Hepatopulmonary Syndrome

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Citation

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Abstract

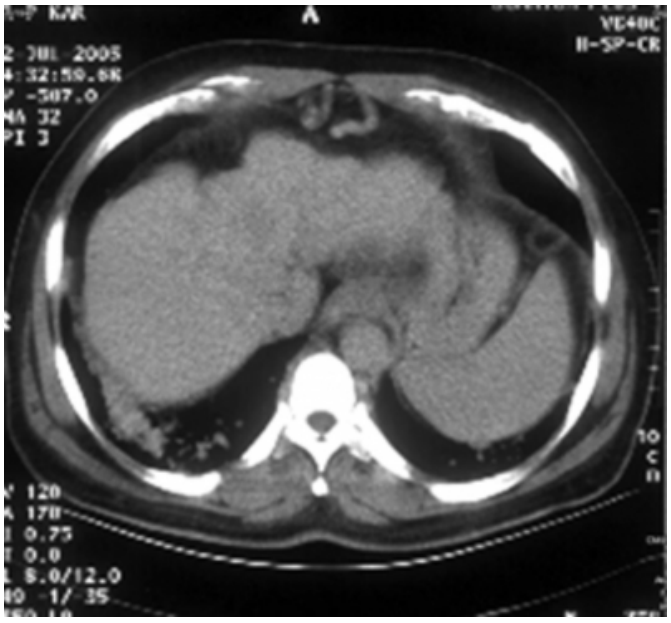
Advanced liver disease and portal hypertension may produce various intrathoracic complications that involve the pleural space, the lung parenchyma and the pulmonary circulation. Dyspnoea and arterial hypoxemia are the most common symptoms and signs in patients with such complications.
Hepatopulmonary syndrome (HP) is the most widely recognized of the processes associated with end stage liver disease.

CASE REPORT

A 49 year old male patient of liver cirrhosis with portal hypertension was admitted to the hospital with progressive dyspnoea. A CT scan of the abdomen revealed an irregular, nodular liver surface consistent with cirrhosis. Evidence of anterior abdominal wall collaterals and perioesophageal thickening s/o oesophageal varices seen (Fig. 1).

Figure 1

Figure 1: Contrast enhanced CT scan of the abdomen showing an irregular nodular liver surface consistent with cirrhosis. There is perioesophageal thickening s/o varices and collaterals seen in the anterior abdominal wall. Evidence of ascites also seen.



Splenomegaly, a dilated splenoportal axis and multiple collaterals at the splenic hilum, anterior abdominal wall, retroperitoneal and gastric bed s/o varices were seen (Fig. 2). Contrast enhanced CT scan of the chest revealed a normal cardiac size (Fig. 3) with normal diameter of the main pulmonary artery (Fig. 4).

Figure 2

Figure 2: CECT abdomen showing splenomegaly, dilated splenoportal axis, collaterals at the splenic hilum, in the anterior abdominal wall and retroperitoneal collaterals.

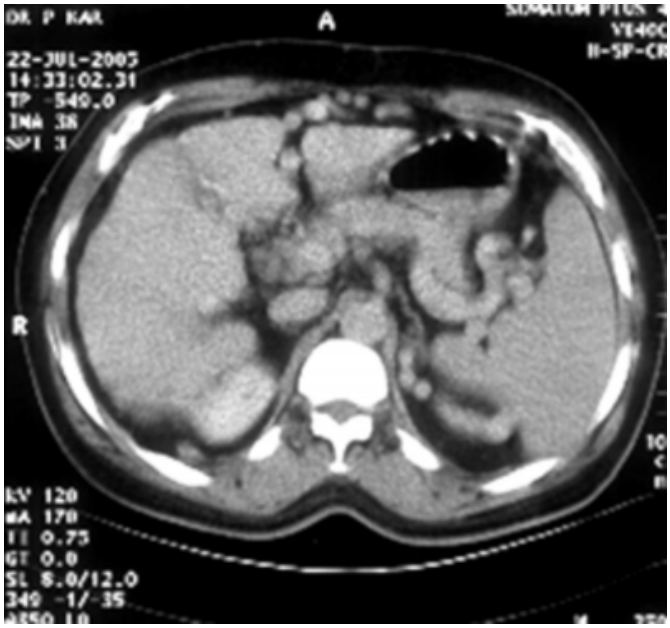


Figure 3

Figure 3: CECT chest showing normal cardiac size with no chamber dilatation.

