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BILATERAL PULMONARY EMBOLI PRESENTING AS PLATYPNEA-ORTHODEOXIA

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ABSTRACT

We are reporting a case of 78 yrs old female presented with platypnea-orthodeoxia, likely caused by bilateral proximal pulmonary emboli. Despite appropriate treatment, she died of respiratory arrest. It is uncommon to have clinical evaluation of platypnea and appropriate documentation of orthodeoxia. Platypnea-orthodeoxia can be a rare but presenting feature of massive pulmonary emboli. Clinicians should have a high index of suspicion, in at risk individuals with refractory hypoxia, prompt evaluation of which may lead to improve outcome.

Key Words: Platypnea-Orthodeoxia, Pulmonary Emboli

INTRODUCTION

Platypnea-orthodeoxia (PO) is a unique syndrome of dyspnea and arterial hypoxemia accentuated on upright position and relieves with recumbency. Platypnea-orthodeoxia is under diagnosed and the exact cause of this "orthostatic cyanosis" is unclear. Most patients presented with PO have evidence of intra-atrial or intra-pulmonary anatomical and physiological shunting facilitated by the erect posture, despite normal pulmonary artery pressure. Rarely, significant ventilation perfusion mismatch secondary to massive pulmonary emboli can present as platypnea-orthodeoxia.

CASE REPORT

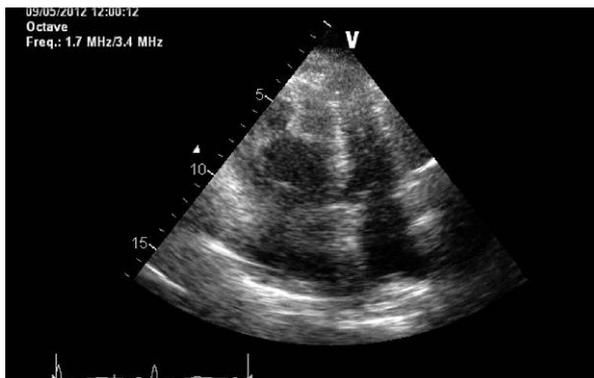
A 78 year old Caucasian female self-presented in emergency department with worsening shortness of breath for three months. She was in New York Heart Association (NYHA) class IV on admission. She had platypnea and orthodeoxia. Her oxygen saturation dropped from 90 to 85% on sitting. Clinically and radiologically she had no heart failure. ECG at the time of her admission revealed sinus tachycardia with right heart strain.

She had raised troponin I of 0.07ng/L (Beckman and Coulter UniCel DxI800 Immunoassay systems, normal 0.01-0.06ng/L). She was referred to cardiology, with a provisional diagnosis of non ST elevation myocardial infarction (NSTEMI)

and impaired left ventricle function.

Transthoracic echocardiography (TTE) revealed dilated and moderately impaired right ventricle (RV) systolic function with preserved left ventricle. She had no evidence of intra-cardiac shunt even on sitting (Figure 1).

Figure 1: Transthoracic Echocardiography Assessment of Right Ventricle

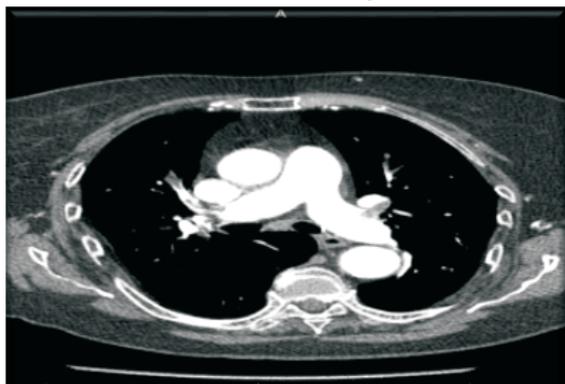


Her computed tomography pulmonary angiogram (CTPA), confirmed proximal bilateral multiple pulmonary emboli (Figure 2). She was anticoagulated with low molecular weight heparin (LMWH) and transitioned to warfarin with target INR of 2-3. She had symptomatic improvement. Despite treatment, she had a respiratory arrest. She was thrombolysed but did not respond to treatment and died 48 hours after presentation.

