CASE REPORT

Platypnea orthodeoxia: a 'laid-back' case of dyspnoea

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SUMMARY

A man with end-stage liver disease who presented with shortness of breath. He exhibited clinical platypnea (worsening shortness of breath on sitting up) and orthodeoxia (oxygen desaturation on sitting up). Follow-up investigations led to the diagnosis of hepatopulmonary syndrome.

BACKGROUND

Dyspnoea exhibited with change from recumbent to seated position is platypnea. The accompanying fall in oxygen saturation is orthodeoxia. While conventionally associated with cardiac and pulmonary aetiologies, cases extrinsic to these have been reported. We describe a case of platypnea and orthodeoxia in a gentleman with end-stage liver disease and review the existing literature. Our goal is twofold: (1) to highlight the association of platypnea–orthodeoxia with extracardiopulmonary sources, specifically with hepatopulmonary syndrome, as this has implications in its management and (2) to obviate its importance for staff who may come across such patients as understanding this process can prevent detrimental positioning of the patient.

CASE PRESENTATION

We report a male with end-stage liver disease who presented with shortness of breath. He was afebrile with heart rate 77, blood pressure 140/94 mm Hg, respirations 16 and O₂ saturation 95% on 3 litre nasal cannula. Examination was significant for scattered spider angiomas. Lungs were clear to auscultation. Abdomen was distended with shifting dullness. Laboratory data revealed normal complete blood count. Metabolic panel was notable for CO₂ 28 meq/l, alkaline phosphatase 216 U/l, aspartate aminotransferase 125 U/l, alanin aminotransferase 72 U/l, total bilirubin 4.1 mg/dl, and albumin 2.9 gm/dl. Chest x-ray did not show focal infiltrates or effusion. Following admission, the patient exhibited dyspnoea which persisted despite large volume paracentesis. Symptoms appeared to worsen on moving from supine to erect position. Simultaneous pulse oximetry showed that the oxygen saturation decreased to less than 70% in the upright position and increased to more than 90% in the recumbent position.

INVESTIGATIONS

- An arterial blood gas obtained upright revealed pH 7.40, PCO₂ 39.7 mm Hg, PO₂ 60.8 mm Hg and HCO₃ 24.6 meq/l. Arterial

blood gas in supine position showed pH 7.44, PCO₂ 37.2 mm Hg, PO₂ 83.5 mm Hg and HCO₃ 24.9 meq/l. This pattern is consistent with platypnea–orthodeoxia.

- A two-dimensional echocardiogram revealed that the left atrium was of normal size. Left ventricular systolic function was preserved at 55% without any obvious regional wall motion abnormality. All valves were normal. There was no significant pericardial effusion.
- Contrast-enhanced transthoracic echocardiography with agitated saline demonstrated saline bubbles in the left heart after four beats, (figure 1A–E) indicating a significant extracardiac right-to-left shunt. This led to the diagnosis of hepatopulmonary syndrome (HPS).

DIFFERENTIAL DIAGNOSIS

Early into evaluation, we wanted to rule out the more common causes of dyspnoea in this patient population. We considered the diagnosis of pneumonia; however, there was no interval history of fevers or sputum production. Moreover, the patient did not exhibit leucocytosis on complete blood panel and his chest x-ray did not produce infiltrates. In the same way, pleural effusions were ruled out. Following admission, we felt that his shortness of breath could be secondary to impulsive ascites and mechanical compression of the diaphragm. However, it was noted by the team that his dyspnoea and real-time oxygen saturation on pulse oximetry improved on lying supine for paracentesis. This was the first clue towards further investigations that followed, including positional arterial blood gases and eventually contrast-enhanced echocardiograms. Eventually, platypnea and orthodeoxia due to an extracardiac shunt were diagnosed. Reported aetiologies to consider

