World Journal of Gastrointestinal Surgery

Online Submissions: http://www.wjgnet.com/1948-9366office
wjgs@wjgnet.com
doi:10.4240/wjgs.v3.i3.39

World J Gastrointest Surg 2011 March 27; 3(3): 39-42
ISSN 1948-9366 (online)

CASE REPORT

A case report of extrahepatic portal vein aneurysm with thrombosis

Ken Ishimura, Tsuyoshi Otani, Hisao Wakabayashi, Keiichi Okano, Fuminori Goda, Yasuyuki Suzuki

Ken Ishimura, Tsuyoshi Otani, Hisao Wakabayashi, Department of Surgery, Kagawakensaiseikai Hospital, 1331-1 Tahikami, Takamatsu, Kagawa, 761-8076, Japan
Keiichi Okano, Fuminori Goda, Yasuyuki Suzuki, Department of Gastroenterological Surgery, Faculty of Medicine, Kagawa University, 1750-1 Miki, Kagawa, 761-0793, Japan
Author contributions: Ishimura K, Otani T, Wakabayashi H, Okano K, Goda F and Suzuki Y designed and performed research; Ishimura K wrote the paper.
Correspondence to: Ken Ishimura, MD, Department of Surgery, Kagawakensaiseikai Hospital, 1331-1 Tahikami, Takamatsu, Kagawa, 761-8076, Japan. ishimura@kms.ac.jp
Telephone: +81-87-8681551 Fax: +81-87-8689733
Received: February 21, 2010 Revised: November 29, 2010
Accepted: December 6, 2010
Published online: March 27, 2011

Abstract

Extrahepatic portal vein aneurysm (PVA) is very rare with only 17 previously reported cases. Methods of treatment include resection, thrombectomy, and portal venous decompression. We report herein the first case of large PVA with thrombosis which has been managed without surgical treatment over a long period. A PVA was detected in a 78-year-old woman by abdominal ultrasonography. Computed tomography revealed an aneurysm of 6 cm in a diameter in the porta hepatis. Portal venography showed obstruction of the portal vein and developed collateral vessels around the aneurysm. Since the patient had no symptoms of portal hypertension, we decided to carefully manage her clinical course without surgical treatment. At present, this patient is healthy and has developed no complications over the 5 years since leaving our hospital. This case suggests that surgical treatment is not required for PVA without portal hypertension.

Key words: Portal vein aneurysm; Thrombosis; Surgical treatment

Peer reviewers: Robert Chamuleau, MD, PhD, Department of Hepatology, Academic Medical Center, University of Amsterdam, Meibergdreef 69-71, Building S, Floor 1, Room 133, 1105 BK Amsterdam, The Netherlands; Marcelo AF Ribeiro, MD, PhD, TCBC, TCBCD, FACS, Department of Surgery, Santo Amaro University, Alameda Gregorio Bogossian Sobrinho, 80 /155, Santana de Parnaiba, SP 06543-385, Brazil

Ishimura K, Otani T, Wakabayashi H, Okano K, Goda F, Suzuki Y. A case report of extrahepatic portal vein aneurysm with thrombosis. *World J Gastrointest Surg* 2011; 3(3): 39-42 Available from: URL: http://www.wjgnet.com/1948-9366/full/v3/i3/39.htm DOI: http://dx.doi.org/10.4240/wjgs.v3.i3.39

INTRODUCTION

Portal vein aneurysm (PVA) is a very rare venous malformation. Barzilai *et al*[1] reported the first case of PVA in 1956. To our knowledge, only 17 proven cases of extrahepatic PVA have previously been reported in the English language, worldwide[1-16]. The etiology of PVA is unknown due to its rarity. PVA is thought to be either of congenital origin, caused by hypoplasia or atresia of the portal vein, or acquired as a result of portal hypertension or trauma. Methods of PVA management include observation, resection, thrombectomy, and portal venous decompression. We report herein a case of large PVA with thrombosis that has been managed without surgical treatment in a long-term.

