

Síndrome de platinea-ortodeoxia en una señora mayor: un desafío diagnóstico

Platypnea-orthodeoxia syndrome in an old lady – a clinical challenge

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ABSTRACT

Introduction: Platypnea-orthodeoxia syndrome (POS) is a rare disorder characterized by dyspnoea and oxygen desaturation in the upright position which improves in the supine position.

Clinical case: 84 years old woman with hypertension and Parkinson disease. She came to the emergency department complaining of dyspnoea. At admission she was polypneic (35 bpm), spO₂ 80%, 157/108mmHg, 115bpm, afebrile, normal pulmonary auscultation and without peripheral oedema. Oxygen supplementation was started with lack of response. The patient, which at first was lying in the bed with the headboard at 45°, recovered the peripheral oxygen saturation to 96-97% when the bed was placed at 0°. Diagnostic tests were performed, excluding cardiac myocardial infarction, pulmonary embolism and pneumonia. Transthoracic echocardiogram revealed an aneurysmatic interauricular septum with transoesophageal echocardiogram showing an interauricular communication ostium secundum type with a septal aneurysm that led to a right to left shunt. The anatomical and clinical features did not allowed percutaneous or surgical closure. After a group discussion we decided for palliative care.

Discussion: POS due to a septal aneurysm with right-to-left shunt is a rare and difficult diagnose because it requires a detailed medical examination and exclusion of any other possible diseases. Besides that, it is a very rare presentation in such an old person.

Keywords: Dyspnea, Shunt, Patent Foramen Ovale

RESUMEN

Introducción: El síndrome de platipnea-ortodesoxia (SPO) es un trastorno poco común caracterizado por disnea y desaturación de oxígeno en posición erguida que mejora en posición supina.

Caso clínico: mujer de 84 años con hipertensión y enfermedad de Parkinson. Acudió al servicio de urgencias quejándose de disnea. Al ingreso se encontraba taquipneica (35cpm), spO₂ 80%, 157/108mmHg, 115bpm, apirética, auscultación pulmonar normal y sin edema periférico. La suplementación con oxígeno se inició con ausencia de respuesta. La paciente, que al principio estaba acostada en la cama con el cabecero a 45°, recuperó la saturación de oxígeno periférico al 96-97% cuando la cama se colocó a 0°. Se realizaron pruebas diagnósticas, excluyendo infarto de miocardio, embolia pulmonar y neumonía. El ecocardiograma transtorácico reveló un septo interauricular aneurismático con ecocardiograma transesofágico que mostraba comunicación interauricular tipo *ostium secundum* con aneurisma septal que conducía a un shunt de derecha a izquierda. Las características anatómicas y clínicas no permitieron el cierre percutáneo o quirúrgico. Después de una discusión en grupo, nos decidimos por los cuidados paliativos.

Discusión: El SPO por aneurisma septal con derivación de derecha a izquierda es un diagnóstico raro y difícil porque requiere un examen médico detallado y la exclusión de cualquier otra posible enfermedad. Además de eso, es una presentación muy rara en una persona tan mayor.

Palabras clave: Dyspnea, Shunt, Patent Foramen Ovale

INTRODUCTION

The *platypnea-orthodeoxia syndrome* (POS) is a clinical condition first described in 1949⁴ and characterized by dyspnoea in the upright position (*platypnea*), associated with oxygen desaturation (*orthodeoxia*); these abnormalities improve or disappear when the patient is placed in the supine position¹. This syndrome is most often due to an interatrial right-to-left shunt associated with 1) an aneurysmatic interauricular septum 2) other atrial septal defects or 3) a patent *foramen ovale*^{1,2}. It can also be due to pulmonary arteriovenous shunts or ventilation/perfusion mismatch in the lungs².

Since 1949 only a few cases were reported, which makes it a rare condition. Reviewing the scientific publications in 2015 only 228 patients were ever identified with this syndrome¹². However, it is possible that the numbers are underestimated². Clinical criteria to diagnose POS have been proposed²: 1) existence of an interatrial communication; 2) a right-to-left shunt; 3) no pulmonary hypertension or elevation of the right atrial pressure; 4) orthodeoxia (sat.O₂<90% or paO₂<60 mmHg in the upright position that normalize in the supine position); 5) platypnea; 6) treatment is based on the closure of the interatrial defect, with resolution of symptoms. So it is important to perform a blood gas analysis preferably in the upright and supine position and compare results; however the diagnosis demands a transesophageal echocardiogram⁷ where the atrial wall defect can be directly seen.