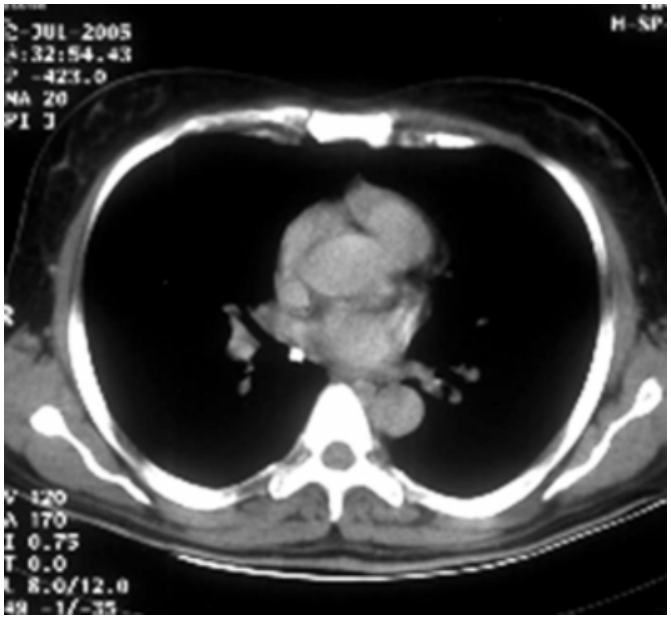
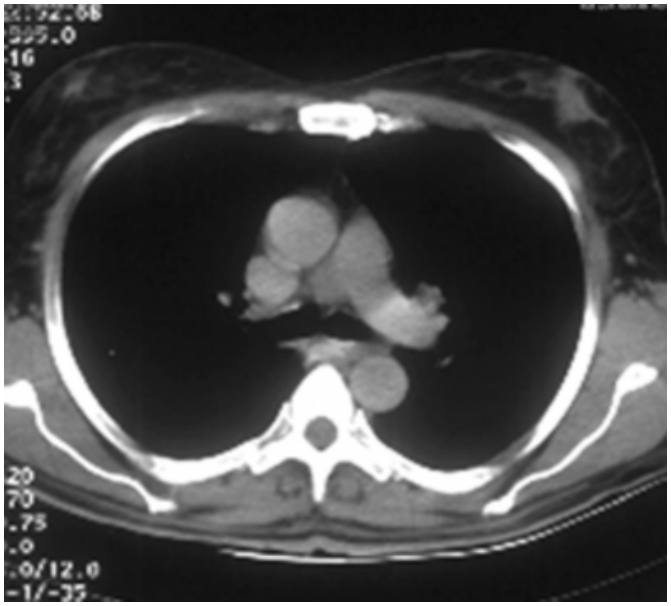


Figure 4

Figure 4: CECT chest showing normal caliber of the central and the right and pulmonary arteries.



Lung parenchymal window settings revealed vascular dilatation in the peripheral pulmonary vessels associated with an abnormally large number of visible terminal artery branches (Fig. 5). HRCT revealed the peripheral vascular branches to be several millimeters in diameter and extending to the pleural surface (Fig. 6).

Figure 5

Figure 5: Lung parenchymal window settings showing bilateral basal predominance of abnormally dilated peripheral pulmonary vessels with an abnormally large number of visible terminal artery branches.

