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Reversible platypnea-orthodeoxia syndrome induced by rapidly progressive interstitial pneumonia in a patient with polymyositis

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Abstract

We report a case of platypnea-orthodeoxia that developed in a 64-year-old Japanese woman during an episode of rapidly progressive interstitial pneumonia with polymyositis. Pulmonary infiltrates were predominant in the bilateral lower lobes. The patient was treated successfully with early administration of immunosuppressive therapies and polymyxin B-immobilized fiber column-direct hemoperfusion, and her platypnea-orthodeoxia improved with resolution of the underlying parenchymal lung disease. Reports of platypnea-orthodeoxia syndrome with interstitial pneumonia are extremely rare. The recognition that platypnea-orthodeoxia syndrome may occur in multiple disease states, including interstitial pneumonia, is crucial to the understanding of this perplexing disorder.

Introduction

Platypnea and orthodeoxia are terms used to describe the rare phenomenon of dyspnea and hypoxemia accentuated by an upright posture and relieved by recumbency [1]. We report these features in a patient with rapidly progressive interstitial pneumonia with polymyositis.

Case Report

The patient was a 64-year-old woman who complained of fever and dyspnea and was diagnosed as having pneumonia at another hospital. The patient deteriorated with administration of antibiotics over 10 days. Afterwards, she experienced rapidly progressive extension of pulmonary infiltration and hypoxemia, and she was transferred to our hospital. Chest X-ray and computed tomography scan revealed consolidations predominantly in the lower lobes. Physical examination revealed femoral muscle atrophy and weakness, and auscultation of the chest identified audible fine crackles. Elevated levels of C-reactive protein of 13.9 mg/dL, KL-6 of 1069 U/mL, and surfactant protein-D of 355 ng/mL were also found. Moreover, anti-threonyl-tRNA synthetase (anti-PL-7) antibody in aminoacyl-tRNA synthetase autoantibody was positive, and electromyography showed abnormalities in the electrical activity of muscles as suspected with inflammation of the muscles. We then diagnosed her as having rapidly progressive interstitial pneumonia with polymyositis. She received noninvasive positive pressure ventilation for severe hypoxemia on admission. Her respiratory distress was refractory to steroid pulse therapy and cyclosporine, and intubation and mechanical ventilation were necessary on the 8th hospital day. Additional therapy, which included intravenous cyclophosphamide pulse (IVCY) therapy and polymyxin B-immobilized fiber column-direct hemoperfusion (PMX-DHP), was administered, and her condition improved

