

• CASE REPORT •

Extrahepatic portal vein aneurysm: Two case reports of surgical intervention

Bi Jin, Yuan Sun, Yi-Qing Li, Yu-Guo Zhao, Chuan-Shan Lai, Xian-Song Feng, Chi-Dan Wan

Bi Jin, Yuan Sun, Yi-Qing Li, Yu-Guo Zhao, Chuan-Shan Lai, Department of Vascular Surgery, Union Hospital, Huazhong Science and Technology University, Wuhan 430022, Hubei Province, China
Xian-Song Feng, Chi-Dan Wan, Department of Portal Hypertension Surgery, Union Hospital, Huazhong Science and Technology University, Wuhan 430022, Hubei Province, China

Correspondence to: Dr. Bi Jin, Department of Vascular Surgery, Union Hospital, Huazhong University of Science and Technology, Wuhan 430022, Hubei Province, China. jinbi2000@hotmail.com
Telephone: +86-27-85726396

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Abstract

We report two cases of extrahepatic portal vein aneurysm, and both of them underwent surgical intervention. The first case had a mild pain in right upper quadrant of the abdomen; the second had no obvious symptoms. Physical examination revealed nothing abnormal. Both of them were diagnosed by magnetic resonance imaging angiography (MRA). One of the aneurysms was located at the main portal vein, the other, at the confluence of the superior mesenteric vein and the splenic vein, and these two places are exactly the most common locations of the extrahepatic portal vein aneurysm reported in the literature (30.7% each site). The first case underwent aneurysmorrhaphy and the second case, aneurysm resection with splenectomy. Both of them recovered soon after the operation, and the symptom of the first case was greatly alleviated. During the follow-up of half a year, no complication and adverse effect of surgical intervention was found and the color Doppler ultrasonography revealed no recurrence of the aneurysmal dilation. We suggest that surgical intervention can alleviate the symptom of the extrahepatic portal vein aneurysm and prevent its complications effectively and safely for low risk patients.

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Key words: Extrahepatic portal vein aneurysm; Surgical intervention; Splenectomy

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INTRODUCTION

Extrahepatic portal vein aneurysm is a rare clinical entity that can be caused by hepatic disease with portal hyperten-

sion or by the malformation of vein^[1], and there are no more than 50 cases reported in the literature. The first entity was reported by Barzilai and Klekner in 1956^[2]. It can happen at any age and with no sexual preference. In most cases it was asymptomatic, so it is not easy to be discovered. It is defined as a focal dilatation of the portal venous system that presents a fusiform or saccular configuration^[3-6]. Ultrasonographic studies have been conducted to evaluate the normal dimensions of the portal vein but considerable variations of the mean portal vein diameter have been reported^[7-9]. Douts and Pearce^[10] reported the maximal diameter of the portal vein as approaching 19 mm. So a diameter greater than 20 mm is therefore regarded as diagnostic standard of extrahepatic aneurysm^[3]. The two most common locations are the main portal vein and the confluence of the superior mesenteric vein and the splenic vein^[11]. In this paper, two cases of extrahepatic portal vein aneurysm and their surgical treatment are presented. One is an aneurysm locating at the truncus of the portal vein; the other is a splenic vein aneurysm at the confluence of the superior mesenteric vein and the splenic vein.

CASE REPORT

Case 1

A 45-year-old woman presented with intermittent right upper quadrant pain that had lasted for 6 mo. Physical examination revealed nothing abnormal. She had a history of hypertension for 10 years. Laboratory analysis was within normal limits except fasting blood glucose, which was 11.8 mmol/L. The magnetic resonance imaging angiography (MRA) revealed an aneurysmal dilation at the truncus of the portal vein, measuring 3.0 cm×3.8 cm in diameter, and the distance between the body of the aneurysm and the bifurcation of the portal vein was 0.8 cm (Figure 1A). To prevent the complications of the aneurysm and relieve the abdominal pain, an operation was performed. During the operation, we carefully separated the aneurysm which was located at the truncus of the portal vein from the hepato-duodenal ligament. The diameter of the aneurysm was about 3 cm×4 cm (Figure 1B). The common hepatic artery and the common bile duct were compressed, but the dimension of the duct above the aneurysm had not increased. No abnormality of the liver and spleen was found. After both sides of the aneurysm were blocked with elastic band, we used a Satinsky claw to divide the body of the aneurysm into two parts and resected the redundant part. The remaining wall of portal vein was sutured, so its diameter had a marked reduction (Figure 1C). In the end, we relaxed the elastic band, and made sure of no hemorrhage of the portal vein.



