

Diaphragmatic herniation due to massive hepatomegaly in a patient with pulmonary arterial hypertension

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DESCRIPTION

A 56-year-old woman with severe pulmonary arterial hypertension (PAH) presented with right upper quadrant abdominal pain. The patient was diagnosed with PAH 5 years prior and started on oral therapy, but due to worsening pulmonary haemodynamics and symptoms, intravenous treprostinil was initiated. Imaging studies demonstrated hepatomegaly and a liver biopsy showed hepatic venous outflow obstruction and findings consistent with nodular regenerative hyperplasia. There was no evidence of hepatic fibrosis. Despite treatment with intravenous treprostinil, the patient had persistent dyspnoea and ongoing symptoms of right heart failure with worsening right upper quadrant abdominal pain. CT scan of the chest, abdomen and pelvis showed marked hepatomegaly with herniation of the caudate lobe into the right hemithorax (figure 1). Liver herniation and cardiomegaly resulted in restrictive pulmonary function with a total lung capacity of 64%.

Survival for patients with PAH is closely associated with right ventricular function.¹ Right heart failure is a late consequence of PAH hypertension that can lead to oedema, ascites and congestive hepatopathy. The underlying pathophysiology involved in hepatic dysfunction from right heart failure involves passive congestion secondary to increased right atrial pressures and/or low cardiac output with decreased arterial perfusion. These deleterious haemodynamics are further amplified by concomitant hypoxia.² Radiological evidence of hepatomegaly or elevated liver enzymes, particularly

bilirubin, is a marker of poor prognosis³ and should prompt referral for advanced treatment options, including lung transplantation.

Learning points

- ▶ Development of right heart failure and congestive hepatopathy are signs of poor prognosis in patients with pulmonary arterial hypertension and should prompt therapeutic modifications and consideration for lung transplantation.
- ▶ In the absence of significant fibrosis in the liver, hepatic dysfunction and hepatomegaly can completely reverse with correction of the right heart failure.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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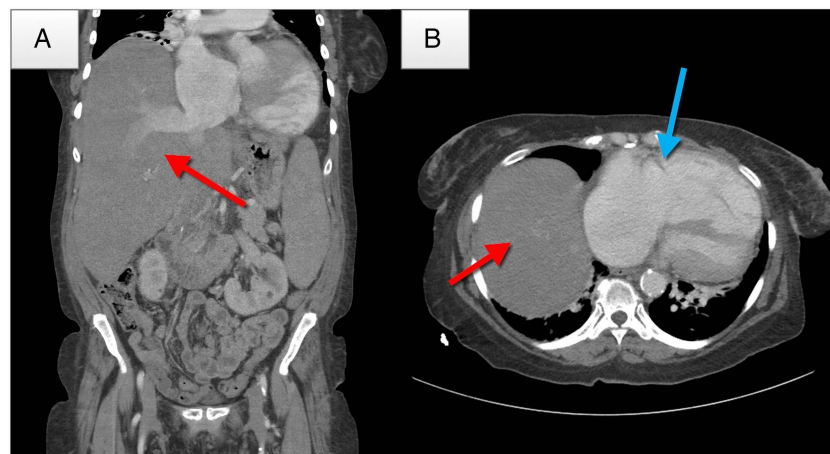


Figure 1 Massive hepatomegaly (red arrows) with herniation of the caudate lobe of the liver through the right hemidiaphragm. Cardiomegaly resulting from right atrial and right ventricular enlargement also present (blue arrow).



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