Case report

Acute tension hydrothorax in chronic liver disease secondary to spontaneous diaphragmatic rupture

Kevin Joseph Tharappel , Nandakrishna Bolanthakodi, Sudha Vidyasagar, Muralidhar Varma

Department of Medicine,
Kasturba Medical College,
Manipal Academy of Higher
Education, Manipal, India

Correspondence to
Dr Nandakrishna Bolanthakodi;
nandaksb@gmail.com

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SUMMARY

Pleural effusion in liver cirrhotics is more commonly transudative. A transudative pleural effusion secondary to ascites in decompensated cirrhosis is also known as hepatic hydrothorax and is usually due to fluid seepage through congenital pores in the diaphragm. The patient, a known case of decompensated chronic liver disease, presented with a massive, left-sided, rapidly accumulating and transudative pleural effusion secondary to spontaneous diaphragmatic rupture. Clinically, he developed sudden onset shortness of breath and became hypotensive. This is a rare entity, and was confirmed on CT thorax revealing a focal segment defect ~1.6 cm over the left hemidiaphragm. Ascites treatment consisting of diuretics with salt restriction and repeated thoracentesis with albumin replacement improved his symptoms and lead to a complete resolution of the effusion.

BACKGROUND

This patient exhibits the key characteristics of hepatic hydrothorax (HH). However, this case is unique as the patient developed a rapid onset, massive effusion and became hypotensive due to spontaneous diaphragmatic rupture, resulting in acute tension hydrothorax. Regardless of how it develops, management is of utmost importance and poses a significant challenge for many clinicians due to its relapsing nature and may explain its poor prognosis. Emerging therapies, such as indwelling pulmonary catheters and novel surgical approaches, are now becoming available, which may see an improved outcome in the future.

CASE PRESENTATION

A 50-year-old man, diagnosed with alcoholic liver disease—decompensated cirrhosis of the liver (Child-Pugh Class C) on torsemide 20 mg OD and spironolactone 100 mg OD for the last 8 months was admitted with complaints of worsening abdominal distension and pedal oedema since 1 week. He had been abstinent from alcohol. He denied any fever or reduced urine output and vitals were stable on the initial presentation. He was icteric, exhibited bilateral gynaecomastia, bilateral pitting pedal oedema and moderate ascites. Other systemic examinations were unremarkable and he had no hepatic encephalopathy. On the second day of admission, the patient developed acute onset dyspnoea and hypotension. Examination revealed reduced breath

sounds over all left lung fields with dull percussion note suggestive of left-sided pleural effusion. Oxygen saturation was 88% on room air. He was treated with an intravenous saline bolus followed by inotropic support to maintain a mean arterial pressure of >65 mm Hg and oxygen inhalation. He was empirically treated with injection piperacillin–tazobactam considering hospital-acquired pneumonia with synpneumonic effusion and septic shock as the potential cause for his sudden deterioration.

INVESTIGATIONS

Routine investigations were sent after admission and were suggestive of chronic liver disease (platelets—77 k/μL, total bilirubin (TB)—2.56 mg/dL, direct bilirubin (DB)—1.29 mg/dL, aspartate transaminase (AST)—37 IU/L, alanine transaminase (ALT)—17 IU/L, alkaline phosphatase (ALP)—102 IU/L, albumin—25 g/L, globulin—46 g/L and international normalised ratio (INR)—1.52). Renal function tests (RFTs) showed blood urea—15 mg/dL, serum creatinine—0.85 mg/dL, Na⁺—133 meq/L and K⁺—3.1 meq/L. Hepatitis B surface antigen (HBsAg) and anti-Hepatitis C virus (HCV) antibodies were negative. Alpha-fetoprotein was within the normal range. A diagnostic ascitic tap revealed a serum ascites albumin gradient of 18 g/L and no evidence of spontaneous bacterial peritonitis (SBP). The chest X-ray was normal on admission ([figure 1](#)).

