CASO CLÍNICO

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Arterioportal Fistula in Cirrhosis: A Case Report

Fístula arterioportal en cirrosis: reporte de un caso

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SUMMARY

Introduction: *Hepatic arterioportal fistula is a rare but treatable condition that may worsen pre-existing portal hypertension in cirrhosis patients. Embolization is the treatment of choice.*
Case Presentation: *A 60-year-old woman with a 6-year history of liver cirrhosis presented with upper gastrointestinal bleeding and an impaired level of consciousness. Endoscopic examination revealed large fundal varices. Computed tomography angiography of the abdomen revealed an incidental arterioportal fistula without prior history of abdominal trauma or liver procedure. Successful embolization was achieved.*
Conclusion: *This case highlighted the need to consider arterioportal fistula as an aggravating factor of existing portal hypertension through precise examination and treatment.*

Keywords*: Fistula, fibrosis, liver.*

RESUMEN

Introducción: *La fístula arterioportal hepática es una condición rara pero tratable que puede empeorar la hipertensión portal preexistente en pacientes con cirrosis. La embolización es el tratamiento de elección.*
Presentación del caso*: Una mujer de 60 años con antecedentes de cirrosis hepática durante 6 años se presentó con hemorragia digestiva alta y alteración del nivel de conciencia. El examen endoscópico reveló grandes venas varicosas en el fondo. La angiografía por tomografía computarizada del abdomen reveló una fístula arterioportal incidental sin antecedentes de trauma abdominal o procedimiento hepático. Se logró una embolización exitosa.*
Conclusión: *Este caso resaltó la necesidad de considerar la fístula arterioportal como un factor agravante de la hipertensión portal existente a través de un examen y tratamiento precisos.*

Palabras clave*: Fístula, fibrosis, hígado.*

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INTRODUCTION

Portal hypertension is a syndrome characterized by the formation of portosystemic collaterals due to cirrhosis or non-cirrhosis causes. The hallmark of portal hypertension is the formation of varices which are commonly found in the esophagus and gastric region, which are at risk of rupture and cause bleeding (1).

Gastric varices are less common (17 %-25 %) than esophageal varices in patients with liver cirrhosis, but bleeding from gastric varices tends to be more severe with an incidence of 16 %-45 % within 3 years and is associated with a high rate of rebleeding, high transfusion requirements, and high rates of higher mortality (1,2). According to Sarin's classification, GOV 2 and IGV 1 types are referred to as fundal varices because they are in the gastric fundus. Fundal varicose veins contribute to nearly 70 % of bleeding from gastric varices (1,3,4).

In cirrhosis patients, an abnormal connection between the hepatic artery and the portal vein termed arterioportal fistula (APF) may aggravate portal vein hypertension. APF can be a rare and reversible cause of pre-sinusoidal portal hypertension (5). The most common causes are abdominal trauma and iatrogenic cause due to interventional hepatic procedures (5). The diagnosis of APF in cirrhosis is often missed because portal hypertension is thought to be due to the natural course of cirrhosis itself (6). Identification of APF is important because it can contribute to lowering portal pressure in patients with cirrhosis if appropriate therapy is given (6-8). Embolization is the first-line therapy in APF. We presented a case of arterioportal fistula without prior trauma/invasive abdominal procedure that underwent embolization.

CASE PRESENTATION

A 60-year-old Javanese woman was admitted to the emergency department with an altered level of consciousness one day before admission. She appeared confused, disoriented and sometimes agitated. She vomited bright red blood and had an episode of black, tarry stool two days before admission. Her medical history included hepatitis B and liver cirrhosis diagnosed six years ago, routinely consuming Tenofovir 300 mg o.d. Patient denied any history of abdominal trauma or interventional hepatic procedure.

On physical examination, her Glasgow Coma Scale was 11 and her hemodynamic parameter was normal. She looked anemic but no blood flowed from the nasogastric tube. During the digital rectal examination, there was no blood on the gloves.

Her blood test revealed hemoglobin (Hb) concentration of 7.4 g/dL, platelets (Plt) 99 000/ μL, partial thromboplastin time (PPT) of 25.4 seconds (control 11.3 seconds), activated partial thromboplastin time (APTT) 36.8 seconds (control 25.3 seconds), and albumin 2.93 g/dL.

