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Refractory Chylous Ascites with Chylothorax and an Umbilical Hernia in a Patient Ineligible for a Transjugular Intrahepatic Portosystemic Shunt

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Female, 21-year-old
Final Diagnosis: Chylous ascites
Symptoms: Pitting edema • shortness of breath
Medication: —
Clinical Procedure: Paracentesis • thoracentesis
Specialty: Gastroenterology and Hepatology • General and Internal Medicine • Palliative Medicine

Objective: Rare disease

Background: Chylous ascites is a rare condition, which is defined by accumulation of a milky fluid due to high triglyceride levels. It is most commonly secondary to malignancy, liver cirrhosis, infection, and tuberculosis.

Case Report: A 21-year-old woman from rural Indonesia, came to the hospital with chronic dyspnea and a history of repeated paracentesis. Six years ago, she was diagnosed with chronic hepatitis B. For the past 2 years, she had complaints of progressive dyspnea and increased abdominal swelling. On examination, there was dullness on chest percussion and decreased breath sounds. Shifting dullness was positive on abdominal examination. Paracentesis and thoracentesis were performed and showed high triglyceride levels. She underwent an abdominal computed tomography scan and was diagnosed with liver cirrhosis, complicated with chylous ascites and chylothorax. Repeated paracentesis was performed as a therapeutic approach; she had strict diet guidelines, and was prescribed octreotide, furosemide, spironolactone, and albumin. Despite this treatment, two years later, she developed an umbilical hernia complicated with ulceration. Hernia repair was not possible due to her comorbidities. She was indicated for a transjugular intrahepatic portosystemic shunt (TIPS) for the refractory chylous ascites. However, this could not be performed as the patient could not afford this expensive procedure, which was not covered by insurance.

Conclusions: Management of refractory chylous ascites is challenging, especially in underdeveloped countries due to socioeconomic problems and limited health care facilities. Although TIPS is indicated in refractory chylous ascites, repeated paracentesis can be useful as an alternative method.

MeSH Keywords: Chylothorax • Chylous Ascites • Hepatitis B • Liver Cirrhosis • Paracentesis

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Background

Chylous ascites is characterized by the accumulation of milky lymphatic fluid within the peritoneal cavity. The diagnosis is confirmed by triglyceride levels of >200 mg/dL [1]. The incidence of chylous ascites was reported to be 1 in 20,000 hospital admissions and this number could increase due to the increased survival rates of patients with chronic liver disease or malignancies[2]. Development of chylous ascites is seen in 0.5% of cirrhosis cases [3]. Treatment for chylous ascites is currently focused on managing the etiologies, dietary adjustment, and pharmacological interventions, such as orlistat, somatostatin, octreotide, and etilefrine [1]. Definitive therapies, such as transjugular intrahepatic portosystemic shunt (TIPS) and liver transplants are preserved for refractory cases or in cases contraindicated for pharmacological therapy. Here, we report the role of palliative therapy with a strict diet, pharmacological interventions, and repeated therapeutic paracentesis in a case of refractory chylous ascites and chylothorax in a patient with cirrhosis.

Case Report

A 21-year-old woman from rural Indonesia presented at the hospital with chronic dyspnea and a history of repeated paracentesis.

Her chief complaint was dyspnea for the past 2 years. She had history of a hepatitis B infection diagnosed 6 years prior. Four years after the initial diagnosis, her condition was complicated by progression to decompensated liver cirrhosis stage 3. During the current visit, the patient complained of progressive dyspnea that occurred throughout the day. It was more prominent while sleeping and therefore, she needed 3 pillows to relieve the dyspnea. Her symptoms were also exacerbated by simple activity, such as walking 200 meters. She had abdominal swelling, which appeared first, followed by the development of edema in her lower extremities. Her past medical history included pulmonary tuberculosis, successfully treated with antituberculosis treatment. The patient had no history of previous surgery. There was no family history of similar illness.