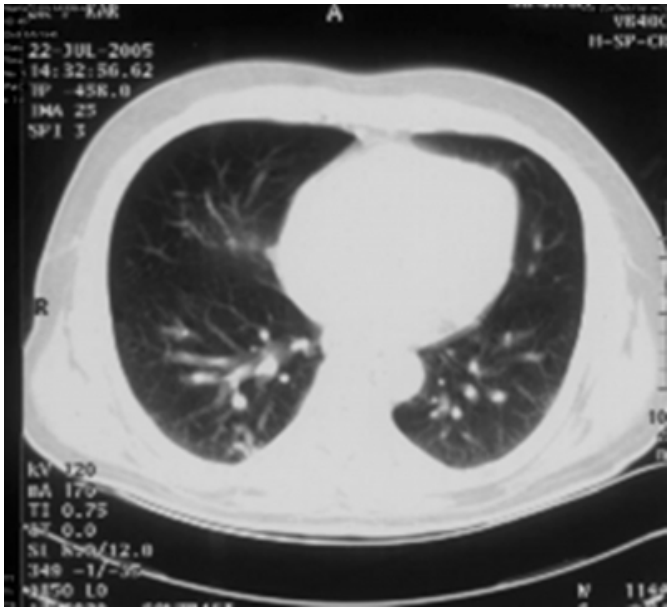
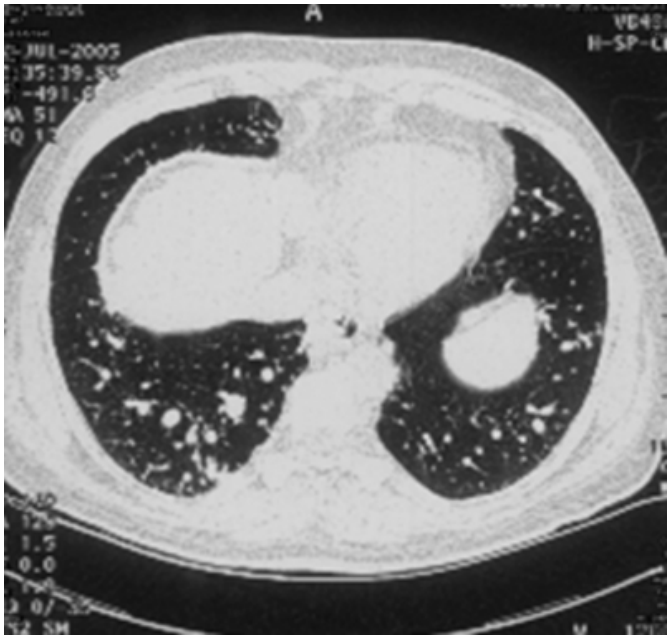


Figure 6

Figure 6: HRCT sections also reveal the peripheral vascular branches to be abnormally dilated extending to the pleural surface giving a spidery appearance.



Based on these findings a diagnosis of hepatopulmonary syndrome was suggested. The patient showed a good response to treatment with 100% oxygen.

DISCUSSION

HEPATOPULMONARY SYNDROME (HPS)

Hepatopulmonary syndrome (HPS) is defined by the triad of hepatic dysfunction, intrapulmonary vascular dilatation and abnormal arterial oxygenation (hypoxemia) [1]. Clinically, HPS typically manifests with progressive dyspnoea and hypoxemia in a patient who has cirrhosis.

Pulmonary complications may occur as a result of end stage liver disease due to a decreased hepatic clearance or increased hepatic production of circulating cytokines and other vascular growth mediators [2].

