Complete spontaneous regression of an extrahepatic portal vein aneurysm

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Primary portal venous aneurysms are rare; however, they are the most common visceral venous aneurysms, and their pathogenesis is not fully understood. Complications include thrombosis, rupture, and mass effect on adjacent structures. The optimal management of these patients is not known. We describe a patient whose large (6-cm) portal vein aneurysm underwent complete spontaneous regression over several years of serial observation. To our knowledge, this observation has not been reported in the English literature. (J Vasc Surg 2011;53:206-8.)

Primary extrahepatic portomesenteric venous aneurysms (PVAs) are rare. The first PVA was reported in 1956 on autopsy in a 21-year-old woman with cirrhosis by Barzilai and Kleckner.1 Since then <200 cases have been reported in the English literature, with most of these diagnosed incidentally on noninvasive abdominal imaging.2 Their etiology, pathophysiology, and natural history are relatively unclear at this time. Symptomatic patients and those with complications of thrombosis, rupture, and compressive effects usually undergo surgical intervention. Prophylactic repair of large asymptomatic PVAs has been recommended for good-risk patients3; however, the optimal management of these asymptomatic venous aneurysms remains unknown.

CASE REPORT

A 57-year-old man was referred to our vascular clinic with an incidental finding of an asymptomatic 6-cm PVA on computed tomography (CT) scanning in 2002. Six years earlier he had suffered a bout of acute alcoholic pancreatitis and an abdominal ultrasound at that time demonstrated a 5-cm portal vein aneurysm; this prompted cessation of alcohol consumption.

At presentation, the patient denied abdominal pain, jaundice, nausea, vomiting, or weight loss, and had no family history of liver disease. Comorbidities included type II diabetes mellitus, hypertension, hyperlipidemia, chronic obstructive pulmonary disease, aortic valve insufficiency, and nonischemic alcoholic cardiomyopathy, with a left ventricular ejection fraction of 25% to 30%. The physical examination was unremarkable, with no stigmata of chronic hepatic disease or portal hypertension. He had no organomegaly on palpation or abdominal bruits. Results of laboratory investigations, including liver function tests and hepatitis profile, were within normal reference ranges at the initial assessment and remained so throughout the follow-up period. The CT scan showed the saccular, 6-cm extrahepatic PVA with no intramural thrombus and no fatty infiltration of the liver; the spleen and pancreas were unremarkable (Fig 1).

Because he was asymptomatic and medically high risk, we elected for serial observation and aspirin therapy. During 5 years (2002 to 2007) of follow-up, he remained asymptomatic, his overall medical status was stable, but he was diagnosed with hyperlipidemia and was prescribed simvastatin in 2006. There was no change in weight, cardiac function (evidenced by echocardiogram showing 25% to 30% ejection fraction in 2007 and 2009), or liver function. The PVA remained stable and did not undergo any change in size on annual CT scanning.

Unknown to us, he was reported by our gastroenterology colleagues as a large (6-cm) stable asymptomatic PVA in 2007.4 Our follow-up CT imaging that same year showed the maximal diameter of the PVA was now 4.7 cm. Another follow-up CT scan 1 year later revealed complete regression of the PVA, with the portal vein now measuring 1.6 cm at its greatest diameter (Fig 2), and it has remained unchanged over the last 2 years of follow-up (2008 to 2010).

DISCUSSION

The portal vein is formed by the junction of the superior mesenteric and splenic veins anterior to the inferior vena cava and posterior to the neck of the pancreas, at the level of the second lumbar vertebra.5 In adults, it measures about 8 cm in length, but its diameter ranges from 0.6 to 1.2 cm on autopsy and up to 1.9 cm on ultrasound imaging.4 There is general agreement that when its diameter reaches or exceeds 2 cm, it is considered aneurysmal in adults.4 The normal pressure of the portal venous system is 5 to 10 mm Hg and maintains the liver blood flow at approximately 1 L/min.6

The etiology and pathogenesis of PVAs remains controversial. Two current theories exist: The first considers congenital factors with the incomplete regression of the right vitelline vein or an inherent weakness in the venous wall resulting in saccular aneurysms, which is supported by a report of the in utero diagnosis of portal PVA by ultrasound imaging. The second theory favors acquired factors, portal hypertension, and compensation of vein wall strength resulting in more fusiform aneurysms.4 Portal hypertension has been reported in about 30% and liver
cirrhosis in 12% to 28% of patients with PVA, and these findings support the hypothesis that portal hypertension is contributory but not essential for the development of PVA.2,7