The treatment through percutaneous approach is preferred because it is effective and has a lower morbidity^{5,6}. Sometimes open heart surgery is necessary.

CLINICAL CASE

We describe the case of an 84-year-old woman, katz D, no cognitive impairment, with hypertension and Parkinson disease. She came to the emergency department (ED) complaining of sudden dyspnoea starting three days before; she denied cough or sputum emission, fever, orthopnea, nocturnal paroxysmal dyspnoea, peripheral oedema or any other complaints. At admission she was agitated, polypneic (respiratory rate of 35 breaths per minute) with ineffective breathing, peripheral oxygen saturation (spO₂) without O₂ supplementation of 80%, hemodynamic stable (blood pressure 157/108 mmHg; heart rate 115 beats per minute), feverless (36.1°C), with normal pulmonary and cardiac auscultation and without peripheral oedema. Oxygen supplementation was immediately placed with no response; even with high concentration mask (HCM) and FiO₂ of nearly 90% the spO₂ did not exceeded 86%. During the medical examination we noticed that the patient, which at first was lying in bed with the headboard at 45°, recovered the peripheral oxygen saturation to 96-97% when the bed was placed at 0°. Some complementary diagnostic tests were performed: a blood gas analysis with FiO₂ 21% revealed pH 7.46, pO₂ 50.3, pCO₂ 32.6, HCO₃-24.7, Lactate 1.2, O₂ saturation 81%; electrocardiogram with sinus tachycardia; blood test without any anomaly (including normal troponin and d-dimers); thoracic x-ray appearing a very discreet pulmonary oedema. With these results we decided to do a computed tomography pulmonary angiogram that excluded pulmonary embolism or any

sign of pulmonary parenchymal infiltrates suggestive of infection. A transthoracic echocardiogram was done in the emergency room and revealed a poorly filled right ventricle and an aneurysmatic interauricular septum (figure 1); an agitated saline bubble test was performed, revealing a right-left shunt. Considering these results, we diagnosed a likely platypnea-orthodeoxia syndrome. Then the patient was submitted to a transesophageal echocardiogram for a better description of the shunt which showed an interauricular communication ostium secundum type with an associated septal aneurysm that led to a bidirectional but predominantly right to left shunt (figure 2). The anatomical features did not allowed a percutaneous closure and the clinical condition and fragility of the patient made it impossible to perform a cardiothoracic surgery to repair the cardiac shunt. After a very intense group discussion we decided for palliative care and the patient ended up dying in a short time.

Figure 1. Transthoracic echocardiogram revealing a poorly filled right ventricle and an aneurysmatic interauricular septum.

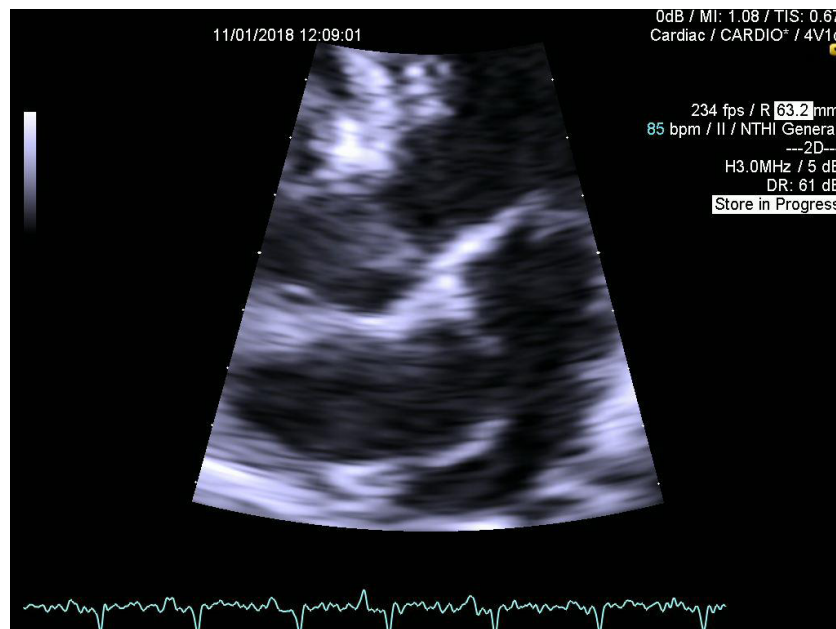
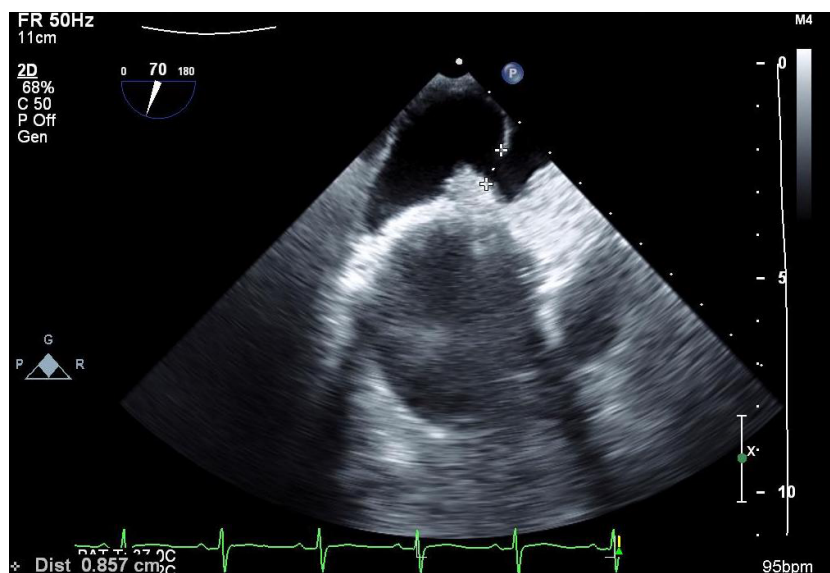