DISCUSSION

Platypnea-orthodeoxia (PO) is an orthostatic worsening of shortness of breath and arterial hypoxia which is relieved by supine position. It can be under recognized as presentation of platypnea is mirage by presence of multiple pathologies. Therefore, it is imperative to evaluate the pattern of dyspnea and hypoxemia to diagnose as well as treat the condition effectively.

Figure 2: CTPA Showing Bilateral Proximal Pulmonary Emboli



Platypnea-orthodeoxia was first described in a patient with an atrial septal defect (ASD) by Burchell et al. in 1949.¹ Intra-cardiac right to left shunt is the most common cause of platypnea, because of prevalence of patent foramen ovale (PFO) and atrial septal defects (ASD) in adult population.^{1,2} Hypoxia caused by intra-cardiac left to right shunt is rare.² In platypnea-orthodeoxia, intra-cardiac right to left shunt despite normal pulmonary artery pressure is facilitated by upright position. Various theories have been postulated to explain this phenomenon including conditions causing atrial septum stretch, flow directed towards PFO by eustachian valve and pathologies causing decrease in right ventricle compliance. Some of these conditions are listed in Table 1.

Contrast transoesophageal echocardiography (TOE) with postural maneuvers is the investigation of choice to demonstrate an intra-cardiac right to left shunt and exclude other structural heart disease.^{3,4}

In absence of cardiac cause of PO, investigations to exclude pulmonary arteriovenous malformation or positional worsening of V/Q mismatch should be considered. In chronic lung disease, architectural destruction with gravitational drop in pulmonary perfusion results in air trapping and decrease perfusion which leads to uncompensated V/Q mismatch resulting in PO.

Vascular pulmonary shunt can be congenital (arteriovenous

Table 1: Reported Conditions Which Can Cause or Present as Platypnea-Orthodeoxia

| |
|---|
| Intracardiac shunt, with decrease right heart compliance or/and stretching of IAS: |
| Pericardial Effusion |
| Aortic Aneurysm or elongation |
| Large hydatid cyst of liver compressing the right ventricle |
| Constrictive pericarditis |
| Eosinophilic endomyo-cardial disease |
| Associated severe skeletal deformity (kyphoscoliosis) |
| Post lobectomy or pneumonectomy |
| Idiopathic hemidiaphragm paralysis |
| Vascular Intrapulmonary right- to- left shunts |
| Pulmonary artero-venous communication |
| Hepatopulmonary syndrome(HPS) |
| Conditions causing significant V/Q mismatch |
| Pulmonary embolism |
| Chronic obstructive airway disease |
| Acute respiratory distress syndrome (ARDS) |
| Upper airway tumour |
| Miscellaneous Cause |
| Autonomic Neuropathy |

* IAS (Intra atrial septum)

malformation AVM) or acquired as in Hepato-pulmonary syndrome (HPS). Increased shunting with upright posture in these patients can present as PO. In HPS, hypoxic vasoconstriction, pleural effusion and diaphragmatic dysfunction cause further worsening of the shunt.

The likely underlying mechanism of platypnea-orthodeoxia (PO) in our patient was significant V/Q mismatch secondary to pulmonary emboli presenting as orthostatic cyanosis. Sitting had not only affected her apical lung perfusion but also dropped her RV filling pressure, resulting in pulmonary perfusion defect leading to platypnea-orthodeoxia.

PO is treated by addressing the underlying cause. In our patient, PO was likely caused by PE and she had symptomatic improvement with anticoagulation. In selected patients with intra-cardiac cause of PO, surgical correction of factors causing anatomic distortion and stretching of intra-atrial septum has been successful.⁶

CONCLUSION

In refractory hypoxia, the diagnosis of platypnea requires a high degree of suspicion. Specific questioning around positional worsening of shortness of breath and documentation of hypoxemia in upright posture is an important step in initial assessment. This also helps in formulating a differential and guides the investigatory

process towards cardiac, pulmonary and hepatic causes.

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