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Case Report

Platypnea-Orthodeoxia Syndrome Associated With Large Right Ventricular Fibroelastoma Successfully Treated by Transcatheter Approach: A Case Report

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Patent foramen ovale (PFO) is the consequence of an incomplete fusion of the septum primum and secundum after birth, leading to the persistence of a communication between the left atrium (LA) and the right atrium (RA) through a tunnel with a flap that opens, depending on the hemodynamics. PFO is present in 25% of the general population and is usually without pathologic significance.[1] PFO may be associated with platypnea-orthodeoxia syndrome (POS),[2-4] a rare condition defined by dyspnea occurring when the patient is in an upright or sitting position, and a drop of > 5% in pulse oximetry or 4 mm Hg in oxygen saturation.[2]

Case

A 71-year-old man was diagnosed, 10 years before his admission to our hospital, with a mass originating from the free wall of the right ventricle. A transjugular biopsy identified a fibroelastoma, which was not resected due to its large size and stability during follow-up. Eight years post-diagnosis, the patient received an internal mammary artery graft to the left anterior descending coronary artery. At this time, grafting the chronically occluded right coronary artery was considered technically infeasible because of the cardiac mass. Another biopsy performed at the time of coronary artery bypass grafting showed similar histopathologic results.

The patient had no symptoms for the following 2 years, after which he was admitted for progressive dyspnea. On admission, his heart rate was 90 beats per minute, his blood pressure was 130/79 mm Hg, and his respiratory rate was 23 breaths per minute. Transcutaneous oxygen measurement was at 82%, and increased to 90% with supplementary oxygen using a face mask (fraction of inspired oxygen 100%) while the patient was in the upright position, in which he presented with a slight dyspnea (Supplemental Table S1). He had no symptoms and needed less oxygen while he was lying down (2 liters per minute via nasal canula). His pulmonary and heart sounds were normal, with no audible heart murmur. We noticed nail clubbing and discrete lower-extremity edema. The electrocardiogram showed a sinus rhythm with no conduction or repolarization abnormalities. Blood tests showed an elevated hemoglobin level (171 g/L, hematocrit 50%), a normal C-reactive protein level, and an elevation of the N-terminal prohormone of brain natriuretic peptide (NT-proBNP) level at 1155 ng/L. Blood gas analysis (performed with the patient lying down) showed significant hypoxemia (partial pressure of oxygen [pO_2] 50.3 mm Hg) with slight respiratory alkalosis (pH 7.45, partial pressure of carbon dioxide [pCO_2] 33.2 mm Hg). When his blood gases were analyzed while he was in supine and sitting positions, respectively, we observed a drop of partial pressure of oxygen in the arterial blood (PaO_2) from 54 to 47 mm Hg. These values confirmed POS. Chest computed tomography confirmed the presence of a large mass originating from the free wall of the right ventricle (RV; Fig. 1), without sign of pulmonary embolism or infection. Transthoracic echocardiography revealed a normal left ventricular ejection fraction, showed the extrinsic mass compressing the RV, and confirmed a large right-to-left shunt seen on color Doppler through the PFO.

Right-heart catheterization excluded pulmonary hypertension (mean pulmonary pressure, 17 mm Hg). His mean RA pressure was slightly elevated (11 mm Hg), as were his RV pressures (Supplemental Table S1). After a multidisciplinary discussion, we decided to close the large PFO under local anesthesia, guided by intracardiac echocardiography (ICE)

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Novel Teaching Points

- A benign tumour of the RV free wall resulting in increased pressure in the RA can cause POS.
- Echocardiography plays an essential role in the diagnosis and follow-up of intracardiac shunts.

and fluoroscopy. The risk of general anesthesia was considered high in the context of the large tumour. The ICE showed the PFO and 2 small atrial septal defects (ASDs; 8 mm and 3 mm in diameter, with unidirectional shunting from right-to-left; Fig. 2) that had not been seen before. A 30-mm GORE CARDIOFORM Septal Occluder device (Gore Medical, Flagstaff, AZ) was deployed and recaptured, because it did not cover the ASDs. A 35-mm Amplatzer Cribriform Septal Occluder (Abbott Vascular, Abbott Park, IL) was able to close the PFO and cover the largest ASD. The second ASD was not covered, and given its very small size, no additional device was implanted. Immediately after the procedure, the oxygen saturation of arterial blood (SaO_2) rose from 82% to 87%. At day 2 post-procedure, his O_2 saturation without oxygen therapy was at 91%-93%, with no difference between an upright and lying position. Clinically, no sign of right-sided heart failure was present. At 12 months, the patient still does not require oxygen.