Saccular extrahepatic PVAs are more common than the fusiform type. These aneurysms are located most commonly at the main portal vein (26.2%), followed by the confluence of the superior mesenteric vein and splenic veins (18.6%) and then the intrahepatic branches (17.1%). There is no gender preponderance, aneurysm diameters ranged from 2.0 to 8.0 cm, and they have been identified in all age groups (range, 0-87 years), with a median age of 52 years.2,8

Differing from arterial aneurysms, physical examination is insufficient to make the diagnosis, and there are no specific laboratory findings. Currently, most patients are diagnosed incidentally while undergoing imaging for other abdominal processes. The prevalence of PVA is 0.6/1000 on ultrasound imaging and 4.3/1000 on multidetector CT scanning.7 Ultrasound imaging, CT, and magnetic resonance imaging can also be used for surveillance or follow-up after treatment. These imaging modalities are readily available today and have made splenoportography nearly obsolete.

Most patients with PVA complain of mild nonspecific abdominal pain (44.7%), others reported symptoms that include jaundice and gastrointestinal bleeding. Reported complications include rupture, thrombosis, mass effect on the bile duct, duodenum or stomach, and inferior vena cava.2,8 Complete thrombosis has occurred in about 14% of reported cases, resulting in two deaths, whereas rupture was uniformly fatal in the two reported patients. Two patients with 2-cm splenic vein aneurysm rupture have also been reported, one in the postpartum period.2

There is no consensus on the use of prophylactic anti-platelet or anticoagulation medications, regardless of the size of the PVA. Most authors advocate serial imaging for patients with asymptomatic small PVAs,7 while patients with thrombus within the PVA are usually treated with surgery or lysis. There is a general tendency for symp-

Fig 1. Coronal and axial views taken in January 2007 show the large, 6-cm saccular, extrahepatic, main portal vein aneurysm, with no mural thrombus or splenomegaly.

Fig 2. Coronal and axial views taken in June 2009 show the extrahepatic main portal vein, with complete regression of the aneurysm.
tomatic patients with PVA or those with complications to undergo surgical intervention. The natural history of extra hepatic PVAs is not well documented, and there is no general consensus on the treatment of asymptomatic PVAs.

Two surgical reports recommend prophylactic surgery in low-risk patients to prevent aneurysm-related complications; however, they cautioned against surgery, especially in patients with portal hypertension and cirrhosis, with a reported mortality of 40% in this patient group. Current surgical therapies have included aneurysmectomy with allograft replacement, aneurysmmorrhaphy alone, liver transplantation when coexisting malignancy exists, and thrombectomy and aneurysmmorrhaphy. In patients who have portal hypertension, shunt procedures combined with splenectomy have been used to decompress the portal venous system. Transhepatic thrombectomy and thrombolysis have also been used in patients with thrombosed aneurysms.

Ozbek et al monitored five patients with serial ultrasound scans and found no enlargement over 2 years. They suggested that in the absence of cirrhosis and portal hypertension, asymptomatic aneurysms can be managed conservatively. A review of follow-up of 53 patients during a mean period of 21 months showed that in 50 (94%) patients, the aneurysm remained stable with no complications, in 2 patients the aneurysm increased in size, and 1 had cavernous transformation. Cho et al, in their series of 6 patients with PVA, observed 2 patients (with 12 and 56 months follow-up) in 1 patient the PVA decreased in size.

We cannot explain why the PVA in our patient underwent a rather rapid, complete regression. Although the starting of a statin medication 2 years before the regression was observed may be totally coincidental, it is an interesting observation. To date, we have the longest follow-up with serial observation in an asymptomatic patient without portal hypertension, and to our knowledge, this is the only reported case with complete spontaneous regression of the PVA.

CONCLUSIONS

Although we cannot draw strong conclusions from this case, we think that observation with periodic imaging of asymptomatic PVAs in medically high-risk patients, especially in those without portal hypertension, may be reasonable until the natural history of these aneurysms is better understood.

REFERENCES


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