On the second day of admission, when the patient became acutely breathless, chest X-ray showed a new-onset moderate left-sided pleural effusion ([figure 2](#)). A repeat chest X-ray performed the next day showed a massive effusion with tracheal shift to the right side which corresponded with a worsening of his symptoms ([figure 3](#)). A therapeutic thoracentesis was done for symptomatic relief, analysis of which showed a total count of 260 white blood cell (WBC)/μL. The fluid was predominantly lymphocytic (70%) and transudative by Light's criteria (pleural protein/serum protein—0.3, pleural lactate dehydrogenase (LDH)/serum LDH—0.2 and LDH—85 IU/L). Pleural fluid cytology and cultures were sterile. Blood cultures were also sterile. A contrast CT chest showed a 1.6 cm defect in the left hemidiaphragm without evidence of bowel herniation ([figure 4](#)). Based on the CT chest report, a diagnosis of acute tension hydrothorax secondary to spontaneous diaphragmatic rupture was considered.

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Figure 1 Chest X-ray taken on day 1 of admission. A normal chest X-ray.

DIFFERENTIAL DIAGNOSIS

Synpneumonic effusion was ruled out as pleural fluid was transudative. Clinical examination findings ruled out the possibility of pneumothorax and pulmonary embolism.

TREATMENT

The patient was given 80 mg intravenous furosemide in two divided doses immediately after developing the effusion on day 3. Following this, daily thoracentesis of 1 L with a 20 g albumin infusion was done for three consecutive days given his breathlessness. Hypotension recovered and inotropes were slowly tapered over 2 days. Dosage of diuretics was gradually increased to 120 mg of furosemide and 200 mg of spironolactone in divided doses over 1 week. Renal function tests were monitored

Figure 2 Chest X-ray on day 2 of admission. This X-ray revealed a moderate left-sided effusion.

Figure 3 Chest X-ray on day 3 of admission. Massive left-sided effusion with tracheal shift to the right side.

and were normal. With these measures, ascites and effusion reduced and hypoxia improved. Repeat chest X-ray done on day 14 showed a moderate effusion (figure 5).

OUTCOME AND FOLLOW-UP

He was discharged on oral furosemide and aldactone. On follow-up after 2 weeks, he was asymptomatic, ascites had reduced and there was a clinical and radiological resolution of the pleural effusion (figure 6). The patient is alive and responding well to treatment.

DISCUSSION

The incidence of HH is 4%–6% in patients with cirrhosis when diagnosed with X-ray; however, it increases to 15% when ultrasound is used.^{1 2} There are several proposed theories as to how it develops. The presence of congenital diaphragmatic defects is often considered the primary mechanism as described by Lieberman *et al* which is mainly concerned with microscopic abnormalities in the collagen structure of the diaphragm.³ Second, lymphatic channels which traverse the diaphragm allows passage of fluid between the abdomen and thorax.⁴ Hypoalbuminemia,

Figure 4 CT on day 6 of admission. Axial and coronal sections showing the defect (white arrows) in left hemidiaphragm with massive left-sided pleural effusion (white star) and ascites.

Figure 5 Chest X-ray on day 14 of admission (last day in the hospital). Chest X-ray showed a resolving, moderate pleural effusion.

which lowers colloidal oncotic pressure, can also lead to pleural fluid accumulation.⁵

Diaphragmatic defects in HH are classically described as four distinct morphological types as described by Huang *et al.*⁶ We would like to emphasise the unique pathophysiology of HH in this patient who has evidence of a sizeable defect that developed spontaneously connecting the pleural and peritoneal cavities. This phenomenon is rarely encountered in clinical practice. Furthermore, the site of the defect is unusual. Most pleural effusions attributed to HH are right-sided due to the increased muscularity and thickness of the left hemidiaphragm. This explains the marked discrepancy in the incidence of right and left pleural effusions in HH. 79.5% are right-sided, 17.5%

Figure 6 Chest X-ray 2 weeks after the patient was discharged. Complete resolution of the effusion when compared with the previous chest X-ray.

left-sided and 3% are bilateral.⁷ In our patient, the effusion was left-sided, rapid in onset and massive requiring repeated thoracentesis. Hence, suspicion of infection or diaphragmatic defect was considered.

