



A Rare Cause of Right Upper Quadrant Pain*

Rafid Al-Mahfoudh MBChB, Derek A. O'Reilly MRCS and Mike G.T. Raraty MB BS PhD FRCS

Department of Surgery, Royal Liverpool University Hospital, Liverpool, UK

Key words: portal vein aneurysm, magnetic resonance imaging, duplex ultrasound

IMAJ 2007;9:171-172

Portal vein aneurysm is a rare vascular anomaly whose cause, clinical manifestations and management have not yet been clarified. It is a rare cause of gastrointestinal symptoms but is likely to be increasingly diagnosed with the growing use of radiologic imaging. Surgical intervention is usually reserved for symptomatic patients or when complications arise. Complications include thrombosis, aneurysmal rupture, complete occlusion of the portal vein, portal-systemic shunt, and pressure effects on adjacent viscera.

Surveillance of portal vein aneurysms is necessary to define their clinical course and appropriate management. We report the case of a 55 year old woman who underwent abdominal ultrasound for right upper quadrant pain. In this patient a conservative approach with regular surveillance was adopted for this asymptomatic portal vein aneurysm. This appears to be a safe strategy.

Patient Description

A 55 year old woman presented to her physician in December 2001 after a single episode of right upper quadrant pain which settled in a few days and has not recurred since. Her medical history included a total abdominal hysterectomy in 1990 and she had been involved in a traffic accident in March 2000. She had been wearing a seatbelt on that occasion but sustained an impact to her right side. Physical examination was normal with no



[A] MRI showing portal vein aneurysm measuring 3.7 cm in maximal diameter (white arrow)

signs of chronic liver disease or portal hypertension; no bruit was heard on abdominal examination. A liver ultrasound to check for gallstones was performed. This showed an aneurysm of the portal vein at its bifurcation, measuring 2 cm in diameter.

She was referred to the Royal Liverpool University Hospital where she has been under surveillance since 2002.

Follow-up imaging for the aneurysm was largely unchanged with similar morphology, as follows: September 2002, magnetic resonance imaging, 3 × 2.5 cm; April 2003, MRI, 3.7 × 3 cm [Figure A]; May 2004, ultrasound, 2.1 cm [Figure B]; June 2005, ultrasound, 2.5 × 2.1 cm. She has remained asymptomatic throughout the period of surveillance.

Comment

Portal vein aneurysm is a rare vascular anomaly whose cause, clinical manifesta-



[B] Doppler ultrasound showing venous flow the portal vein aneurysm (white arrow)

tions and management have not yet been clarified. Portal vein aneurysm was first described by Barzilai in 1956, in a patient with hepatic cirrhosis. Since then, fewer than 60 cases of extrahepatic portal vein aneurysms have been reported in the literature [1-3]. The frequent application of radiologic imaging for diagnosis and screening of abdominal disorders accounts for the increasing number of case reports in recent years [1].

The portal vein is a unique vessel because of the presence of capillaries on both ends and the absence of valves. The diameter of the portal vein varies considerably, but a segmental expansion of over 2 cm can, as a rule, be considered an aneurysm [4]. Aneurysm of the portal vein may occur at the main portal vein, at the junction of the superior mesenteric and splenic veins, and at the branching portion of the left and right portal veins in the porta hepatis. The two most common locations are the main portal vein and the confluence of the superior mesenteric vein and the splenic vein [5]. Multiple

* Presented at the 18th Israeli Medical Association World Fellowship International Conference

intrahepatic portal vein aneurysms in the same patient have also been reported.

The cause of portal vein aneurysm remains controversial but may be either congenital or acquired. Congenital causes include developmental anomaly, abnormality of the internal walls of the vessel, or failure of regression of portions of the vitelline veins and their anastomoses [5]. The congenital theory hypothesis is strongly supported by the *in utero* diagnosis of portal vein aneurysm. Portal hypertension and chronic hepatic disease have been identified as predisposing factors for portal aneurysm, but the association is now more doubtful as the expanded use of medical imaging has increased the number of patients with portal vein aneurysm without portal hypertension or chronic liver disease [2,3]. Other reported acquired causes include necrotizing pancreatitis, surgery, abdominal trauma, and as a late complication of liver transplantation. In our case, there is history of a traffic accident 20 months prior to the patient's diagnosis, but whether this was causative is a matter of speculation.

