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Case Report

Transjugular intrahepatic portosystemic shunt for chylous ascites in a patient with recurrent cirrhosis following liver transplantation

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ABSTRACT

Chylous ascites (CA) is the extravasation of lipid-rich lymphatic fluid into the peritoneal space following trauma or obstruction of the lymphatic system. Refractory cases of cirrhosis-related CA may be amendable to transjugular intrahepatic portosystemic shunting (TIPS). We present a case of TIPS in the setting of refractory CA secondary to cirrhosis of a transplanted liver graft. Following TIPS, the patient reported immediate improvement in abdominal pain and no longer requires paracentesis. Our case suggests TIPS to be a safe and effective treatment option for CA in liver transplant patients with cirrhosis.

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Introduction

Chylous ascites (CA) is a rare form of ascites defined as the extravasation of lipid-rich lymphatic fluid into the peritoneal cavity [1]. Disruption of the lymphatic system secondary to traumatic injury or obstruction produces CA, which is turbid or milky in appearance from high triglyceride levels, usually more than 1.10 kg/m³ [1,2]. Intra-abdominal malignancies and cirrhosis account for two-thirds of all cases in developed countries, whereas infection (e.g., tuberculosis) is the predominant cause in the developing world [3]. CA can also

occur following orthotopic liver transplantation (LT), particularly in patients with preoperative ascites [4]. Treatment of the underlying cause is crucial. Conservative medical management of CA includes a high-protein low-fat diet and therapeutic paracentesis [1]. Resistant cases may be amendable to peritoneovenous shunting and transjugular intrahepatic portosystemic shunting (TIPS). TIPS is reserved for cirrhosisrelated CA; however, its use in cirrhosis of a transplanted liver graft has not been well documented. A patient is reported with refractory CA and cirrhosis after LT, in whom the CA was treated with TIPS.

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Case report

A 55-year-old male with a history of orthotopic LT 10 years ago for hepatitis C virus (HCV)-induced cirrhosis was referred to interventional radiology for management of resistant ascites. He developed recurrent HCV cirrhosis of the transplanted liver which was confirmed by liver biopsy (grade 2 stage III cirrhosis). Subsequently, the patient developed persistent CA. Splenic artery embolization was performed 3 years ago to control the ascites, but the patient had only minimal improvement in ascites volume. Two years ago, he underwent hepatic lymphangiography which demonstrated an amorphous area communicating with the hepatic veins and lymphatic channels (Fig. 1). The area was embolized with approximately 0.3 mL of the Onyx Liquid Embolization System (Medtronic, Minneapolis, MN). Notwithstanding, he continued to require weekly therapeutic paracenteses and presented multiple times to the emergency department with intractable abdominal pain. Ascitic fluid was always milky and consistently showed normal lymphocyte, neutrophil, and erythrocyte counts; and elevated triglyceride levels-ranging from 2.39–3.06 kg/m³—diagnostic of CA. Additionally, the ascites to plasma triglyceride ratio was consistently elevated (greater than 5:1). Furosemide, aldactone, and a low-fat diet supplemented with medium-chain triglycerides were tried but failed to alleviate the CA. He was ultimately referred for TIPS creation.

Before TIPS placement, paracentesis was done and drained 3 L of CA. Next, the right internal jugular vein was accessed, and the right hepatic vein was cannulated. The venous pressures were measured, and portosystemic gradient was 13 mm Hg. A CO₂-wedged porto-venogram showed a patent PV. The right PV was next accessed with a Rosch-Uchida needle (Cook Medical, Bloomington, IN). The parenchymal tract was dilated with an 8 mm \times 8 cm balloon and a 10 mm \times 9 cm Viatorr stent



Fig. 1 – Hepatic lymphangiogram showing amorphous collection lymphatic structures (bottom arrow) communicating with hepatic veins (top arrow) and lymphatic channels.

(Gore, Newark, DE) was then placed (Fig. 2). Porto-venogram after stent placement demonstrated brisk shunting between the PV and the right atrium. Post-TIPS portosystemic gradient was 1 mm Hg. After the procedure, the patient reported improvement in abdominal pain and fullness. Weekly paracenteses were performed up to 2 months after TIPS, but the volumes consistently decreased. Three months after the procedure, the patient did not require therapeutic paracentesis.

Discussion

CA is uncommon, accounting for only 0.5%—1% of cirrhosisrelated ascites [5]. Spontaneous rupture of the serosal lymphatic channels, secondary to excessive lymph flow, is thought to cause CA in cirrhosis [5]. Hepatic lymph flow increases in cirrhosis due to increased pressure gradient between the capillaries and interstitial compartments of the liver [5]. The pressure gradient itself increases secondary to cirrhosis-induced sinusoidal and post-sinusoidal portal hypertension (PH) [5]. Moreover, PH results in splanchnic venous congestion, which increases gastrointestinal lymphatic flow [5].

The incidence of CA in LT patients is similar to that of non-LT patients. In a study of 516 LT patients, 24 developed CA (0.5%) [4]. CA occurs after LT through two separate, or combined, mechanisms. One mechanism, as previously described, involves increased lymph production and flow secondary to cirrhosis [4]. The second mechanism pertains to intraoperative injury of periportal and retrohepatic lymphatic vessels and inadequate ligation of the damaged vessels [4]. In



Fig. 2 – Portal venogram demonstrating patency of the intraparenchymal tract and visualization of Viatorr stent. Note donor inferior vena cava stump (arrow).

addition, preoperative ascites and the use of a LigaSure (Medtronic, Minneapolis, MN) vessel sealing system (as opposed to conventional suture ligation) are independent risk factors for the development of CA after LT [4].

Studies addressing the optimal treatment of CA are limited [3]. However, a stepwise approach—escalating from conservative medical management to invasive procedures-is the consensus [5]. Most patients respond initially to a high-protein and low-fat diet with medium-chain triglycerides (MCT) [3]. The restriction of long-chain triglycerides minimizes the breakdown of these into monoglycerides and free fatty acids (FFAs), which are carried in chylomicrons through the intestinal lymphatic ducts [3]. In contrast, MCTs are converted to FFAs and glycerol and transported directly from the intestines to the liver via the portal vein, never entering the lymphatic system [3]. Thus, a low-fat diet supplemented with MCTs decreases the production and flow of chyle [3]. Cirrhosis-induced CA is also amendable to sodium restriction and diuretics [3]. If patients do not respond, bowel rest and total parenteral nutrition may be necessary [3]. Additionally, therapeutic paracentesis can provide immediate and significant relief of dyspnea and abdominal pain, although prolonged drainage can result in electrolyte imbalances and infection [3,5].

If cirrhosis-related CA fails to respond to the above measures, TIPS creation may provide symptomatic improvement [5]. The mechanism by which TIPS ameliorates CA is unclear, although it may be the result of diminished hepatic and gastrointestinal lymph flow secondary to decreased portal venous pressures [5]. Tsauo et al. reported a case of TIPS for the treatment of CA (and chylothorax) and conducted a systematic review of the literature; all ten patients reported in the study experienced improvement in CA after TIPS. However, none of the reported patients had undergone liver transplantation.

Our case is the first reported of TIPS for the treatment of CA following LT. TIPS creation was a technical success without any postoperative complications, and improved CA significantly, eliminating the need for weekly therapeutic paracentesis. TIPS was most likely successful because it addressed cirrhosis-related CA. It is unclear whether TIPS would alleviate CA resulting from damage to the lymphatic system during transplant surgery. For such cases, percutaneous lymph vessel embolization may be more appropriate. To conclude, TIPS may be an effective and safe treatment for medically refractory CA in LT patients with cirrhosis.

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