

Hepatic Arterioportal Fistula Presenting as Gastric Variceal Hemorrhage

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ABSTRACT

Hepatic arterioportal fistulae (APF) are abnormal communications between the hepatic artery and the portal vein. In this report, we present the second case in the literature of a symptomatic APF presenting as a gastric variceal bleeding. A 55-year-old female presented to our facility with hematemesis. Upper endoscopy revealed a bleeding gastric varix. A computed tomography scan identified a large left hepatic lobe APF between the left hepatic artery and the left portal vein. Through angiography coil embolization was performed and with resultant loss of arterial flow, the APF was decompressed. On hospital day 3, the patient developed new melena. Portovenogram was performed and a TIPS stent was deployed. The patient subsequently did well. Hepatic arterioportal fistulae can result in portal hypertension secondary to arterial blood flowing directly into the portal vein bypassing the hepatic sinusoids. Iatrogenic causes (e.g. percutaneous liver biopsy) represent more than 50% of published cases of APFs. Most APFs resolve spontaneously as they are small and peripherally located. In rare instances, when APFs are centrally located, clinical symptoms develop. There have been 30 reported cases of symptomatic intrahepatic APFs following percutaneous liver biopsy. Of those, only one case presented as a gastric variceal bleed. Digital subtraction angiography is the gold standard in the diagnosis and treatment of APFs. In addition to initial embolization, we elected to treat the patient with TIPS due to the magnitude of her bleed. Although rare, intrahepatic APF should be kept on the differential of a patient presenting with isolated gastric varices.

Key words: hepatic arterioportal fistula – gastric varix – portal hypertension

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INTRODUCTION

Portal hypertension is defined as an elevation in pressure in the portal venous system characterized by an increase in portal blood flow or an increase in hepatic resistance. Typically, portal hypertension is caused by structural changes in the liver attendant to cirrhosis; however, both pre and post-hepatic causes are also well described [1]. Hepatic arterioportal fistulae (APF) are abnormal communications between the hepatic artery and the portal vein that may arise spontaneously, through genetic disorders, or from trauma [2]. Arterioportal fistulae are theorized to cause portal hypertension by creation

of an inflow block (resulting from the interruption of portal venous flow by the inflow of arterial blood) with subsequent increased pressure in portal vein radicals [3]. Hepatic APFs are usually small and self-limiting; however, in rare cases fistulae can grow in size and become clinically symptomatic (e.g. esophageal varices and ascites) [2]. In this report, we present a patient with a gastric variceal hemorrhage secondary to an APF. This case highlights the importance of consideration of hepatic APFs in the differential diagnosis of isolated gastric varices.

CASE REPORT

A 55-year-old female with a history of primary biliary cirrhosis (PBC) (biopsy established two years prior with stage two fibrosis) maintained on ursodiol (15mg/kg) presented to our facility with hematemesis and melena. Upon admission her pulse rate was 71 beats/min and arterial blood pressure was 119/70 mmHg. Laboratory tests revealed: hemoglobin, 7.1 g/dL; hematocrit, 21.0 %; platelet, 191 K/uL; prothrombin time, 15.0 s; BUN, 16 mg/dL; creatinine, 1.2 mg/dL; total bilirubin, 1.2 mg/dL, aspartate aminotransferase, 36 U/L; alanine aminotransferase, 29 U/L; alkaline phosphatase, 146 U/L;

albumin 3.6, g/dl. The gastroenterology service was consulted for evaluation and an upper endoscopy was performed which revealed: normal esophagus, blood pooled in the fundus, and a pigmented protuberance with platelet plug in the gastric cardia consistent with a recently bleeding gastric varix (Fig. 1).

The patient's hemodynamics remained stable post-procedure and a computerized tomography (CT) scan was performed. CT identified a large left hepatic lobe APF between the left hepatic artery and the left portal vein (Fig. 2). Angiography was then employed to characterize the APF.



Fig. 1. Upper endoscopic image showing blood pooled in the fundus and a pigmented protuberance with a platelet plug in the gastric cardia consistent with a bleeding gastric varix.

