

Platypnea-orthodeoxia Syndrome Induced by an Infected Giant Hepatic Cyst

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Abstract

An 83-year-old man was admitted with a chief complaint of exacerbation of dyspnea. His blood oxygen saturation was 90% in the recumbent position despite oxygen therapy, and it dropped to less than 80% when the patient attempted to sit upright. A computed tomography scan revealed a giant hepatic cyst compressing the right atrium and the inferior vena cava. After percutaneous drainage, the oxygen saturation improved and did not change with alteration of the patient's positions from recumbent to sitting or standing. This case report describes a patient with the platypnea-orthodeoxia syndrome due to a giant hepatic cyst successfully managed by percutaneous drainage.

Key words: platypnea-orthodeoxia syndrome, giant hepatic cyst, ventilation-perfusion mismatch

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Introduction

Platypnea-orthodeoxia (PO) is an uncommon syndrome characterized by dyspnea and hypoxemia accompanying a change from the recumbent position to sitting or standing (1). Although the mechanism for the occurrence of hypoxemia in this syndrome is not well understood, most reported cases of PO syndrome are associated with intracardiac shunts, intrapulmonary vascular shunts or severe ventilation/perfusion (V/Q) mismatch (2-5). We herein report a case of severe platypnea and orthodeoxia with evidence of severe V/Q mismatch secondary to a compressive effect on the right ventricle, inferior vena cava (IVC) and diaphragm by a giant hepatic cyst.

Case Report

An 83-year-old man was admitted to our hospital due to the exacerbation of dyspnea without any other respiratory symptoms. He denied a history of chest pains, palpitations, dizziness and hematochezia. Over the five years preceding

this admission, he had experienced recurrent episodes of oxygen desaturation, with oxygen saturation levels less than 85% in the upright position that improved to more than 90% in the recumbent position.

The patient had a history of a colonic arteriovenous malformation, hypertension, hyperuricemia and a hemorrhagic gastric ulcer. He took febuxostat 10 mg and lansoprazole 15 mg both once daily. He was a non-smoker and consumed no alcohol. There was no history of any drug or food allergies. His family history and social history were otherwise non-contributory.

On a physical examination, he appeared mildly unwell. He was afebrile with a tachycardia of 107 beats per minute, a blood pressure of 159/99 mmHg, a body temperature of 37°C and tachypnea with a respiratory rate of 24 breaths per minute in the recumbent position. Pulse oximetry on continuous oxygen therapy at a rate of 10 L/min demonstrated an oxygen saturation above 90% in the recumbent position, which dropped to less than 80% when sitting upright. No conjunctival pallor, icterus, cyanosis, digital clubbing or spider nevi were detectable on the physical examination. A cardiovascular examination revealed a normal jugular venous

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Table 1. Laboratory Data on Admission Day.

Complete Blood Count		Normal range
WBC	42 ×10 ² /μL	30 - 97 ×10 ² /μL
Neut	76.9 %	36.6-79.9 %
Hb	11.5 g/dL	13.1-17.6 g/dL
Ht	35.0 %	38.1-50.8 %
MCV	90.7 fL	84.6-100.6 fL
Plt	22.6 ×10 ⁴ /μL	12.4-30.5 ×10 ⁴ /μL
PT-INR	0.9 %	77.8-130.0 %
APTT	25.7 Sec	23.6-31.3 Sec
D-dimer	14.9 μg/mL	0.0-1.0 μg/mL
Biochemistry		Normal range
CK	7 IU/L	52-192 IU/L
T.bil	1.4 mg/dL	0.1-1.2 mg/dL
AST	23 IU/L	12-35 IU/L
ALT	13 IU/L	6-40 IU/L
LDH	157 IU/L	119-229 IU/L
γ-GTP	50 IU/L	0-48 IU/L
ALP	422 IU/L	115-359 IU/L
BUN	45.2 mg/dL	7.4-19.5 mg/dL
Cre	0.9 mg/dL	0.5-1.2 mg/dL
TP	6.3 g/dL	6.4-8.3 g/dL
Alb	2.2 g/dL	3.8-5.2 g/dL
CRP	26.1 mg/dL	0-0.5 mg/dL
BNP	70.5 pg/mL	0-18.4 pg/mL