Discussion

POS was first reported in 1949 by Burchell et al.,[5] who were describing how closure of a PFO can resolve hypoxemia. The need for an additional anatomic or functional modification to explain right-to-left shunt in patients with interatrial communication is well established, given the high rate of PFO in the general population. This phenomenon can be explained by a change in blood flow in the RA in situations in which RA pressure is normal, as in POS due to an intracardiac tumour. In this condition, blood flow is redirected toward the atrial septum, causing right-to-left shunt. Such case reports have involved RA tumour. Other classical predisposing factors include kyphoscoliosis, pneumectomy, pleural effusion, diaphragmatic paralysis and ascension, hepatomegaly, ascending aorta enlargement, and aneurysm.[4] An interesting point to note is that in our patient, the RA and RV pressures were increased. The difference between this case and the cases reported in the literature can be explained by the location of the tumour—the RV fibroelastoma did not redirect the RA blood flow but caused an obstruction and thus an upstream increase of pressure, favouring the shunt. Of note, the unidirectional right-to-left shunt of the ASDs seen on ICE suggests, in our case, a contribution to POS of at least the largest ASD, in addition to the PFO. To our knowledge, this case is the first report of an intracardiac tumour causing POS due to increased RA pressure.

In our patient, we feared that PFO closure would cause an acute right-sided overload. However, as the desaturation was severe, we decided to attempt the procedure. We also took into consideration the stability of the tumour over years and its inoperability. The success of the procedure in our patient shows that PFO and atrial septal defect closure is feasible, effective, and safe when such intracardiac communications are responsible for POS.[6] Nevertheless, data on safety when RV pressure is increased are scarce.

Conclusion

POS is a rare pathology that must be suspected in the presence of a position-dependent hypoxemia, especially when it occurs when the patient is standing. Echocardiography plays a crucial role in the evaluation of cardiac involvement in POS. Our case illustrates both the role of an intracardiac shunt in the development of POS, and that shunt closure can resolve POS. This closure can be performed safely even when the RV pressure is increased, in selected cases.

Ethics Statement

Our institution does not require ethical approval for reporting individual cases. The authors confirm that a patient consent form has been obtained for this article.

Patient Consent

The authors confirm that a patient consent form has been obtained for this article, even though it is a retrospective case report using deindentified data.

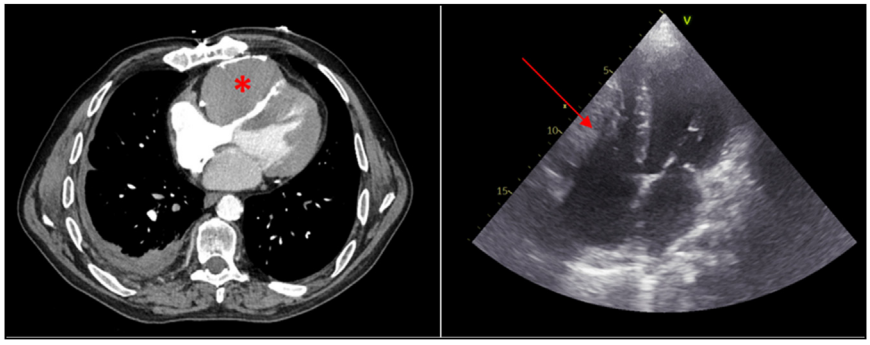

Figure 1. Thoracic computed tomography at admission (**left**) and transthoracic echocardiogram (**right**). These exams show compression of right ventricle cavity by a fibroelastoma originating from the right ventricle free wall (indicated by **red asterisk** and **red arrow**).

