Portal Vein Aneurysm: Case Report and Review of the Literature

Shazia A. Rafiq, MD
Michael D. Sitrin, MD
Division of Gastroenterology, Hepatology, and Nutrition
State University of New York at Buffalo

A portal vein aneurysm is an unusual condition that is now being recognized more frequently with the regular use of abdominal imaging. Since the first case described by Barzilai and Kleckner in 1956, only approximately 50 cases have been reported in the English-language literature. The relative rarity of this condition has limited our understanding of the natural history of the disease. We describe a case of a portal vein aneurysm that has been followed for over 10 years.

Case Report

A 57-year-old white man was referred to us for evaluation of a portal vein aneurysm that had increased slightly in size on computed tomography (CT) scan. The patient had been initially diagnosed with a portal vein aneurysm on a CT scan 6 years prior, when he was admitted for mild alcoholic pancreatitis. At that time, the aneurysm measured 5 cm in diameter, and the patient was managed conservatively. Follow-up scan 6 years later showed that the aneurysm had increased to 6 cm, and the patient was, therefore, referred to us for a gastroenterologic evaluation. The patient reported that he was continuing to do well, without any symptoms of abdominal pain, nausea, vomiting, or early satiety, and reported a good appetite and a stable weight. He did not have any history of jaundice, fever, chills, liver disease, gastrointestinal bleeding, or abdominal or vascular surgery. His medical history did, however, include diabetes mellitus, hypertension, and hypercholesterolemia. The patient admitted to heavy alcohol abuse in the past but had quit drinking 6 years ago after being diagnosed with alcoholic pancreatitis.

Physical examination was significant for the absence of chronic liver disease stigmata or signs of portal hypertension. His abdomen was soft and nontender without organomegaly or palpable abdominal masses. Arterial and venous examinations were normal, and there were no abdominal bruits.

Complete blood count and metabolic panel including liver function tests were within normal limits. CT scan showed a 6-cm aneurysm at its origin that had increased slightly in size since the CT scan 6 years previous (Figure 1). No intramural thrombus was seen, and the rest of the abdominal vasculature was normal. The liver showed fatty infiltration, and the spleen and pancreas were unremarkable. Abdominal Doppler ultrasound confirmed the presence of the aneurysm (Figure 2), and magnetic resonance angiography revealed no thrombus in the aneurysm.

Management options were discussed with the patient, who chose conservative care with serial imaging to monitor the size of the aneurysm. He has been followed for 4 years with serial ultrasounds and CT examinations. The aneurysm size has remained stable, and there have been no associated complications.
Discusssion

Barzilai and Kleckner first reported a case of a main portal vein aneurysm in 1956.1 In that patient, the aneurysm ruptured and was diagnosed at autopsy. Fewer than 50 cases have since been reported in the English-language literature, and many of them have been incidental findings in patients having abdominal imaging for unrelated complaints.2-4

The portal vein is a unique vessel because of the presence of capillaries on both ends and the absence of valves. Autopsy and ultrasonographic studies have been conducted to evaluate the normal dimensions of the portal vein, and considerable variation of the portal vein diameter has been reported. Douglass and associates 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.5 In an ultrasonographic study by Doust and Pearce, the maximum portal vein diameter never exceeded 1.5 cm in normal patients and 1.9 cm in cirrhotic patients.6 A diameter of greater than 2 cm is, therefore, usually regarded as aneurysmal. The intrahepatic portal branches are generally considered aneurysmal if they measure more than 0.7 cm in normal patients and 0.85 cm in cirrhotic patients, especially if the venous outline assumes a fusiform or saccular configuration.6

Some types of portal venous system aneurysms appear to be congenital in origin. The portal venous system develops from the vitelline and umbilical veins that drain the intestinal blood of the embryo. It has been postulated that during embryologic development, there is a regression failure in the right primitive vitelline vein. The diverticular remnant of the vitelline vein is believed to enlarge, forming a saccular aneurysm of the portal vein later in life.2 Other researchers attribute the development of a portal vein aneurysm to an inherent weakness of the vein wall.2 The congenital origin hypothesis for some types of portal vein aneurysms is strongly supported by a report of the in-utero diagnosis of portal vein aneurysm by ultrasonography;7 the occurrence of this condition in children and young adults with no history of liver disease or other predisposing factors;8 reports of liver biopsies showing normal histologic evidence and normal portal venous pressures in patients with extrahepatic portal vein aneurysms;9 and a report of a portal vein aneurysm associated with hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu disease), in which a multifaceted inherited vascular disease has been postulated.9

Other patients have portal vein aneurysms that appear to be acquired. Portal hypertension secondary to cirrhosis or other hepatic pathologies is the most frequently reported cause of acquired portal vein aneurysms.2,4,10 With portal hypertension, there is intimal thickening and compensatory medial hypertrophy of the portal vein. With time, the medial hypertrophy is replaced by fibrous tissue that weakens the tensile strength of the vein wall, making it susceptible to aneurysmal dilatation.11 Portal vein aneurysms, however, are rarely encountered in patients with known cirrhosis. Some researchers, therefore, have suggested that aneurysmal dilation occurs when portal hypertension develops in an individual with a hereditary weakness or a developmental anomaly of the portal vein.2

