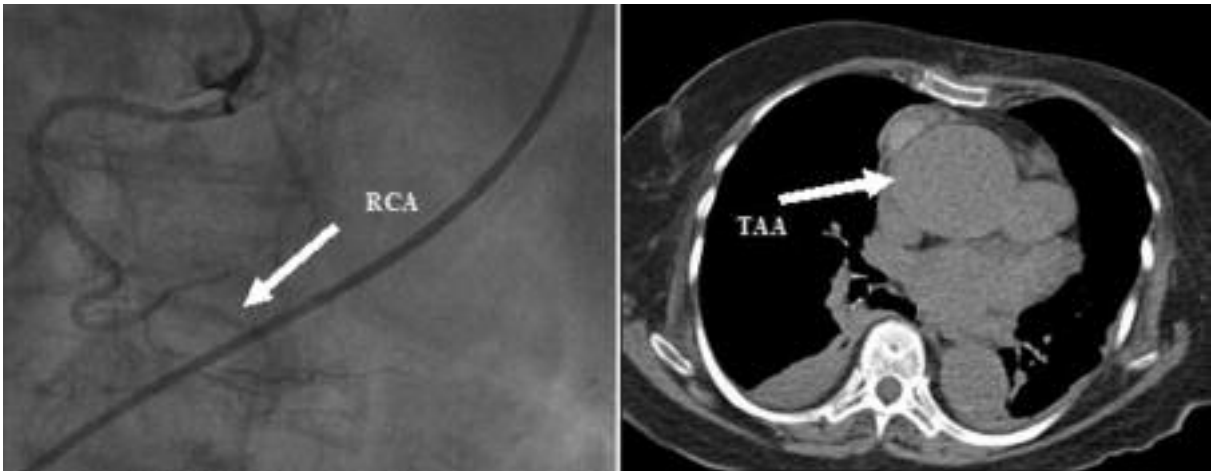

Figure 1. RCA: distal right coronary artery occlusion (on admission before PTCA). TAA: Thoracic Aortic Aneurysm (60 mm) on thoracic CT scan.

Therefore a transoesophagal echocardiography (TEE) with color Doppler was performed in supine position and revealed an interatrial communication with a right-to-left (RL) shunt through a patent foramen ovale (Figure 2) associated with an interatrial septal aneurysm (described as an interatrial tunnel: 8 mm to RA, 6 mm to LA, with an estimated 13 mm total length).

The RL intracardiac shunt was also confirmed with the injection of contrast medium microbubbles. Cerebral lesions observed on MNR examination were compatible with embolic sequels and embolic coronary occlusion could be, therefore, also possible. Surgical approach was refused by the patient. A percutaneous closure of the patent foramen ovale was performed using Amplatzer PFO Occluder 25/35 (Saint-Jude Medical, MN, USA) (Figure 2). After this procedure the patient became asymptomatic and hypoxemia induced by orthostatic conditions did not appear again. TEE also confirmed the complete closure. After a normal postprocedural course, the patient was transferred for revalidation without complications. Antiplatelet therapy (aspirin and clopidogrel) was continued afterward.

