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Single Case

Repeated Plastic Stentings of Common Hepatic Duct for Portal Vein Aneurysm Compression in a Patient Unsuitable for Surgery

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Keywords
Portal vein aneurysm · Intraductal ultrasonography · Common hepatic duct · Intrahepatic duct · Biliary stent

Abstract
Portal vein aneurysms are rare vascular malformations with unclear etiologies and optimal treatment guidelines. Although Doppler ultrasonography is the most commonly used diagnostic tool, there is no gold standard imaging modality. Despite recommendations of surgical treatment for symptomatic aneurysms, there are limited options in the management of portal vein aneurysm-related complications in patients unfit for surgical intervention. We describe an 85-year-old man who presented with abdominal pain and low-grade fever with clinical signs consistent with cholangitis. Endoscopic retrograde cholangiopancreatography revealed a common hepatic duct stricture and concomitant intraductal ultrasonography identified adjacent aneurysmal portal vein dilatation. The final diagnosis of portal vein aneurysm was made using contrast computerized tomography scan. The patient was considered unsuitable for surgery due to his advanced age and multiple comorbidities. Instead, an endoscopic biliary plastic stent

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was inserted as a therapeutic alternative, which successfully achieved complete resolution of symptoms 3 days after the procedure. The patient was regularly followed at the outpatient clinic with repeated stent replacements every 3 to 4 months. After a follow-up of over 3.5 years, the patient remained symptom-free without signs of portal vein aneurysm compression. The result suggests that repeated stent replacements may be a therapeutic option for biliary compression by portal vein aneurysm in patients contraindicated for surgical intervention.

Introduction

Portal vein aneurysm (PVA) is defined as a saccular or fusiform dilatation of a portal vein segment exceeding predetermined normal calibers. PVAs have a reported incidence of 0.43% [1, 2], making up less than 3% of all aneurysms of venous origin [1]. The low clinical incidence of this condition has led to ambiguity and difficulty in generating widely accepted management guidelines. These aneurysms are most commonly classified via location of occurrence [3] (intrahepatic and extrahepatic) or etiology (either acquired or congenital) [4]. Up to one-third of patients may be asymptomatic, with PVA being an incidental finding during routine scanning [5]. Symptomatic PVAs commonly manifest as abdominal or back pain with signs of gastrointestinal bleeding, fever, jaundice, abdominal swelling, malaise, and weight loss being reported in some cases [1]. Most PVAs are acquired and often associated with liver disease and portal hypertension [6], so patients may also present clinically with signs of these respective complications. Further complications include elevated risk of thrombosis; manifestations due to compression against adjacent anatomical structures, most notably the common bile duct, duodenum, and inferior vena cava [1]; and spontaneous rupture [3, 5], albeit rarely.

The current preferred treatment option for complicated PVA is surgical, with indications being manifestation of symptoms or evidence of thrombosis or rupture [5, 7]; however, surgical guidelines are still unclear. In suitable patients, minimally invasive procedures such as stenting [6] may also be considered. To date, the long-term result of repeated biliary plastic stentings as a definitive treatment option has not been reported. This article reports a case of PVA diagnosed with intraductal ultrasonography (IDUS) in a patient unsuitable for surgery who was successfully treated with repeated biliary plastic stent exchanges every 3 to 4 months over a period of 3.5 years.

Case Report

An 85-year-old man with a background of dementia, type 2 diabetes mellitus, and peptic ulcer disease presented to the emergency department with complaints of low-grade fever since the morning of the hospital visit. The patient also reported presence of mild intermittent cough and poor activity levels, which he first noticed 6 days ago, as well as dull upper abdominal pain, which has persisted for 1 week, and an accidental fall with head injury to the occipital region. On clinical examination, the patient's temperature was 37.9°C, heart rate 100 beats per minute, respiratory rate 20 breaths per minute, and blood pressure 148/82 mm Hg. The patient's abdomen was soft with normoactive bowel sounds and mild epigastric tenderness without muscle guarding, organomegaly, or any other peripheral stigmata suggestive of any pathology. Initial laboratory tests showed normal hemoglobin 14.7 g/dL (normal: 13–18 g/dL), platelets 221 × 1,000/μL (normal: 140–450 × 1,000/μL), and PT 12.1 s (normal: 9.4–

