Case Report

Platypnea-orthodeoxia syndrome in idiopathic pulmonary fibrosis with *Pneumocystis jiroveci* pneumonia

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ABSTRACT

Platypnea-orthodeoxia syndrome (POS), the occurrence of dyspnea and arterial desaturation on changing from supine to sitting position, is a very rare phenomenon. Most case reports have been in association with an intra-atrial communication, and observation of this unusual physiological abnormality in pulmonary conditions has been extremely rare. We present a case of idiopathic pulmonary fibrosis where the patient developed characteristic symptoms of POS when it was complicated by *Pneumocystis jiroveci* pneumonia. This was objectively confirmed. A similar association has not been reported earlier in literature.

KEY WORDS: Idiopathic pulmonary fibrosis, platypnea-orthodeoxia syndrome, *Pneumocystis jiroveci* pneumonia

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INTRODUCTION

Platypnea-orthodeoxia syndrome (POS) is a very rare clinical syndrome characterized by dyspnea and arterial deoxygenation accompanying a change from a recumbent to sitting or standing position.[1,2] Most case reports have been in association with an intra-atrial communication such as an atrial septal defect (ASD) or a patent foramen ovale (PFO).[2] Observation of this unusual physiological abnormality in pulmonary conditions has been extremely rare, such reports being in association with emphysema, arteriovenous malformation, amiodarone toxicity,[3] rarely in interstitial lung disease,[4,5] and none in idiopathic pulmonary fibrosis (IPF). There are also two reports of POS occurring during pneumonia due to *Pneumocystis jiroveci*.[6,7] We report a case of a patient with IPF who developed the characteristic symptoms of POS when he acquired *P. jiroveci* pneumonia (PJP). The aim of the report is to describe the clinical scenario of this very rare association.

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CASE REPORT

A 48-year-old male hypertensive patient with no significant smoking history or alcohol abuse presented with complaints of cough and progressive exertional dyspnea for 3 years. Examination revealed clubbing and fine bibasilar "velcro" crepitations. The resting pulse oximetry saturation was 80%. Pulmonary function studies revealed a severe restrictive impairment with a severely reduced diffusion capacity. A high-resolution computed tomography (HRCT) of the chest showed predominantly bilateral basal subpleural interstitial thickening with honeycombing and traction bronchiectasis, a picture diagnostic of IPF [Figure 1].[8] A transthoracic echocardiography revealed normal chamber sizes, left ventricular ejection fraction of 62%, Grade I diastolic dysfunction, moderate tricuspid regurgitation, and an estimated mean pulmonary artery pressure of 50 mmHg. There was no evidence of an ASD

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or a PFO. He was advised to start oral prednisolone and was followed regularly but showed little symptomatic improvement. Therefore, it was gradually tapered off and he was given symptomatic and supportive treatment.

One year later, he presented with rapidly worsening dyspnea and cough over the past week with mucoid expectoration of about half a cup per day associated with low-grade fever. He had also noticed a worsening of breathlessness on assuming an upright position and felt relieved in a supine position. There was no pedal edema, raised jugular venous pressure, or other signs of heart failure. Examination revealed respiratory distress with cyanosis and a respiratory rate of 32/min with rapid and shallow breathing, a pulse rate of 130 min, and a normal blood pressure. The patient was afebrile and required oxygen with 50% venturi and additional nasal prongs to barely maintain a pulse oximetry saturation between 85 and 90%. Arterial blood gas analysis revealed a pH 7.39, $PaCO_2$ 43 mmHg, PaO_2 45 mmHg, and HCO^{3-} 26.1 mmol/L with normal serum electrolytes.

Laboratory investigations including hemogram, kidney biochemistry profile, and coagulation parameters were in the normal range. Liver function tests revealed normal bilirubin and alkaline phosphatase levels but elevated other liver enzymes (alanine aminotransferase 230 IU/L, aspartate aminotransferase 121 IU/L). Hepatitis panel – HbsAg, hepatitis C virus (IgM) and hepatitis A virus (IgM) – and HIV (serology) were negative. The electrocardiogram showed sinus tachycardia.

