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ANESTHESIA ORTHODEOXIA-PLATYPNEA SYNDROME ASSOCIATED WITH ATRIAL SEPTAL DEFECT: CASE REPORT

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ABSTRACT

Orthodeoxia-platypnea syndrome (POS) is a rare clinical condition, with few reports in the literature. Characterized by the presence of a pulmonary arteriovenous shunt (arteriovenous malformation and hepatopulmonary syndrome) or intracardiac right-to-left shunt (patent foramen ovale and/or related interatrial septal defects). Atrial septal defect (ASD) is the most prevalent cause of POS, representing around 87% of cases. The treatment of this syndrome varies according to its etiology. In this report, a POS situation associated with the presence of an ASD is presented, and the diagnostic strategy and approach used to treat this condition are discussed.

Keywords: Dyspnea, Interatrial communication, Hypoxia, Cardiology, Rare Syndromes.

INTRODUCTION

The Orthodeoxia-platypnea syndrome (POS) is a rare condition characterized by the presence of desaturation (orthodeoxia) and dyspnea (platypnea) in the upright position, with improvement of the symptoms in the supine position.[1] This phenomenon is caused by the presence of a pulmonary arteriovenous shunt (arteriovenous malformation and hepatopulmonary syndrome) or an intracardiac right-to-left shunt (patent foramen ovale and/or related interatrial septal defects).[2] Although its prevalence is underestimated, POS has relatively simple tests for diagnosis, such as comparing arterial blood gas measurements taken in different patient positions or evaluating the intracardiac impact of the right-to-left shunt through the use of a transesophageal echocardiogram with contrast in both the supine and upright positions.[1]

The first case of POS was described by Burchell and associates in 1949, with few cases reported since then.[3] A meta-analysis of 150 articles comprising 239 patients between 1949 and 2016 showed that interatrial communication was the most prevalent cause of POS in the studies, found in 208 patients (87%). The Patent Foramen Ovale (PFO) was the most commonly responsible for the intracardiac

shunt. In addition to PFO, defects in the atrial septum (ASD) and atrial septal aneurysm (ASA) were reported. Extracardiac causes of POS included intrapulmonary arteriovenous malformations (9.2%) and pulmonary parenchymal diseases (3.7%) (4).

Transesophageal echocardiography represents the primary diagnostic modality, providing good visualization of any defects or aneurysms that may be present in the atrial septum[4]. The definitive treatment of POS secondary to an intracardiac shunt involves closing the interatrial defect, carefully considering not only the severity of the patient's symptoms but also the patient's underlying medical conditions. The individual's ability to tolerate an invasive procedure must also be considered when making the decision. Recently, percutaneous closure has surpassed cardiac surgery in the treatment of ASD and PFO, due to reduced morbidity, mortality, and costs[5]. As for extracardiac causes, the approach is individualized for each etiology, such as pulmonary artery embolization for the treatment of symptomatic primary pulmonary arteriovenous malformation; liver transplantation for hepatopulmonary syndrome; treatment of the underlying pulmonary condition for patients with primary parenchymal lung disease, among other treatment modalities.[4]

The case report describes the percutaneous closure of an ASD as treatment for POS in a patient with low functional capacity.

CASE REPORT

A The patient is an 81-year-old female with a recent history of pulmonary embolism (PE), admitted due to desaturation, asthenia, and a non-productive cough. A chest CT angiogram performed at admission ruled out a new PE and the presence of opacities. The patient was transferred to the intensive care unit (ICU) for persistent severe hypoxemia at rest. In the ICU, it was observed that hypoxia worsened in the supine position but improved in the lateral decubitus positions. Due to suspicion of interatrial shunt, a transthoracic echocardiogram was performed, which showed an ejection fraction of 70%, septal wall: 10 mm, posterior wall: 9 mm, diastolic diameter: 45 ml, systolic diameter: 27 ml, systolic pulmonary artery pressure: 32 mmHg, left ventricle (LV) with preserved global and segmental systolic function at rest, and signs of redundancy in its leaflets with passage of microbubbles on agitated contrast, suggestive of a patent foramen ovale (PFO). The patient showed improvement in hypoxemia and was discharged from the ICU. On the ward, she experienced a fall from her own height, which resulted in the return of hypoxemia, especially when sitting on the bed, and she was again transferred to the ICU. The hypothesis of hepatorenal syndrome was raised, and an abdominal CT angiogram was performed, revealing narrowing of the inferior vena cava without flow obstruction. She underwent sequential balloon catheter angioplasty of the intrahepatic segment of the inferior vena cava. However, the patient continued to experience hypoxemia.

Subsequently, in the hemodynamics department, the patient underwent pulmonary arteriography and right heart catheterization, which revealed the absence of pulmonary arterial hypertension, with significant transvalvular gradients and right chamber overload. Easy passage of the catheter from the right atrium to the left atrium was observed at the location of the oval fossa. A suspicion of an atrial septal defect (ASD) or patent foramen ovale (PFO) was raised.