Figure 2. Transesophageal echocardiogram revealing an interauricular communication ostium secundum type with an associated septal aneurysm with bidirectional but predominantly right to left shunt.



DISCUSSION

We described a clinical case of an 84 years patient with good cognitive function and few diseases but an important functional limitation. She presented to our ED very uncomfortable, with some findings that worried us. It was possible to rapidly get the correct diagnosis through a good physical examination. In fact, after excluding other diseases like acute heart failure, myocardial infarction, pulmonary thromboembolism or pneumonia, it was the physical examination that led us to the correct diagnose. We noticed that our patient presented oxygen desaturation and dyspnoea in the upright position that did not respond to oxygen supplementation and that improved in the supine position. First a transthoracic and then a transoesophageal echocardiogram led us to this difficult diagnose: *platypnea-orthodeoxia syndrome* (POS).

As stated before, this syndrome is probably underdiagnosed² as it requires a high clinical suspicion. However simple tests like a blood gas analysis in the upright and supine positions or a good physical examination can lead us to the right diagnose⁸; the transoesophageal echocardiogram is readily available, relatively easy to perform and has a good sensibility⁷.

The platypnea seen in this condition normally is due to a right-to-left shunt through an interatrial septum defect¹ in the absence of pulmonary hypertension. Interatrial defects are relatively common; for example, *patent foramen ovale* is seen in 20-34% of the general population⁹ and it is usually asymptomatic because the communication only opens when the intrathoracic pressure rises. POS is a different situation because there is a continuous communication and deoxygenated blood goes from right to left atrium in certain body positions; this communication is due to, for example, an aneurysmatic defect. It is not well known why this shunt only appears in some body positions; in POS there is not pulmonary hypertension, but the upright position allows that some anatomic changes occur and make the blood go from right to left side¹⁰. Besides all of this, an aneurysmatic defect which only appears at the age of 84 years is very uncommon.

When in the presence of POS, it is indicated to close the interatrial communication⁹. Percutaneous closure is a safe and effective way to perform it¹¹ especially for *ostium secundum* type communications with appropriate morphology¹³. When this type of procedure is not possible or appropriate, surgical closure must be done¹³.

In the described case it was not possible to perform a percutaneous closure because the morphology was not adequate. Surgical closure was considered as an alternative treatment; however, a multidisciplinary team considered that anatomical features did not allowed this approach. The patient ended up in palliative care as this was the best option considering all the clinical data.

Platypnea-orthodeoxia syndrome (POS) is a rare syndrome especially with the clinical presentation that we described before. In fact it is not common that an 84 years old woman develops acute symptoms of POS, demonstrating that probably this was due an aneurysmatic rupture of a previously normal interatrial septum. Unfortunately it was not possible to perform any type of treatment to the patient but it was possible to ensure her a good death.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest in this work.

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This research had no funding sources.

ETHICAL ASPECTS

All participants submitted a consent form to be included in this study.

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