On physical examination, the patient appeared weak and cachectic. There was an increase in jugular venous pressure. On chest examination, there were decreased breath sounds accompanied by dullness on percussion over the lung field up to the second intercostal space. The patient's abdomen was distended with positive shifting dullness. Her lower extremities had pitting edema. Other physical findings were unremarkable.

A chest X-ray was performed because of her dyspnea and abnormal chest examination findings. The results showed bilateral effusion up to the fourth and fifth intercostal spaces. Contrast chest and abdominal computed tomography (CT) scans were performed for confirmation of the effusion. The chest CT scans showed massive pleural effusion on the left lung with compressive atelectasis on the lower lobe. There was infiltration in the upper right lobe, and the mediastinum was pushed to the right side. Liver cirrhosis was seen on the abdominal CT scan. It was complicated by esophageal varices, splenomegaly, massive ascites, and bilateral portal vein occlusion with dilation of the major portal vein, splenic vein, and coronary vein. There was no sign of a space-occupying lesion or hepatoma. Echocardiography was performed and showed no abnormalities.

Paracentesis and thoracentesis were performed and the fluids analyzed for the first time. Fluid analysis from the paracentesis showed a white cloudy liquid with triglyceride levels of 954 mg/dL, a lactic acid dehydrogenase (LDH) ratio of 0.17, and protein ratio of 0.19. Pleural fluid from the thoracentesis appeared brown and cloudy, with triglyceride levels of 1130 mg/dL, an LDH ratio of 0.52, and a protein ratio of 0.27. Both fluids were transudative. Cytology from the fluids showed no signs of malignancy. HBsAg was positive-reactive and the aspartate transaminase (AST) and alanine transaminase (ALT) levels were within normal limits.

She was then admitted to the ward with a diagnosis of concomitant chylous ascites and chylothorax. She was administered octreotide (2×100 mcg/per day), albumin, and paracentesis was managed with a chest tube placement. After improvement of the dyspnea, furosemide (3×40 mg) and spironolactone (2×100 mg) were prescribed and she was discharged from hospital after 4 days.

Within a year, despite her adherence to medications and diet, she complained of several episodes of exacerbated dyspnea. Therefore, she was managed with therapeutic paracentesis and a chest tube. At the one year follow-up, thoracentesis was performed, despite her exacerbating symptoms of shortness of breath (Figure 1). The result yielded 600 cc of red-colored fluid with 89% mononuclear cells and a positive Rivalta test, which indicated exudative fluid.

Two years later, her dyspnea had progressively worsened, and she was admitted again to the hospital. She was managed conservatively with medications and repeated therapeutic paracentesis, and after some improvement, she was discharged after 4 days. In addition, her umbilicus enlarged within 2 weeks of her last hospital discharge, and there was a yellowish-white fluid oozing out from the ulcerated umbilicus. After consultation with the general surgeon, the patient was diagnosed with an umbilical hernia and scheduled for elective hernia repair. This was delayed because of her comorbidities. At the time of admission, paracentesis and thoracentesis were

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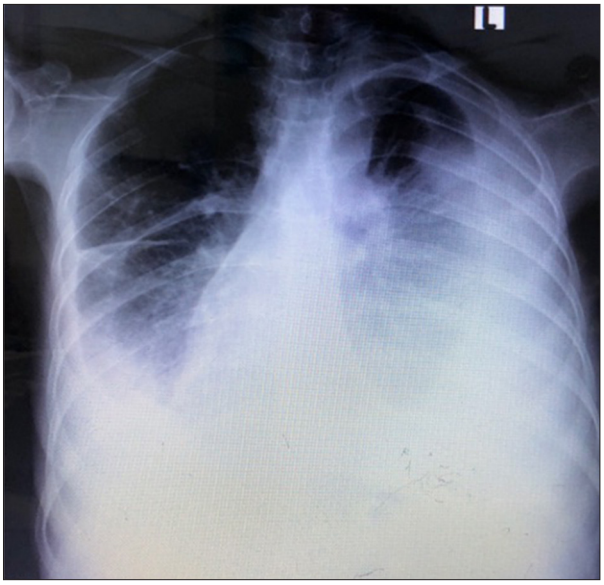

Figure 1. Chest X-ray at the one-year follow-up showed bilateral pleural effusion.

