Hepatopulmonary syndrome (HPS), 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.

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 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.

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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.

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References