12.5 s). Abnormal laboratory findings included elevated total serum bilirubin 2.35 mg/dL (normal: 0.3–1.2 mg/dL), alanine aminotransferase (ALT) 365 U/L (normal: 10–50 U/L), aspartate aminotransferase (AST) 724 U/L (normal: 5–35 U/L), white blood cells 10.3 × 10^3/mm^3 (normal: 3.8–10 × 10^3/mm^3), segmented neutrophils 88.3% (normal: 47–75%), and lymphocytes 6.0% (normal: 20–55%). Viral hepatitis markers were negative for HBsAg and Anti-HCV. No other abnormalities were detected in other blood parameters including creatinine and lipase. After surgical evaluation of the head injury, no remarkable sequelae were found, and the patient was given supportive treatment. Considering elevated serum bilirubin, ALT, and AST and normal hemoglobin in conjunction with reported fever and epigastric pain, cholangitis was suspected, for which abdominal sonography demonstrated bilateral intrahepatic duct (IHD) dilatation. Visualization of adjacent structures could not be demonstrated due to air artifact and the patient's inability to cooperate due to underlying dementia. Non-contrast computerized tomography (CT) scan also showed findings consistent with those of ultrasonography (Fig. 1a). Endoscopic retrograde cholangiopancreatography (ERCP) (JF260V, Olympus, Tokyo, Japan) arranged after informed consent revealed indentation of the common hepatic duct (CHD) with upstream bilateral IHD dilatations (Fig. 1b). IDUS (UM-G20-29R, 20 MHz, Olympus, Tokyo, Japan) was performed, which showed a lobulated hypoechoic mass with widest diameter of 11 mm containing mobile echogenic substance outside the CHD stricture, highly suggestive of a vascular lesion (Fig. 1c). Endoscopic sphincterotomy was done and a biliary plastic stent (Cotton-Leung Biliary Stent, 10 Fr, 9 cm, Cook Group Inc., USA) was inserted through the common bile duct, bypassing the CHD stricture resulting in successful drainage (Fig. 2a, b). Contrast CT scan performed 6 h post-ERCP revealed lobulated aneurysmal dilatation of the main portal vein with largest diameter measuring 3.2 cm (Fig. 2c, d), confirming the diagnosis of extrahepatic PVA causing CHD compression. Total serum bilirubin normalized (0.64 U/L) after 3 days, along with resolution of presenting symptoms. The patient was discharged unremarkably 9 days after stenting. He was regularly followed at the outpatient clinic with repeated stent exchanges every 3–4 months over a period of 3.5 years, without any complications arising from the aneurysm or the stent between exchanges. The number of stent exchanges totaled 15 times until the patient passed away from unrelated respiratory failure due to pneumonia.

Discussion

PVA is a rare vascular phenomenon first described by Barzilai and Kleckner in 1956 [8], later defined by Doust and Pearce in 1976 as a portal vein extrahepatic diameter of >2.0 cm and intrahepatic diameter of >0.9 cm [9–11]. Intrahepatic PVAs are often smaller than their extrahepatic counterparts as intrahepatic dilatation is limited anatomically by surrounding liver parenchyma [3]. In our case, the extrahepatic aneurysmal portal vein diameter measured 3.2 cm, consistent with proposed diagnostic criteria.