Substances implicated in liver lung interaction [2]

Figure 7

Substance	Site of production	Type of imbalance	Suspected mechanism of interaction	Result
Glucagon	Pancreatic alpha cells	Decreased hepatic clearance	Primarily glucose and carbohydrate metabolism; vasodilator	HPS
Calcitonin gene-related peptide	Hepatocytes	Decreased hepatic clearance	Potent vasodilator	HPS
Vasoactive intestinal peptide	Neurotransmitter	Decreased hepatic clearance	Potent vasodilator	HPS
Atrial natriuretic factor	Atrial myocytes	Decreased hepatic, renal, pulmonary clearance	Diuretic, natriuretic, and vasodilative effects	HPS
Substance P	Gastrointestinal tract	Decreased hepatic clearance	Potent pulmonary vasodilator mediated by nitric oxide synthesis	...
Platelet-activating factor, prostaglanding E_2 and I_2	...	...	Possibly pulmonary vasodilatation	HPS
Nitric oxide	Endotoxin stimulated	Portosystemic shunts decrease clearance	Endothelial cell-dependent vasodilatation and smooth muscle relaxation	HPS

As shown in the table, the serum concentrations of many circulating mediators are elevated in cirrhotic patients and such elevations are known to cause pulmonary vasodilatation. Because the principal vasoactive substance has not been identified, no effective pharmacologic intervention is available and the treatment consists of supplemental oxygen therapy or liver transplantation [3].

Hypoxemia in patients who have HPS is thought to occur primarily because of vascular dilatation (diffusion - perfusion impairment). Hypoxia is believed to result from an inability of oxygen to diffuse to the centre of the massively dilated peripheral vessels resulting in a right to left shunt. These vessels which are normally 8-15 μm in diameter have been demonstrated to dilate to 15-500 μm [1].

The presence of intrapulmonary vascular dilatation can be established with imaging. Chest radiographs demonstrate basilar nodules or reticulonodular areas of increased opacity in 5-13.8% of patients with chronic liver disease and 46-100% of patients with hepatopulmonary syndrome [4]. Lung volumes are preserved. Computed tomography (CT) may demonstrate dilated vessels with an increased number of terminal branches extending to the pleura bilaterally, predominantly in lower lobes and can be useful in distinguishing hepatopulmonary syndrome from other causes of hypoxemia such as pulmonary fibrosis. Also of importance in the CT diagnosis of HPS is the recognition of associated hepatic disease with findings of cirrhosis, splenomegaly, varices and ascites [4].

Two patterns of vascular abnormality are seen on pulmonary angiography in this syndrome. The lesions of HPS have been classified into two types [1]. The type 1 (minimal) pattern is most common (85%), it is associated with spidery appearance of peripheral vessels and usually a good response to treatment with 100% oxygen. Type 2 lesions (15%) represent small, discrete pulmonary arteriovenous fistulae, type 2 lesions are associated with a poor response to 100% oxygen.

The presence of intrapulmonary right to left shunt can be demonstrated by contrast echocardiography or Tc-99m macroaggregated albumin lung scanning [5]. The rationale for both the diagnostic tests in hepatopulmonary syndrome is that microbubbles (at least 15 μm) used for contrast echocardiography or the aggregated albumin (20 to 60 μm) for Tc-99m macroaggregated albumin lung scanning can pass through dilated but not normal pulmonary capillaries (less than 8 μ in diameter).

Thus awareness of the clinical and radiographic spectrum of the abnormalities that can occur in patients with advanced liver disease who also have respiratory symptoms assists the radiologist in reaching a meaningful differential diagnosis.

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References

1. Castro M, Krowka MJ. Hepatopulmonary syndrome a pulmonary vascular complication of liver disease. Clin Chest Med 1996; 17: 35-48.
2. King PD, Rumbaut R, Sanchez C. Pulmonary manifestations of chronic liver disease. Dig. Dis 1996; 14: 73-82.
3. Lange PA, Stoller JK. The hepatopulmonary syndrome. Ann Intern Med 1995; 122: 521-529.

4. McAdams HP, Erasmus J, Crocketl R, Mitchell J, Godwin JD, McDermott VG. The hepatopulmonary syndrome : radiologic findings in 10 patients. Am J Roentgenol 1996; 166: 1379-1385.

5. Vedrinne JM, Duperret S, Bizollon T et al. Comparison of transesophageal and transthoracic contrast echocardiography for detection of an intrapulmonary shunt in liver disease. Chest 1997; 111: 1236-40.

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