

Background

- Arterioportal fistula (APF) is a rare cause of non-cirrhotic portal hypertension.
- This is a case of a hepatic artery-portal vein fistula presenting with recurrent variceal hemorrhage and ascites in a patient with no known liver disease.

Methods

- Case report and literature review

Case

- 61-year-old woman with history of aplastic anemia on cyclosporine, remote colon cancer s/p sigmoid resection with work up including a percutaneous liver biopsy to evaluate a benign liver lesion as a possible metastasis about 10 years prior to this current presentation.
- She initially presented to an outside hospital with hematemesis, as well as a new onset of increasing abdominal girth and lower extremity edema.
- EGD showed bleeding esophageal varices that were successfully band ligated. Over the next 6-weeks, she required multiple EGDs with band ligation for recurrent variceal bleeding.
- She also had evidence of ascites and diagnostic paracentesis demonstrated a high serum ascites albumin gradient (SAAG) of 3.1. Total Protein in ascitic fluid was 2.6.
- She was ultimately referred to our center for evaluation of her candidacy for a possible transjugular intrahepatic portosystemic shunt (TIPS).
- CT scan was notable for right hepatic lobe atrophy, with marked enlargement of the right hepatic artery and portal vein.
- Subsequent MRI identified a right hepatic lobe arteriovenous malformation between the right hepatic artery and portal vein with evidence of portal hypertension.

Case (continued)

- Transjugular liver biopsy with pressures was notable for a hepatic venous pressure gradient (HVPG) of 7.5 mmHg.
- Liver biopsy showed perisinusoidal fibrosis in zone three without cirrhosis, and intimal hyperplasia of portal vein (arteriolization) consistent with arterioportal fistula (Figure 1).
- Rather than pursuing TIPS, we elected to perform coil embolization of the two right hepatic arteries supplying the fistula (Figure 2).
- Despite technical success, a small fistula persisted and repeat embolizations were required. She has not needed paracentesis nor has any bleeding during the 1 year follow-up.

Figure 1: Liver biopsy showing arteriolization of the portal vein

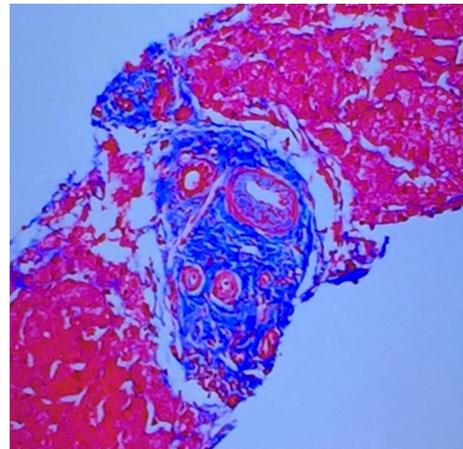


Figure 2: Hepatic artery-portal vein fistula before and after coil embolization



Discussion

- Non-cirrhotic portal hypertension can be classified as pre-hepatic, intrahepatic, and post-hepatic in origin.
- APFs are a rare cause of pre-hepatic non-cirrhotic portal hypertension, and they can be either congenital or acquired.
- Acquired etiologies include penetrating hepatic trauma, splanchnic artery aneurysms, surgical complications, or they may occur spontaneously in the setting of cirrhosis or liver tumors.
- Large, more central APFs are most likely to present with portal hypertension.
- Small APFs can occur following percutaneous liver biopsies, but large APFs complicating a remote percutaneous biopsy, as in our case, are rare.
- Congenital APFs are intrahepatic and diffuse, and typically present in children. However, there are some case reports of adult onset congenital APFs in the literature.
- APF should be considered in patients with non-cirrhotic portal hypertension and, when identified, treatment is embolization or surgery, not TIPS.

References

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