Whilst there is ambiguity over the etiology of PVAs, the majority are of acquired origins, of which many occur in conjunction with advanced hepatocellular pathology and associated portal hypertension [10]. The combination of hyperdynamic blood flow and increased portal pressure induces thickening of the vessel intima, leading to compensatory medial hypertrophy of the vessel [1, 9]. This is gradually replaced by fibrous connective tissue, which compromises the structural integrity of the venous wall, causing a reduction in tensile strength, ultimately increasing susceptibility to aneurysmal dilatation [5, 9]. There is also speculation of multiple other acquired causes, namely severe pancreatitis, trauma [3, 9], and malignant

invasion of the portal vein [12]. Despite established correlation between portal hypertension and the presence of PVAs, a retrospective review showed that the incidences of cirrhosis and those of portal hypertension in patients with PVA were merely 12 and 32%, respectively [2]. It has also been proposed that an inherent portal vein wall weakness is required along with portal hypertension for the development of PVA. This suggests a congenital component in the etiology, which is further supported by cases in children in the absence of liver disease or portal hypertension [3, 5], as well as a case of in utero diagnosis in a 37-week gestational fetus [13]. The etiology of PVA in our patient was likely weakening of the portal vein vessel wall, taking into consideration the patient's old age and the absence of liver cirrhosis and signs of portal hypertension.

Most reported cases of extrahepatic PVAs tend to occur at the main portal vein and at the junction between the superior mesenteric vein and the splenic vein, whereas intrahepatic PVAs tend to originate at venous bifurcation sites [2, 3]. In our patient, the location of PVA was extrahepatic and was located at the main portal vein, matching previously established common sites. Due to advances in imaging technology, most detected PVAs are asymptomatic and identified incidentally during investigation of unrelated pathology [2, 14]. Most symptomatic PVAs present with non-specific abdominal pain and few patients report this with gastrointestinal variceal bleeding [5] despite background portal hypertension. Symptoms of complications such as thrombosis [2], rupture, and external pressure on adjacent anatomical structures are even rarer [1]. Our reported patient also presented with abdominal pain; however, our case was of the minority due to the PVA resulting in CHD compression ultimately causing hyperbilirubinemia, IHD dilatation, and acute obstructive cholangitis.

It is important to be aware of the appearance of PVA over multiple imaging modalities because PVAs can mimic solid, cystic, and hypervascular abdominal masses [14]. Imaging modalities utilized to achieve a diagnostic picture include Duplex ultrasound, color Doppler, and contrast abdominal CT, with only few reported cases of magnetic resonance imaging (MRI) being used [5, 6]. Whilst there is no gold standard imaging method for diagnosing PVAs because of the rarity of the condition, the convenience of Doppler ultrasonography makes it the most widely used modality [7]. Ultrasounds allow easy non-invasive detection and identification of aneurysms without exposure to radiation and at low monetary cost. Ultrasounds also have the ability to distinguish PVAs from other hypervascular masses [7]. Other more specific imaging techniques including venous phasic mesenteric angiography and hepatic scintigraphy allow complete visualization of the local vasculature, accurately delineating and outlining the borders of its anatomy, but are largely restricted to cases requiring surgery [3]. Usage of CT and MRI can provide more detailed anatomical outline of aneurysms and implementation can be especially effective when surgical intervention is considered [1, 7, 14]. In our case, IDUS, performed in tandem with ERCP, was used as an initial diagnostic tool, revealing IHD compression by a vascular lesion. This diagnostic strategy successfully avoided invasive procedures such as intraductal biopsy, which may lead to vessel rupture and disastrous hemorrhage. To our knowledge, this is the first reported case of the utilization of IDUS to identify the possible presence of a PVA. The final diagnosis of PVA was established via enhanced abdominal CT scan.

Whilst there is still discussion over the optimal treatment of PVAs, it is generally accepted that intervention is indicated in expanding PVAs or in symptomatic cases [5, 7]. Another author has also suggested that presence of non-thrombotic PVAs >3 cm should be considered for surgical intervention [15]. Surgical approaches differ between PVAs with and without portal hypertension. PVAs without portal hypertension are often attributed to congenital etiology [4] with aneurysmorrhaphy or aneurysmectomy as proposed definitive treatment options in

an effort to restore laminar flow in the portal vein [5]. Surgical shunt procedures and liver transplantation [5, 14] are preferred for PVAs occurring in conjunction with extensive liver disease and portal hypertension with the intention of preventing further dilatation. Examples of other proposed alternatives in treating PVA-related complications include cases of PVA-induced thrombosis treated successfully with percutaneous thrombolysis [10, 14] or thrombolectomy [2]. For patients who are unsuitable for operations, biliary stenting has been proposed to be a treatment alternative for PVA causing biliary compression [6]. However, the approach of repeated periodical biliary stent replacements as in our case has not been documented. Due to the patient's advanced age, multiple comorbidities and elevated risk of surgical complications, biliary stenting was utilized with palliative intention. Repeated stent exchanges ensured maintenance of stent patency for the prevention of occlusive complications of the bile duct in the patient during a follow-up of over 3.5 years.