She was assessed initially with grade III hepatic encephalopathy, hematemesis melena due to variceal rupture, Child-Pugh B liver cirrhosis associated with Hepatitis B virus infection, and anemia. She was treated with oxygen, proton pump inhibitor injection, vitamin K injection, octreotide injection, L-ornithine-L-aspartate injection, and planned packed red cell transfusion.

On her 4th day of hospitalization, her consciousness had recovered and an esophago-gastroduodenal (EGD) examination was performed with the result of moderate portal hypertensive gastropathy with ulcers and fundal varices (Figure 1).

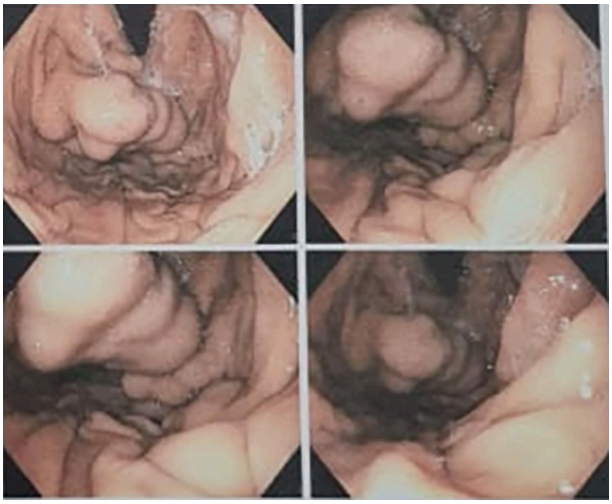

Figure 1. EGD revealed moderate portal hypertensive gastropathy with ulcers and fundal varices.

Abdominal Computed Tomography Angiography (CTA) examination was performed and revealed signs of portal hypertension (dilated portal vein diameter ± 17 mm, normal <13 mm) and an arterioportal fistula. Embolization of the right hepatic artery with polyvinyl alcohol (PVA) of 500-710 was performed by an interventional radiologist (Figure 2).

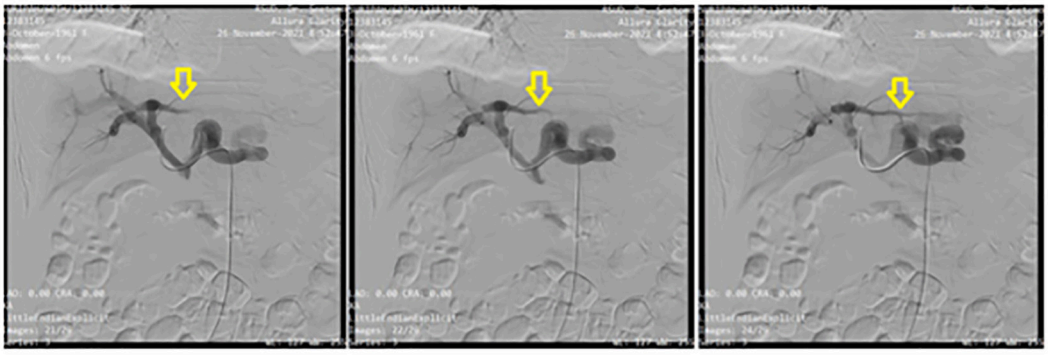

Figure 2. CTA revealed right hepatic artery fistulation to the portal vein.

After embolization, the right hepatic artery fistulation to the portal vein was no longer visible (Figure 3). Two days after embolization, she was discharged with Tenofovir 300 mg o.d. and propranolol 5 mg b.i.d. Despite the clinical improvement when discharged, the patient did not return to the outpatient clinic for further evaluation.