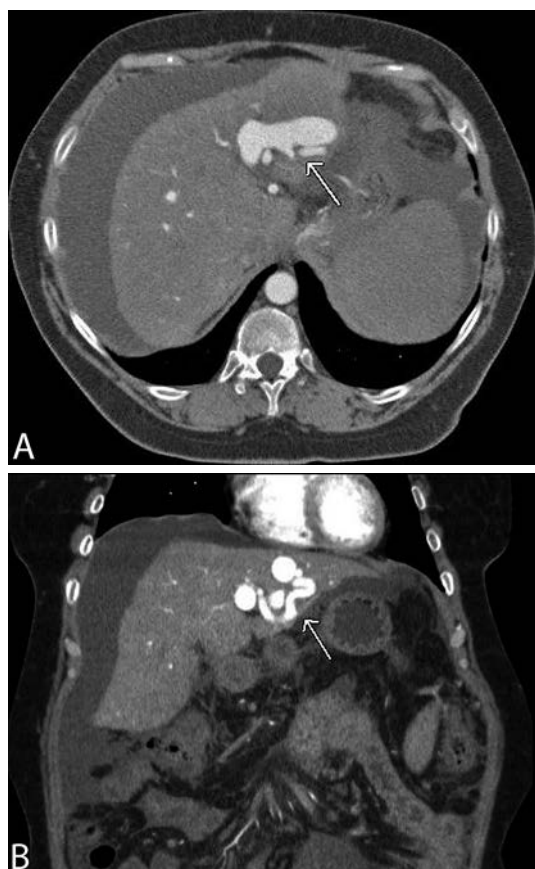


Fig. 2. CT abdomen arterial phase, axial (A) and coronal (B) views showing left hepatic artery (arrow) with early enhancement of left portal vein suggestive of arterial venous fistula.

Using ultrasound guidance, a 19 gauge needle was used to access the right common femoral artery. Celiac artery angiogram demonstrated a hypertrophied common and left hepatic artery; a left hepatic artery to left portal vein APF was confirmed (Fig. 3A). The left portal vein was dilated (Fig. 3B) with contrast filling in a retrograde manner into the portal vein, splenic vein, and subsequently, a gastric varix. Coil embolization was performed and with loss of arterial flow the APF was “depressurized” (containing now only portal inflow) and decompressed.

On hospital day 3, the patient developed recurrent melena and a drop in hemoglobin from 8.9 g/dl to 7.4 g/dl. CT of the abdomen was repeated and although there was a decrease in the size of the APF, the resultant gastric varix was still present. Portovenogram was performed: a 10-French sheath was passed through the intrahepatic tract to the portal vein. A Viatorr TIPS stent was deployed a portion within the proximal portal vein and spanning the intrahepatic into the distal hepatic vein/inferior vena cava junction. The deployed stent was balloon dilated to 7 mm. A Fathom 016 wire was then used to successfully select the rounded proximal aneurysmal portion of the left portal vein supplied by the APF. Embolization was performed utilizing Ruby coils. Stasis was achieved within the portal vein segment. A Simmons 1 catheter was then used to select the proximal main splenic vein. Coil embolization was performed within the distal portion of the branch, proximal

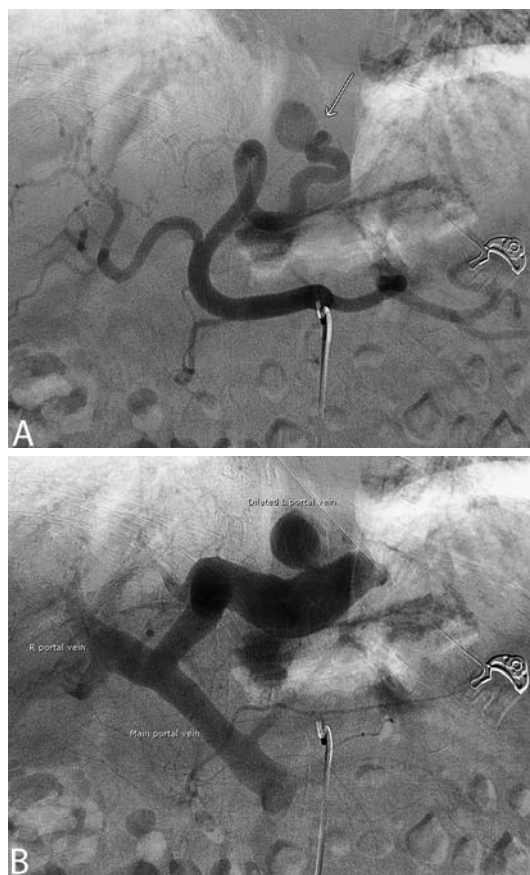
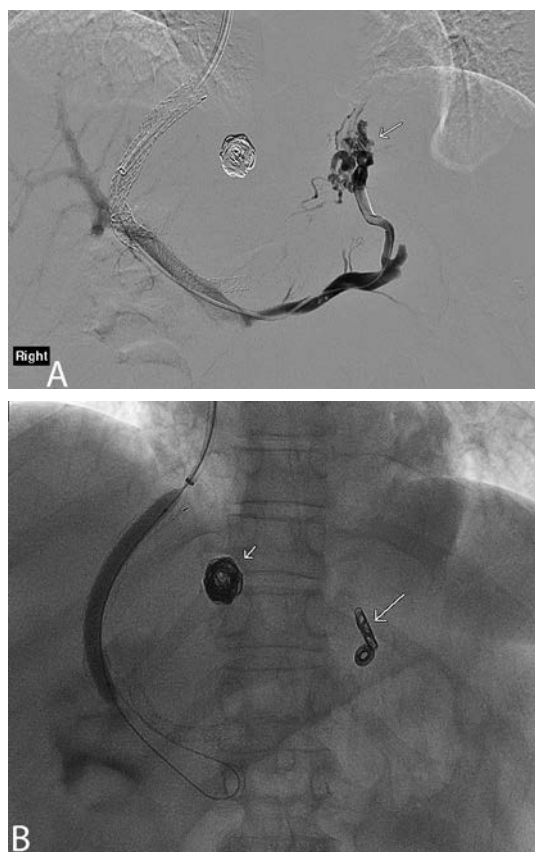