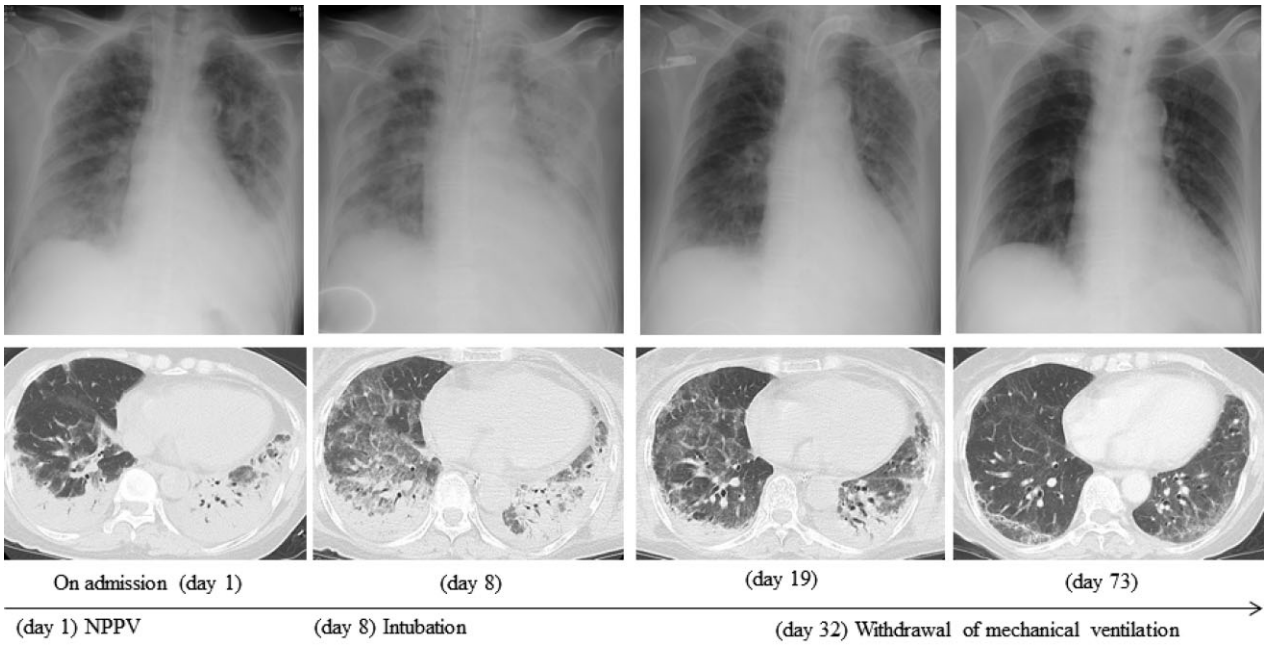

Figure 1. Clinical course as depicted by radiological imaging. Chest X-ray and computed tomographic images showed bilateral consolidations predominantly in the lower lobes at initial presentation (day 1). Deterioration was observed radiologically on hospital day 8, and intubation and mechanical ventilation were performed. Afterwards, the lesions had improved slightly by day 19, and mechanical ventilation was withdrawal on hospital day 32. Radiological improvement with no hypoxemia in the sitting position was observed on hospital day 73.

Table 1. Serial arterial blood gas results.

Hospital day	Day 1	Day 6		Day 8	Day 19		Day 59		Day 73	
Position	Supine	Supine	Seated	Supine	Supine	Seated	Supine	Seated	Supine	Seated
FiO_2	0.6	0.8	0.8	1.0	0.4	0.4	0.21	0.21	0.21	0.21
pH	7.46	7.48	7.49	7.47	7.47	7.51	7.45	7.46	7.41	7.43
PaO_2 (mmHg)	87.7	90.8 >	62.9	68.3	92.7 >	72.4	80.5 >	67.5	78.8 <	82.4
$PaCO_2$ (mmHg)	39.3	43.2	41.4	45.8	47.4	43.8	37.4	39.8	37.6	36.3

FiO_2, fraction of inspired oxygen; $PaCO_2$, partial pressure of carbon dioxide; PaO_2, partial pressure of oxygen.

slightly. Mechanical ventilation was withdrawn on the 32nd hospital day (Fig. 1).

On the 6th hospital day, we noticed that she experienced dyspnea and hypoxemia when in a sitting position. We suspected platypnea-orthodeoxia syndrome (POS) because there was no evidence of liver disease, hepatopulmonary syndrome, or pulmonary arteriovenous malformations. On the 54th hospital day, transesophageal echocardiography was performed in the prone and sitting positions. Microbubble opacification of the left atrium following intravenous administration of normal saline was not detected after either two or six cycles, thus excluding the presence of intracardiac or intrapulmonary shunts (Fig. 2). Pulmonary perfusion imaging with ^{99m}Tc-macroaggregated albumin was also performed, and the result did not suggest the presence of a right-to-left shunt. Radiological improvement was observed on the 73rd hospital day (Fig. 1), and she experienced no hypoxemia in the sitting position (Table 1). Therefore, we diagnosed POS, which resulted from the pulmonary parenchymal ventilation/perfusion mismatch caused by the interstitial pneumonia.

Discussion

We describe a case of polymyositis complicated by rapidly progressive interstitial pneumonia. The patient was treated with early administration of immunosuppressive therapies including steroid pulse therapy and cyclosporine, which resulted in deterioration on radiological imaging and respiratory failure that required mechanical ventilation. Therefore, although we did not investigate the lung tissue pathologically in our patient, we thought that her

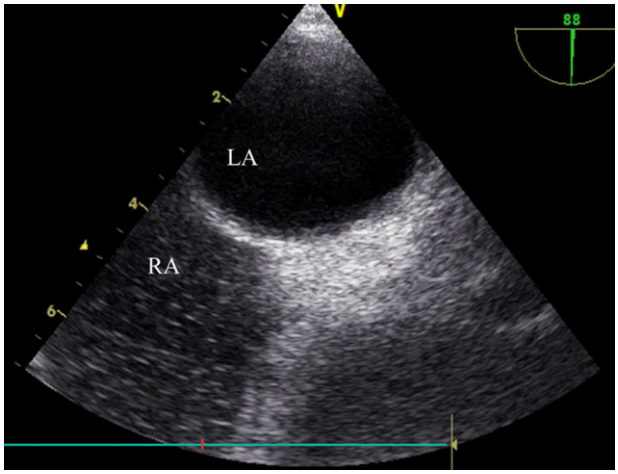

Figure 2. Image from the transesophageal echocardiographic study conducted on the 54th hospital day. Transesophageal echocardiography was performed in the prone and sitting positions. Microbubble opacification after intravenously administered normal saline was not detected in the left atrium after either two or six cycles. LA, left atrium; RA, right atrium.

respiratory failure showed the presence of a diffuse alveolar damage pattern. Recent reports have suggested that PMX-DHP might improve oxygenation in patients with acute lung injury/acute respiratory distress syndrome associated with fulminant interstitial pneumonia or rapidly progressive interstitial pneumonia with polymyositis/dermatomyositis [2]. Moreover, early intervention with aggressive combination therapy including IVCY and cyclosporine might be life saving as a treatment for rapidly progressive interstitial pneumonia polymyositis/dermatomyositis [3]. Treatment with these immunosuppressive therapies and PMX-DHP was successful in our patient.