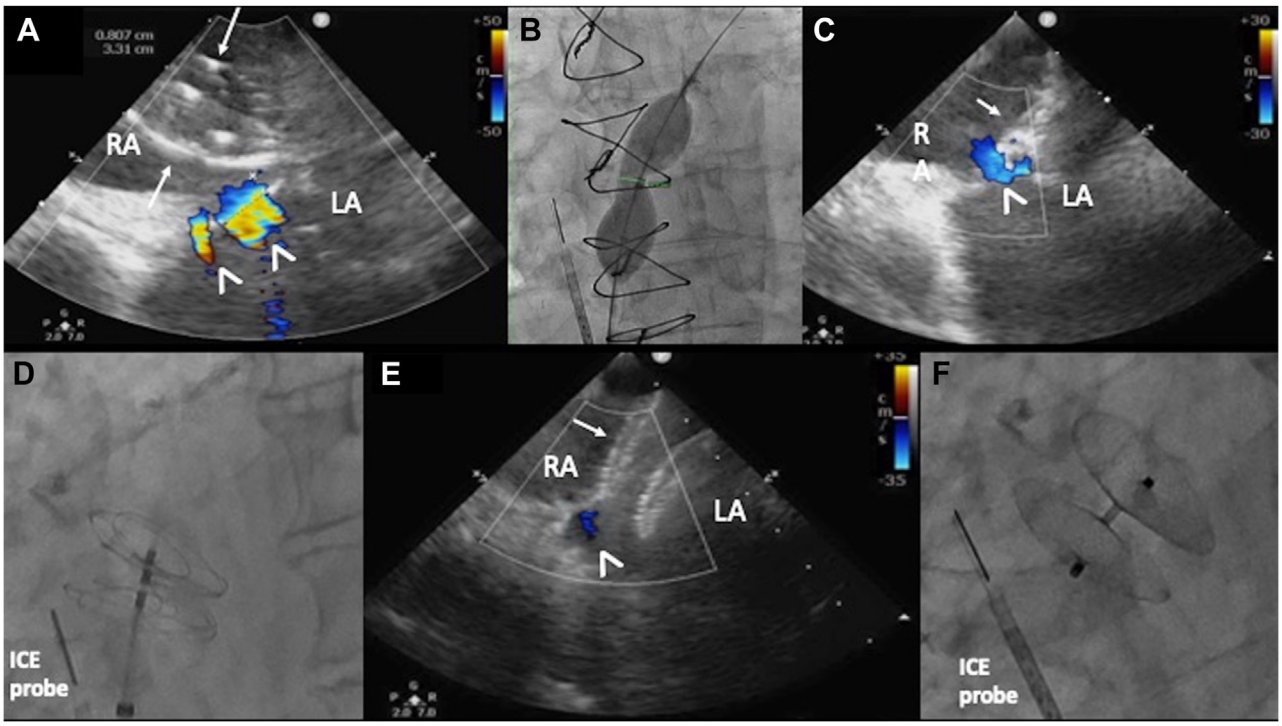

Figure 2. Intra-cardiac echocardiographic (ICE) and fluoscopic images during the procedure. (**A**) ICE showing balloon occlusion of the patent foramen ovale (PFO) (between **arrows**), **arrowheads** showing 2 atrial septal defects (8 mm and 3 mm in diameter). (**B**) Fluoroscopic images showing balloon occlusion of the PFO (8 mm in diameter). (**C**) ICE images showing the 30-mm GORE CARDIOFORM Septal Occluder (Gore Medical, Flagstaff, AZ) (**arrow**) and a residual shunt through the atrial septal defects (**arrowheads**). (**D**) Floroscopic image showing the 30-mm GORE CARDIOFORM Septal Occluder. (**E**) ICE images showing the 35-mm Cribriform Amplatzer Septal Occluder (Abbott Vascular, Abbott Park, IL) (**arrow**) and a minor residual shunt through the small atrial septal defect (**arrowhead**). The larger septal defect is covered by the device. (**F**) Fluoroscopic image showing the 35-mm Cribriform Amplatzer Septal Occluder. LA, left atrium; RA, right atrium.

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Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit *CJC Open* at https://www.cjcopen.ca/ and at https://doi.org/10.1016/j.cjco.2023.07.014.