Spontaneous rupture of diaphragm (SRD) in a cirrhotic resulting in acute tension HH is an extremely rare occurrence. In chronic liver disease, the most likely explanation is a sudden increase in intra-abdominal pressure due to tense ascites. However, this patient was on diuretics for 8 months prior to admission with only moderate ascites on examination. In general, SRD is commonly due to raised abdominal pressure as seen with squatting and prolonged coughing.^{8,9} Ehlers-Danlos—hypermobile type, in rare cases, can also cause spontaneous rupture due to inherent weakness in the muscularity of the diaphragm.¹⁰ Losanoff *et al* performed a world literature review on SRD collecting 28 reports between 1953 and 2009.⁹ Cough and physical exercise were the inciting factors in the majority of cases. 68% were left-sided and most were located peripherally. This could explain the left-sided diaphragmatic defect in our patient and consequent seepage of fluid into the pleural space due to the pressure difference between the two cavities. Most likely a combination of factors was responsible for developing the defect in this patient: his ascites, a ‘weak’ diaphragm and possibly, a sudden bout of coughing, straining or change in position could have favoured rupture formation.

Medical management is the mainstay of therapy for most patients with HH. Nevertheless, 21%–26% of patients do not respond to initial therapy consisting of salt restriction with diuretics.¹¹ The patient continued to have a persistent effusion on chest X-ray with initial medical therapy. For this reason, he underwent thoracentesis on three occasions where ~1 L of fluid was removed each time. CT thorax, which illustrated the diaphragmatic rent, gave a possible explanation as to why the pleural effusion had not subsided on initial diuretic therapy. As a result, he was started on dual diuretics, furosemide and spironolactone, which he took in divided doses and continued until discharge. With this regimen, he showed marked improvement.

HH is a chronic, relapsing complication of advanced liver disease and therefore it is important to be aware of alternative therapies if initial medical management fails due to poor compliance or refractoriness. Repair of the diaphragmatic defect may be a reasonable option in this patient due to its prominence on CT scan which is remarkable as they are rarely detected by non-invasive imaging.¹² Most diaphragmatic repairs are not performed as the defect cannot be visualised, with one study citing that in 41 patients in whom diaphragmatic repair was considered, the defect could only be visualised in 5.¹³ However, if directly observed, a surgical closure is a viable option. Since the patient responded well to medical therapy along with repeated thoracentesis and showed no evidence of bowel herniation on CT, we did not pursue a surgical line of treatment.

Chemical pleurodesis involves the instillation of a sclerosing agent such as Talc into the pleural cavity which results in pleural membrane irritation and subsequent obliteration of the pleural space. It is a procedure that has variable success and is associated with a high recurrence rate. Placement of an indwelling pulmonary catheter is an emerging prospect that can also be considered a ‘bridge’ to liver transplantation. Light, Richard L reviewed the work of Chen and colleagues who conducted a pilot study on the use of indwelling tunnelled pleural catheters in managing HH.¹⁴ The 25 patients who underwent the procedure did not require a subsequent one and more than a third achieved spontaneous pleurodesis. This is in stark contrast to the use of a chest tube

which had a high complication rate as described by Orman *et al.*¹⁵

The Transjugular Intrahepatic Portosystemic Shunt (TIPS) procedure is relatively safe and it is important to assess whether this patient would be a viable candidate for the procedure in the future. Refractory HH patients with Child Turcotte Pugh score (CTP) grade A or B, MELD score <15 and those aged <60 with no cardiac complications may benefit from the procedure.^{16–18} Although the patient presented in this case is <60, his MELD score (19) and CTP grade (C) make him a poor candidate. Liver transplantation is still the ultimate goal; since most patients are poor candidates for surgery, it is often not possible. This places emphasis on alternative therapies.

To conclude, SRD and HH are classically described as two distinct pathologies. We now know they are not mutually exclusive. This case adds to the current medical literature on HH as it provides another mechanism of how HH develops and also highlights several rare occurrences; the patient has a left-sided, massive effusion which was detected with non-invasive imaging.

Learning points

- Pleural effusion in a patient with chronic liver disease has a wide differential diagnosis.
- Hepatic hydrothorax (HH) manifesting as a left-sided, massive pleural effusion detected by non-invasive imaging is exceedingly rare.
- Medical management is the primary mode of therapy in treating HH.
- TIPS procedure acts as a bridge to liver transplantation in refractory HH and is an effective treatment in viable candidates.