The clinical significance of portal vein aneurysm arises mainly due to the development of complications, such as thrombosis, aneurysmal rupture, complete occlusion of the portal vein, portal-systemic shunt, and pressure effects on adjacent

viscera. Acute thrombosis of the portal vein aneurysm can cause severe life-threatening portal hypertension with rupture of the aneurysm, gastrointestinal bleeding or bowel infarction [1,3]. However, in the past two decades no mortality as a consequence of portal vein aneurysm has been reported [3].

Duplex Doppler ultrasonography is accurate for diagnosis and follow-up in the majority of cases. Enhanced dynamic helical computerized tomography may also demonstrate the anatomy of a portal vein aneurysm [1]. MRI and MR angiography are useful for confirming the diagnosis and may have a potential role in determining a surgical or conservative approach. The literature mainly advocates conservative management with regular follow-up for asymptomatic aneurysms. Surgical intervention (aneurysmorrhaphy) is usually reserved for symptomatic patients or when complications arise [3]. For thrombosis of a portal vein aneurysm there are various therapeutic options, including surgical or percutaneous thrombectomy, systemic anticoagulation or local thrombolysis.

In conclusion, portal vein aneurysms are a rare cause of gastrointestinal symptoms but are likely to be increasingly diagnosed. Surveillance of portal vein aneurysms is needed to define their clinical course and appropriate management. At

present, the management should be on a case-by-case basis, depending on the size and anatomy of the aneurysm, presence of symptoms, patient fitness, and the development of complications. As this case illustrates, conservative management with regular surveillance appears to be a safe strategy.

References

1. Lau H, Chew DK, Belkin M. Extrahepatic portal vein aneurysm: a case report and review of the literature. *Cardiovasc Surg* 2002;10:58–61.
2. 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.
3. 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.
4. Dognini L, Carreri AI, Moscatelli G. Aneurysm of the portal vein: ultrasound and computed tomography identification. *J Clin Ultrasound* 1991;19:178–82.
5. Lopez-Machado E, Mallorquin-Jimenez F, Medina-Benitez A, Ruiz-Carazo E, Cubero-Garcia M. Aneurysms of the portal venous system: ultrasonography and CT findings. *Eur J Radiol* 1998;26:210–14.

Correspondence: Dr. R. Al-Mahfoudh, Dept. of Surgery, Royal Liverpool University Hospital, Prescot Street, Liverpool, L7 8XP, UK.
email: rafidalmahfoudh@yahoo.com

Capsule

Rapid advice guideline panel on avian influenza

The recent spread of avian influenza A (H5N1) virus to poultry and wild birds has increased the threat of human infections with H5N1 virus worldwide. Despite international agreement to stockpile antivirals, evidence-based guidelines for their use do not exist. The WHO assembled an international multidisciplinary panel to develop rapid advice for the pharmacological management of human H5N1 virus infection in the current pandemic alert period. A transparent methodological guideline process on the basis of the Grading Recommendations, Assessment, Development and Evaluation (GRADE) approach was used to develop evidence-based guidelines. Our development of specific recommendations for treatment and chemoprophylaxis of sporadic H5N1 infection resulted from the benefits, harms, burden, and cost of interventions in several patient and exposure groups.

Overall, the quality of the underlying evidence for all recommendations was rated as very low because it was based on small case series of H5N1 patients, on extrapolation from preclinical studies, and high quality studies of seasonal influenza. A strong recommendation to treat H5N1 patients with oseltamivir was made in part because of the severity of the disease. Similarly, strong recommendations were made to use neuraminidase inhibitors as chemoprophylaxis in high risk exposure populations. Emergence of other novel influenza A viral subtypes with pandemic potential, or changes in the pathogenicity of H5N1 virus strains, will require an update of these guidelines and the WHO will be monitoring this closely.

Lancet Infect Dis 2007;7:21

Eitan Israeli