WBC: white blood cells, Neut: neutrophils, Hb: haemoglobin, Ht: haematocrit, MCV: mean cell volume, Plt: platelets, PT-INR: prothrombin time-international normalized ratio, APTT: activated partial thromboplastin time, HBsAg: hepatitis B surface antigen, HCV-Ab: hepatitis C virus antibody, CK: creatinine kinase, T.bil: total bilirubin, γ-GTP: gamma-glutamyl transpeptase, ALP: alkaline phosphatase, AST: aspartate aminotransferase, ALT: alanine aminotransferase, LDH: lactate dehydrogenase, BUN: blood-urea-nitrogen, Cre: creatinine, TP: total protein, Alb: albumin, CRP: C-reactive protein, BNP: B-type natriuretic peptide, RBC: red blood cells



Figure 1. An ultrasound scan reveals a giant hepatic cyst. Echogenic substances are visible at the bottom of the cyst leading to a “fluid-fluid” level.

pressure and heart sounds with no detectable murmurs. A respiratory examination revealed a short trachea and diminished breath sounds in the right lung base. The abdomen

Table 2. Blood Gas Data on Admission Day.

In the recumbent position on continuous oxygen therapy at a rate of 10 L/min	
pH	7.535
P _a CO ₂	25.8 mmHg
P _a O ₂	41.6 mmHg
HCO ₃	21.3 mmol/L
Base Excess	-0.2 mmol/L
Anion Gap	12.8 mmol/L
Lac	2.36 mmol/L



Figure 2. A computed tomography scan showing a giant hepatic cyst in the right lobe of the liver compressing the adjacent diaphragm, right ventricle and the inferior vena cava (arrow heads).

was non-distended and without scars. The liver span was approximately 10 cm with no detectable ascites. The liver was palpable 3 cm below the costal margin without tenderness. There were no peripheral stigmata of systemic diseases. Examinations of the genitourinary, neurological, dermatological and musculoskeletal systems were all normal.

The laboratory data at admission are shown in Tables 1 and 2. Laboratory tests revealed a normal complete blood count. The metabolic panel revealed mildly abnormal liver function, a high level of C-reactive protein (CRP) and hypoalbuminemia. The electrocardiogram showed sinus rhythm with a normal axis and no acute changes. A chest radiograph showed the right cupola of the diaphragm to be at a higher level than that of the left. A transthoracic echocardiogram revealed a normal-sized right atrium without any right-to-left shunt. Abdominal ultrasonography revealed a 67 ×49×47 mm giant hepatic cyst filled with multiple hyperechoic spots (Fig. 1). A contrast computed tomography (CT) scan of the chest and abdomen showed a giant hepatic cyst in the right lobe of the liver elevating the right hemidiaphragm while at the same time compressing the right atrium and the IVC (Fig. 2). Spiral contrast CT showed no evi-

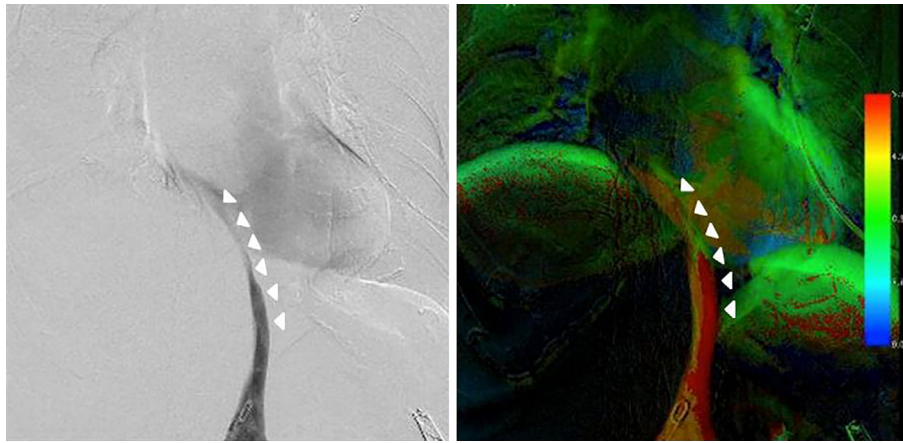


Figure 3. Angiography imaging and flow studies of the inferior vena cava (IVC) before the drainage procedure. The IVC pressure was 23 mmH₂O, and the vessel was compressed and curved by the liver cyst (arrow heads). The bilateral pulmonary arteries were easily visible. The oxygen saturation was <70% on 35 L/min high O₂ flow.

