Giant Intrahepatic Portal Vein Aneurysm: Leave it or Treat it?

Amit Shrivastava [*], Jagdeesh S. Rampal [†], D. Nageshwar Reddy [‡]

[*]McGill University, Montreal, Canada, [†]Chief, Department of Intervention Radiology, Asian Institute of Gastroenterology, Hyderabad, India and [‡]Director, Department of Medical Gastroenterology, Asian Institute of Gastroenterology, Hyderabad, India

Portal vein aneurysm (PVA) is a rare vascular dilatation of the portal vein. It is a rare vascular anomaly representing less than 3% of all visceral aneurysms and is not well understood. Usually, PVA are incidental findings, are asymptomatic, and clinical symptoms are proportionally related to size. Patients present with nonspecific epigastric pain or gastrointestinal bleeding with underlying portal hypertension. PVA may be associated with various complications such as biliary tract compression, portal vein thrombosis/rupture, duodenal compression, gastrointestinal bleeding, and inferior vena cava obstruction. Differential diagnoses of portal vein aneurysms are solid, cystic, and hypervascular abdominal masses, and it is important that the radiologists be aware of their multi-modality appearance; hence, the aim of this article was to provide an overview of the available literature to better simplify various aspects of this rare entity and diagnostic appearance on different modality with available treatment options. In our case, a 55-year-old male patient came to the gastroenterology OPD for further management of pancreatitis with portal hypertension and biliary obstruction with plastic stents in CBD and PD for the same. In this article, we have reported a case of largest intrahepatic portal vein aneurysm and its management by endovascular technique. As per our knowledge, this is the largest intrahepatic portal vein aneurysm and first case where the endovascular technique was used for the treatment of the same. (J Clin Exp Hepatol 2017;7:71–76)

The portal vein has distinctive characters of the presence of capillaries on both ends and the absence of valves. Portal vein aneurysm (PVA) is a rare vascular dilatation of the portal vein, which was first described by Barzilai and Kleckner.[1] Since then, 190 cases of PVA have been reported in 96 reports in the literature, mainly as case reports or surgical series.[2] Douglass et al. studied 92 autopsies and found the diameter of the portal vein to be 0.64–1.21 cm in patients without cirrhosis or portal hypertension.[3] PVA is defined as a portal vein diameter greater than 1.9 cm in cirrhotic patients and 1.5 cm in normal liver. In 1976, Doust and Pearce conducted extensive vascular study among 53 patients to assess the size of the portal vein and underlying liver status through abdominal ultrasound and they detected that the maximum caliber of the portal vein was 1.9 cm in cirrhotic patients and 1.5 cm in patients with normal livers.[4] Subsequently, all portal veins caliber in antero-posterior diameter beyond such measures were considered pathological and with a diameter of >2.0 cm considered aneurismal.[5] No universally accepted data are available to quantify intrahepatic portal vein branches as aneurysmal. Doust and Pearce characterized an intrahepatic portal vein aneurysm if its diameter measures larger than 0.7 cm in normal patients and 0.85 cm in cirrhotic patients.[4]

PVA is very rarely diagnosed disorder that has been observed in 0.067% of the patients by ultrasonography[6] and after introduction of MDCT, the prevalence of portal venous system aneurysms was 0.43%.[5]

Because portal vein aneurysms can mimic solid, cystic, and hypervascular abdominal masses, it is important that the radiologist be aware of their multi-modality appearances. Hence, the aim of this article was to provide an overview of the available literature to better simplify various aspects of this rare entity and treatment options available. In this article, we have reported a case of largest intrahepatic portal vein aneurysm and its management by endovascular technique. As per our knowledge, this is the largest intrahepatic portal vein aneurysm and first case where the endovascular technique was used for the treatment of the same.

CASE REPORT

In our case, a 55-year-old male patient came to the gastroenterology OPD for further management of pancreatitis with portal hypertension and biliary obstruction with

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Address for correspondence: Amit Shrivastava, Post Doctoral Fellow, Abdominal Imaging and Non-vascular Intervention, McGill University, Montreal, Canada. Fax: +1 514 934 8263.
E-mail: dr.amitsrivastava@gmail.com
Abbreviations: CBD: common bile duct; DSA: Digital Subtraction Angiography; MDCT: multi detector computed tomography; MRI: magnetic resonance imaging; PD: pancreatic duct; PVA: portal vein aneurysm; USG: ultrasonography
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plastic stents in CBD and PD for the same. On USG examination, an anechoic area was noted in the segment VI of the liver on the gray scale (Figure 1A). On color Doppler, complete filling of an anechoic area was noted which showed the characteristic monophasic waveform on duplex Doppler ultrasonography (Figure 1B). CT scan of the patient examined was obtained on a 16 slice – MDCT scanner (Siemens Medical Solutions). CECT scan revealed a well-circumscribed hypodense area in the segment VI of the liver. No contrast enhancement was seen in the arterial phase (Figure 1C) and complete filling of the lesion in the portal phase (Figure 1D and E). The density of the contrast was similar to the density of the portal vein on delayed phase. The size of the lesion was 6.2 cm × 4.3 cm. Digital Subtraction Angiography (Siemens artis U) was done by the right femoral approach to completely exclude the possibility of the arterial feeder.