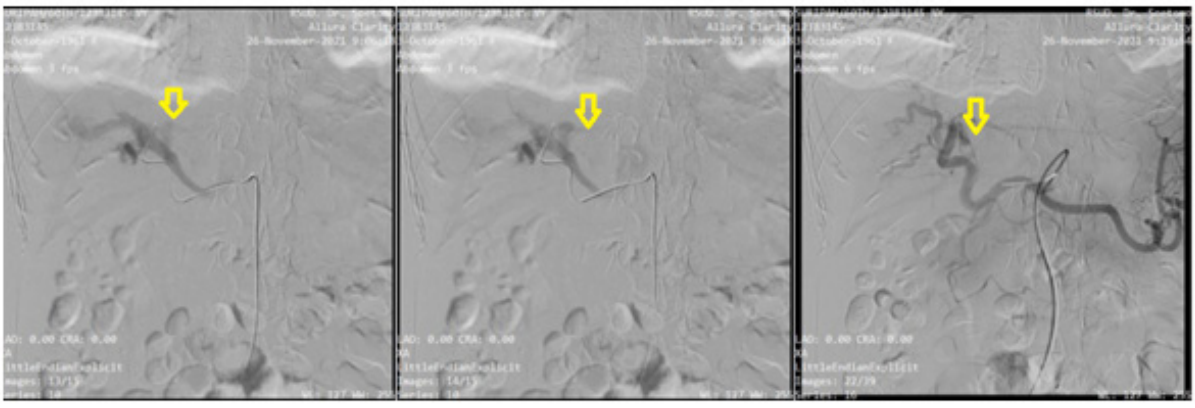

Figure 3. Angiography after embolization of the right hepatic artery with PVA 500-710, the fistula was no longer visible.

DISCUSSION

Portal hypertension can be defined as a portal pressure gradient (pressure difference between the portal vein and hepatic vein) of more than 5 mmHg. The pressure can be measured indirectly using the hepatic portal venous gradient (HVPG). HVPG is clinically significant if the value > 10 mmHg and may bleed when the pressure gradient is exceeding 12 mmHg (9).

In cirrhosis patients, increased liver resistance to portal blood flow due to structural and functional changes of the liver will culminate in increased portal vein pressure and cause typical portal hypertension clinical manifestations such as ascites (due to fluid escape into the peritoneal cavity) and development of collateral vessels (varices). Other factors that increased flow into the portal vein will further increase portal pressure and may worsen the degree of varices and subsequently gastrointestinal bleeding. To determine the cause of portal hypertension in symptomatic patients, risk factors for the underlying cause must be sought out (10).

The cause of portal hypertension is classified as prehepatic, hepatic, or post-hepatic depending on the location of the primary obstruction to portal blood flow. In hepatic (sinusoidal) causes such as cirrhosis, portal hypertension is caused by increased resistance to portal flow due to fibrosis and regenerative nodules and hyperdynamic flow of blood entering the portal vein. Pre-hepatic causes may include a portal or splenic vein thrombosis and arteriovenous fistula. Post-hepatic causes are Budd-Chiari syndrome or congestive heart failure. Non-cirrhosis causes of portal hypertension may include intra-hepatic or pre-hepatic lesions in the absence of liver cirrhosis, such as schistosomiasis, biliary cirrhosis, hepatoportal sclerosis, and congenital liver fibrosis (9,11,12).

One cause that may worsen portal hypertension that could go unnoticed in cirrhosis patients is APF because portal hypertension is already present in cirrhosis patients and clinical deterioration may be misinterpreted for the natural history of cirrhosis itself. An APF is an abnormal connection between the hepatic artery and the portal vein which can be a rare and reversible cause of pre-sinusoidal portal hypertension. APF can appear spontaneously in liver cirrhosis (5). In patients with liver cirrhosis, the diagnosis of APF is often underestimated because APF can be small and asymptomatic. APF is often caused by procedures that cause injury to the hepatic artery and portal vein such as percutaneous liver biopsy or penetrating liver trauma (6,7).

The majority of patients with APF are asymptomatic, however, when symptoms occur, it is usually due to the effects of increased portal pressure. The most common symptoms were gastrointestinal bleeding (33 %), ascites (26 %), congestive heart failure (4.5 %), and diarrhea (4.5 %). Bruits or thrills can also be detected in about 33 % of patients and are an early sign of APF. Thrill usually only appears when the diameter of the fistula is > 4 mm (5).

APF is classified based on its etiology, size, anatomy, and location. They can be classified as congenital or acquired, large or small, intra-hepatic or extra-hepatic, central or peripheral, and traumatic or spontaneous. Commonly, APF is classified into 3 types. Type 1 APF is usually small, peripheral, and intra-hepatic fistulas, usually asymptomatic, often occur after percutaneous liver biopsy, and may close spontaneously within 1 month. If the fistula remains open within 1 month and symptoms develop, then embolization therapy is needed. Type 2 APF is a larger, more central fistula, and occurs after penetrating abdominal trauma. Type 2 APF can cause portal hypertension and should be treated with embolization or surgery in cases where endovascular therapy fails or endovascular therapy is not available. Type 3 APF is a rare congenital fistula, usually intra-hepatic and diffuse, and can cause severe portal hypertension in childhood. Treatment options are hepatic artery ligation, embolization, hepatectomy, or liver transplantation (5).