CASE REPORT

A 78-year-old woman was admitted to our surgical department with a PVA. Her chief complaint was abdominal discomfort. The patient had no history of abdominal surgery, liver biopsy, trauma, or hepatitis. She had already been diagnosed 6 years previously at another medical institution with an aneurysm of 3 cm in a diameter located in the main portal trunk. However, she had not under-

Table 1 Reported extrahepatic portal vein aneurysms

Author (year)	Age (yr)	Sex	Size(cm)	Liver disease	Portal hypertension	Treatment
Barzilai *et al*[1] (1956)	21	F	2	Liver cirrhosis	+	Splenectomy
Leonsins *et al*[2] (1960)	52	M	8	Liver cirrhosis	+	Splenectomy
Sedgwick[3] (1960)	25	F	5	Liver cirrhosis	+	Cholecystojejunostomy
Hermann *et al*[4] (1965)	26	F	6	Portal fibrosis	+	Portocaval shunt
Liebowitz *et al*[5] (1967)	55	F	8	-	-	Splenectomy
Thomas[6] (1967)	18	M	8	Obstructive jaundice	+	Died
Thomas *et al*[6] (1967)	13	F	3	-	+	Portocaval shunt
Vine *et al*[7] (1979)	50	F	3	Hepatic parenchymal abnormality	-	Observation
Boyez *et al*[8] (1986)	57	F	4	-	-	Observation
Thompson *et al*[9] (1986)	21	F	6	-	-	Cholecystectomy
Andoh *et al*[10] (1988)	57	F	8	-	-	Partial resection PVA Splenectomy
Lee *et al*[11] (1989)	5	M	1.9	-	-	Observation
Baker *et al*[12] (1990)	34	F	8	-	-	Resection PVA Splenectomy
Hagiwara *et al*[13] (1991)	34	M	2.7	-	-	Observation
Dognini *et al*[14] (1991)	67	F	2.4	-	-	Sphincterotomy
Glazer *et al*[15] (1991)	26	F	7	-	+	Thrombectomy Aneurysmorrhaphy
Brock *et al*[16] (1997)	72	F	6	-	-	Resection PVA
Present case	78	F	6	-	-	Observation

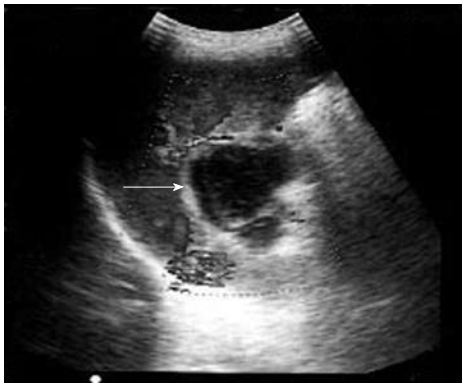

Figure 1 Ultrasound oblique scan through the long axis of the portal vein.

gone further examination until this hospitalization. On admission, the patient was 137 cm in height and weighed 40 kg, having exhibited no weight loss. Her consciousness was alert. Results of a general physical examination were otherwise normal; the palpebral conjunctiva was not anemic and the bulbar conjunctiva was not icteric. The liver and spleen were not palpable, and the abdomen was soft and flat, with no palpable tumor. No vascular bruit was heard. Laboratory test results included aspartate transaminase of 22 U/L, alanine transaminase of 63 U/L, and alkaline phosphatase of 430 U/L. Abdominal ultrasonogram showed a hypoechoic mass connected with the main portal trunk (Figure 1). Computed tomography scan revealed a mass of 6 cm in a diameter in the porta hepatis (Figure 2A). Developed collateral vessels around the mass were detected on the delay phase by intravenous contrast material, suggesting thrombosis of the portal aneurysm (Figure 2B). The venous phase of a superior mesenteric artery angiogram showed obstruction of the portal vein, and that intrahepatic portal flow was maintained by collateral supply (Figure 3). There was no arterial or venous fistula. As the patient's liver function was almost within normal limits and she showed no symptoms of portal hypertension, we decided not to intervene surgically but to carefully monitor her clinical course. She was well when discharged, and has remained symptom-free for 5 years since leaving the hospital.

DISCUSSION

Since the natural history of PVA is not clear, it is difficult to determine the strategy of treatment for this disease.

The 18 reported cases of extrahepatic PVA, including our case[1-16], are listed in Table 1. The age of patients in these cases varies from 5 to 78 years old, with our patient being oldest. The PVA ranged from 1.9 to 8 cm in a diameter, with an average diameter of 5.3 cm. Six of the 18 cases revealed underlying liver disease[1-4,6,7], and seven were associated with portal hypertension[1-4,6,15]. The advance of radiological diagnosis has resulted in the identification of a number of PVA without liver disease since 1986. Surgical treatments were performed on 13 out of the 18 cases, and direct surgery for PVA was carried out in four[10,12,15,16]. Resection of PVA was done in three cases, and thrombectomy and aneurysmorrhaphy were performed on one case with thrombus. On the other hand, five cases including our case, were managed by observation[7,8,11,13]. A PVA of 4 cm in diameter monitored for 2 years did not change in size[8] and aneurysms reported by Lee and Hagiwara did not change in size after 5 years of observation[11,13]. These aneurysms were smaller than the one observed in our patient.

