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Case Report

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Flood Syndrome and Portal Vein Thrombosis: An Unusual Complication of Liver Cirrhosis

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ABSTRACT

Spontaneous paracentesis is a life-threatening complication of liver cirrhosis. Importance of precise and early identification cannot be overstated, as early treatment improves the prognosis and survival. A 78-year-old woman with cirrhosis presented with ascitic fluid gushing out from an umbilical hernia. The patient received intravascular repletion with intravenous albumin, antibiotics, and rapid umbilical herniorrhaphy. Post-operatively, the patient experienced acute kidney injury and portal vein thrombosis, which was corrected with electrolyte replacement and transjugular intrahepatic portosystemic shunt with thrombectomy. The patient recovered completely and was discharged without difficulty.

Introduction

Flood syndrome, also referred to as spontaneous paracentesis, is an extremely rare cirrhosis complication in which ascitic fluid ruptures from the abdomen through a spontaneously perforated umbilical hernia. This uncommon condition can be life-threatening; however, due to its rarity, there is no established or recommended management strategy. This report presents a case of Flood syndrome in a 78-year-old woman with cirrhosis who received intravascular repletion with albumin and had a rapid umbilical herniorrhaphy. The clinical course was complicated by acute kidney injury (AKI) and portal vein thrombosis. After undergoing a transjugular intrahepatic portosystemic shunt (TIPS) with thrombectomy to manage the thrombus, the patient recovered without incident.

Case Presentation

A 78-year-old woman with a history of cryptogenic cirrhosis presented to the emergency room with an ascitic fluid gushing out from an umbilical hernia. The patient was diagnosed with liver cirrhosis roughly one-year prior with no etiology identified; the disease course was complicated by esophageal varix (grade I) and ascites that were managed with diuretics and paracentesis. Several days prior to admission, the

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patient noticed increasing abdominal distension and a formation of a small protuberance at the umbilicus. The bulge progressed to become erythematous and warm, as the abdomen became enlarged (Figure 1).

Then, the day prior to admission, the patient noticed leakage of a straw-colored fluid from the umbilical bulge. The patient instantly filled four cups of the fluid (estimated to be greater than 2 liters); however, the patient did not have associated abdominal pain, nausea, vomiting, lightheadedness, or other systemic signs of illness. Upon evaluation, the patient was hemodynamically stable, afebrile, and saturated 100% on room air without respiratory distress. Physical examination of the patient's abdomen was soft and non-tender with bowel sounds present; in addition, the umbilical region was erythematous, and a straw-colored liquid was leaking from the left side of the umbilicus.

Laboratory values were noted and depicted in Table 1. Liver enzymes like serum alanine aminotransferase (ALT), serum aspartate aminotransferase (AST), and total bilirubin were also documented. Table 1 further depicts the patients' complete blood count (CBC) measuring several components and features of the patient's blood, including white blood cell count (WBC), hemoglobin, and platelets.

Computed tomography (CT) scan of the abdomen and pelvis with intravenous contrast revealed a cirrhotic liver with various portosystemic collaterals, including distal esophageal varices, mesenteric edema with small bowel thickening, and a significant amount of