The patient was referred for percutaneous closure of the ASD. A new transesophageal echocardiogram (TEE) prior to the procedure showed the presence of an ASD in the fossa ovalis region (ostium secundum), measuring approximately 5-6 mm, with exclusive left-to-right flow. The interatrial septum was hypermobile, with an aneurysmal appearance. The left atrium was catheterized using a Multipurpose

catheter, with a 0.035 long guide, 3 mm J-tip, and rigid. The catheter was removed, leaving the guide in place, over which a 10 French (FR) sheath-introducer set was advanced. Once the sheath reached the left atrium (LA), the 0.035 guide and introducer were removed. A 28LA/28RA prosthesis was chosen, which was advanced through the sheath into the LA. The 28 disk was opened in the LA, then the set was pulled towards the interatrial septum (IAS), and its position was corrected using TEE and fluoroscopy. Once positioned parallel to the septum, the 28 disk was released in the right atrium (RA), with full deployment of the prosthesis (Figure 01). The procedure was completed, and all material was removed for hemostatic compression. During the procedure, the patient experienced temporary ST segment elevation on the monitor, and coronary angiography was performed via the left femoral artery with 6 FR, revealing chronic changes similar to a previous exam. The most likely cause of this complication was suspected coronary gas embolization. After the procedure, the patient showed significant improvement in their hypoxia, remaining stable, with no change in peripheral oxygen saturation in the supine position, while sitting, and in orthostasis.

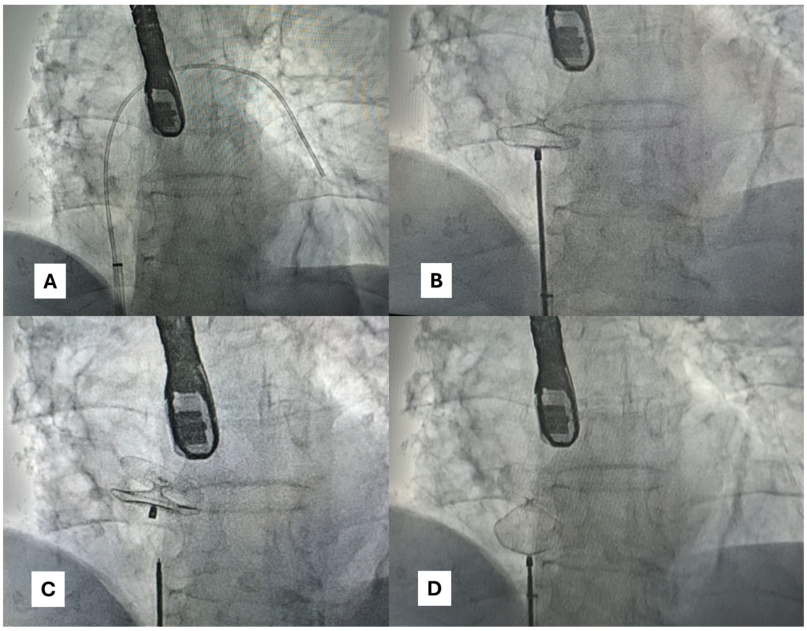

Figure 01. Images of the atrial septal defect (ASD) closure procedure. A) Crossing the ASD with the MP-multipurpose catheter, positioned in the left pulmonary vein; B) Placement of the prosthesis; C) Release of the prosthesis; D) Final appearance after the procedure.

DISCUSSION

Orthodeoxia-platypnea syndrome (POS) is a condition characterized by the presence of dyspnea associated with desaturation in the orthostatic or sitting position, with improvement of the condition in the supine position.[6] The primary mechanisms are related to intracardiac or extracardiac abnormalities, and various etiologies. In the presence of the suspected clinical picture, the etiology associated with the identified condition should be investigated; even so, POS may occur without identifiable pulmonary or cardiac disease in 13% to 47% of cases. POS secondary to

an atrial septal defect (ASD) has been most commonly reported in situations where the ventricular filling pressure is capable of generating a transient pressure gradient from the right atrium to the left in the orthostatic position, as cited in our report.[7]

The diagnostic test of choice for this syndrome is the transthoracic echocardiogram, which should be performed with agitated saline contrast, improving diagnostic performance. If the results of the transthoracic echocardiogram are inconclusive or negative, with a high index of suspicion remaining, a transesophageal echocardiogram (TEE) is recommended. At the same time, sources of intrapulmonary shunt can be investigated with agitated saline injection. Perfusion scintigraphy and pulmonary arteriography should also be considered in this context.[7]

The patent foramen ovale (PFO) is relatively prevalent in the general population (around 25-30%, depending on the individual's age). However, most people with a PFO never develop symptoms of orthodeoxia-platypnea syndrome (POS) because the left atrial pressure is 5-8 mmHg higher than the right atrial pressure, resulting in functional closure without shunt. Therefore, for POS to occur, in addition to the anatomical septal interruption between the two heart chambers, a second anatomical/functional phenomenon is necessary to direct the blood flow from the right to the left through the interatrial communication (ASD).[7]

As reported by Knapper and colleagues, percutaneous closure of the interatrial defect is the gold standard treatment for POS in the context of intracardiac shunt, showing symptomatic improvement in more than 95% of patients, with rare adverse events and a good prognosis.[5] Similarly, in our case, other therapeutic measures were undertaken without improvement of the condition. Only after the percutaneous closure of the ASD did the patient show significant reversal of hypoxia in orthostasis.

CONCLUSION

In the case now reported, percutaneous closure of the ASD was the procedure of choice for the treatment of POS, with successful outcomes, and no recurrence of the hypoxia.

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