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Case Report

Intrahepatic portal-venous shunts during PVE

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ABSTRACT

Portal venous embolization (PVE) is a well-validated technique to promote contralateral liver lobe hypertrophy prior to hepatic resection. We present a case of a patient with Type IV cholangiocarcinoma undergoing PVE prior to hepatic surgical resection. However, intrahepatic portal-venous shunts were incidentally found during the procedure and were subsequently embolized using embolic coils and N-butyl cyanoacrylate. While most patients with congenital portal-venous shunts remain asymptomatic, an unrecognized shunt during PVE could have resulted in a devastating complication secondary to nontarget embolization through the fistula.

To our knowledge, this is the first reported case of a portal-venous shunt being discovered during a PVE.

Introduction

Portal venous embolization (PVE) is an image-guided technique used to induce parenchymal hypertrophy on one side of the liver prior to hepatic resection on the other side, in order to create an adequate future liver remnant (FLR). This technique redirects blood flow from the targeted portal veins toward the segments of FLR [1]. The size of FLR is typically determined by multiphase contrast enhanced computed tomography (CT), and is subsequently standardized to patient size through a ratio of FLR to total functional liver volume—also known as standardized FLR [1]. This percentage is utilized to determine if a PVE is indicated in a given patient [2]. PVE performed in patients with hepatobiliary malignancies has been shown to be safe and effective in inducing hypertrophy of FLR prior to hepatic resection [3]. Embolic coils, dehydrated alcohol, and spherical microparticles have been widely utilized for PVE. N-butyl-cryanoacrylate is a newer liquid embolic agent that can also be used for PVE and while available data are still limited, it has been shown to be superior to spherical microparticles and coils leading to a greater percentage of left lobe hypertrophy [4]. Absolute contraindications to the procedure include tumor thrombus and clinically significant portal hypertension [1]. Complications of PVE include pneumothorax, cholangitis, FLR injury, hemoperitoneum, and rarely biliary-pleural fistula in the setting of cholangiocarcinoma and obstructive jaundice [1,5,6].

Case report

The patient is a 66-year-old female who presented with abdominal discomfort, bloating, and weight loss of 3 months

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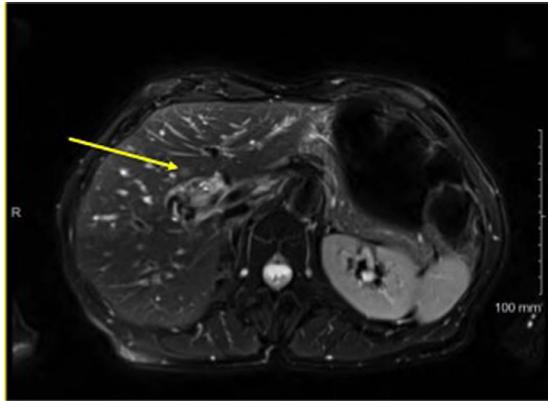
Fig. 1 – MR abdomen was obtained due to clinical elevation in liver function tests and biliary duct dilatation. T2-weighted and T1-weighted postcontrast MRI images showing T2 hyperintense perihilar lesion with delayed contrast enhancement on T1 weighted image (shown by yellow arrows). MR, magnetic resonance; MRI, magnetic resonance imaging. (Color version of figure is available online.)

Fig. 2 – MR abdomen was obtained due to clinical elevation in liver function tests and biliary duct dilatation. T2-weighted and T1-weighted postcontrast MRI images showing T2 hyperintense perihilar lesion with delayed contrast enhancement on T1-weighted image (shown by yellow arrows). MR, magnetic resonance; MRI, magnetic resonance imaging. (Color version of figure is available online.)

duration at an outpatient clinic. She had elevated liver function tests and CT scan of abdomen that demonstrated intraductal dilation at an outside institution. The patient was then referred to our medical center for endoscopic ultrasound (EUS), and her initial workup included magnetic resonance imaging (MRI) of abdomen and magnetic resonance cholangiopancreatography (MRCP) followed by EUS and endoscopic retrograde cholangiopancreatography (ERCP).

The magnetic resonance cholangiopancreatography demonstrated a Bismuth-Corlette Type IV cholangiocarcinoma (Figs. 1-2). A roughly 3.5 cm ill-defined tumor was centered on the proximal hepatic duct extending into the hepatic hilum. There is evidence of periductal tumor extension along the right and left hepatic ducts to involve the first and second order ducts. The anterior division of the right portal vein was diminutive due to tumor involvement. The EUS displayed a 1.5-2 cm isoechoic poorly demarcated mass with significant intrahepatic biliary dilatation upstream. The mass was subsequently biopsied using a 25G needle with preliminary cytology revealing adenocarcinoma. A random liver biopsy was also performed and was unremarkable with no significant fibrosis or liver fatty infiltration.