Identification of APF is important because it may lower portal pressure in liver cirrhosis patients with appropriate therapy. The simplest screening is by color Doppler ultrasound examination. APF may be detected when there is a decrease in the resistive index and pulsatility index of at least 30 %-40 % in one lobe compared to the index in the other lobe and the blood flow in the intrahepatic branch of the portal vein of that lobe is opposite to the flow in the normal lobe (hepatopetal) (6,7).

Arteriography is the gold standard for diagnostics in APF. Pathognomonic signs include early visualization of the portal vein during aortic or celiac artery injection. Angiography often shows a single fistula with hepatopedal flow without evidence of portal hypertension, although cirrhosis may present with hepatofugal

flow. CTA shows premature filling of the veins during the arterial phase and a conspicuous focus on the hepatic arterial phase (5,8).

Our patient presented with a history of vomiting bright red blood and an episode of black, tarry stool caused by ruptured fundal varices. A possible cause of her gastrointestinal bleeding symptoms is increased portal hypertension due to liver cirrhosis (hepatic) and arterioportal fistula (prehepatic). Interestingly, CT angiography was initially performed to assess the afferent and efferent vessels of her fundal varices but instead found an arterioportal fistula. From the abdominal CT scan, we can also exclude additional prehepatic and post-hepatic causes of portal hypertension. The patient had no prior history of abdominal trauma or invasive abdominal procedure so APF in this patient may be occurred spontaneously related to her cirrhosis condition.

The initial management of acute bleeding from gastric varices is not different from that of esophageal varices, which includes fluid resuscitation, correction of coagulopathy, medical therapy with antibiotics and vasoactive drugs, and early intervention with endoscopy. Radiological management, balloon tamponade, and surgery are used in cases of hemostatic failure after endoscopic intervention and pharmacological therapy (4,13,14).

Endoscopic interventions include tissue adhesive with endoscopic cyanoacrylate glue injection (ECI), fibrin and thrombin therapy, endoscopic band ligation (EBL) and sclerosants including alcohol. ECI is the treatment of choice according to the Baveno VI consensus and is most often used in the management of gastric varices with a hemostatic rate of 91 %-100 % and a rebleeding rate of about 7 %-28 % (4). Although there are no strong recommendations and the risk of embolization, ECI can be used as primary and secondary prophylaxis of fundal variceal bleeding (13-15).

The choices of treatment for APF include surgery and minimally invasive interventions such as trans-catheter arterial embolization (TAE). TAE had lower morbidity and lower cost than surgery, therefore it became the first-line treatment for APF. Various embolic agents may be used with their advantages and disadvantages. Lipiodol agents may be useful in poor blood shunt but may easily occlude small blood vessels and induce liver ischemia. Polyvinyl alcohol (PVA) has to be combined with a contrast agent but is effective for long-term occlusion effects. Spring steel coils are typically used for long-term simple shunts because they may not reach the small distal vascular that are difficult to reach. Gelatin sponge particles are typically reabsorbed within 2-4 weeks, therefore had a high recanalization rate (16).

In a retrospective analysis of 97 cases of patients with hepatic arterioportal fistulas in China Hospital from January 2010 to January 2020, Cao et al. revealed that 64,9 % of APF patients treated with TACE showed comparable efficacy between the embolization agents such as polyvinyl alcohol, lipiodol combined with gelatin sponge, and spring steel ring (16). In our patient, we chose polyvinyl alcohol for embolization agents.

Complications that may occur with the embolization procedure are agent (coil) migration, vascular injury, liver failure, infection and subsequent abscess, portal thrombosis, and bile duct stricture (8,17). In cases where embolization fails, the choice is surgery with hepatic artery ligation or fistula ligation (8). In most patients with acquired APF, the prognosis is good due to the minimal physiologic changes associated with isolated APF and the currently effective treatments available (5,18). Our patient underwent embolization with no visible fistulation of the right hepatic artery to the portal vein.

CONCLUSION

APF is a rare condition that may cause increased portal pressure, mainly in a patient with underlying portal hypertension such as liver cirrhosis. Assessment of vascularity in gastric varices (including fundal varices) and identification of arterioportal fistulas are important to determine the appropriate management of the patient to stop active bleeding and prevent rebleeding. Embolization therapy is the treatment of choice for APF.

Academic Collaborations of the Authors

ZNH collected the data and wrote the manuscript, and BW conducted, supervised, and supported the project.

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Conflict of Interest

The author stated there is no conflict of interest.

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