CASE REPORT - OPEN ACCESS

International Journal of Gastroenterology Hepatology and Endoscopy DOI: https://dx.doi.org/10.51521/IJGHE.2024.1208

Contents lists available at bostonsciencepublishing.us

International Journal of Gastroenterology Hepatology and Endoscopy

Recurrent Hepatic Hydrothorax in the bsence of Ascites in Liver Cirrhosis – A Challenging Condition to Diagnose and Treat: Case and Review of Literature

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ARTICLE INFO

Article history:
Received 04 December 2023
Revised 29 January 2024
Accepted 05 January 2024
Published 07 January 2024

KEYWORDS:
Hepatic hydrothorax,
Liver cirrhosis,
Pleural effusion.

ABSTRACT

Hepatic hydrothorax is a rare manifestation of chronic liver disease occurring in patients with advanced cirrhosis, portal hypertension and ascites. Hepatic hydrothorax is the excessive (>500 ml) accumulation of transudative fluid in the pleural cavity in patients with decompensated liver cirrhosis in the absence of cardiopulmonary, kidney and pleural diseases. We describe a case of liver cirrhosis who presented with rapidly accumulating right pleural effusion and on evaluation was found to have a transudate. There was no ascites on clinical examination or medical imaging. On thorough investigation for cardiac pulmonary and renal disease as well as malignancy the cause of effusion could not be ascertained. Finally, a diagnosis of hepatic hydrothorax was made based on existing liver cirrhosis and features of portal hypertension (esophageal varices). Hepatic hydrothorax in the absence of ascites is very rare but known entity and is believed to occur due to high absorptive capacity of peritoneum which surpasses the secretory rate. Presence of hepatic hydrothorax portends a poor prognosis and is associated with increased mortality. Treatment involves restricting salt, water intake & use of diuretics. Therapeutic thoracentesis is required in cases of respiratory depression. In resistant cases TIPS or an indwelling pleural catheter is placed and patients manage symptoms through intermittent drainage of pleural fluid. Definitive treatment is orthoptic liver transplantation..

INTRODUCTION

Hepatic hydrothorax is the accumulation of transudative fluid in pleural cavity usually in excess of 500 ml in a patient with decompensated liver cirrhosis that is not due to cardiopulmonary, renal or pleural/respiratory diseases. It is localized mostly to right side as almost 80-85% cases have right sided pleural effusion & only approximately 13% of cases have left sided pleural effusion whereas just 2% have fluid on both sides. Hepatic hydrothorax is observed rarely & depending on the diagnostic method is found in 5-10 % of patients constituting only 2-3% of all cases of pleural effusion. Hepatic hydrothorax is associated with poor prognosis and increased mortality. It causes respiratory failure which aggravates the clinical course of liver cirrhosis and the emergence of spontaneous bacterial pleural empyema leading to death in many cases [1]. The median survival of patients in presence of hepatic hydrothorax is noted to be only 8-12 months [2].

CASE PRESENTATION

A 50-year-old female, known case of chronic liver disease (CTP class C) related to Non-alcoholic fatty liver disease presented to our Outpatient department with progressive shortness of breath of 2 weeks' duration. She did not have any symptoms of chest pain, palpitations or abdominal distention. On examination she had dull note over right side of chest with markedly decreased vocal resonance & breath sounds. Cardiac examination did not reveal any abnormal findings and there were no clinical signs of ascites. On investigations chest X-ray was done which revealed massive pleural effusion on right side fig 1. USG Abdomen revealed features of coarse liver echo-pattern but there was no ascites. On evaluation for pleural effusion the analysis revealed Transudative effusion with a mixture of neutrophils, lymphocytes and mesothelial cells. Pleural fluid cytology was negative for malignant cells. Her Echocardiography showed normal left and right heart functions with no tricuspid regurgitation. There was no diastolic dysfunction. and serum BNP was also normal for age. Kidney functions as well as urine examination for proteinuria were also unremarkable. Having ruled out all causes of Transudative pleural effusion a possible Hepatic Hydrothorax was suspected as the cause for her pleural effusion. To confirm the suspicion, we needed a radioisotope 99mTc-sulfur colloid injection in peritoneal cavity and to assess the positive uptake of radioactivity within the right pleural cavity. However, these facilities were not available at our hospital. She was subjected to therapeutic paracentesis and on follow up after 2 weeks she again developed pleural effusion with no apparent Ascites. She was advised installation of pleural catheter for self intermittent drainage but the family planned to go for liver transplantation at a designated center.

