Platypnea orthodeoxia syndrome in a patient with patent foramen ovale and normal atrial pressure. Case report and presentation of underlying pathophysiological mechanisms

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Abstract

Platypnea-orthodeoxia syndrome is characterized by clinically significant postural hypoxia. The full spectrum of the syndrome includes intracardial and extracardial abnormalities with R to L shunt. Various concurrent underlying physiological abnormalities are usually encountered that require thorough clinical and laboratory evaluation. A high clinical suspicion in patients with unexplained dyspnea is also required to reach a firm diagnosis. We herein present a rare case of an 82-years-old patient with episodic unexplained dyspnea, patent foramen ovale with normal pulmonary pressures and we review the underlying physiologic mechanisms.

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Introduction

Platypnea-orthodeoxia (POD) is a rare medical condition characterized by positional hypoxemia (orthodeoxia) and postural dyspnea (platypnea) in the upright position that is corrected in recumbency. Clinically it is the exact opposite of orthopnea that is usually characterizing heart failure. Underlying mechanisms of POD syndrome include intrathoracic intracardial, intrathoracic extracardial and various other abnormalities, leading to right to left shunt (R->L) [1]. Fundamental prerequisites for platypnea-orthodeoxia development are two components: an anatomical component leading to an interatrial communication and a functional component producing a deformity in the atrial septum that leads to a redirection of shunt flow with the assumption of an upright posture.

POD syndrome is defined as PO_2>4mmHg and/or SpO_2> 5% drop in the upright position, compared to the supine one that is usually restored to baseline in supine position. The hypoxemia in the upright position is caused by a R->L anatomic and/or dynamic functional shunt, where venous blood is mixed with arterial and usually is not corrected or even alleviated with supplementary oxygen therapy [2]. In most intrathoracic intracardiac cases a trans-atrial R->L is diagnosed, where patent foramen ovale (PFO) constitutes the predominant diagnosis [3].

Case Report

An 82-year-old female never smoker patient was referred to the Respiratory Department, University Hospital of Patras, following a prolonged (60 days) hospitalization to a secondary care facility. The patient reported episodic dyspnea, with concurrent considerable desaturation, initially on exertion and later at rest. No profound cause was reported. Presyncopal episodes were also frequently encountered. Past medical history included transient ischemic attack, arterial hypertension, depression, and hypothyroidism. She was under treatment with acetylsalicylic acid, olmesartan, furosemide, paroxetine and levothyroxine.

Diagnostic workout on the admission in the secondary care hospital included full blood count and comprehensive metabolic panel, which were within normal limits. Computerized tomography pulmonary angiography was also normal, with no apparent pulmonary embolism, or other parenchymal or vascular abnormalities. Cardiac Doppler ultrasound (U/S) was also within normal limits, age adjusted, with an ejection fraction of 50%, normal ventricular dimensions and no indirect signs of pulmonary hypertension. Arterial blood gases (ABGs) were PO_2=88 mmHg PCO_2=35 mmHg, PH=7.41, (HCO_3^-) = 22 mmol/l on room air. During her hospitalization she developed episodic severe hypoxemia requiring high mixtures of inspired oxygen (FiO_2=60%).

At the time of the admission to our department, on top of the above-mentioned diagnostic workout, Pulmonary Function Tests were performed and were within normal limits [FVC=2.65 lt (97%), FEV_1=2.21lt (106%), FEV_1/FVC=0.84, DLCO=16 ml/mmHg/min (84%)]. Lung perfusion scintigraphy scan was normal and intra-pulmonary shunt and chronic thromboembolic disease were also excluded. Full body contrast enhanced CT did not reveal any extrathoracic R->L communications or liver abnormalities. After a thorough clinical and laboratory reevaluation of the patient a postural hypoxemia/dyspnea was noticed. ABGs on upright/supine positions where PO_2=49/79 mmHg, PCO_2=33/34 mmHg, pH=7.46/7.43 HCO_3=25/24 mmol/l SpO_2=85/95% (FiO_2=21%), respectively, leading to the provisional diagnosis of POD syndrome. A contrast enhanced, with agitated saline, cardiac transesophageal U/S with the use of the Valsalva maneuver was performed, where R->L shunt within two cardiac cycles was noticed, showing a PFO and establishing the diagnosis of the POD syndrome (Figure 1a and Video 1).