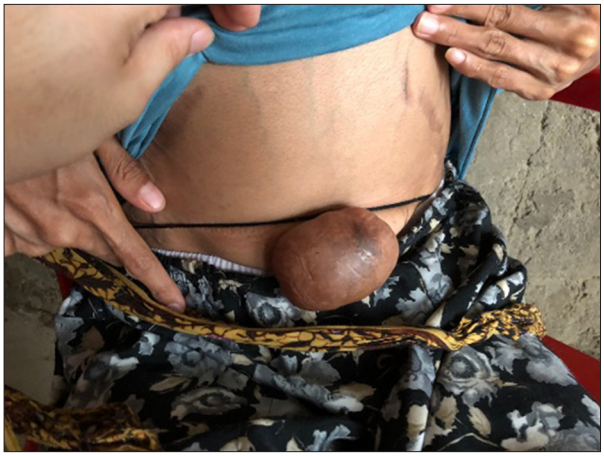

Figure 2. Follow-up 6 months after diagnosis of the umbilical hernia showed an improvement in ulceration on the hernia with no discharge.

performed. Paracentesis showed triglyceride levels decreased to 100 mg/dL, an LDH ratio 0.17, and protein ratio of 0.16. Thoracentesis showed an LDH ratio of 2.02 and protein ratio of 0.58. The Ziehl-Neelsen test was performed to rule out tuberculosis as the cause of the exudative fluid and the result was negative. She was managed once again with the placement of a chest tube, therapeutic paracentesis, octreotide, and albumin. She was discharged once more with the same prescribed dose of furosemide and spironolactone as before.

Six months after the initial development of the umbilical hernia, there was an improvement in the ulceration and decrease in size of the hernia (Figure 2). Although there was an improvement in her ascites, edema and umbilical hernia, she still had dyspnea recur throughout the day, exacerbated by simple activities.

The patient gave verbal consent only for publication of this case report in a scientific journal and publication of photographs in the scientific journal without revealing her identity. The hospital's institutional review board approved publication of this case report in a scientific journal without revealing the patient's identity.

Discussion

Chylous ascites is diagnosed by gross examination and presence of high levels of triglycerides in the fluid from the paracentesis. Establishing the etiology is important as chylous ascites is mainly secondary to other conditions. In developed countries, chylous ascites is most commonly caused by malignancy and cirrhosis; while other etiologies are infection, trauma, and iatrogenic causes [1,4]. In this patient, cirrhosis was a possible cause of the chylous ascites, while malignancy and infection were ruled out as secondary causes. Chylous ascites is a rare condition and presents as a secondary to chronic disorders with liver cirrhosis as a causal factor in 0.5% cases [3–5].

Chylothorax is characterized by the accumulation of lymphatic fluid in pleural space. It can be caused by nontraumatic conditions, such as lung cancer, infection, and obstruction or by a traumatic event [6]. While chylothorax can appear as a single condition, it can be associated with chylous ascites [7]. It is mainly caused by extravasation of ascitic fluid from chylous ascites to the pleural space via a microscopic defect in the diaphragm [7].

Diagnosis of chylous ascites in this patient was confirmed by the presence of high triglycerides (954 mg/dL) and the characteristic appearance from the paracentesis. The development of chylous ascites was mainly secondary to cirrhosis based on the CT scan findings. Cirrhosis contributes to an increase in portal pressure, leading to excessive hepatic and gastrointestinal lymphatic flow [2]. This patient presented with dyspnea as the chief complaint due to an increase in intra-abdominal pressure because of the accumulation of ascitic fluid [1]. In addition to the dyspnea, the patient also presented with classic findings of liver cirrhosis, such as ascites, abdominal swelling, and edema of the lower extremities. Other etiologies, such as malignancy, tumor, and tuberculosis infection were ruled out based on the abdominal CT scan, cytology of the paracentesis fluid, and culture of the fluid. This patient had a positive HbsAg corresponding to an active hepatitis B infection, which explains the development of liver cirrhosis in this patient.