POS is a rare clinical phenomenon that is associated with normal oxygen saturation in the supine position (platypnea) and arterial oxygen desaturation in an upright position (orthodeoxia). The exact underlying mechanisms leading to this unusual condition are not fully understood but most probably derive from one of the following three mechanisms: intracardiac shunting, pulmonary vascular shunting, or ventilation/perfusion mismatch or a combination of these mechanisms [4].

In our patient, anatomic shunts were sought using both microbubble injection during transesophageal echocardiography and pulmonary perfusion imaging with ^{99m}Tc-macroaggregated albumin, but none could be identified. In addition, the POS in our patient improved spontaneously over a 2-month period along with radiological improvement of the consolidation located predominantly in the bilateral lower lobes. Therefore, we diagnosed POS because of pulmonary parenchymal ventilation/perfusion mismatch caused by the interstitial pneumonia. Parenchymal lung diseases, such as emphysema, can also be present occasionally with POS [4]. When the patient stands upright, right ventricular preload diminishes, resulting in decreased output to the pulmonary arteries such that alveolar pressures exceed arterial and venous pressures. Moreover, gravity promotes preferential blood flow to the basal areas of the lung, and apical areas tend to act as dead space, which contributes to an increase in ventilation/perfusion mismatch and increased dyspnea [4]. However, reports of cases of POS in patients with interstitial pneumonia are extremely rare [5–7]. Tenholder et al. suggested that in severe fibrosis, the air spaces may be so far separated from the pulmonary vasculature as to seriously impair diffusion of oxygen [5]. The gravitational effect of the upright posture presumably aggravates this problem by increasing blood flow through this physiological shunt. Likely, however, other factors are involved because other patients with apparently similar basal infiltration do not manifest this phenomenon. Moreover, it has been reported that this phenomenon remains unclear for a failure of the hypoxic pulmonary vasoconstriction in interstitial pneumonia [5]. The POS in two of these reported cases was not reversible [5, 6]. To the best our knowledge, only one case of reversible POS in interstitial pneumonia has been previously reported [7]. Therefore, further analysis of ventilation/perfusion mismatch relationships in interstitial pneumonia may help to define the mechanisms for POS.

In conclusion, we describe a case of reversible POS induced by rapidly progressive interstitial pneumonia with polymyositis. POS is a rare and usually underestimated syndrome. Intracardiac shunts and anatomic pulmonary vascular shunts are the most common etiologic associations. However, severe ventilation/perfusion mismatch should also be considered as a probable explanation. The recognition that POS may occur in multiple disease states, including interstitial pneumonia, is crucial to the understanding of this perplexing disorder.

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Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

References

1. Robin ED, Laman D, Horn BR, et al. 1976. Platypnea related to orthodeoxia caused by true vascular lung shunts. N. Engl. J. Med. 22:941–943.
2. Ichiyasu H, Horio Y, Tsumura S, et al. 2014. Favorable outcome with hemoperfusion of polymyxin B-immobilized fiber column for rapidly progressive interstitial pneumonia associated with clinically amyopathic dermatomyositis: report of three cases. Mod. Rheumatol. 24:361–365.
3. Suzuki A, Shoji N, Kikuchi E, et al. 2013. Successful combination therapy with corticosteroids, biweekly intravenous pulse cyclophosphamide and cyclosporin A for acute interstitial pneumonia in patients with dermatomyositis: report of three cases. Jpn. J. Clin. Immunol. 36:122–128.
4. Rodrigues P, Palma P, and Sousa-Pereira L. 2012. Platypnea-orthodeoxia syndrome in review: defining a new disease? Cardiology 123:15–23.
5. Tenholder MF, Russell MD, Knight E, et al. 1987. Orthodeoxia: a new finding in interstitial fibrosis. Am. Rev. Respir. Dis. 136:170–173.
6. Bourke SJ, Munro NC, White JE, et al. 1995. Platypnoea-orthodeoxia in cryptogenic fibrosing alveolitis. Respir. Med. 89:387–389.
7. Naccache JM, Faure O, Loire R, et al. 2000. Severe hypoxemia with orthodeoxia due to right to left shunt in idiopathic bronchiolitis obliterans organizing pneumonia. Rev. Mal. Respir. 17:113–116, (in French).