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ORCID iD

Kevin Joseph Tharappel <http://orcid.org/0000-0002-6476-1832>

REFERENCES

- 1 Johnston RF, LOO RV. Hepatic hydrothorax; studies to determine the source of the fluid and report of thirteen cases. *Ann Intern Med* 1964;61:385–401.
- 2 Chen T-A, Lo G-H, Lai K-H. Risk factors for spontaneous bacterial empyema in cirrhotic patients with hydrothorax. *J Chin Med Assoc* 2003;66:579–86.
- 3 Lieberman FL, Hidemura R, Peters RL, *et al.* Pathogenesis and treatment of hydrothorax complicating cirrhosis with ascites. *Ann Intern Med* 1966;64:341–51.
- 4 Dumont AE, Mulholland JH. Flow rate and composition of thoracic-duct lymph in patients with cirrhosis. *N Engl J Med* 1960;263:471–4.
- 5 Higgins G, Kelsall AR, O'Brien JRP, *et al.* Witts LJ: ascites in chronic disease of the liver. *QJ Med* 1947;16:263–74.
- 6 Huang P-M, Chang Y-L, Yang C-Y, *et al.* The morphology of diaphragmatic defects in hepatic hydrothorax: thorascopic finding. *J Thorac Cardiovasc Surg* 2005;130:141–5.
- 7 Castellote J, Xiol X. Hepatic hydrothorax. In: Light RW, YCG L, eds. *Textbook of pleural diseases*. 2nd ed. London: Hodder Arnold Pub, 2008: 402–9.
- 8 Kim SW, Lee S. Massive spontaneous diaphragmatic rupture induced by a squatting position. *Korean J Thorac Cardiovasc Surg* 2013;46:230–3.
- 9 Losanoff JE, Edelman DA, Salwen WA, *et al.* Spontaneous rupture of the diaphragm: case report and comprehensive review of the world literature. *J Thorac Cardiovasc Surg* 2010;139:e127–8.
- 10 Amin R, Waibel BH. Spontaneous diaphragmatic rupture in hypermobile type Ehlers-Danlos syndrome. *Case Rep Surg* 2017;2017:1–3.
- 11 Esteve M, Xiol X, Fernandez F, *et al.* Baliellas C: treatment and outcome of hydrothorax in liver cirrhosis. *J Clin Nutr Gastroenterol* 1986;1:139–44.
- 12 Zenda T, Miyamoto S, Murata S, *et al.* Detection of diaphragmatic defect as the cause of severe hepatic hydrothorax with magnetic resonance imaging. *Am J Gastroenterol* 1998;93:2288–9.
- 13 Cerfolio RJ, Bryant AS. Efficacy of video-assisted thoracoscopic surgery with talc pleurodesis for porous diaphragm syndrome in patients with refractory hepatic hydrothorax. *Ann Thorac Surg* 2006;82:457–9.
- 14 Chen A, Massoni J, Jung D, *et al.* Indwelling tunneled pleural catheters for the management of hepatic hydrothorax. A pilot study. *Ann Am Thorac Soc* 2016;13:862–6.
- 15 Orman ES, Lok ASF. Outcomes of patients with chest tube insertion for hepatic hydrothorax. *Hepatol Int* 2009;3:582–6.
- 16 Siegertetter V, Deibert P, Ochs A, *et al.* Treatment of refractory hepatic hydrothorax with transjugular intrahepatic portosystemic shunt: long-term results in 40 patients. *Eur J Gastroenterol Hepatol* 2001;13:529–34.
- 17 Spencer EB, Cohen DT, Darcy MD. Safety and efficacy of transjugular intrahepatic portosystemic shunt creation for the treatment of hepatic hydrothorax. *Journal of Vascular and Interventional Radiology* 2002;13:385–90.
- 18 Dhanasekaran R, West JK, Gonzales PC, *et al.* Transjugular intrahepatic portosystemic shunt for symptomatic refractory hepatic hydrothorax in patients with cirrhosis. *Am J Gastroenterol* 2010;105:635–41.

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