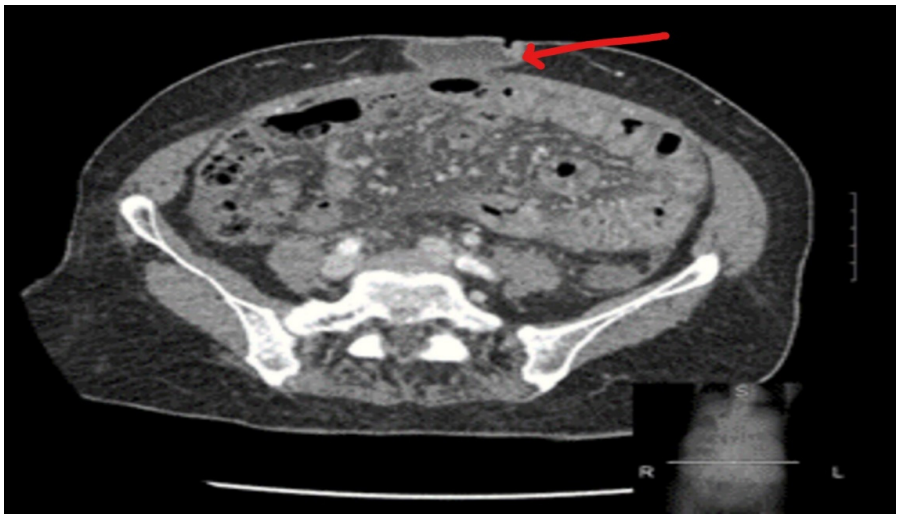

Fig. 1. Radiological findings of the abdomen showing umbilical hernia with an opening, a weakness that developed in the abdominal wall through and around the abdominal muscles.

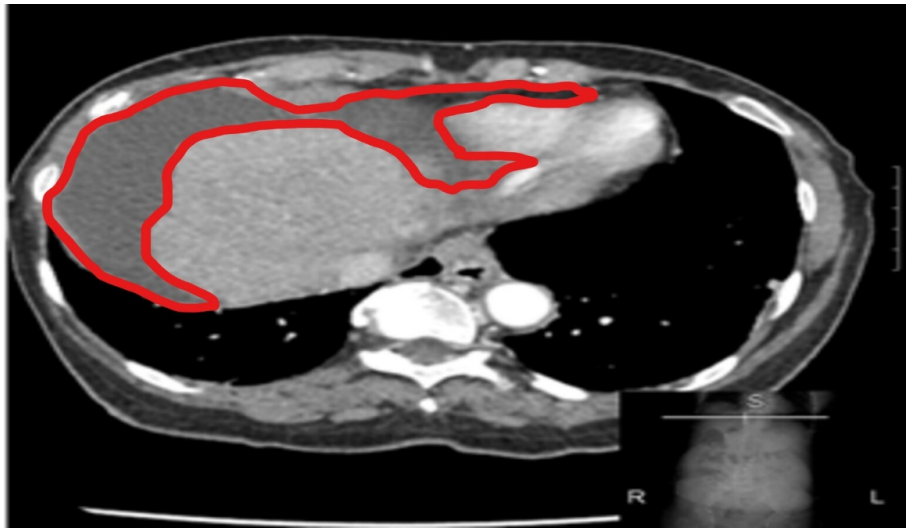

Fig. 2. Radiological findings of the abdomen showing liver with ascitic fluid, resulting from high pressure in the blood vessels of the liver.

Table 1. Patient's laboratory findings upon evaluation in the emergency department.

Laboratory parameter	Reference range and unit	Patient's value
Sodium	136-145 mEq/L	135 mEq/L
Potassium	3.5-5.0 mEq/L	4.1 mEq/L
Chloride	95-105 mEq/L	106 mEq/L
Carbon dioxide	33-45 mmHg	25 mmHg
Blood urea nitrogen (BUN)	7-18 mg/dL	11 mEq/L
Creatinine	0.6-1.2 mg/dL	1.08 mEq/L
Glomerular filtration rate (GFR)	> 60 mL/min/1.73 m^2	49 mL/min/1.73 m^2
Alanine aminotransferase (ALT)	8-20 U/L	11 U/L
Aspartate aminotransferase (AST)	8-20 U/L	44 U/L
Total bilirubin	0.1-1.0 mg/dL	1.2 g/L
White blood cell (WBC)	4500-11,000 / mm^3	7.4 x 10^3 uL
Hemoglobin	Female: 12.0-16.0 g/dL	11.6 g/dL
Platelets	150,000-400,000 /mm^3	248 x 10^3 uL

ascitic fluid around the liver (Figure 2).

