Clinical Case Report Based Study

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Multiple Atrial Communications presenting as Platypnea-Orthodeoxia Syndrome

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ABSTRACT

We report a case of platypnea-orthodeoxia syndrome resulting from multiple inter-atrial communications with right-to-left shunting in a 73 year old woman. Our patient presented with progressive shortness of breath over the last eighteen months. She was on 10 liters of high flow oxygen at time of presentation to our hospital and was bed bound owing to severe dyspnea. Her history of chronic pulmonary emboli despite therapeutic coumadin levels and ulcerative colitis initially led to an exhaustive hepatopulmonary work-up. On clinical exam, she was noted to have normal oxygen saturation with lying down, but with sitting up or walking she developed significant hypoxia and respiratory distress. With no evidence of lung parenchymal or vascular diseases, right heart catheterization was performed and revealed normal right sided pressures. Intracardiac echocardiography confirmed a typical appearing patent foramen ovale (PFO) with atrial septal aneurysm and bidirectional shunting, and a small inferior secundum atrial septal defect (ASD). Although the ASD was congenital, aortic dilation with ageing could have altered the location of the atrial septum which facilitated laminar blood flow from the inferior vena cava to the left atrium. We proceeded to close both defects with an Amplatzer Cribiform Occluder. Her symptoms dramatically improved 48 hours following closure and she was ambulating on room air at time of discharge. One month follow-up was notable for no recurrence of symptoms. PFOs and small ASDs are generally innocuous but could cause right to left shunting even in the absence of elevated right sided pressures. Excluding other causes of platypnea-orthodeoxia, and closing the atrial communications can lead to resolution of symptoms.

Keywords: Multiple Atrial Communications, Platypnea-Orthodeoxia Syndrome.

INTRODUCTION

Platypnea-Orthodeoxia Syndrome (POS) is an uncommon clinical syndrome which is characterized by dyspnea and deoxygenation on sitting or standing from recumbent position[1]. It is caused by significant right to left shunting which is considerably increased in the sitting, standing position or walking.

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POS has been described in patients with hepatic cirrhosis, hereditary hemorrhagic telangiectasia, porto-pulmonary syndrome, pulmonary hypertension, and interatrial communications[2–4]. In general, poor right ventricular (RV) compliance leads to right to left shunting at the atrial level. Poor RV compliance and elevated RV filling pressures, in turn, may be caused by pulmonary hypertension, pulmonary stenosis, pulmonary insufficiency, RV infarct, tricuspid insufficiency, congenital (Ebstein anomaly, hypoplastic RV), and cardiomyopathies.

Inter-atrial communications are common, reported as high as 25–35% in the form of patent foramen ovale (PFO), but only a small number of people present with platypnea-orthodeoxia. Thus platypnea-orthodeoxia disease (POD) was proposed as a specific category within

POS[5], representing an interesting group of patients with POS who have an atrial communication with intracardiac right-to-left shunt, without abnormalities of RV compliance.

We report a 73 year old woman with multiple atrial communications who presented with POS with dramatic improvement in symptoms after device closure of the defects.

Case presentation:

A 73-year-old woman with history of coronary artery disease status post left circumflex artery stent four years ago, recurrent pulmonary emboli now on enoxaparin therapy, ulcerative colitis in remission was referred to our institution for progressive dyspnea on exertion that was progressively worsening over eighteen months. Prior to her presentation, she has a recent negative stress test and transesophageal echocardiogram which showed a PFO, atrial septal aneurysm, and a small inferiorly located secundum atrial septal defect (ASD) with bidirectional shunting. She was referred to our institution for further work-up of the dyspnea.

On arrival she was afebrile, her blood pressure was 113/83 mm Hg, pulse rate 80/minute, respiration rate 22, oxygenation 96% with 2 liters of nasal cannula while supine. However on sitting up, her oxygen saturation would precipitously drop to the low 80s and she would require 10 liters of oxygen to maintain oxygen saturation over 90%. Chest auscultation revealed normal rhythm and rate without significant murmurs and lung exam revealed bilateral air entry with no adventitious sounds. There was no evidence of peripheral cyanosis or digital clubbing. Of note, patient was unable to ambulate without getting significantly dyspneic. The electrocardiogram showed sinus rhythm with occasional premature atrial complexes (Figure 1A). Chest radiography revealed did not show cardiomegaly, congestion, or emphysematous changes (Figure 1B).