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In this case, the pleural effusion was seen on the chest X-ray. Thoracentesis was performed for diagnostic purposes, and yielded a high triglyceride level of 1130 mg/dL. This similarity to chylous ascites might explain the extravasation of ascitic fluid from the abdominal cavity to the pleural cavity. Given the patient's history of pulmonary tuberculosis, the pleural fluid was cultured in order to exclude a tuberculosis reinfection as the primary cause.

In addition to the chylous ascites and chylothorax, complications to the abdominal wall hernia occur in 20% to 40% patients with liver cirrhosis and ascites [8]. This patient had a complicated umbilical hernia mainly due to increased intra-abdominal pressure and weakened abdominal muscles associated with malnutrition[8]. An abdominal wall hernia is associated with refractory ascites and increased complications, such as incarceration, evisceration, ascites drainage, and peritonitis [8]. In addition, umbilical hernia repair in patients with liver cirrhosis has a high mortality rate [9]. Therapeutic paracentesis can reduce ascites and have some benefit in preventing recurrence of the hernia [10]. On the other hand, emergent surgical repair is indicated in a patient with an umbilical hernia and refractory ascites due to a higher risk of incarceration caused by repeated large volume paracentesis [11]. This complication can develop due to decompression of ascitic fluid that leads to entrapment of the small bowel in the hernia ring. Surgical repair may be done in patients with liver cirrhosis after improvement of the condition [11,12]. However, in this case, the surgeon deferred the hernia repair due to comorbidities and the refractory condition.

Recommended management of chylous ascites is by dietary adjustment with a low-fat and high-protein diet in addition to octreotide [1,13]. This patient was managed with octreotide and albumin during hospitalization. A chest tube placement and paracentesis were performed for diagnostic and therapeutic purposes. She had repeated paracentesis due to exacerbation of her symptoms. Administration of octreotide in this case showed an improvement in the chylous ascites that is explained by an improvement in the triglyceride levels on subsequent paracentesis. Despite improvement in the chylous ascites, she still complained about persistent dyspnea. While other symptoms can accompany ascites, the patient's symptom of dyspnea is refractory despite medications and repeated therapeutic paracentesis. It is explained by minimal improvement of symptoms reported by her even at rest and doing daily activities, as well as development of the umbilical hernia as a complication. Because of these refractory symptoms, this patient is indicated for definitive therapy [14]. TIPS is indicated, as definitive therapy, for patients with refractory conditions, who have had repeated paracentesis without any significant improvement [15]. However, as there were socioeconomic limitations, she underwent repeated paracentesis as long-term management, in addition to octreotide and albumin replacement. Even with minimal improvement in symptoms and quality of life, at this point, conservative measures such as repeated therapeutic paracentesis with furosemide and spironolactone, are the best modalities available, in cases where TIPS is not possible, due to socioeconomic problems and/or limited facilities [16–18]. However, long-term management must be adjusted with repeated paracentesis and medications due to the presence of the umbilical hernia as it might increase morbidity and mortality in this patient.

While there is no guideline regarding therapeutic paracentesis for patients with chylous ascites who have refractory symptoms, further studies are needed to determine the benefit and harm in long-term management of therapeutic paracentesis in patients with refractory chylous ascites complicated with chylothorax and umbilical hernia.

Conclusions

Management of a patient with refractory chylous ascites due to liver cirrhosis is challenging, especially in association with socioeconomic problems and comorbidities. Its treatment and outcome are tailored to the patient's condition and depend on the experience of the medical team. Repeated paracentesis can be useful in managing patients with refractory chylous ascites. Although this approach is not the optimal treatment, it can be one of the many modalities, especially in underdeveloped countries. The palliative term should be weighed on the risk and benefit of invasive procedures, and the patient must be carefully observed through strict follow-ups to prevent comorbidities and lower the mortality rate.

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Department and Institution where work was done

Department of Internal Medicine, Siloam General Hospital, Tangerang, Indonesia.

Conflict of interests

None.

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