Figure 1 A: MRA revealed a fusiform dilation of the portal vein, measuring 3.0 cm×3.8 cm in diameter. B: During the operation, the aneurysm was exposed. C: After the aneurysm was resected, the dimension of the portal vein was greatly reduced.

Pathological examination diagnosed the specimen of the aneurysm as congenital amyoplasia. After the operation, anticoagulant was administered for a week. The patient recovered soon and the symptom was greatly alleviated. During half a year's follow-up with Doppler ultrasonography, no stricture and dilation of portal vein was detected, and there was no sign of thrombosis. The symptom has not recurred.

Case 2

A 34-year-old woman without clinical manifestation was detected of an aneurysmal dilation of portal vein in a routine physical examination by the abdominal Doppler ultrasonography. Physical examination and the laboratory analysis including liver-associated enzymes were unremarkable. She suffered from schistosomiasis 20 years ago. MRA revealed an aneurysmal dilation at the confluence of the superior mesenteric vein and the splenic vein. The maximal diameter of the aneurysmal dilation was 4 cm approximately. No other abnormality was revealed (Figure 2 A). A surgical intervention was performed to prevent the complications of the aneurysm. On celiotomy, the aneurysm which measured about 3 cm×4 cm was found situated at the confluence of the splenic vein and the superior mesenteric vein. It had a pedicle connected to the distal extremity of the splenic vein (Figure 2B). There was no obvious abnormality of the liver and spleen. We resected the spleen at first, then separated the pedicle of the aneurysm and ligated it with

silk thread. So the aneurysm could be resected without great difficulty (Figure 2C). Pathologic diagnosis confirmed the specimen to be a true aneurysm of the splenic vein. She had no discomfort during a half year's follow-up, and no abnormality of the portal vein was observed by the Doppler ultrasonography.

DISCUSSION

The portal vein is a unique vessel because of the presence of capillaries on both ends of the system and the absence of valves. The etiology is considered to be congenital; secondary to portal hypertension, chronic hepatic disease or associated with abnormal weakness of the vein wall^[5]. And there were some other rare causes reported: Yang *et al*^[12], reported a portal vein aneurysm caused by gastric adenocarcinoma invading the portal venous system. Tolgonay *et al*^[13], reported a case of a splenic vein aneurysm that appeared and regressed in parallel with splenic size during the course of a systemic infection in a patient with leukemia. In this case, the portal blood flow which is associated with most forms of inflammatory diseases was considered to be related to the cause of portal vein aneurysm. Though various causes were discussed in the literature, the etiology still remains controversial, and the cause in majority of cases is difficult to determine. Expanded use of noninvasive medical imaging techniques has increased the number of patients with portal vein aneurysm but without portal hypertension



Figure 2 A: MRA revealed an aneurysmal dilation at the confluence of the splenic vein and the superior mesenteric vein. The maximal diameter of the aneurysm was 4 cm approximately. B: The aneurysm which measured about 3.0 cm×4.0 cm was exposed. C: The splenic aneurysm was resected.

or chronic liver disease. Portal hypertension was reported in 10 of 12 cases (83%) before 1985 in the literature but in only 2 of 11 cases (18%) after 1985. Chronic liver disease was reported in 9 of 12 cases (75%) before 1985 but in only 2 of 11 cases (18%) after 1985. These data suggest that neither the relationship between portal aneurysm and portal hypertension nor the relationship between portal aneurysm and chronic hepatic disease^[2] is as strong as previously understood. The affirmative etiology still needs to be investigated for a long time with analyses of more entities. Neither of our patients had history or clinical evidence of underlying liver disease, portal hypertension, or other disease states that would predispose them to the development of aneurysms. Though one of the cases had a history of schistosomiasis, but during the operation no abnormality of the liver and no signs of portal hypertension were found.