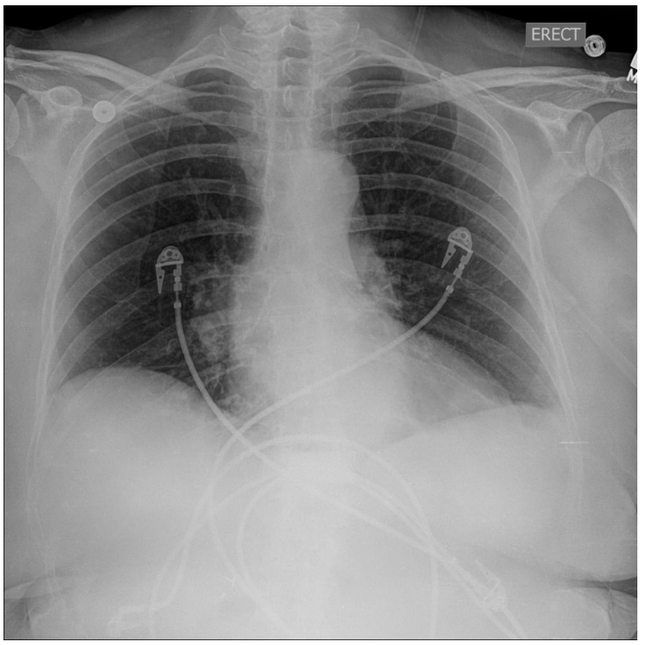

Figure 1B: Chest radiography showed prominent aortic arch with unfolding of the aorta

Since the patient's presentation was consistent with platypnea-orthodeoxia syndrome, she underwent an extensive hepato-pulmonary evaluation. Review of outside hospital computed tomography scan to rule out pulmonary emboli did not reveal pulmonary emboli. A high resolution chest coronary tomography scan did not show evidence of interstitial lung disease (Figure 1C). Her pulmonary function tests were normal. Shunt fraction study was positive with PaO2 of only 65 and SaO2 = 94% after 100% oxygen in upright position. This study confirmed the diagnosis of POS. Abdominal imaging showed no evidence of cirrhosis and her liver function tests were normal

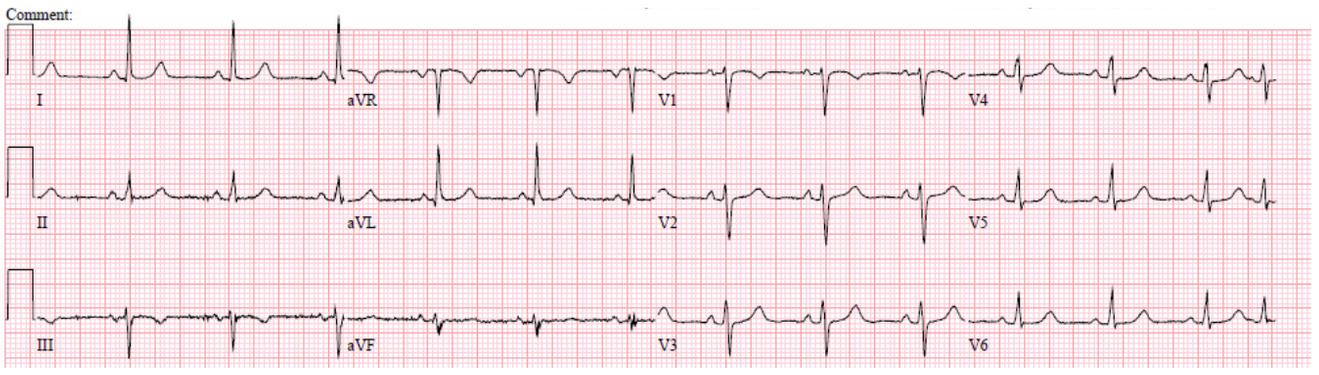

Figure 1A: Electrocardiogram showed sinus rhythm with occasional premature atrial complexes