In our case, the proposed etiology of PVA was portal hypertension and pancreatitis. Under USG guidance, the portal vein system was accessed by puncturing the segment VI branch of portal vein by 18 gauze needle and negotiated the Terumo wire 0.035 through the needle (Figure 2A). The punctured site of the portal vein was just close to the portal vein aneurysm, so sheath was not placed. Then, the needle was withdrawn and 5F slip catheter was placed over the Terumo guide wire. Further detailed anatomy of the PVA and its branches was evaluated by Portogram. Subsequently, the identified branches of the PVA were selectively cannulated and embolized with Nester coils (Figure 2B). After coiling of PVA and its branches, we tried to completely embolize the PVA with 1:1 combination of Glue and Lipidol. Under USG guidance and fluoroscopy guidance, the PVA was directly punctured with 18 G needle and embolized with Lipiodol-Glue combination. As complications with the use of Glue are well established like incomplete embolization, catheter damage, trapped catheter, and loss of access, PVA was directly punctured (Figure 2C). Follow-up USG after 24 h showed non-vascular echogenic area in place of PVA (Figure 2D). Follow-up CT scan after one month revealed the non-filling of the aneurysm by contrast and no residual/recurrences had been observed in PVA (Figure 2E). After 5 days, the patient was discharged uneventfully. The patient remains symptom-free in the 12-month follow-up visit.

DISCUSSION

PVA is a rare vascular anomaly representing less than 3% of all visceral aneurysms.[5] Etiology of PVA is not well understood. Two categories of PVA have been proposed – acquired and congenital. As the portal vein develops from the vitelline and umbilical veins, it was anticipated that the absence of regression of the right primitive vitelline vein leads to congenital aneurysms of the portal vein and developing a diverticulum from the vitelline vein remnant. This diverticulum enlarges over time, and later with increasing portal vein pressures, it results in a saccular portal vein aneurysm.[7] Chronic liver disease, portal hypertension, pancreatitis, trauma, and surgery are the acquired causes of PVA. In a study, Koc et al. found that portal vein aneurysms were associated with cirrhosis in 12% of patients and portal hypertension as the most common etiology in 32% of the cases.[5] Therefore, it has been recommended that both developmental portal vein wall weakness and portal hypertension are obligatory for portal vein aneurysm development in the setting of cirrhosis. Other causes of PVA are pancreatitis, trauma, and invasive malignancy.[5,8,9] Portal venous system aneurysms were frequently (63%) extrahepatic in location compared to intrahepatic location. There appears to be no gender predilection and the mean age at diagnosis is 53 years.[5]

Usually PVA are incidental findings, are asymptomatic, and clinical symptoms are proportionally related to size. Some patients present with nonspecific epigastric pain or gastrointestinal bleeding with underlying sequelae of portal hypertension. Large portal venous system aneurysms can cause mass effect resulting in compression of the duodenum or bile ducts, which results in obstructive jaundice, direct hyperbilirubinemia, and duodenal obstruction.[7] Symptoms of thrombosis in PVA are nausea, abdominal pain, and fever.

PVA may be associated with various complications such as biliary tract compression, portal vein thrombosis/rupture, mass effect over duodenum, gastrointestinal bleeding, and inferior vena cava obstruction.[2] Thrombosis is the most frequent complication, with complete thrombosis and non-occlusive thrombus occurring in 13.6% and 6%, respectively.[4] Spontaneous rupture of the portal trunk or one of its branches is reported in only 2 cases.[2,4]

There is a restricted understanding of this entity as it is relatively rare, and often an incidental finding unrelated to the patient's preliminary complaint. Ultrasound is a favorable modality to follow the caliber of portal vein aneurysms due to lack of radiation and inexpensive nature. Color Doppler ultrasonography showed color flow in the lesion and duplex Doppler ultrasonography displayed the characteristic monophasic waveform and constant hepatopetal flow along the aneurysmal wall. Ultrasound is also useful to distinguish a portal vein aneurysm from a hypervascular mass. Contrast-enhanced CT and MR are both useful in the setting of ambiguous sonographic findings, in particular when differentiating slow flow from thrombosis. On computed tomography (CT)/MRI scan, portal vein aneurysm is appreciated as a well-defined contrast enhanced mass communicating with the portal vein in portal phase. Complications associated with the PVA and planning of surgery/intervention procedures are better evaluated on cross-sectional imaging. The use of portal venography is often restricted when interventional procedures are necessary.[7]