Albumin infusion and piperacillin-tazobactam were given to the patient immediately. Surgery was consulted, and the patient had an umbilical hernia suture repair with a Jackson-Pratt (JP) surgical drain. The patient was observed for the post-operative course for the next two days (a total of four days after the original hospital admission) before being discharged home with the JP drain in place. Two weeks later, the patient called the office for feeling short of breath and having observed a significant abdominal JP drain output (i.e., when the drainage is 30 mL or less over 24 hours). On evaluation, the patient was found to have laboratory findings consistent with AKI (creatinine increase to 1.3 mEq/L from baseline 1.0 mEq/L), hyperkalemia (potassium of 5.5 mEq/L), and hyponatremia (sodium of 128 mEq/L). The patient was admitted and underwent a CT scan of the abdomen/pelvis, which revealed a near complete thrombosis of the intrahepatic portal vein (including right and left portal branches distal to the portal conference).

To assess for signs of esophageal varices with elevated stigmata, an esophagogastroduodenoscopy (EGD) was performed. The EGD indicated grade 2 esophageal varices; the patient was deemed not a candidate for systemic anticoagulation. To treat the portal vein thrombosis the patient underwent TIPS with thrombectomy. The procedure was successful; however, hospitalization was complicated by the development of a fever, the source of which was later determined to be an abdominal infection (JP drain culture was positive for methicillin-susceptible *Staphylococcus aureus*), presumed to be secondary to the recent abdominal procedure. The patient received intravenous antibiotics and recovered uneventfully. Laboratory derangement had slowly stabilized to acceptable ranges. The JP drain was removed as there was no longer a significant output. The patient was discharged home with a recommendation for further monitoring as an outpatient follow-up.

Discussion

Flood syndrome is perhaps aptly named after its unusual and yet impactful presentation of an ascitic fluid rupturing through an umbilical hernia and "flooding out." This phenomenon was first reported in 1959 under the title of "Spontaneous Abdominal Paracentesis" [1]. It is then presumed to be officially coined after Dr. Frank Flood's description of five cases in 1961, all of which had died after suffering from a spontaneous perforation of the umbilical hernia [2]. The pathogenesis of this fatal disease first involves a rapidly formed umbilical hernia in cirrhotic patients; umbilical hernia is present in 20% of patients affected with end-stage liver disease, compared to only 2%

in the general population [3,4]. This is thought to be related to increased intra-abdominal pressure caused by ascites, which causes the abdominal contents to protrude through a probable umbilicus defect [5-7]. In addition, recanalization, dilation, and variceal formation from portal hypertension can precipitate a supraumbilical fascial defect; moreover, poor nutritional status can contribute to a weakening of the abdominal wall muscle [8,9]. As the ascitic fluid builds up in the peritoneal cavity, it poses pressure on the weakened portion of the abdominal wall at the linea alba and leads to the dehiscence of the umbilical hernia. Then the umbilical hernia perforates and the ascitic fluids surge out, giving rise to the name "spontaneous paracentesis."

In today's literature, there is very little information about Flood syndrome [10]. Certainly, an anomaly of the complications of cirrhosis, Flood syndrome can present abruptly, and it can be fatal if not addressed promptly. Complications can include evisceration of the small intestine with possible incarceration, hypotension, resultant end-organ damage such as renal failure (due to sudden large volume removal), cellulitis, peritonitis, and sepsis [11]. Skin ulceration or necrosis may occur prior to the rupture of the umbilical hernia in many cases, and this should be taken seriously [12]. Rupture of the umbilical hernia may follow a sudden increase in the intra-abdominal pressure such as coughing, vomiting, or straining. Management strategy is up for debate as there are no established guidelines or recommendations. The mainstay of initial focus should be on prompt medical evaluation of the hemodynamic stability, as patients can rapidly deteriorate after losing such a large volume in a short period of time.