1. Cardiovascular–intracardiac shunts,¹ bioprosthetic tricuspid valve stenosis,² transposition of the great arteries,³ eosinophilic endomyocardial disease,⁴ constrictive pericarditis,⁵ atrial myxoma⁶ aortic aneurysm.⁷
2. Pulmonary postpneumonectomy,⁸ amiodarone-induced pulmonary toxicity⁹ recurrent pulmonary emboli,¹⁰ adult respiratory distress syndrome.¹¹
3. Hepatic hepatopulmonary syndrome, portopulmonary hypertension.
4. Infectious hydatid cyst,¹² cytomegavirus and pneumocystis jiroveci infection.¹³

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Figure 1 (A–E) Contrast echocardiogram of our patient demonstrating the introduction of agitated saline which appears in the right side of the heart (right atrium and right ventricle with the first cardiac beat. Saline does not appear to the left side of the heart (left atrium and left ventricle) until after the third cardiac beat. This signifies the presence of an extracardiac shunt.

5. Neurological Parkinson's disease,¹⁴ diabetic autonomic neuropathy.¹⁵
6. Miscellaneous laryngeal carcinoma,¹⁶ unilateral paralysis of the diaphragm,¹⁷ blunt chest wall trauma,¹⁸ new onset ileus,¹⁹ radiation-induced bronchial stenosis,²⁰ bronchopleural fistula²¹ fat embolism,²² propafenone overdose in Ebstein anomaly.²³

OUTCOME AND FOLLOW-UP

Our patient was followed through subsequent examinations and chart reviews. Unfortunately, he passed away before a liver transplant could be performed.

DISCUSSION

The clinical signs of platypnea and orthodeoxia are underdiagnosed. The dyspnoea exhibited with a change in position from recumbency to seating is termed *Platypnea*. The accompanying fall in oxygen saturation on erect posture is termed *Orthodeoxia*. This decrease in PaO₂ should be greater than or equal to 5% or greater than 4 mm Hg when posturing from the supine to the upright position. Both the dyspnoea and desaturation improve on assuming the recumbent position. Pulmonary pressures, right heart pressures, chest x-ray and ECG tend to be normal.

It was first described by Burchell *et al* in 1949 as a reflex orthostatic dyspnoea occurring in positional association to pulmonary hypertension.²⁴ The terminologies of 'Platypnea' and 'Orthodeoxia' were formally accepted in 1969 and 1976, respectively. The true incidence has not been reported, as it is not standard practice of care to evaluate for it unless there is a high index of suspicion. The clinical existence of orthodeoxia

and platypnea is shrouded by traditional understanding of cardiopulmonary dyspnoea. The obstacles in its diagnosis are the relative rarity of the syndrome, unawareness of its presence and misleading (false-negative) diagnostic testing in the supine position. Moreover, it is often mismanaged owing to known and practised standards of care. When dealing with patients exhibiting dyspnoea, the response of healthcare staff is to seat them up. In a great majority of patients with dyspnoea, this action met with favourable improvement in comfort, breathing and oxygenation. However, this same action may be detrimental in certain cases. Consequently, these patients benefit from lying in the supine or recumbent position.

Most reported cases of platypnea–orthodeoxia deal with cardiac and pulmonary aetiologies. The more commonly reported association is with intracardiac shunts, as has often been described with patent foramen ovale and atrial septal defects. It is thought that standing erect may distort the underlying defects to allow greater deoxygenated blood to flow across anatomical defects and mix with oxygenated blood returning to the heart. These shunts are accentuated by orthostatic changes, sometimes only after valsalva manoeuvres. The pulmonary causes are secondary to intrapulmonary vascular shunts or severe ventilation perfusion abnormalities. Altman *et al*²⁵ described the existence of preferential increased basilar blood flow compared with the apical regions secondary to effects of gravity. Comparatively, alveolar pressure remains constant. When the ventilatory mechanics become impaired, alveolar pressures can become substantially elevated. This phenomenon is apparent in apical segments and is exacerbated on assumption of upright postures. Ultimately, this can result in a decrease in pulmonary artery pressures and pulmonary capillary compression resulting in

respiratory death through a decrease in blood flow. Additionally, infectious, neurological and isolated aetiologies have been described in the literature.