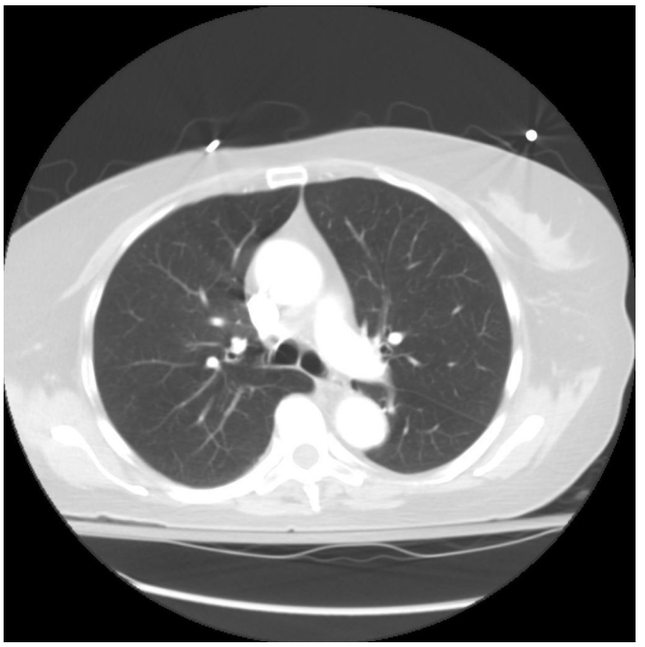

Figure 1C: High resolution chest coronary tomography scan did not show evidence of interstitial lung disease

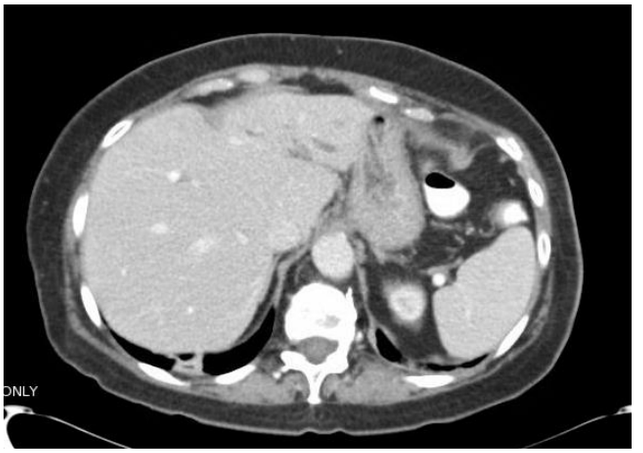

Figure 1D: Abdominal imaging showed no evidence of cirrhosis

(Figure 1D). Transthoracic echocardiogram in the supine position using a lower extremity intravenous access was positive for trivial amount of right-to-left shunting. She underwent a right heart catheterization which revealed normal right sided pressures. However pulmonary vein saturation was normal in all the pulmonary veins except a saturation of 77% in the left lower pulmonary vein. This raised the possibility of a pulmonary arterio-venous malformation (AVM). However, there was no evidence of arterio-venous malformation on review of the computed tomography scan. To ensure that AVM was not missed, injection of agitated saline in pulmonary arteries was done with intracardiac echocardiographic monitoring of left atrium. Lack of return of agitated saline on selective injections of left, right and main pulmonary arteries

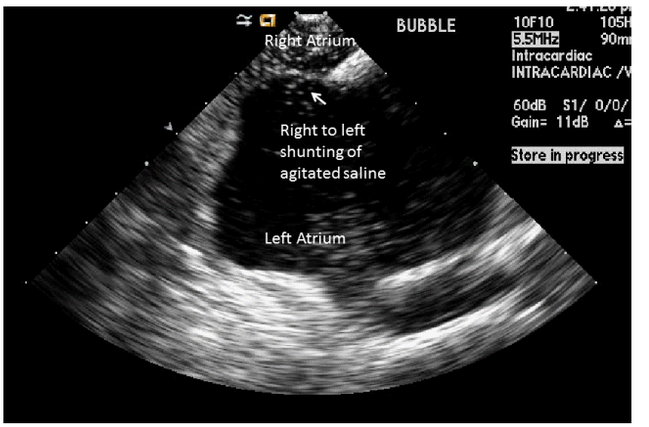

Figure 2A: Intracardiac right to left shunting with agitated saline

confirmed absence of pulmonary AVMs. Injection of agitated saline in the inferior vena cava confirmed the presence of a significant right to left shunt. (Figure 2A). A 35 mm cribriform Amplatzer Septal Occluder was deployed across the inferior ASD with successful closure of both defects.

