American Journal of Emergency Medicine 37 (2019) 175.e1–175.e2

Contents lists available at ScienceDirect

American Journal of Emergency Medicine

journal homepage: www.elsevier.com/locate/ajem

Case Report

Hepatopulmonary syndrome-attributed extreme hypoxemia and polycythemia revealing liver cirrhosis☆

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ARTICLE INFO

Article history:
Received 10 September 2018
Accepted 25 September 2018

Keywords:
Acute respiratory distress
Acute respiratory failure
Intrapulmonary arteriovenous shunt

ABSTRACT

We report an unusual case of severe hepatopulmonary syndrome with previously unrecognized cirrhosis, presenting with acute on chronic dyspnoea, extreme hypoxemia, secondary polycythemia as well as direct identification of arteriovenous communications on computed tomography angiography. Hepatopulmonary syndrome, defined as the combination of hepatopathy, arterial deoxygenation and pulmonary vascular dilatation, is increasingly recognized as a life-threatening complication in advanced liver disease and transplant candidacy. It is usually diagnosed in chronic liver disease patients following pre-transplant evaluation or mild dyspnea investigation. Diagnosis relies on the indirect evidence of pulmonary arteriovenous communications suggested by echocardiography with a bubble study. Clinicians need to be aware of this rare but potential acute presentation at the emergency room.

Hepatopulmonary syndrome (HPS), defined as the combination of hepatopathy, arterial deoxygenation and pulmonary vascular dilatation, is increasingly recognized as life-threatening complication in advanced liver disease and transplant candidacy [1]. Usually, HPS is diagnosed in chronic liver disease patients following pre-transplant evaluation or mild dyspnea investigation. Diagnosis relies on the indirect evidence of pulmonary arteriovenous communications suggested by echocardiography with a bubble study. We report the unusual acute presentation of severe HPS in a patient with previously unrecognized cirrhosis.

A 73-year-old African female presented at the emergency room for extreme fatigue and dyspnea that dramatically worsened over the past few years. The patient had refused to consult any doctor so far. On admission, she was conscious, afebrile, with increased heart rate (108/min), normal blood pressure (139/83 mmHg), increased respiratory frequency (28/min) and extremely low SpO_2 (31%). Clinical examination was unremarkable. She reported no history of smoking. Arterial blood gases while breathing on room air showed pH 7.39, PaO_2 24 mmHg, $PaCO_2$ 28 mmHg and HCO_3^- 17 mmol/L, with lactate concentration (3.0 mmol/L; N, 1.0–2.0), non-elevated carboxyhemoglobinemia (0.9%) and methemoglobinemia (0.3%). Massive increase in alveolar-arterial oxygen gradient (91 mmHg; N < 23 considering her age) was demonstrated. The patient promptly received high-flow nasal oxygen (FiO_2~100%) but was poorly responsive with SpO_2 71% and PaO_2 44 mmHg. Laboratory tests showed hemoglobin 21.4 g/dL, hematocrit 67%, leukocytes 4.2 G/L, platelets 180 G/L, international normalized ratio 1.19, serum creatinine 97 μmol/L (N ⟨120), albumin 32 g/L (N > 35), alanine aminotransferase 12 UI/L (N < 40), γ-glutamyltransferase 182 UI/L (N < 58), bilirubin 28 μmol/L (N < 17) and C-reactive protein 20 mg/L (N < 5).

Computed tomography (CT) angiography with contrast media injection showed diffuse distal dilatation of pulmonary arteries and veins, predominating at the lung bases (Fig. 1) without evidence of arteriovenous malformation, pulmonary embolism or primary lung disease. Transthoracic contrast-enhanced echocardiography with agitated saline detected microbubbles in the left heart chamber within 3–4 cardiac cycles after right atrial passage, suggesting intrapulmonary arteriovenous shunt according to the guidelines [2]. No evidence of cardiac disease, specifically no atrial or ventricular septal defects were found.

Abdominal CT-scan showed a dysmorphic liver with nodular contours and evidence of portal hypertensive collaterals but no tumour or ascites, consistent with the diagnosis of cirrhosis and portal hypertension. Ongoing alcohol abuse, viral hepatitis B recovery and active chronic viral hepatitis C were found but no other cause of liver disease including metabolic and auto-immune disorders. In particular, serum ferritin was normal and human immunodeficiency virus, anti–liver-

☆ Informed consent was obtained from the patient for publication.