Chest radiograph [Figure 2] revealed bilateral volume reduction with dense reticulonodular opacities in all the three zones and honeycombing in the lower zones. HRCT scan of the chest [Figure 3] revealed bilateral intralobular and interlobular septal thickening and subpleural honeycombing with diffuse ground glass haze in both lower lobes. In view of his refractory hypoxemia and ground glass haze on CT scan, *P. jiroveci* infection was suspected and subsequently confirmed by Gomori methenamine silver stain done on sputum. Sputum was negative for acid-fast bacilli on two occasions. No other pathogen was isolated from sputum cultures. After a detailed discussion on the likely outcomes, the family opted against mechanical ventilation. Treatment was started with co-trimoxazole.

Due to a definite postural association of symptoms, cardiorespiratory parameters were monitored in the supine position, 1 min after assuming sitting position, and then again after returning to the supine position [Table 1]. Oxygen administration was continued as above. Breathlessness was rated using the modified Borg scale. Assumption of sitting position resulted in tachycardia, tachypnea, and substantial desaturation associated with marked increase in breathlessness. Resumption of the supine position led to recovery toward baseline.

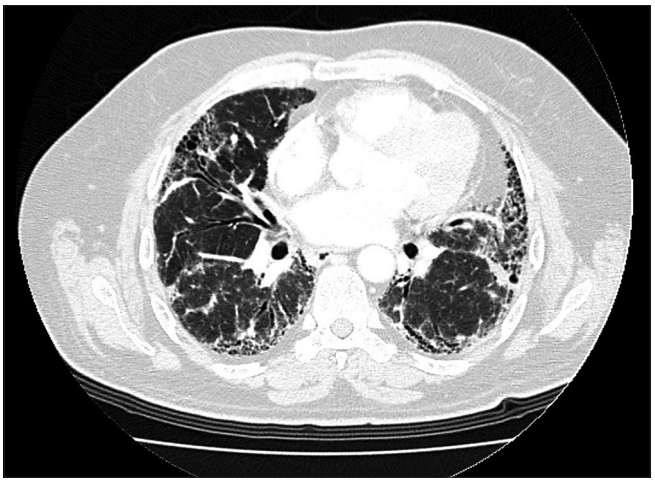

Figure 1: High-resolution computed tomography of the chest on initial presentation showing bilateral subpleural interstitial thickening with honeycombing and traction bronchiectasis, diagnostic of idiopathic pulmonary fibrosis

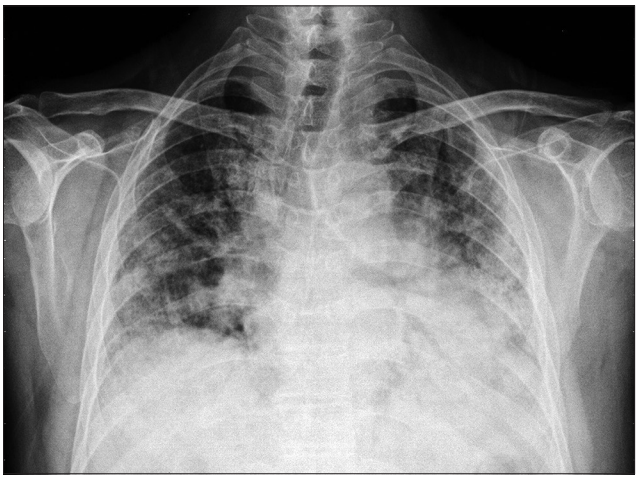

Figure 2: Chest radiograph (posteroanterior view) showing bilaterally dense reticulonodular shadows and ill-defined heart borders

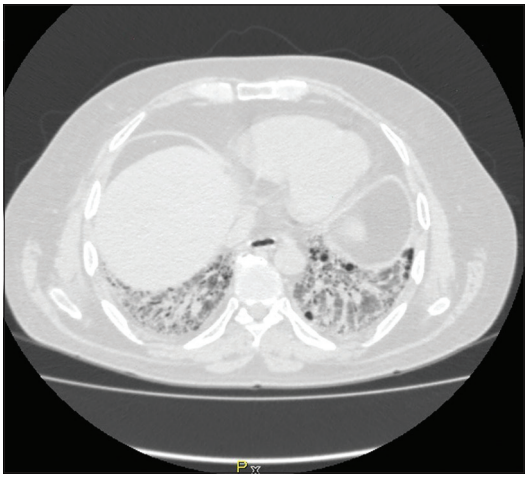

Figure 3: High-resolution computed tomography of the chest (lower lobe cuts) showing bilateral intralobular and interlobular septal thickening with subpleural honeycombing and diffuse ground glass haze during the episode of *Pneumocystis jiroveci* pneumonia

Table 1: Cardiorespiratory monitoring in different positions

Cardiorespiratory Parameters	Baseline (Supine)	Sitting Position	Resuming Supine Position
Pulse (bpm)	128	180	150
Blood Pressure (mmHg)	130/80	120/78	110/78
Respiratory rate/min	31	46	36
Borg Score (0-10)	3	10	5
Pulse oximetry, SpO_2 (%)	92	78 to 84 after 1 minute	92 (after 8 min)

A final diagnosis of IPF with hypertension with PJP with Type I respiratory failure with POS was made. The patient succumbed to his illness after a week.