To conclude, we report the first successful experience in the initial identification of extrahepatic PVA through IDUS procedure. This is also the first report of effective long-term biliary plastic stenting with periodical replacements in the palliative treatment of PVA. Our results not only demonstrate the effectiveness of IDUS for diagnosing vascular lesions surrounding the biliary tree, but also suggest that long-term stenting may be a feasible therapeutic approach for PVA-related biliary obstruction in patients unsuitable for surgical intervention.

Statement of Ethics

Written informed consent for publication was obtained from the patient.

Disclosure Statement

The authors declare no conflicts of interest.

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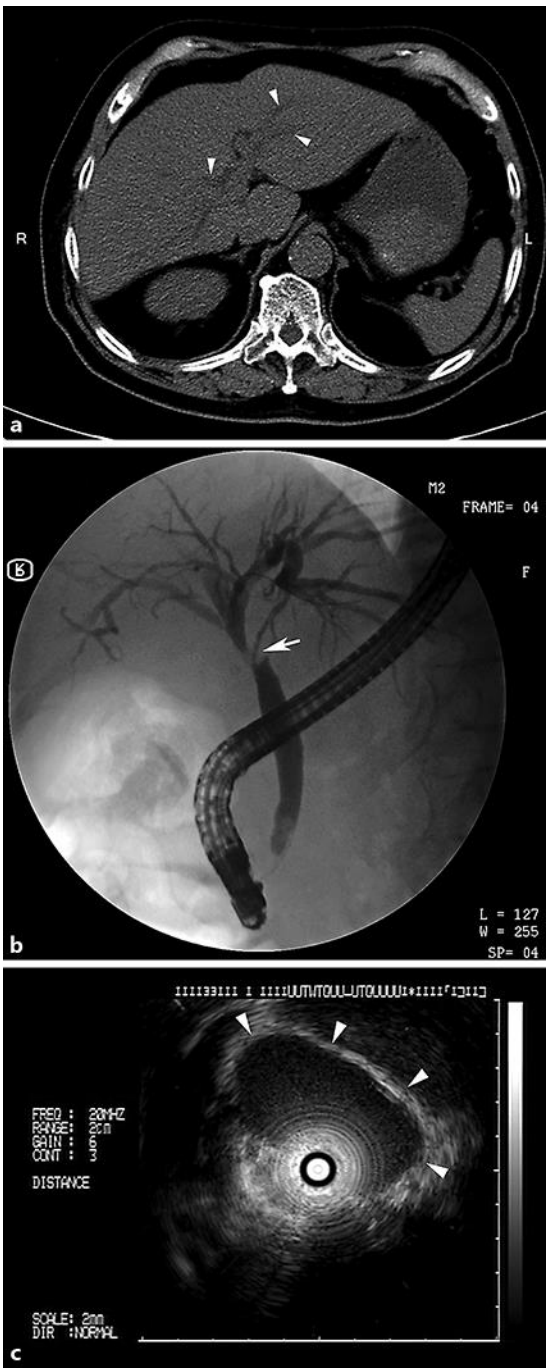

Fig. 1. **a** Non-enhanced abdominal computed tomographic scan showing bilateral intrahepatic ductal dilatations (arrowheads). **b** Endoscopic retrograde cholangiopancreatography demonstrating indentation of the common hepatic duct (arrow) with upstream bilateral IHD dilatations. **c** Intraductal ultrasonography revealing a lobulated hypoechoic mass with widest diameter of 11 mm containing mobile echogenic content outside the ductal stricture, highly suggestive of a vascular lesion.

