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Intractable Hepatic Hydrothorax: A Successful Outcome following CPAP Treatment

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ABSTRACT

Hepatic hydrothorax is an uncommon complication in patients with end-stage liver disease. It may result in dyspnoea, hypoxia and infection, and carries a poor prognosis. Initial treatment is based on a sodium-free diet together with diuretics. In case of recurrent hydrothorax, a transjugular intrahepatic portosystemic shunt (TIPS) or liver transplant should be considered. Here we describe an 80-year-old woman with decompensated liver cirrhosis related to NASH who presented with refractory hepatic hydrothorax. Treatment with CPAP resulted in a marked improvement in her pleural effusion.

LEARNING POINTS

- Hydrothorax is uncommon, occurring in up to 4–6% of all patients with cirrhosis and in up to 10% of patients with decompensated cirrhosis.
- Hepatic hydrothorax can be refractory to diuretics and salt restriction.
- CPAP may be an interesting alternative treatment.

KEYWORDS

Cirrhosis, hepatic hydrothorax, CPAP

INTRODUCTION

In the literature, pleural effusion is often associated with ascites, usually in connection with solid ovarian tumours, Demons-Meigs syndrome or hepatic cirrhosis. Hepatic hydrothorax is defined as the accumulation of significant pleural effusion in a cirrhotic patient without primary pulmonary or cardiac disease. Initial treatment is based on a sodium-free diet together with diuretics. In case of recurrent hydrothorax, liver transplantation should be considered. When possible, the placement of a transjugular intrahepatic portosystemic shunt (TIPS) may be sufficient or could serve as a temporary treatment while the patient awaits a liver transplant [1].

CASE DESCRIPTION

An 80-year-old woman was admitted to our unit for shortness of breath. She was known to have liver cirrhosis related to NASH (Child-Pugh 7-B, MELD 11). About 6 months previously, she had developed ascites and treatment with salt restriction, furosemide and spironolactone was started. Gastroscopy later showed stage 1 oesophageal varices. Two months after this, she was admitted to the respiratory medicine

unit for massive right pleural effusion. Investigation revealed she had hepatic hydrothorax, which was treated with thoracentesis that removed 4000 ml of transudative fluid. Treatment with diuretics was maintained. One month later, she was again admitted for recurrent pleural effusion. The abdomen was mildly distended, but ascites was not evident. The age of the patient precluded a liver transplant and the presence of portal vein thrombosis contraindicated TIPS.

The patient was again treated with thoracentesis which removed 4000 ml of transudative fluid. A search of the medical literature revealed some rare, similar cases where CPAP had significantly reduced pleural effusion recurrence. Consequently, CPAP was instituted and 6 months later, the patient has not had recurrence of her effusion and her quality of life has markedly improved.

DISCUSSION

Hepatic hydrothorax is defined as the accumulation of significant pleural effusion in a cirrhotic patient without primary pulmonary or cardiac disease. Hydrothorax is uncommon, occurring in up to 4–6% of all patients with cirrhosis and in up to 10% of patients with decompensated cirrhosis. Hepatic hydrothorax is usually seen on the right side (65–87% of reported cases). The formation of hepatic hydrothorax is linked to unidirectional transfer of ascites from the peritoneum to the pleural cavity through diaphragmatic defects. Diagnosis can be confirmed by peritoneal scintigraphy, ultrasonography and magnetic resonance imaging [1].

These diaphragmatic defects have been demonstrated macroscopically and microscopically in patients with hepatic hydrothorax. They have also been visualized intraoperatively, during thoracoscopy and during autopsy studies. The size of these defects varies from 0.03 to 5–6 mm in diameter, which makes their identification difficult. The ascites may be absent or minimal in 20% of patients with hepatic hydrothorax. Initial treatment is based on a sodium-free diet together with diuretics. However, hepatic hydrothorax is refractory to salt restriction and diuretics in approximately 25% of cases. In case of recurrent hydrothorax, liver transplantation should be considered. When possible, the placement of a TIPS may be sufficient or could be used as temporary treatment while the patient awaits a liver transplant. Other surgical approaches have also been used in the management of refractory hepatic hydrothorax [1–3].

Recently, some rare cases of resistant hepatic hydrothorax have been successfully treated using CPAP. Takahashi et al. described a 62-year-old patient with resistant hepatic hydrothorax and obstructive sleep apnoea. Surprisingly, CPAP initiation resulted in a marked improvement in pleural effusion. The pleural effusion returned after CPAP was stopped and improved after CPAP was reinstated [4]. Saito et al. reported two cases of intractable hepatic hydrothorax successfully treated with nasal CPAP [5]. In 2015, Yamamoto et al. described a case of refractory hepatic hydrothorax that improved with a combination of thoracocentesis and CPAP [6].

The mechanism underlying the improvement in these patients may have been the intrathoracic positive pressure induced by CPAP. CPAP treatment does not carry any risks, and its use in hepatic hydrothorax should be investigated in a large series of patients.

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