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Table 1: Laboratory profile of the patients.

Variables	Value	Reference range
Hemoglobin	12.4 g/dl	13-16 g/dl
Albumin creatinine ratio	20 mg/gm	< 30 mg/gm
BNP	42 pg/ml	100 pg/ml
NT-PRO BNP	102 pg/ml	400 pg/ml
Bilirubin	0.8 mg/dl	0.8-1.2 mg/dl
ALT	58 IU/l	<45 IU/l
AST	50 IU/l	<45 IU/l
ALP	125 IU/l	<250 IU/l
Total protein	6.8 g/dl	6-8 g/dl
Albumin	2.3 g/dl	3.5-5 g/dl
Globulin	4.5 g/dl	2.3.5 g/dl
PT	20 s	12-16 s
INR	1.7	<1.5
BUN	23 mg/dl	7-20 mg/dl
Creatinine	0.9 mg/dl	0.5-1.5 mg/dl
Uric acid	4 mg/dl	4-6 mg/dl
Echo EF	60 %	50-70%
TAPSE	18 mm	<70

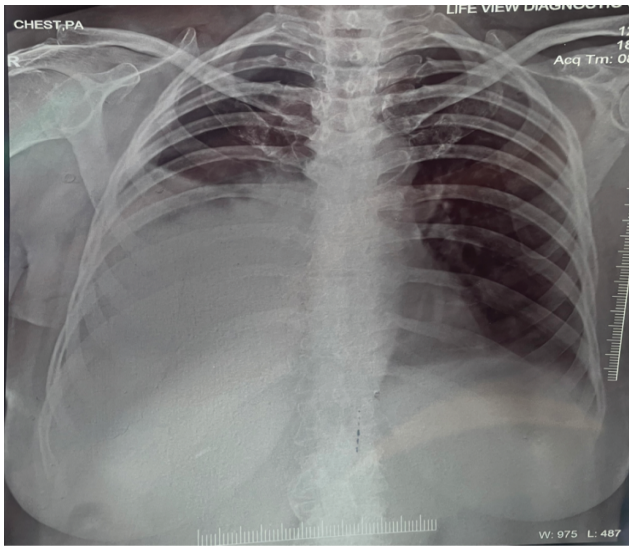

Figure 1: Chest X-ray showing large pleural effusion on right side.

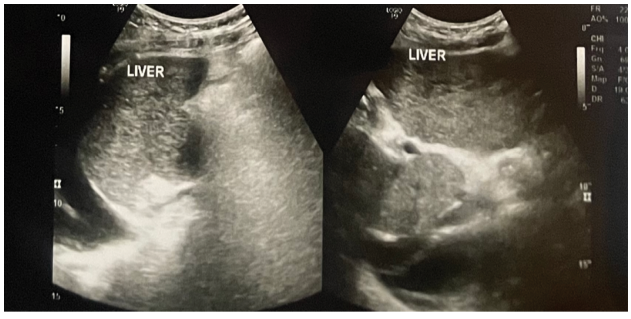

Figure 2: Bransabdominal ultrasound showing coarse liver echo pattern. No ascites is seen.

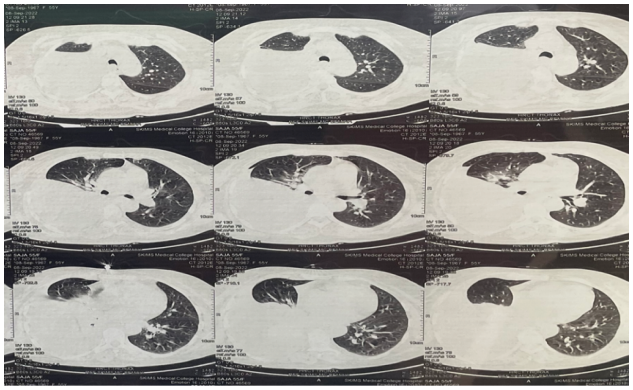

Figure 3: CT showing moderate right pleural effusion with passive collapse of right lower lobe.