Other causes of acquired portal vein aneurysm include severe pancreatitis causing digestion and inflammation of the portal vein,2 trauma,2 and invasion of the portal vein by various malignancies.12 An iatrogenic cause of a portal vein aneurysm is the arterialization of the portal vein in conjunction with a portacaval shunt, which creates an A-V fistula between the hepatic end of the portal vein and the arterial system to preserve hepatic flow and to reduce postoperative encephalopathy following a portosystemic shunt.13,14

Aneurysms of the portal venous system occur mainly at sites of bifurcation or confluence, and can be intrahepatic or extrahepatic. Aneurysms in the small or peripheral intrahepatic portal branches are rare but have been reported in association with portal-hepatic venous shunts.15 Most patients with portal vein aneurysms are asymptomatic or have unrelated gastrointestinal complaints that prompt abdominal imaging. Complications of portal vein aneurysms include thrombosis, rupture, and symptoms from pressure on adjacent structures. Acute thrombosis of the portal vein aneurysm can result in severe, life-threatening portal hypertension.2,4,16 Recurrent or chronic thrombosis may result in the complete occlusion of the portal vein and manifestations of extrahepatic pulmonary
hypertension such as varices and bleeding.\textsuperscript{2,4} Rupture of portal vein aneurysms is unusual because of low portal venous pressure, but the risk of rupture rises in the presence of portal hypertension.\textsuperscript{1,17} Large extraportal vein aneurysms can cause symptoms by pressing on adjacent viscera. Deviation, compression, and obstruction of the common bile duct can cause recurrent abdominal pain, cholestasis, and obstructive jaundice. Obstruction of the duodenum, compression of the portal vein by the aneurysm causing thrombosis and portal hypertension, and compression of the vena cava have also been reported.\textsuperscript{2,4}

Several noninvasive imaging modalities are used to diagnose portal vein aneurysms.\textsuperscript{18,19} The typical finding on B-mode ultrasonography is an anechoic cyst-like lesion near the porta hepatis. Doppler or color flow study helps to confirm the diagnosis by demonstrating the presence of blood flow with a nonpulsatile monophasic waveform within the lesion, differentiating it from a liver cyst. The aneurysm, however, may be echogenic due to the presence of a mural thrombus and may mimic a tumor. Ultrasonography with Doppler is an excellent method for determining the size of an aneurysm and monitoring for expansion. Contrast CT and magnetic resonance angiography can be helpful in patients with equivocal ultrasonographic findings or to delineate the lesion when surgical intervention is contemplated. Portography and mesenteric angiography can also help define the vascular anatomy.

The diverse patient populations in which portal vein aneurysms occur makes it difficult to draw firm conclusions about the natural history and optimal management of these lesions. In general, the risk of complications appears to be low.\textsuperscript{2,4} Patients are often asymptomatic, and, as in our patient, the aneurysm may remain relatively stable for many years. A progressive increase in aneurysm size can indicate the presence of portal hypertension and warrants further evaluation.

The management of portal vein aneurysms remains somewhat controversial. Asymptomatic, small aneurysms in patients without portal hypertension or cirrhosis are often advised to be managed conservatively, with regular follow-ups of aneurysm size and monitoring for new symptoms.\textsuperscript{2-4} Calligaro and colleagues, however, recommended prophylactic surgery for abdominal venous aneurysms in low-risk patients to prevent potential serious complications.\textsuperscript{20} Conservative management certainly seems justified if the aneurysm is in a location that is difficult to approach surgically.

Surgical intervention should, however, be considered if the portal vein aneurysm causes symptoms, or if the aneurysm expands and/or presents a high risk of thrombosis or rupture. Patients with portal hypertension and portal vein thrombosis have generally been candidates for shunt surgery.\textsuperscript{2} Portocaval or mesocaval shunts reduce the portal pressure and may prevent the progressive dilation of the portal vein aneurysm. Those with thrombosis that extends into the superior mesenteric and splenic veins should undergo thrombectomy, if possible. For patients without portal hypertension, aneureysmorrhaphy is preferred, as it preserves the portal blood circulation and restores laminar flow in the portal vein, avoiding stasis of blood and subsequent thrombosis.\textsuperscript{2,3} The risk of rupture is also reduced because lateral wall tension is decreased by diminution of the size of the lesion.

Our patient had a large portal vein aneurysm that had remained relatively stable and asymptomatic for 10 years, one of the longest follow-ups reported in the literature. The aneurysm was likely of congenital origin, as the patient had no signs of liver disease or portal hypertension and had only one episode of mild alcoholic pancreatitis and no other predisposing causes.

References