Rare disease

Enlarged extrahepatic portal vein aneurysm in a non-cirrhotic patient: a therapeutic dilemma

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Summary

The authors report a case of a 75-year-old woman who was seen about 5 years ago and found to have asymptomatic gallstone and extrahepatic portal vein aneurysm (PVA) adjacent to the gallbladder. Further investigations revealed no evidence of cirrhosis. A follow-up ultrasound revealed that the PVA increased in diameter from 20 to 25 mm over 5 years and surgical intervention was recommended. Cholecystectomy and omental wrapping around PVA were performed. Postoperative follow-up investigations for 3 years revealed no increases in PVA diameter. Although PVA is clinically rare, the authors believe that their case report presents an overview of related literature, discusses indications for surgery in PVA and describes their surgical method for managing PVA.

BACKGROUND

Unlike arterial aneurysms, primary venous aneurysms are uncommon. Among them, extrahepatic portal vein aneurysm (PVA) is a relatively rare clinical entity, especially in a non-cirrhotic patient. The management of PVA should be on a case-by-case basis, depending on symptom, presence of cirrhotic status and size and anatomy of PVA.

CASE PRESENTATION

A 75-year-old Japanese woman who had a 5-year history of asymptomatic gallstone and PVA was investigated for medical follow-up. Physical examination and

the laboratory analysis including liver-associated enzymes were unremarkable. Abdominal ultrasonography showed a dilatation in the extrahepatic portal vein (figure 1), which was revealed to be slightly enlarged during this 5 years (from 2.0 to 2.5 cm in diameter). Subsequent abdominal CT (figure 2) revealed an aneurysmal saccular dilation at the truncus of the portal vein, measuring 2.6×2.6 cm in diameter. The aneurysm was significantly enhanced in the portal phase but there was no thrombus in PVA. The liver parenchyma was homogeneous, and the spleen was of normal size without any signs of portal hypertension. There were neither esophageal nor gastric varices.

Figure 1 (A) Abdominal B-mode ultrasonography showed a saccular dilatation in the extrahepatic portal vein. (B) A Doppler ultrasonography showed turbulence of the portal venous flow inside the portal vein aneurysm.

Figure 2 Multi-detector CT of the abdomen revealed an aneurysmal saccular dilation at the truncus of the portal vein adjacent to bifurcation, measuring 2.6×2.6 cm in diameter (white arrows). The aneurysm was significantly enhanced in the portal phase and showed no thrombus inside. A gallbladder with gallstones was located close to the aneurysm (a black dotted arrow). (A, B) enhanced axial images, (B) is slightly more caudal image than that shown in (A). (C, D) enhanced coronal images, (D) is slightly more ventral image than that shown in (C).

Figure 3 During the operation, the portal vein aneurysm, measuring about 3×2.5 cm in diameter, was found situated at the right side of the truncus of the portal vein (white arrows). The common hepatic artery and the common bile duct were mildly compressed but the duct above the aneurysm had not dilated. The wall of the aneurysm was so thin that turbulence of the portal venous flow could be seen. The thin wall composed approximately three fourths of entire wall. CBD, common bile duct; PHA, proper hepatic artery.

Figure 4 Follow-up enhanced CT of the abdomen revealed no alteration of portal venous aneurysm in size (white arrows) during a long period. The aneurysms were accompanied by the wrapped omentum (a black dotted arrows). (A) An image after 1 year since operation. (B) An image after 3 years since operation.

TREATMENT

A diagnosis of enlarged PVA in the non-cirrhotic patient was made and a surgical intervention was performed to prevent complications of PVA. On celiotomy, the aneurysm, measuring about 3×2.5 cm in diameter, was found situated at the right side of the truncus of the portal vein. The common bile duct was only mildly compressed by PVA. A thin wall of PVA composed approximately three fourths of entire wall (figure 3), and aneurysmorrhaphy seemed contraindicated because the procedure may induce stenosis of the portal vein. As an alternative procedure, cholecystectomy and omental wrapping around PVA were performed to prevent possible adjacent inflammation from future spreading to PVA.

OUTCOME AND FOLLOW-UP

The patient recovered soon postoperatively. She remained asymptomatic and her periodic imaging tests showed no alteration in PVA for 3 years since her operation (figure 4).