Fig. 3. A) Celiac artery angiogram in early arterial phase showing the enlarged left hepatic artery and the arterial-portal fistula (arrow). B) Portal venous phase showing a dilated left portal vein compared to the normal sized right portal vein. The enlarged vessels are due to long-standing process.



Fi. 4. A) Portography through TIPS access showing gastric varices from the splenic vein (arrow). B) Coil embolization of the arterio-portal fistula (short arrow) and gastric varices (long arrow).

to the varix, utilizing Ruby coils. A final portovenogram was performed (Fig. 4). The patient was monitored for 48 hours with stable hemodynamics and subsequently discharged on hospital day 5. An upper endoscopy performed one month post discharge was normal with no gastric varices or portal hypertensive related changes in the upper gastrointestinal tract.

DISCUSSION

The existence of an APF was first reported in 1889 by Goodhart [4]. Arterioportal fistulae can result in portal hypertension secondary to arterial blood flowing directly into the portal vein bypassing the hepatic sinusoids. The common presentations of symptomatic intrahepatic APFs include gastrointestinal bleeding, ascites, congestive heart failure, abdominal pain, and diarrhea [2].

Spontaneous APFs are usually associated with congenital vascular malformations (e.g. hereditary hemorrhagic telangiectasia), benign neoplasms, cirrhosis, and hepatocellular carcinoma. Blunt or penetrating trauma can cause APFs and are usually diagnosed within 2 years [5]. Iatrogenic causes (percutaneous liver biopsy, transjugular liver biopsy, percutaneous transhepatic biliary drainage, TIPS) represent more than 50% of published cases of APFs [6].

Preger first described an APF resulting from a liver biopsy in 1967 [7]. The frequency of APFs following liver biopsy was reported to be 5.4-10 %. Most APFs resolve spontaneously

within 12 weeks as they are small and peripherally located. In rare cases, when APFs are centrally located and grow larger, clinical symptoms develop [8]. Since 1977 there have been 30 reported cases of symptomatic intrahepatic APFs following percutaneous liver biopsy. Of those cases, 13 presented with gastrointestinal bleeding secondary to esophageal varices or hemobilia and only 1 case presented as a gastric variceal bleed [9].

On physical examination, the presence of an abdominal bruit can suggest APF. Ultrasound with Doppler, CT, magnetic resonance imaging (MRI), digital subtraction angiography (DSA) can be employed in the diagnosis. Doppler ultrasound is typically employed as an initial screen, with high-flow velocities and arterial waveforms in the portal vein. Turbulence with reversal of flow may also be present in the portal vein. Confirmation of these findings with CT or MRI is necessary and often shows early and prolonged enhancement of the portal vein during arterial phase imaging [2]. Digital subtraction angiography is the gold standard in the diagnosis, treatment planning, and follow-up of APFs [10].

Urgent intervention may be needed when an APF causes luminal gastrointestinal bleeding. Elective treatment is otherwise considered to prevent the development of portal hypertension or manage its associated complications (e.g. ascites). In the past, surgical ligation of the supplying artery was performed; however, now the treatment of choice is endovascular transcatheter arterial embolization. The goal of embolization is not only selective fistula closure but preservation of adjacent normal vasculature which mandates characterization of collateral sources of blood supply to hepatic segments [11]. Various embolizing tools for fistula closure have been tried and include: mechanical agents such as stainless-steel coils, detachable balloons, as well as liquid agents such as onyx and cyanoacrylate glue [12].

We postulate that our patient's APF may have occurred as a consequence of percutaneous liver biopsy in 2010. We elected to treat the patient with TIPS in addition to repeating embolization, due to the magnitude of her bleed as well as to treat any contributing portal hypertension that may have resulted from her underlying PBC (which may cause portal hypertension in the absence of cirrhosis).

CONCLUSION

We describe a case of gastric variceal bleeding secondary to an APF successfully treated with coil embolization. Although rare, intrahepatic APF should be kept on the differential of a patient presenting with isolated gastric varices. Cross sectional imaging and interventional radiologic techniques can be successful in characterizing and treating luminal gastrointestinal tract bleeding from APF.

Conflicts of interest. None to declare.

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