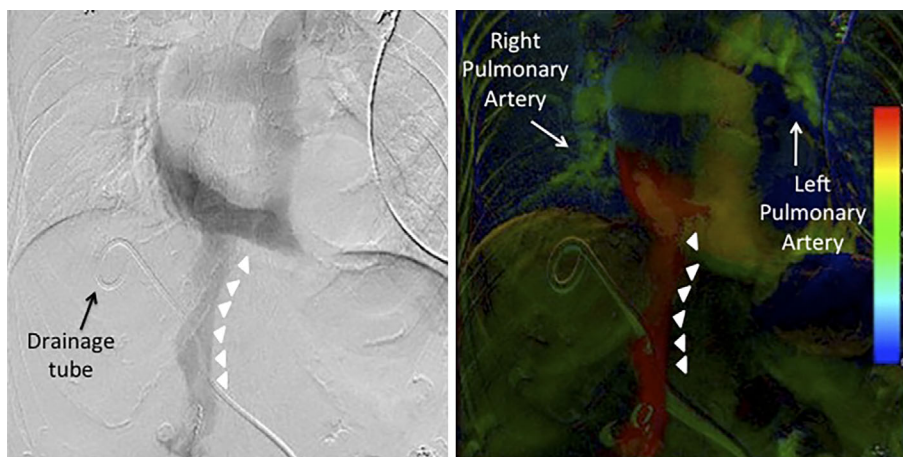


Figure 4. Angiography imaging and flow studies of the inferior vena cava (IVC) after the drainage procedure. The IVC pressure was 13 mmH₂O, and the IVC drained directly into the right atrium (arrow heads). The pulmonary artery outflow tract and bilateral pulmonary arteries were clearly visible (white arrow). The oxygen saturation levels improved to more than 90% breathing ambient room air.

dence of thromboembolism or pulmonary arteriovenous malformation. Following hospital admission, angiography demonstrated severe tapering of the segmental pulmonary arteries in both lungs without any intracardiac or intrapulmonary shunts (Fig. 3). We diagnosed the patient with severe PO syndrome induced by a giant hepatic cyst, which caused secondary compression of the right ventricle, the IVC and the diaphragm.

To decompress the right atrium and IVC, the patient underwent percutaneous transhepatic drainage of the giant hepatic cyst. The procedure was performed without any complications, and the drainage fluid was brown in colour, which suggested a possible infection. Angiography that was repeated after the drainage procedure showed a reduction in the calibre of the IVC stenosis (Fig. 4). The patient reported significant improvement in his symptoms post-procedure, and his oxygen saturations in the recumbent position improved from 80% to 97% breathing ambient room air. Fur-

thermore, his oxygen saturation did not change from the recumbent position to the sitting or standing positions after treatment (Table 3). On the 8th day of hospital admission, *Klebsiella pneumoniae* was isolated from the fluid drained from the giant hepatic cyst. Although the initial assessment of the antimicrobial sensitivity of the organism revealed it to be susceptible to ampicillin-sulbactam, the patient's fever and abnormal values did not improve. On the 16th day of admission, he underwent fenestration and prophylactic resection of the giant hepatic cyst. In this procedure, the cyst roof was opened, and 250 mL of the milky cyst fluid and contents were evacuated by negative pressure. The small gap between the liver parenchyma and outer membrane of the cyst of the liver was separated and completely peeled away from the cyst of the liver. Post-treatment, his fever, C-reactive protein levels and hypoalbuminemia significantly improved. The patient achieved a full recovery, and he was discharged from the hospital on the 50th day.

