

Case Report

Giant Portal Vein Extra Hepatic Aneurysm: An Uncommon Finding

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Abstract

Portal vein aneurysm is a localized dilation of the venous wall above its normal size. Portal aneurysms are rare lesions, with less than 200 cases published world wide. Usually, they are asymptomatic and incidentally discovered by imaging tests. When symptoms appear cause thrombosis, bleeding, porto-systemic shunts or compression of biliary duct, duodenum or inferior vena cava. Follow-up with ultrasound is the management of choice in most of the cases. Surgical technique depends on size, location, liver disease and complications. With portal thrombosis, anticoagulant treatment is also required. Aneurysmorrhaphy, the leading treatment for saccular aneurysms, is the most frequent treatment. Aneurysmectomy is the first choice for fusiform aneurysms. The portal wall may be replaced by prosthesis or cadaveric donor vein. When there is an associated portal hypertension the better treatment is a portocaval shunt with splenectomy. Patients with cirrhosis and portal hypertension have an important surgical risk.

Introduction

Visceral venous aneurysm are rare lesions, most frequently located at the hepatic portal system, though these account for less than 3% of all venous aneurysms [1-14]. Barzilai and Klechner reported the first case in 1956, with less than 200 cases reported since [1-6,9,11,12].

Portal Vein Aneurysm (PVA) is a localized dilation of the venous wall above its normal size. Normal portal vein diameter depends on patient age and portal pressure. PVA's are usually asymptomatic and incidentally discovered [2,4,13,15], which explain the increasing number of cases diagnosed as doppler ultrasounds (Doppler-US), Computer Tomography (CT) scans and Magnetic Resonance Imaging (MRI) are increasingly used [1-5,7,8,10-13,15]. Etiology of PVA's is unknown although they may be secondary to advanced portal hypertension, pancreatitis, trauma, endo-phleboscrosis, liver transplant surgery or, less frequently, to a congenital weakness of the portal wall [1-4,6-15]. They may be intrahepatic but more usual they are extrahepatic. They may also have a fusiform or saccular shape. The later are less prevalent but more prone to complicate [9]. Symptoms appear when PVA's cause thrombosis,

bleeding, porto-systemic shunts or compression of biliary duct, duodenum or inferior vena cava. Abdominal pain (44%-54%) is the most frequent symptom, occasionally associated with gastrointestinal bleeding (9.8%) or rarely with fever, abdominal distension, dyspepsia, nausea, vomiting or jaundice [1-4,6,7,9-12,14,15].

Papers on this subject describe isolated cases and individualized treatments, so there is no a consensus on the surgical technique and timing [1-3,8,9,12-14]. Most of the authors agree to follow-up asymptomatic PVA's patients, since 88% of the aneurysms will keep the same size and will present no symptoms or complications. When portal thrombosis is present anticoagulant treatment is warranted. Aneurysmorrhaphy is the standard technique for PVA's repair, while portocaval shunt is the preferred choice when portal hypertension is present [1,3,4,7,10-14].

Case report

A 74 years old white woman complained of postprandial dyspepsia unremitting with proton pump inhibitors. Her medical record included arterial hypertension, type 2 diabetes mellitus, dyslipidemia, and fibromyalgia, repair of lumbar medullary canal steno-

sis secondary to degenerative scoliosis, cholecystectomy and total hysterectomy for benign lesions. Her liver function tests were normal and her viral hepatitis blood tests were negative. Abdominal US showed a 53x44x34 mm lesion at the liver hilum. Angio-CT scan found a portal vein of 56 mm in diameter (Figure 1).

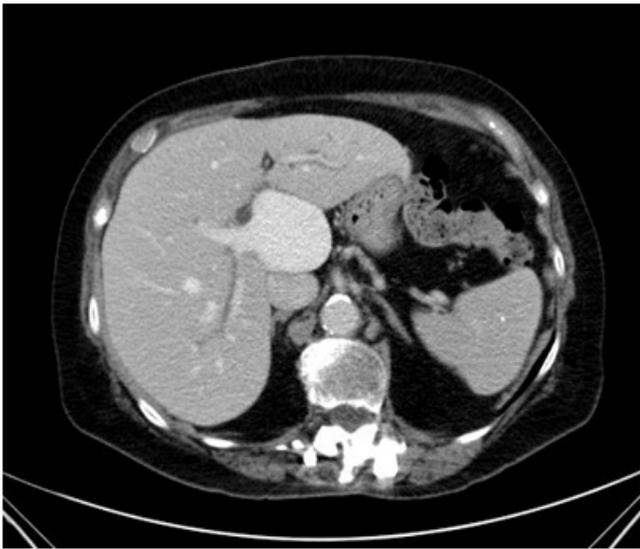


Figure 1: Portal vein extra hepatic aneurysm of 56 mm in diameter.

Intra operative US visualized a 60 mm PVA that was treated by Aneurysmorrhaphy. She was discharged on 7th postoperative day after an uneventful recovery. Pathology report described a normal vein wall with localized none specific regressive changes.

One month later, abdominal US and doppler-US showed a normal diameter portal vein with a normal flow. Patient remained without symptoms 15 months after surgery.

Discussion

PVA's are the most frequent visceral venous aneurysms. Their prevalence was 0.43% in Koc et al. report obtained after reviewing 4186 abdominal CT scans. There were no significant differences regarding age or sex [15]. The largest study was reported by Sfyoreas et al. who collected 191 cases [1].