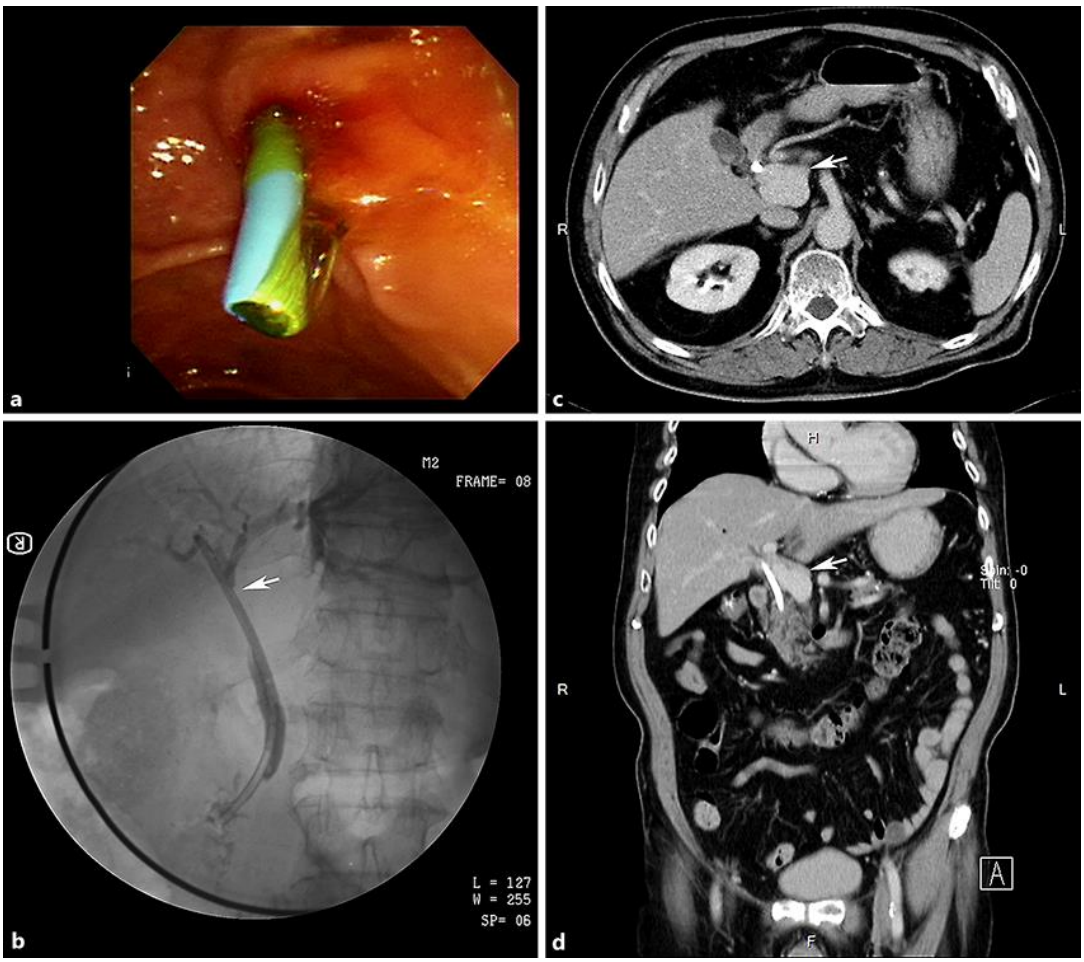

Fig. 2. **a** Insertion of a biliary plastic stent into the common bile duct. **b** Fluoroscopy showing successful biliary tree drainage after stenting bypassing the stricture (arrow). **c** Contrast computed tomographic scan 6 h after endoscopic retrograde cholangiopancreatography revealing lobulated aneurysmal dilatation of the main portal vein with largest diameter measuring 3.2 cm (arrow). **d** Coronal section of enhanced CT scan demonstrating consistent finding of portal vein aneurysm (arrow).