Table 3. Changes in Oxygen Saturation.

	Before drainage	After drainage
Sitting position	70-80% on high flow 35L	96-98% on room air
Supine position	85-92% on room air	96-98% on room air

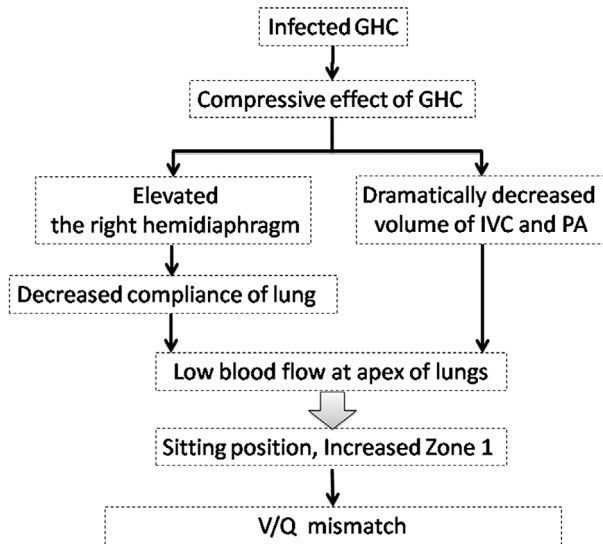


Figure 5. Diagram of platypnea and orthodeoxia with ventilation-perfusion mismatch. GHC: giant hepatic cyst, IVC: inferior vena cava, PA: pulmonary artery, V/Q: ventilation-perfusion

Discussion

The reported case is a rare presentation of PO syndrome owing to an infected giant hepatic cyst. Although only a few cases of a giant hepatic cyst compressing the IVC have been reported, to our knowledge, this is the first report confirming PO syndrome with evidence of severe V/Q mismatch due to an infected giant hepatic cyst.

In 1949, Burchell et al. (6) first reported reflex orthostatic dyspnea occurring in association with pulmonary hypertension. The precise underlying mechanisms were unclear at the time, despite the reported association of several diseases with PO syndrome. In an analysis by Rodrigues et al. (7), more than 80% of cases were associated with interatrial communications, including atrial septal defects or patent foramen ovale, and several cases presented with V/Q mismatch, which included emphysema, chronic obstructive pulmonary disease, other diffuse parenchymal lung diseases, amiodarone pulmonary toxicity (8) and hepatopulmonary syndrome (9).

In our patient, we suspected the main etiology of hypoxemia to be mechanical, via compression of the right atrium, the IVC and the diaphragm by the infected giant hepatic cyst. Infected hepatic cysts are relatively rare, and cholestasis due to compression of the bile ducts by giant hepatic cysts may lead to the infection of the cyst with *Klebsiella*

pneumoniae (10, 11). Angiography in the present case revealed the right atrium and the IVC to be compressed, thereby impeding outflow to the pulmonary vascular bed. Simultaneously, right diaphragmatic splinting by the liver cyst may have exacerbated the pulmonary vascular resistance and right ventricular afterload. These changes may have adversely affected the pulmonary gaseous exchange, thereby resulting in an increase in the lung areas with a high V/Q ratio (12-15).

The change in oxygenation when moving from a recumbent position to a sitting position is assumed to be because the standing position results in an enlargement of the lung areas with high V/Q ratios, thereby exacerbating the existing V/Q mismatch (Fig. 5) (9). Ventilation-perfusion zones of the lung help clarify the influence of the sitting position on the degree of alveolar distention and blood flow. Zone 1 is where the alveolar air pressure is greater than either the pulmonary arterial or venous pressures. Zone 2 is where the alveolar air pressure is less than the pulmonary arterial pressure, but greater than the pulmonary venous pressure. Zone 3 is where the alveolar air pressure is less than both the pulmonary arterial and venous pressures (16). The pulmonary blood flow is directed preferentially to the lung bases versus the apical regions secondary to the effects of gravity and lung compliance in the sitting position. While the pulmonary artery pressure exceeds the alveolar pressure in the gravitational middle (Zone 2) and lower parts of the lung fields (Zone 3), the alveolar pressure at the uppermost parts of the lung may exceed the pulmonary artery pressure. These vessels therefore account for the dead space with high V/Q ratios (Zone 1) (17). Although Zone 1 is almost non-existent in normal healthy lungs (Fig. 6A), extremely low pulmonary arteriolar flow expands this dead space.