A surgical examination for herniorrhaphy is also recommended, while the time of operation is debatable. Given the significant morbidity and postoperative problems associated with cirrhotic patients, such as bleeding from coagulopathy, protracted wound infection, encephalopathy, and rapid decompensation of liver function, some specialists advise against urgent surgical repair. Other anatomic or functional variables must be considered as well, for instance, if the umbilical vein is still intact, umbilical herniorrhaphy would be contraindicated as ligation of the umbilical vein can lead to an obstruction of the portal circulation outflow, causing an acute portal vein thrombosis and acute liver failure [13]. However, it must also be noted that sole conservative measures are also associated with a high risk of spontaneous bacterial peritonitis. Therefore, clinical judgment is required in the management of Flood syndrome which should be individualized because the anatomy of collateral vessels significantly differs among patients.

Many experts from case series seem to voice the same recommendation for the initial management: careful patient monitoring and stabilization followed by medical peritoneal diversion. Some experts achieved this through cautious drainage of ascites, peritoneal dialysis, or employing a portosystemic shunt system such as TIPS. The latter approach seems to have been utilized frequently among the experts. One group of clinicians used splenic embolization and percutaneous peritoneal drainage with a good outcome [14]. Volume replacement with intravenous solution and prevention of infectious complications through broad-spectrum antibiotics also seemed to confer morbidity benefits [15]. Then, after medical stabilization is achieved, surgical herniorrhaphy is to be pursued. In our case, the patient was managed with volume replacement with albumin, local wound treatment, and broad-spectrum antibiotics. This patient was then referred to surgery, who deemed the patient an adequate candidate for umbilical herniorrhaphy. The patient developed acute renal injury and an electrolyte imbalance that could be caused by over-drainage of ascites via JP drain catheter. In addition, the patient was found to have a portal vein thrombosis on subsequent imaging and was eventually treated with thrombectomy via TIPS. The patient made a full recovery without any difficulties.

Study limitations

The case has some limitations which includes the inability to retrieve the albumin and prothrombin values for the patient; however, we believe that the albumin value is below 3.4 grams per deciliter (g/dL) (normal range is 3.4 to 5.4 g/dL (34 to 54 g/L) due to significant loss of fluid and liver impairment resulting to albumin infusion that was given to the patient. Also, we believe that the prothrombin time is longer than the normal range (11 to 13.5 seconds) or (normal INR of 0.8 to 1.1) due to the liver impairment. Also, we could not obtain the initial laboratory findings of the ascites before JP drainage and after i.e., cell d/c, albumin, LDH, PMN count, glucose, protein, culture etc.; however, we also believe that a positive culture of the ascites could have resulted to piperacillin-tazobactam being given to the patient before the JP drainage and the positive culture with methicillin-susceptible *Staphylococcus aureus* reported for the ascites after JP drainage.

Other unknowns include the exact dosage of intravenous albumin, whether albumin was infused

during whole hospitalization period, whether umbilical varix was ligated during surgery, whether the patient was on diuretics, whether the patient was dehydrated after drainage, information on discharge medication and information on the long-term follow-up results of the patient regarding TIPS patency and portal vein thrombosis recurrence could not be obtained. Lastly, image of TIPS with thrombectomy is also unavailable. Thus, results obtained in the study should be interpreted as presented.

Conclusion

In summary, intravascular repletion with intravenous albumin and rapid umbilical herniorrhaphy is an effective approach to providing support to patients with spontaneous paracentesis. Its application in patients with liver cirrhosis is an infrequently reported, yet potentially life-saving management strategy. However, when additional complications arise, a rapid multidisciplinary approach (i.e., electrolyte correction and TIPS) may be required to control the sequelae of AKI and portal vein thrombosis. Also, management of Flood syndrome should be individualized because the anatomy of collateral vessels significantly differs among patients. This case study demonstrates the potential value of intravascular repletion with intravenous albumin and prompt therapy of umbilical herniorrhaphy, as well as the ongoing necessity of contingency planning should new issues arise.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors have no conflict of interest to declare.

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