DISCUSSION

Platypnea implies breathlessness perceived in an upright posture and relieved on assuming a supine position. The drop in saturation or the partial pressure of arterial oxygen accompanying such posture change is known as orthodeoxia. Although this phenomenon was first reported by Burchell *et al.*,[9] the term "platypnea" was first coined by Altman and Robin[10] to describe this phenomenon in patients with severe pulmonary emphysema. Its development requires the presence of an anatomical intra-atrial communications such as an ASD or a PFO[2] and a functional component deforming the atrial septum and resulting in a redirected right-to-left shunt on assumption of an upright posture.[1] The proposed mechanism is that in the presence of an anatomical defect, assuming an upright posture may stretch the septum thereby augmenting the venous flow from right to left even when not associated with increased right intracardiac pressure.[1] Further, it has been proposed that although approximately 25% of the general population is born with a PFO, POS, if it develops, is observed only during adulthood due to an acquired modification of the right atrial chamber anatomy.

However, POS may occur even without an obvious intracardiac shunt though it is very unusual and extremely rare in pulmonary diseases. Patients in whom this phenomenon has been documented in case reports had advanced emphysema, postpneumonectomy state,[3] amiodarone toxicity,[11] acute respiratory distress syndrome,[12] and recurrent pulmonary embolism.[13] Two cases of POS have been reported in patients with interstitial lung disease[4,5] and in two cases of pneumonia caused by opportunistic organisms such as *P. jiroveci* and cytomegalovirus.[6,7] A review of the literature revealed that POS has never been reported in a case of IPF or IPF complicated by PJP. The present case is the first to report such an association.

The true mechanism underlying the occurrence of POS in pulmonary conditions is not known. As it is extremely unusual even in conditions where it has been reported, it remains a curiosity. It has been attributed to an exaggerated ventilation-perfusion mismatch on assuming an upright posture due to gravity-dependent shift in perfusion to the lung bases.[3,5,6,12] The shunt in pulmonary conditions is thus intrapulmonary made worse with positional change to upright position.

As platypnea and orthodeoxia are not features of IPF, and also because the patient did not have these complaints earlier, it is likely that PJP was the triggering event for POS. IPF characteristically involves lower more than the upper lobes. We postulate that infection further worsened the ventilation-perfusion mismatch at lung bases converting these areas into virtual right-to-left shunts on assumption of an upright posture with gravity-dependent blood flow increase to basal areas. A return to supine position redistributed blood to the relatively better ventilated upper zones, thus reducing the right-to-left shunt and improved oxygenation. This phenomenon was objectively demonstrated in our patient. Moving to sitting position led to tachycardia, tachypnea, and substantial desaturation and marked increase in breathlessness that returned toward baseline on resumption of the supine position.

Bronchoalveolar lavage is the most common invasive procedure used to diagnose PJP because of its high sensitivity. However, the severe hypoxemia in the present case as well as a refusal of mechanical ventilation precluded bronchoscopy. Maskell *et al.*[14] have shown that oral glucocorticoid therapy is an independent risk factor for colonization with *P. jiroveci*. Detection in expectorated sputum in our patient can therefore only be taken to suggest a presumptive diagnosis. However, whether colonization by *P. jiroveci* leads to PCP directly is not established. While an acute exacerbation of IPF would also cause refractory hypoxemia with severe ventilation-perfusion mismatching and HRCT features noted in the present case, the presence of fever pointed to an infectious etiology. No other pathogen was isolated. The absence of orthopnea and clinical signs of heart failure left PJP as the only likely pathology.

Recognition of platypnea and orthodeoxia has implications for nursing of the patient. In contrast to the usual positioning of the breathless patient in the semi-recumbent position, a supine position should be the optimal nursing position for these patients if POS is observed.

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Conflicts of interest
There are no conflicts of interest.

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