Most authors considered as PVA's those with a diameter greater than 2 cm. Extrahepatic aneurysms are 63% of total PVA's and may be found at the main portal vein (52%), at the superior mesenteric-splenic vein confluence (44%) or at the right or left portal vein (3-4%) [1,2,5-7,10,11,15]. Although some authors like Oleske et al. and others found smaller diameter in intrahepatic than in extra hepatic PVA's, others have not found significant differences regarding their site [1,2,4,8,10,15].

The congenital origin of PVA's appeal to an abnormal development of the portal system during the complex course of involution of the right and left yolk veins surrounding the forthcoming duodenum. Others resort to an increased portal flow due to arterio-portal fistulas or porto-hepatic shunts [6]. This congenital background is based on the finding of PVA's in children and young adults, even in intrauterine fetuses, without portal hypertension or associated liver disease. Congenital PVA's are considered stable and their management consist of imaging follow-up.

For some authors, the pressure of portal hypertension on the relatively thin portal venous wall mainly causes acquired PVA's. Portal thrombosis, associated with up to 30% of cases with PVA's, may be considered causative because of the resultant increased venous pressure. Thus, patients with thrombophilia are at greater risk of developing PVA's [1,6,8-11,15].

In general, symptoms depend on aneurysm size. The biggest ones may cause symptoms in a third of the patients due to complications. Sfyoreas et al. reported complete thrombosis in 13.6% and partial thrombosis in 3.4% of the cases. More than half of the patients (54.1%) present nonspecific abdominal pain as the sole symptom, 9.8% present gastrointestinal tract bleeding, mainly due to esophageal varicose veins, 11.5% show even less remarkable symptoms like fever, abdominal distension, weight loss, general malaise or jaundice while the remaining 24% are incidental findings. Additionally, 30.8% of the cases are associated with portal hypertension and 28.3% with liver cirrhosis [1-4,6,7,9-12,14,15].

Incidental PVA's appear in 0.6/1000 abdominal US studies and in 4.3/1000 abdominal CT scans as reported by Lall et al. Doppler-US, venous phase mesenteric angiogram, MRI and liver scintigraphy may be used [2,6,7,11,15]. CT scan gives the better anatomical view, showing a well-defined mass, its size and location. Doppler-US may render more data about the surrounding veins. Mesenteric angiogram may be disturbed by thrombosis. Transjugular hepatic venography can measure venous pressure and take liver biopsies; although it is an invasive technique, its anatomical definition of the portal anatomy is valuable [2,6,15].

Follow-up with US is the management of choice in 88% of the cases, which showed a stable size and no complications after Sfyoreas et al. [1,7,14]. Surgical technique depends on size, location, liver disease and complications. Aneurysmorrhaphy, aneurysmectomy, both with or without splenectomy, aneurysmectomy with splenectomy and portocaval shunt, Aneurysmorrhaphy and portocaval shunt, liver transplant in patients with liver disease or radiologic approaches like tranship aticthrombectomy or thrombolysis have been described (Figure 2) [1,4,7,11,12]. With portal thrombosis, anticoagulant treatment is also required. Aneurysmorrhaphy, the leading treatment for sacular PVA's, is the most frequent

treatment. After resection of the redundant vein wall a normal portal diameter and flow should be obtained. Aneurysmectomy is the first choice for fusiform PVA's. The portal wall may be replaced by prosthesis or cadaveric donor vein [1,3]. When there is an associated portal hypertension the better treatment is a portocaval shunt with splenectomy. Although it decreases portal pressure, it does not treat the aneurysm and may give way to liver encephalopathy [7]. Patients with cirrhosis and portal hypertension have an important surgical risk, with a postoperative mortality of up to 40%, so that the better management (surgery vs. follow-up) is to be established. Savadkahi et al. propose surgical treatment for low risk patients, thus avoiding possible life-threatening complications. If follow-up is decided, frequent doppler-US or CT scans should be done to be aware of the portal diameter and flow [1-3,7,8,11,12].

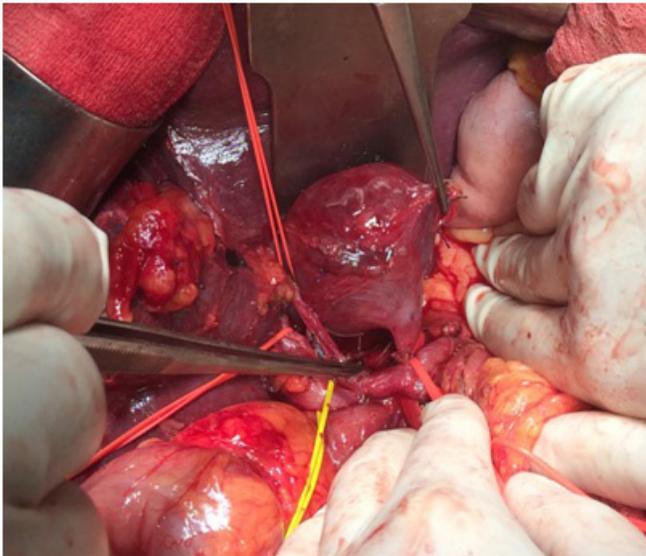


Figure 2: Dissection of portal aneurysm for aneurysmorrhaphy.

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