Excluding our case, three out of 17 patients had throm-

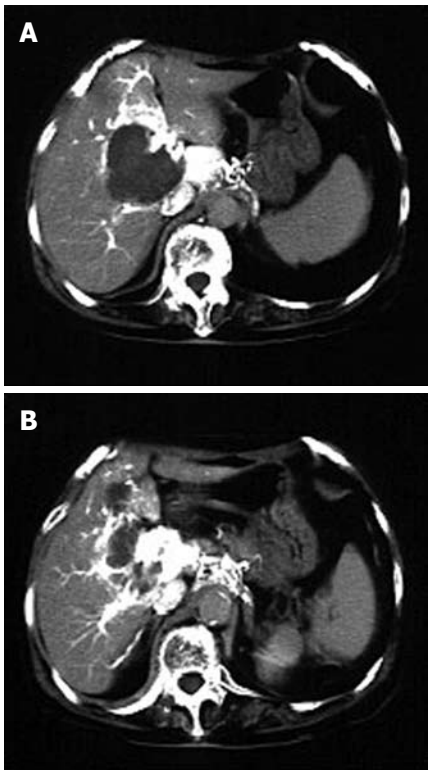

Figure 2 Abdominal computed tomography scan with contrast. A: A hypovascular mass in the porta hepatis; B: Developed collateral vessels around the mass.

bus[1,6,15]. Two of these died of aneurysm rupture[1,6]. Both were less than 30 years old. Their PVA etiology seemed to be acquired, as their PVA was the result of liver disease. On the other hand, the remaining patient was treated by a surgical procedure, and remained symptom free for 10 years after operation[15]. The origin of this case was also acquired. However it is unclear whether operation was needed in this case, given that the PVA contained a large amount of organized clot and the wall of the aneurysm showed normal venous structure with no atrophy of the muscle. Our patient had chronic progress, and PVA etiology was suspected to be of congenital origin. She was saved from rupture of the PVA as collateral vessels had developed around it. We inferred that the patient was symptom free because of the formation of sufficient collaterals. We believe that after short follow-up asymptomatic aneurysm with thrombus can be successfully managed by observation alone.

The decision on surgical treatment depends on the size, anatomy of PVA as well as the symptoms and condition of the patient. In the past, large PVAs over 4 cm in diameter had been operated on. Ours seems to be the first case reported without operative treatment for PVAs over 5 cm in a diameter. Miyauchi *et al*[17] concluded that the indications for surgical interventions in the treatment of PVA with porta hepatic venous fistula were as follows: (1) Patients with symptoms and large shunts; (2) Patients with enlarging fistulae; and (3) Patients with multiple fistulae where angiography shows that the lesions are sufficiently localized that the volume of the shunt cannot be reduced by conservative therapy. Moreover, patients who have biliary tract obstruction and hemobilia caused by PVAs also require operation.

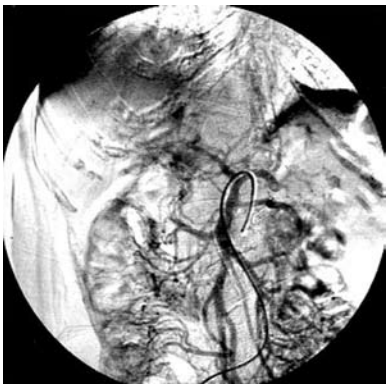

Figure 3 Portal venogram (digital subtraction angiogram) after selective superior mesenteric arteriography.

Since the natural history and incidence of PVA is not well known, it is difficult to decide the best treatment. Prognosis in symptomatic patients treated with surgery is dependent on the underlying liver disease. In a case reported by Brock, the patient underwent PVA resection because of a lack of experience for judging whether the large uncomplicated saccular PVA in his patient should be resected[16]. However, our case indicated a natural history of PVA without arterial or venous fistula, and suggests that surgical treatment is not required for PVA in the absence of portal hypertension. Moreover, our case also suggests that it might be possible to decide PVA treatment based on etiology.

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S- Editor Wang JL **L- Editor** Hughes D **E- Editor** Zheng XM