In our case, the symptoms were due to HPS. This is characterised by cirrhosis and/or portal hypertension and hypoxaemia from intrapulmonary vascular dilation (IVPD) and right-to-left blood shunting. Upright posture facilitates a gravitational increase in blood flow through the basal regions of both lungs, where the dilated vessels and arteriovenous communications are usually located. This results in ventilation-perfusion mismatch and clinical dyspnoea, which is often debilitating. Platypnea and orthodeoxia in the presence of cirrhosis is suggestive of HPS as the probable diagnosis. This independently worsens prognosis of cirrhosis. The diagnosis of HPS carries important implications with regard to candidacy for liver transplantation also. Liver transplantation candidacy is determined by Model for End Stage Liver Disease (MELD) scores, which in turn is based on creatine, bilirubin and international normalised ratio values. In the presence of HPS, candidates for liver transplant accumulate higher MELD scores. This is because of the higher death rates associated with HPS. Furthermore, no medical therapy has been shown to be effective. Supplemental oxygen may at best provide some symptomatic relief. As a result, the urgency with which these patients require a liver transplant is more defined. In patients with severe and refractory hypoxaemia, liver transplantation is the only therapeutic option of proven benefit. It can result in substantial improvement in gas exchange abnormalities, although the mechanism of pulmonary vasculature remodelling after transplantation is not clearly understood.

Learning points

- This is a short review of a case of platypnea and orthodeoxia: its causes, common and rare, pathophysiology and clinical importance in the way of diagnosis and appropriate management.
- The obstacles in its diagnosis are the relative rarity of the syndrome, unawareness of its presence and misleading (false-negative) diagnostic testing in the supine position.
- It is often mismanaged owing to the known and practiced standards of care. The response of healthcare staff when dealing with patients exhibiting dyspnoea is to seat them up. In a great majority of patients with dyspnoea, this action met with favourable improvement in comfort, breathing and oxygenation. However, this same action may be detrimental in certain cases. Staff must be educated in regard to supine positioning for clinical benefit and relief of dyspnoea.
- The hepatopulmonary syndrome must be considered in any patient with cirrhosis who has evidence of oxygen desaturation, even more so if this tailors to a positional pattern, as it may yield. In the presence of hepatopulmonary syndrome (HPS), candidates for liver transplant accumulate higher MELD scores. This is because of the higher death rates associated with HPS. Furthermore, no medical therapy has been shown to be effective. Supplemental oxygen may, at best, provide some symptomatic relief. As a result, the urgency with which these patients require a liver transplant is more defined.
- Therapeutically, modification of positioning may be all that is required for symptomatic relief. For other situations, management is more cause-specific.

Regardless of the specific cause and pathophysiology of the symptoms, a high index of suspicion is needed to make the diagnosis. Moreover, routine testing in the supine position can be misleading. Initially, positional pulse oximetry will demonstrate orthostatically favoured saturations; this, coupled with a detailed history and examination, is the key to diagnosis. Supplementary studies include technetium-microlabelled albumin to delineate pulmonary intravascular dilatations and/or right heart catheterisation to rule in normal pulmonary pressures. Contrast-enhanced transthoracic echocardiography after injection of agitated normal saline provides a sensitive and non-invasive tool for the detection of IVPDs and represents the diagnostic gold standard for HPS. Microbubbles are physiologically trapped and absorbed by alveoli during the first pass and should not appear in the left atrium, whereas a late appearance of saline microbubbles in the left heart chambers (visualised after >3 cardiac cycles) suggests passage through abnormally dilated lung vessels. Conversely, an immediate appearance of saline microbubbles in the left atrium raises the suspicion of an intracardiac right-to-left shunt. In our case described above, a contrast echocardiogram was utilised and revealed the appearance of agitated saline to the left side of the heart after three cardiac cycles, thereby signifying an extracardiac shunt.

Competing interests None.

Patient consent Obtained.

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