She improved dramatically after the closure of the ASD and did not require oxygen. She was discharged four days later. She was placed on aspirin and Plavix. She was advised to take antibiotic prophylaxis for prevention of bacterial endocarditis for six months. Follow-up echocardiogram done six weeks after the closure showed no residual shunt across the ASD. (Figure 2B).

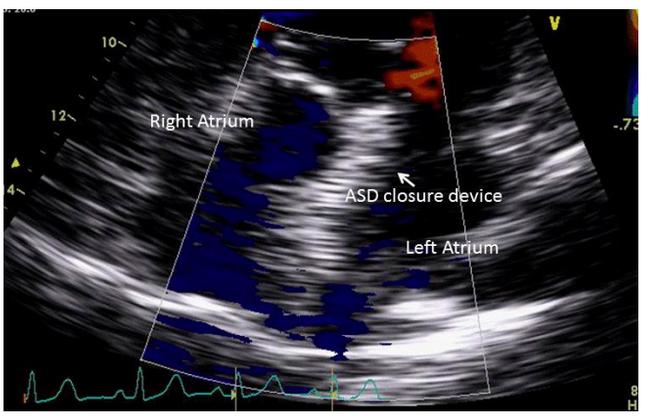

Figure 2B: Atrial septal defect (ASD) closure device with no residual shunt noted with color Doppler

Discussion

Platypnea-Orthodeoxia Syndrome (POS) is an uncommon clinical syndrome which is characterized by dyspnea and deoxygenation on sitting or standing from recumbent position. It was first described by Burchell et al in 1949 as a reflex orthostatic dyspnea occurring in positional association to pulmonary hypertension[6]. The terms "platypnea" and "orthodeoxia" were formally accepted in 1969 and 1976, respectively.

Mechanistically deoxygenation is a consequence of right to left shunting seen with pulmonary arteriovenous malformation or abnormal RV compliance driving right to left shunting through patent foramen ovale. However the most challenging pathophysiology revolves around evidence of right to left shunting and intracardiac communication but without elevated right-sided pressures, significant valvular disease, or an abnormal appearing RV, to explain a right-to-left shunt.

In the presence of an inter-atrial communication, the right-to-left shunt can be caused by a transient pressure elevation in the right chambers (pressure-related) or by the socalled 'flow' phenomenon, when there is a distortion in the anatomy that favors the passage of the blood that enters the right atrium into the interatrial communication[7]. Some authors advocate a combination of both mechanisms[8].

New-onset right to left shunting in late adulthood through an interatrial communication is thought to result from aortic dilatation. Review of literature reveals that elongated/tortuous aortic root can stretch out the inter-atrial defects and facilitate right to left shunting[9–11]. It is possible that in our patient, aortic root unfolding with aging gradually altered the shape of the atrium, leading to the distortion of the position of the atrial septum relative to the direction of blood flow from the inferior vena cava[12]. One can hypothesize that in the recumbent position the anatomical location of the atrial communication prevents venous return from entering the left atrium. However on sitting or standing, increase in venous pressure in the face of a distorted atrial septum, laminar venous flow enters the left atrium directly leading to a right to left shunt.

Although POS is a rare disease, it is important to be careful to pay attention to the dyspnea pattern with detailed history taking. POD is a disease of exclusion and hepatopulmonary causes should be carefully ruled out before hasty closure of the atrial septal defects. For our patient, the lower oxygen saturation in the left pulmonary vein could have raised the concern of a left lower lobe AVM. Theoretically left lower lobe AVM can have a gravitational increase in flow in the sitting position leading to POS[13]. However review of computed tomography and the gold standard technique of injecting the pulmonary arteries conclusively excluded this possibility. Also multiple pulmonary emboli could have caused an infraction in the left lower lobe and could possibly explain this incidental finding.

Thorough investigation with positional evaluation for right to left shunting should be undertaken. Premature closure of innocuous atrial septal defects would lead to insertion of unnecessary intracardiac hardware with no resolution of symptoms. Once the diagnosis is confirmed, closure of the atrial communication leads to remarkable improvement in symptoms[14].

Disclosures

We do not have financial interest to disclose.

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