Discussion

Our report highlighted that POD is an under-recognized syndrome able to lead to fruitful clinical implications including the diagnosis of PFO. PFO is quite common in the general population and up to 27% of adults may have some intra-atrial communication [4], with no other clinical signs of postural hypoxemia and symptoms that constitute the POD syndrome. It is of great interest that a "clinically silent" PFO might explain paradoxical strokes and systemic emboli in young adults [5], so its diagnosis is of paramount significance, having in mind that even an "normal" cardiac echo might miss a PFO defect and a transesophageal echocardiography is mandatory [6] in order to confirm the diagnosis, as in our case. Common extracardiac etiologies of POD syndrome include liver cirrhosis and accompanied hepatopulmonary syndrome with arteriovenous fistulae and intrapulmonary shunt, pulmonary arterio-venous malformation, Rendu-Osler-Weber syndrome, and numerous others [2].

Of note, PFO is usually not solely accountable for the full spectrum of POD syndrome [7] , which is commonly accompanied by other abnormalities that can lead to the full syndrome, by exacerbating intracardial shunting in specific physiological conditions. Atrial and flow related physiologic driving forces may lead an anatomic, not clinically significant PFO, to a clinically significant defect. Under normal conditions the L atrial pressures are higher than the R ones, so a significant pulmonary hypertension is usually required for a clinically significant R->L communication in a patient with PFO.

Many cases with an intracardial R->L shunt can have a concurrent anatomical or functional defect, with normal R pulmonary and atrial pressures [2,3]. In these special cases, changes in the anatomic relationship between the vena cava (VC) and the atrial septum, along with physiological adaptations, may lead to R->L shunt with normal pulmonary pressures. These anatomical changes can lead to anatomical stretching and narrowing of the intra-atrial septum. At the same time blood flow from VC is directed against the thin atrial wall (with a concurrent PFO) that can cause a dynamic R->L blood flow in the upright position, the so called "flow phenomenon". In that case, the blood stream is directed towards a stretched and provisionally thinner atrial septum. Concurrent physiological adaptations can lead to altered compliance in both cardiac chambers, a condition that is frequently exacerbated by age, as might have happened in our case. In both these physiological adaptations right chambers become stiffer compared to the left ones [8]. Moreover, an early a transient reversal in the L to R pressure gradient has been recorded during in early diastole in some elderly patients that could also contribute to the R->L shunt in the absence of true pulmonary hypertension [9].

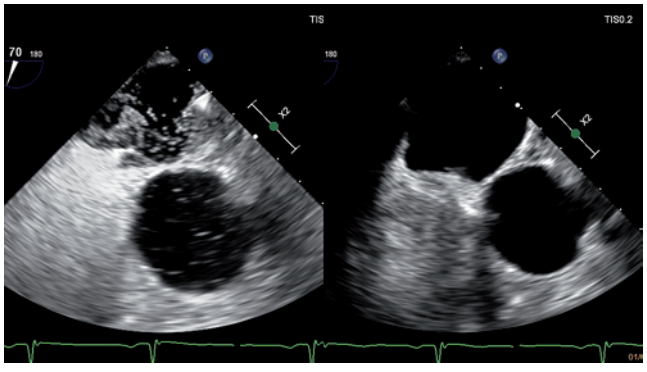

Figure 1. a) Transesophageal echocardiogram with agitated saline contrast demonstrating significant R->L shunting through the patent foramen ovale. b) Transesophageal echocardiogram demonstrating the full restoration of the septum integrity.

Clinical course

In our patient a thorough clinical and laboratory evaluation was performed and no other underlying anatomical intra or extracardial R->L communication was discovered. No pulmonary hypertension was diagnosed, and heart catheterization was also normal. Changing position from supine to upright in this patient with PFO could have led to an atrial "flow phenomenon" and altered compliance of atrial chambers mechanisms, thus explaining the dynamic, transient/episodic POD syndrome, an undoubtably rare phenomenon. An Amplatzer vascular plug was placed with good septal closing (Figure 1b and Video 2). Invasive septal restoration was successful and postural hypoxemia was alleviated and not relapsed up today (Figure 1 a,b). Clinical course and rehabilitation of the patient was uncomplicated and rapid.

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