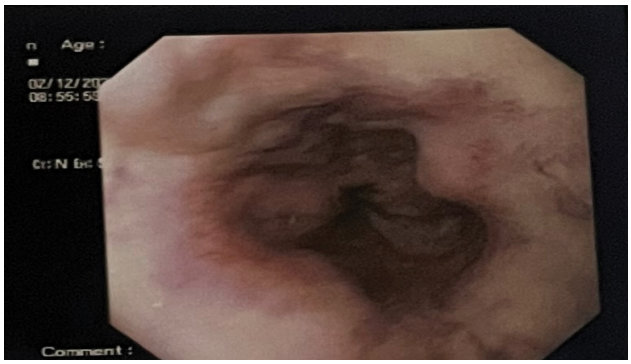

Figure 4: Endoscopy showing high grade esophageal varices.

DISCUSSION

Hepatic hydrothorax is defined as the transudative fluid that accumulates in pleural cavity in patients with decompensated liver disease [1]. The most probable pathogenesis of hepatic hydrothorax is the increase in intrahepatic vascular resistance due to massive structural changes associated with fibrosis and increased vascular tone in the hepatic microcirculation leading to portal hypertension [3]. This eventually causes splanchnic and systemic vasodilation as well as activation of various neurohormonal signaling pathways leading to hepatorenal syndrome and hence decreased sodium and water excretion as well as glomerular filtration rate (GFR) [1]. All these changes lead to accumulation of fluid in peritoneal cavity called as ascites. However, owing to some small defects located mainly on the right side of diaphragmatic tendon usually as well as the negative intra-thoracic pressure, the ascitic fluid at times moves rapidly from peritoneal cavity into the pleural space unidirectional due to piston effect. Also due to higher absorptive capacity of peritoneum and lymphatic drainage than pleura only pleural effusion may be detected with absence of concomitant ascites. This condition of hepatic hydrothorax in the absence of ascitis is seen rarely and its occurrence carries a poor prognosis with increased mortality. In an undiagnosed case of liver cirrhosis, diagnosis of hepatic hydrothorax in the absence of clinical ascites may be difficult and is a diagnosis of exclusion [4]. In patients suffering from liver cirrhosis clinical examination may reveal pleural effusion & allow preliminary diagnosis of hepatic hydrothorax [1]. The localization of fluid is usually on the right side . If fluid is detected on the left side along with respiratory symptoms & fever, other diseases including spontaneous bacterial pleural empyema should be excluded [1]. A pleural puncture is performed and the pleural fluid is analyzed which comes out to be a transudative fluid. The other causes of transudative pleural effusion like heart failure, nephrotic syndrome, myxedema, SVC obstruction should be ruled out especially in the absence of ascitis. In case the diagnosis is doubtful, Radioisotope techniques are used in which we inject 99mTc-labelled microspheres of human serum albumin or a Sulphur colloid into the peritoneal cavity & check for its migration into the pleural cavity in which case the radioactivity is usually detected in the right side