DISCUSSION

Extrahepatic PVA is a rare clinical entity, with no more than 70 cases described in the English literature to the best of our knowledge.^{1 2} It is defined as a focal dilatation of portal vein that presents an either fusiform or saccular

configuration,^{3 4} and a diameter greater than 20 mm is regarded as diagnostic threshold of extrahepatic PVA.⁵ The first case of PVA, reported by Barzilai and Klekner in 1956, was described as the necropsy of a cirrhotic patient with PVA and thrombus that ruptured into the biliary system.¹ PVA can happen at any age with no sexual preference.⁶ It can be caused by hepatic disease with portal hypertension or by the malformation of vein.⁷ The two most common locations are the main portal vein and the confluence of the superior mesenteric vein and the splenic vein.⁸ Before the 1980s, gastrointestinal bleeding was a typical sign of PVA. Recently, however, with a progress in new imaging modes such as ultrasonography, CT, MRI and angiography, the number of diagnosed cases, especially of the asymptomatic ones, have increased.^{2 9 10}

The treatment of PVA depends basically on the patient's initial clinical symptoms and on the presence or absence of associated portal hypertension. For asymptomatic PVA without any evidence of portal hypertension, conservative management is recommended because most PVAs may remain stable in size during a long follow-up period.^{4 6 11} Surgical intervention is suitable for patients with unstable symptoms or increase in size during observation.^{9 10} The aim of surgical intervention is to relieve compression symptoms and to prevent the complications including rupture and thrombosis. For patients with portal hypertension, a splenectomy and porto-systemic shunt operation is suitable and the direct approach to the aneurysm should be avoided due to the high risk of bleeding.^{10 12} For patients without portal hypertension, the surgical management includes aneurysmorrhaphy and removal or distance of adjacent inflammatory origin.^{3 4 9} Dissection or mobilisation of the aneurysm should be avoided, since it may be injurious and will bring great difficulty to the operation like the case in the current study.⁹ Omental wrapping is a relatively safe and less invasive procedure for infected aneurysms, and can be used to separate aneurysm from surrounding inflammatory lesions.^{13 14} In our case, cholecystectomy and omental wrapping around PVA were performed to prevent possible adjacent inflammation from future spreading to PVA instead of radical aneurysmorrhaphy, and there has been no alteration in PVA for 3 years after surgery. This surgical intervention can be considered as one of the options for the treatment of PVA patients like the current case.

Learning points

- ▶ Extrahepatic PVA is a relatively rare clinical entity, but recently the reported cases increase with a progress in diagnostic imaging techniques.
- ▶ PVA can occur in cirrhotics and non-cirrhotic patients.
- ▶ The management of PVA should be on a case-by-case basis, depending on symptom, size and anatomy of PVA.
- ▶ In case of PVA with gallstones, less invasive surgical intervention including cholecystectomy and omental wrapping procedure can be considered as one of the treatment options.

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Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Barzilai R**, Kleckner MS Jr. Hemocholycyst following ruptured aneurysm of portal vein; report of a case. *AMA Arch Surg* 1956;**72**:725–7.
2. **Andraus W**, Amico EC, Machado MA, *et al*. Portal vein aneurysm. *Clinics (Sao Paulo)* 2007;**62**:203–5.
3. **Dognini L**, Carreri AL, Moscatelli G. Aneurysm of the portal vein: ultrasound and computed tomography identification. *J Clin Ultrasound* 1991;**19**:178–82.
4. **Ohnami Y**, Ishida H, Konno K, *et al*. Portal vein aneurysm: report of six cases and review of the literature. *Abdom Imaging* 1997;**22**:281–6.
5. **Doust BD**, Pearce JD. Gray-scale ultrasonic properties of the normal and inflamed pancreas. *Radiology* 1976;**120**:653–7.
6. **Mhanna T**, Bernard P, Pilleul F, *et al*. Portal vein aneurysm: report of two cases. *Hepatogastroenterology* 2004;**51**:1162–4.
7. **Glazer S**, Gaspar MR, Esposito V, *et al*. Extrahepatic portal vein aneurysm: report of a case treated by thrombectomy and aneurysmorrhaphy. *Ann Vasc Surg* 1992;**6**:338–43.
8. **López-Machado E**, Mallorquín-Jiménez F, Medina-Benítez A, *et al*. Aneurysms of the portal venous system: ultrasonography and CT findings. *Eur J Radiol* 1998;**26**:210–4.
9. **Jin B**, Sun Y, Li YQ, *et al*. Extrahepatic portal vein aneurysm: two case reports of surgical intervention. *World J Gastroenterol* 2005;**11**:2206–9.
10. **Luo HF**, Wang HJ, Li B, *et al*. Diagnosis and management of extrahepatic portal vein aneurysm: a case report. *HBPD INT* 2006;**5**:311–3.
11. **Itoh Y**, Kawasaki T, Nishikawa H, *et al*. A case of extrahepatic portal vein aneurysm accompanying lupoid hepatitis. *J Clin Ultrasound* 1995;**23**:374–8.
12. **Lau H**, Chew DK, Belkin M. Extrahepatic portal vein aneurysm: a case report and review of the literature. *Cardiovasc Surg* 2002;**10**:58–61.
13. **Kitamura T**, Morota T, Motomura N, *et al*. Management of infected grafts and aneurysms of the aorta. *Ann Vasc Surg* 2005;**19**:335–42.
14. **Inafuku H**, Senaha S, Morishima Y, *et al*. Infected thoracoabdominal aortic aneurysms including the major abdominal branches in 4 cases. *Ann Thorac Cardiovasc Surg* 2008;**14**:196–9.

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