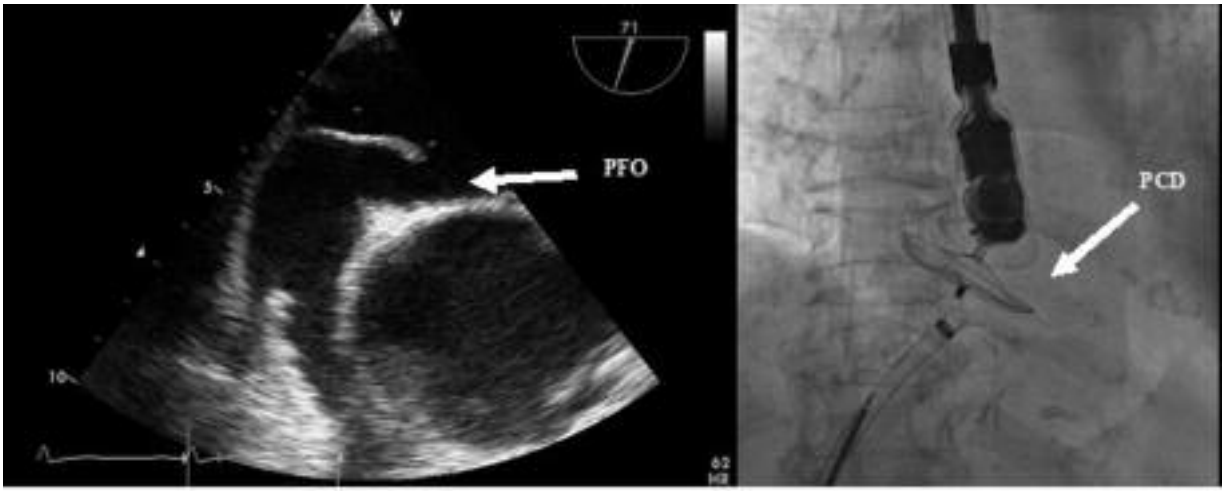

Figure 2. PFO: Patent Foramen Ovale. Intracardiac shunt diagnosed on TEE color Doppler (and confirmed by microbubbles injection). PCD: Percutaneous Closure Device (Amplatzer PFO Occluder 25/35; Saint-Jude Medical, Minnesota, USA) successfully inserted in PFO.

Comment

Platypnea-orthodeoxia syndrome is a clinically striking syndrome of postural hypoxemia with breathlessness[1,2]. Several factors could coexist for this syndrome to develop: the anatomical factor may be an interatrial communication via an atrial septal defect, PFO, or fenestrated atrial septal aneurysm; and the functional factor may be cardiac, such as constrictive pericarditis; pulmonary, such as pulmonary emphysema, recurrent pulmonary embolism, arteriovenous malformation, or previous pneumonectomy; abdominal, such as liver cirrhosis; or vascular, such as an aortic aneurysm or elongation, all of which might cause a deformity of the atrial septum[3,4].

Patent Foramen Ovale (PFO) is a rare cause of platypnea and orthodeoxia and could be responsible for paradoxal embolism as for example in coronary vessels.

Several causes of RL shunting have been hypothesized, such as redirection of shunt flow or a stretch of an interatrial hole that occurs with a postural change and a decrease in right ventricular compliance accentuating the interatrial gradient with respiration and the Valsalva maneuver[1,4]. In this case, the RL shunt was provoked by a passive postural change and by sitting, especially during inspiration, and abdominal compression. Accordingly, the compression of the right atrium by aortic dilation, the positional relationship between the inferior vena cava and the PFO, and an increase in venous return to the right atrium could account for RL shunting via the PFO. Right-to-left interatrial shunt is a rare but important cause of profound hypoxemia and could be responsible for systemic embolism as in coronary vessels.

Another mechanism may consist of the development of abnormal anatomic relationships between the vena cava and the atrial septum, increasing in the upright position, directing preferentially the venous blood flow from the inferior vena cava through a PFO or a small atrial defect into the left atrium[4,5]. It is called the "flow phenomenon" (e.g., a preferential blood flow streaming from the inferior vena cava toward the atrial septum as a part of the remnant prenatal circulatory pattern) which could be majored in presence of an interatrial septal aneurysm like in our case.

TEE is the test of choice in visualizing microbubbles in the left chambers of the heart in all of these patients. Right-sided heart angiography has been progressively replaced by contrast TTE or TEE, which was performed in the present case (only in the upright position). TTE may not clearly visualize the atrial septum because of its posterior location in the thorax. TEE is superior to TTE for the detection of shunts[5-10]. Intravenous injection of saline mixed with microbubbles of air greatly enhances the diagnosis yield of right-to-left shunts by TTE or TEE by permitting visualization of the shunt between atria. The sensitivity of this technique may be augmented by having the patient cough or perform a Valsalva manoeuvre, thereby, increasing intrathoracic pressure and right-to-left shunting. This technique could be performed in different positions, such as in the upright position. This was due to a direct blood flow from the inferior vena cava to the PFO, consequently opening it. Right-to-left shunting may thus be detected through small defects, such as a PFO, even in the setting of normal right-sided heart pressures.

Conclusions

Platypnea-orthodeoxia syndrome is recognized as a relatively uncommon condition and might be overlooked. In a typical clinical context, it is important to search for a PFO. TTE and TEE allow also elimination of other causes of cyanosis, such as tricuspid insufficiency or arterial pulmonary hypertension. When the diagnosis of PFO is made, several therapeutic options exist. Treatment by percutaneous closure is the option of choice; this technique is minimally invasive. Antiplatelet therapy is required for at least 6 months afterward.

This syndrome should be kept in mind in patients presenting acute coronary syndrome and aortic aneurysm with dyspnea and postural hypoxemia. The presence of an interatrial septal aneurysm could be considered as a factor majoring paradoxal embolism and development of severe hypoxemia.

Conflict of Interest

The Authors declare that they have no conflict of interests.

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