of pleural cavity. This confirms the diagnosis & the rate of isotope movement gives the idea about the size of defect in the diaphragm. [5] Therapeutic measures start from controlling ascites because the fluid preferentially keeps on accumulating in the pleural cavity and therefore we need to decrease the rate of ascitic fluid production. In these patients there is slight decrease in sodium excretion so sodium intake should be limited and stimulate its excretion by aid of diuretics. The consumption should be reduced to 80-120 mmol/day which corresponds to 4.6-6.9 g of salt/day [1]. In addition patients should also take either spironolactone or amiloride at initial dose of 50-200 mg/day & 5-10 mg/day respectively. Dose of spironolactone should be gradually increased by 100mg every 7 days to maximum dose is 400mg/ day. Treatment is effective if a body weight reduction of at least 2kg/week occurs. If monotherapy with spironolactone is inefficient or if hyperkalemia develops, furosemide at a starting dose of 40 mg/ day with gradual increase to a maximum of 160 mg/day. In case of Hepatic hydrothorax & tense ascites large volume paracentesis under albumin cover should be done in order to prevent circulatory collapse (8 g/l of ascitic fluid). There is an improvement in symptoms of Hepatic hydrothorax within 2 hrs. of large volume paracentesis. It also causes improvement of PaO_2/FiO_2 & lung volume at the end of exhalation [6,7]. Octreotide can play a beneficial role in the management of Hepatic hydrothorax owing to its ability to suppress the activation of RAAS induced by diuretics and increased excretion of sodium & water [8]. Barreales, *et al*., used Octreotide after ineffective use of diuretics and a low sodium diet at a rate of 25ug/h on 1st day, 50ug/h on 2nd & 100ug/h for next 5 days. The amount of fluid in pleural cavity decreased after 5th day. During a 6-month observation there was no relapse of Hepatic hydrothorax [9]. In another case a good result was achieved after a 5day course of Terlipressin therapy in combination with albumin infusion given to a patient with decompensated liver cirrhosis who had type 1 hepatorenal syndrome and hepatic hydrothorax. Repeated thoracocentesis is the routine procedure to remove fluid from pleural space in refractory Hepatic hydrothorax. This procedure is relatively safe even in patients who are at an increased risk of bleeding [10]. To avoid re-expansion pulmonary edema it is recommended to remove only 1L of transudate at a time. However, a study by Feller-kopman, *et al*., which included a study sample of 185 patients who underwent large volume thoracocentesis (1-3 L) did not find clinical and radiological signs of re-expansion pulmonary edema and even proposed to revise the recommendations & suggested to stop thoracocentesis only if there are unpleasant sensations in chest or decreased in pleural pressure to less than -20 mm H_2O at end of exhalation [12]. Drainage of pleural cavity by putting drains into the pleural cavity is one of the methods of draining the fluid. However, it is not recommended in refractory hepatic hydrothorax as it can lead to complications like pneumothorax, pleural empyema, occlusion of catheters & a large loss of fluid which leads to renal dysfunction and electrolyte imbalance. The trans jugular intrahepatic portosystemic shunt (TIPS) has been proposed as an alternative to paracentesis. TIPS reduces the rate of ascites recurrence mainly due to the reduction in the filtration pressure. In addition, TIPS results in a positive effect on renal function including hepatorenal syndrome, demonstrated by a rapid increase in urinary sodium excretion, urinary volume, and improvement in plasma creatinine concentration. Furthermore, plasma renin activity, aldosterone, and noradrenaline concentrations improve gradually after TIPS insertion suggesting a positive effect on systemic under filling, the factor of hepatorenal syndrome [13]. Pleurovenous shunting has also been described & used in patients for treatment of malignant pleural effusion. However, it is rarely used in hepatic hydrothorax. Pleurodesis can serve as a treatment modality in case of unsuccessful repeated thoracocentesis. Chemical Pleurodesis should be performed after removal of ascitic fluid. Some experts recommend to combine it with constant positive pressure in airways which decreases negative pressure in pleural cavity & hence prevents ascitic fluid flow in pleural cavity rendering it dry for longer periods [14]. The most common chemicals used for chemical Pleurodesis are talc, tetracycline, doxycycline, bleomycin, povidone-iodine & picibanil with or without minocycline [15]. Thoracoscopic mesh repair of diaphragmatic defects with or without Pleurodesis are also effective treatment options to control accumulation of fluid in pleural cavity. Most of the patients with hepatic hydrothorax are at a terminal stage of liver cirrhosis & hence are potential candidates for orthoptic liver transplant [16]. Liver transplantation is ultimately the only definitive treatment. Patients with hepatic hydrothorax did not require therapeutic paracentesis after transplantation [17].

CONCLUSION

Hepatic hydrothorax is rare but well known entity in advanced liver disease with gross ascites. However, hydrothorax in the absence of ascites has very rarely been reported in literature and occurs due to high absorptive surface of peritoneum as compared to pleura in addition to piston effect of diaphragm. It is a diagnosis of exclusion and all causes of transudative pleural effusion should be ruled out especially heart, renal and pleural disorders. Presence of hepatic hydrothorax in chronic liver disease occurs in advanced cirrhosis therefore harbinger of bad prognosis and is associated with worse short term mortality. Management is same as of ascitis with definitive treatment being orthoptic liver transplant.

Acknowledgement: Authors are grateful to the patient for his cooperation throughout follow up.

Conflict of interests: None declared

Source of funding: Nil

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