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https://doi.org/10.1016/j.ajem.2018.09.044
0735-6757/

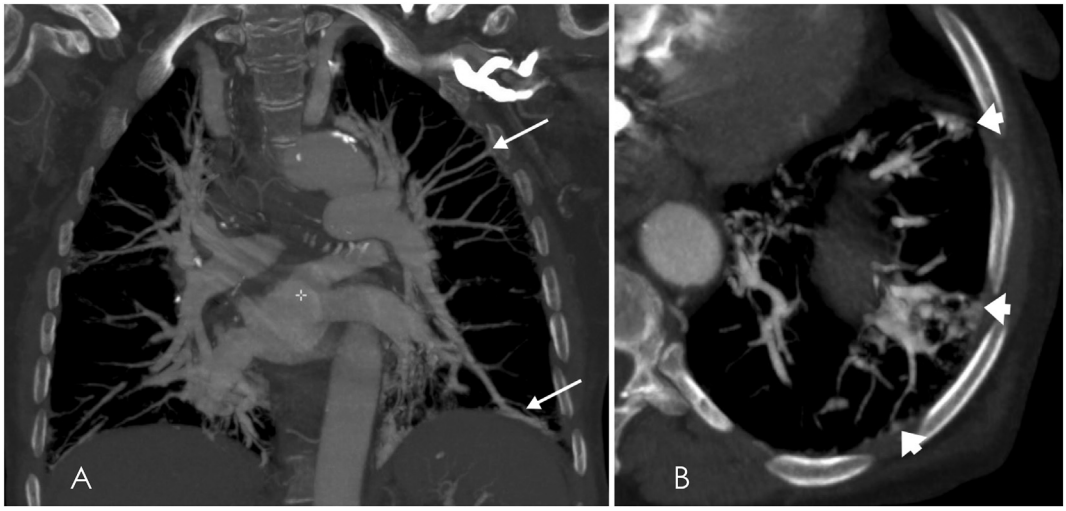

Fig. 1. Chest computed tomography angiography with maximum intensity projection in (A) oblique coronal view showing dilation of peripheral lung vessels extending to the sub-pleural region with typical non-tapered aspect (white arrow); (B) transversal view zoomed on the basal region of the left lung showing dilated distal lung vessels extending to the pleura (white arrowheads) with subpleural opacities resembling telangiectasia.

kidney microsomal, antimitochondrial, antinuclear and anti-smooth-muscle antibodies were negative. Model of end-stage liver disease (MELD) score was 11.

The patient was deemed non-eligible for liver transplant owing to her age, general status and the ongoing alcohol abuse. As a result, transjugular liver biopsy with pressure measurement was not considered. Due to the absence of pulmonary arteriovenous malformation, she was not amenable to therapeutic embolization. Subsequently, she was discharged home with oxygen and best supportive care. At 6-month follow-up, she was alive with stable dyspnea on mild exertion.

This patient's HPS presentation was remarkable. Liver cirrhosis evidenced at severely symptomatic end-stage HPS is uncommon. Severe dyspnea as the revealing symptom of liver disease has rarely been reported and when reported, usually combined with marked erythrocytosis attributed to the compensation for severe hypoxemia [3]. In our patient, the delayed medical consultation clearly explained the observed extreme hypoxemia and polycythemia.

Diagnosis relies on indirect assessment using contrast-enhanced echocardiography or technetium-99 m-labeled macro-aggregated albumin scanning [1]. Confirmingly, direct identification of arteriovenous communications on CT angiography is rare. Using micropaque-gelatin angiograms in autopsies, Berthelot et al. first reported precapillary arteriovenous communications and dilated capillaries of 500 μm (<15 in healthy conditions), peripherally distributed along the pleural surface of the lower lobes with spider angiomata appearance [4].

Finally, as shown in our patient, HPS time-course is independent of the underlying liver disease with progressive worsening in the absence of liver transplantation [5].

Conflict of interest

The authors declare no conflict of interest.

Financial disclosure

None.

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