The EUS was followed by an ERCP with placement of 2 intrahepatic stents for hyperbilirubinemia. Prior to performing ERCP, her AST and/or ALT ratio blood urea nitrogen and/or creatinine ratio, and hemoglobin were 0.74, 24, and 12.7 g/dL respectively. The ERCP demonstrated a high-grade, malignant-appearing, and hilar stricture extending to the origins of right and left hepatic ducts with upstream dilatation. Two 10 Fr plastic stents were deployed bilaterally across the hilum (Fig. 3). Her diagnostic laparoscopy and cytology from peritoneal washings was negative.

The patient was diagnosed with Stage IIIB intrahepatic cholangiocarcinoma given the invasion of one of the portal vein branches and no enlarged nodes seen on MRI. The goal

Fig. 3 – ERCP guided bilateral biliary stent placement (as depicted by blue arrows). ERCP, endoscopic retrograde cholangiopancreatography. (Color version of figure is available online.)

of her treatment was curative intent after PVE prior to resection followed by neoadjuvant chemotherapy with gemcitabine and cisplatin.

Patient was planned for PVE via ipsilateral approach. A 22-gauge Chiba needle was used for percutaneous puncture of right hepatic lobe. After subsequent catheterization of a peripheral right portal vein using a 5 Fr reverse curve catheter
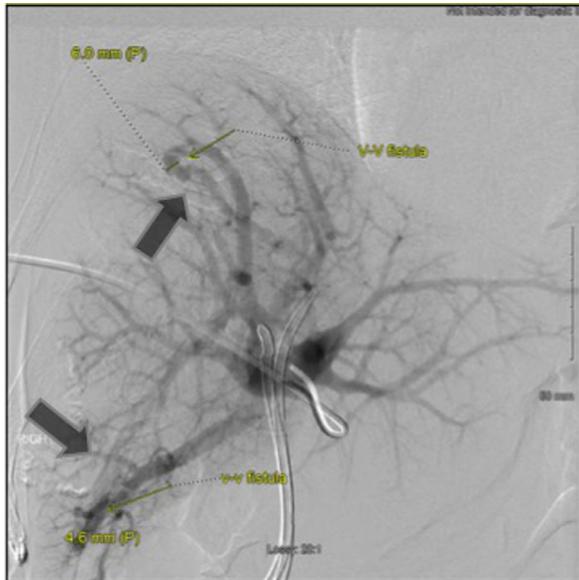
Fig. 4 – Subtracted angiographic images of the portal vein demonstrating the 2 intrahepatic porto-venous shunts at the time of PVE. Gray arrows pointing toward the 2 porto-venous shunts. PVE, Portal venous embolization.

Fig. 5 – Subtracted angiographic images of the portal vein demonstrating the 2 intrahepatic porto-venous shunts at the time of PVE. Gray arrows pointing toward the 2 porto-venous shunts. PVE, Portal venous embolization.

Fig. 6 – Maximum intensity projection (MIP) CT images depicting the 2 right lobe intrahepatic porto-venous shunt. Black arrows show the conjoining of the venous and portal system. CT, computed tomography.

Fig. 7 – Maximum intensity projection (MIP) CT images depicting the 2 right lobe intrahepatic porto-venous shunt. Black arrows show the conjoining of the venous and portal system. CT, computed tomography.

via a 5 Fr vascular sheath, a 2.8 Fr microcatheter system was used to select anterior and posterior divisions of right portal vein supplying segments V-VIII. Intraprocedurally, 2 previously unknown portal-venous—hepatic vein fistulas were discovered (Figs. 4-7). The largest of which was in hepatic segment VIII, measured 6 mm in diameter at level of fistulization and demonstrated slightly delayed flow communication

with right hepatic vein drainage into inferior vena cava. This portal-venous fistula was embolized with high viscosity 1:1 ratio of N-butyl cyanoacrylate to ethiodol to avoid nontarget embolization via the shunt, followed by 5:1 ratio of N-butyl cyanoacrylate to ethiodol embolization of segments V and/or VIII portal vein branches. The second portal-venous fistula was seen in Segment VI, measuring 5 mm in diameter at level of fistulization and demonstrated rapid flow communication into the right hepatic vein. Distal embolization of the fistula was performed using 8 mm detachable microcoils prior to embolization of Segment VI and/or VII portal vein branches using N-butyl cyanoacrylate to ethiodol 5:1 ratio. Final portal vein angiography was performed to ascertain optimal right portal vein embolization with preservation of left portal vasculature (Fig. 8). The total elapsed time for the procedure was

Fig. 8 – Postembolization parenchymagram showing complete embolization of the right portal vein and a perfused left hepatic lobe. Black arrows indicate areas of embolization.