Though most of the patients with portal vein aneurysm were asymptomatic or presented with mild abdominal pain^[2,11,14,15], some patients presented with jaundice and gastrointestinal bleeding^[6,16-19]. Complications of portal vein aneurysm include thrombosis, aneurysmal rupture, complete occlusion of the portal vein, portal-systemic shunt and pressure effects on adjacent viscera. Large extrahepatic portal vein aneurysms can cause obstruction of the common bile duct and duodenum. Patients may present with recurrent abdominal pain and obstructive jaundice. Acute thrombosis of the portal vein aneurysm can result in severe life-threatening portal hypertension^[5,11,20-22]. One of our two patients had compression symptoms, the other was free from any discomfort, and neither of them had jaundice and gastrointestinal symptoms.

The two cases were both discovered by color Doppler ultrasonography, and then confirmed by MRA (Figures 1A and 2A). Portal vein aneurysm can easily and confidently be established by color Doppler ultrasonography, and it is not very expensive. So color Doppler ultrasonography can be used in follow-up of most of the patients. Computerized tomography angiography, MRA can also determine the location and its relation of the adjacent organs. It is most important that they play a potential role in determining the surgical or therapeutic approach because they can supply adequate information for the surgeons. All of them are non-invasive methods. Angiography is mandatory only in determining surgical approach or when a portacaval fistula is present^[3].

In the past two decades no mortalities of portal vein aneurysm have been reported. Most of the patients keep well with the aneurysm remaining stable in size during follow-up^[4,15,23-25]. It is received mostly that for asymptomatic patients without evidence of portal hypertension or cirrhosis, conservative management with regular follow-up is advisable^[6,11]. But not everyone agreed with this opinion, the more current surgery literature notes that prophylactic surgery for these aneurysms is cautiously recommended for low-risk patients^[26]. We think surgical intervention should be considered once symptoms arise or the aneurysm expands, indicating an increased risk of thrombosis or rupture. In the literature, seven patients underwent surgical interventions for their extrahepatic portal vein aneurysms^[4,5,17,27].

Both the patients underwent surgical intervention. For

the main portal vein aneurysm, an aneurysmorrhaphy operation was performed. The splenic vein aneurysm was resected after splenectomy. The aim of surgical intervention is to relieve the compression symptoms of the aneurysm and prevent the occurrence of the complications. As for the aneurysms located at the main portal vein, the methods of surgical intervention included aneurysmorrhaphy, meso-caval shunt and portacaval shunt. All were documented as having good clinical outcomes during the early postoperative period^[28]. For patients with portal hypertension, a portal-systemic shunt operation is recommended. It avoids dissection or mobilization of the aneurysm, which may be injurious and will bring great difficulty to the operation^[17]. A shunting procedure helps to reduce the portal venous pressure, which may prevent the progressive dilatation of the portal vein aneurysm^[27]. For patients without portal hypertension, aneurysmorrhaphy is advocated as it can preserve the portal blood flow without the complications of portal-systemic shunting, such as encephalopathy. And laminar flow in the portal vein is restored, so stasis of portal blood flow which tends to result in thrombosis can be avoided. Risk of rupture is also reduced as the lateral wall tension is diminished by reduction of the radius after aneurysmorrhaphy^[28]. The surgical treatment of splenic vein aneurysm includes aneurysm resection with splenectomy, splenorenal shunt, and distal pancreatectomy^[29,30], all of which can procure good results.

For patients with greatly impaired hepatic function or those intolerant to shunting or devascularization surgery, non-surgical and less invasive treatments could be considered. Some held the opinion that the increased blood flow in portal hypertension come mainly from the splenic vein. So partial splenic embolization can decrease the blood flow and pressure of the main portal vein, equivalent to the conjoint effects of splenectomy and devascularization^[31].

Before operation, somatostatin can be used in the patients with high portal hypertension for it can effectively lower the portal venous pressure with little side effect. Anticoagulant should be used during the postoperative period to avoid thrombosis.

After careful evaluation of our two patients, surgical interventions were performed successfully and they both experienced an uncomplicated postoperative course. No recurrence of aneurysm was discovered by color Doppler ultrasonography in half a year's follow-up. We suggest that the surgical intervention can relieve the symptoms and prevent the occurrence of the complications effectively and safely for low risk patients. And it should be greatly emphasized that the management should be on a case-by-case basis, depending on the size and anatomy of the aneurysm, as well as the symptoms and clinical condition of the patient^[28]. In summary, extrahepatic portal vein aneurysm is a rare condition, which will be increasingly recognized in the practise of vascular surgery.

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