In the present case, the compressive effect of a giant hepatic cyst failed to compensate for the reduced right ventricular ejection fraction by a postural change from the supine to the sitting position. In the upright position, gravity and reduced lung compliance affected the blood flow in the apical regions. Under these circumstances, blood vessels can become completely collapsed by the alveolar pressure, resulting in a high V/Q ratio and low oxygen saturation (Fig. 6B). We further believe that the influence of postural changes on the deformity of the IVC and right atrium resulted in the reduction of the right ventricular ejection fraction. Although imaging was not performed with the patient in different body positions, the postural change from a recumbent position to a sitting position may have been accompanied by an exacerbation of mechanical compression and a reduction in the venous return volume.

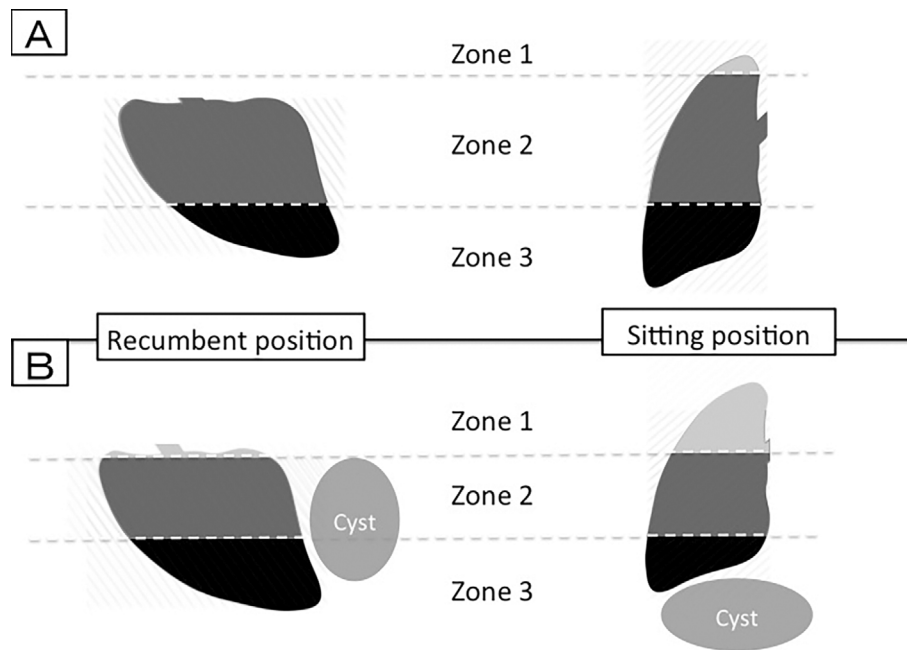


Figure 6. A) Diagram of the influence of gravity on the degree of alveolar distention and blood flow under normal, healthy conditions. B) Diagram of the influence of gravity on the degree of alveolar distention and blood flow in this patient's case. Ventilation-perfusion zones of the lung based and the relationship between the pressure in the alveoli (PA), in the arteries (Pa), and in the veins (Pv). Zone 1: $PA > Pa > Pv$; Zone 2: $Pa > PA > Pv$; Zone 3: $Pa > Pv > PA$.

This case report describes a patient with PO syndrome caused by the extrinsic compression of the right ventricle, the IVC and the diaphragm by a giant hepatic cyst, which was successfully managed by percutaneous drainage. The patient's PO syndrome is believed to have occurred due to V/Q mismatch exacerbated in the upright position by the enlargement of the lung areas with a high V/Q ratio due to a reduction in the apical flow.

The authors state that they have no Conflict of Interest (COI).

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