68 minutes. The patient was discharged the following day after pain control adjustments with oral oxycodone.

The patient was subsequently started on neoadjuvant chemotherapy regimen including gemcitabine and cisplatin. She underwent right trisegmentectomy with roux-en-Y hepaticojejunostomy for her hilar cholangiocarcinoma after MRI demonstrated adequate hypertrophy of the left hemiliver. During follow-up imaging, she was found to have a metachronous lung nodule biopsy proven to be a squamous cell carcinoma that was treated with stereotactic body radiation therapy and neoadjuvant chemotherapy.

Discussion

Increasing number of patients with congenital portosystemic venous shunts are being identified given the improvement in cross-sectional imaging techniques [7]. There are major types of congenital hepatic venous shunts, extrahepatic, and intrahepatic. A direct conduit between systemic vein and portal-mesenteric vessel upstream of the portal vein usually leads to the formation of an extrahepatic shunt [8]. Extrahepatic shunts include Type I shunts (end to side shunts) and Type II shunts (side to side shunts). Type I shunts are formed when portal blood channels into systemic circulation eliminating the need for intrahepatic portalvenous supply. They are more severe, and if patients are symptomatic, liver transplant is the definitive treatment as the shunt is the only drainage pathway for splenic and mesenteric blood [8,9]. Type II shunts are formed when branches of portal vein drain directly into the systemic vasculature. They vary in clinical significance ranging from asymptomatic to hepatic

encephalopathy and can be treated surgically or be embolized [8].

On the other hand, a significant amount of variability exists in the presentation of intrahepatic portosystemic shunts. The patient presentation is variable ranging from asymptomatic to severe hepatic pathology such as hepatic encephalopathy, hypergalactosemia without enzyme deficiencies, and portal aneurysms with some patients identified during their childhood [10]. In patients with shunt ratios >30%, hepatic encephalopathy could develop at any point in time [10]. When the ratio exceeds 60%, patients are prophylactically treated to avoid hepatic encephalopathy [11]. The etiology can be congenital or acquired; age at presentation can be from birth to elderly; furthermore, they may be diagnosed incidentally as they can be asymptomatic. Treatment options include surgical ligation, resection, and endovascular embolization [12]. Complications of intrahepatic portal shunts can range from none to portal hypertension, congestive heart failure, hepatic encephalopathy, cirrhosis, membranoproliferative glomerulonephritis, and cholangitis [13–15].

Intrahepatic portal-venous shunts are divided into 4 morphologic types: Type I—single channel of equal diameter that connects the right portal vein to inferior vena cava; Type II—single or multiple communications between the peripheral branches of portal vein and hepatic vein in one segment of liver; Type III—connection of hepatic and portal veins through an aneurysm, and finally Type IV—single or multiple communications between the peripheral branches of portal and hepatic vein involving both hepatic lobes [16] (Fig. 9). The communication between umbilical vein and vitelline duct through diffusion aided by pressure gradients and increased flow is one example of various theories postulated regarding the etiology of the congenital origin of intrahepatic shunts [17].

The incidence of intrahepatic shunts in one study was found to be 6/25,579 asymptomatic patients with no evidence of liver damage, hyperplasia, or cirrhosis detected by ultrasound [18]. Underlying liver disease directly correlates with an increase in the incidence of intrahepatic shunts. Another study reported the identification of 134 patients with intrahepatic vascular shunts (venous to venous and venous to arterial) in a total of 3143 patients (most of whom had cirrhosis, hepatocellular carcinoma, or traumatic liver injury) [19]. While test methods and disease states clearly play a role in its identification, the difference in incidences between the 2 studies reiterates the variable nature of intrahepatic vascular shunts.

Extrahepatic shunts and large intrahepatic shunts tend to persist through adulthood, whereas intrahepatic shunts tend to resolve when patients are <1 year old [20]. The risk of hepatic encephalopathy increases with age in the presence of these shunts [20,21]. In both cases of intrahepatic and extrahepatic shunts, asymptomatic patients were incidentally diagnosed via radiological studies—Doppler ultrasonography and CT abdominal angiography. Although a consensus is present in treating symptomatic patients, whom to treat is a complicated decision [21]. Our patient had no evidence of underlying liver disease or history of hepatitis. The intrahepatic shunts were asymptomatic and were only identified
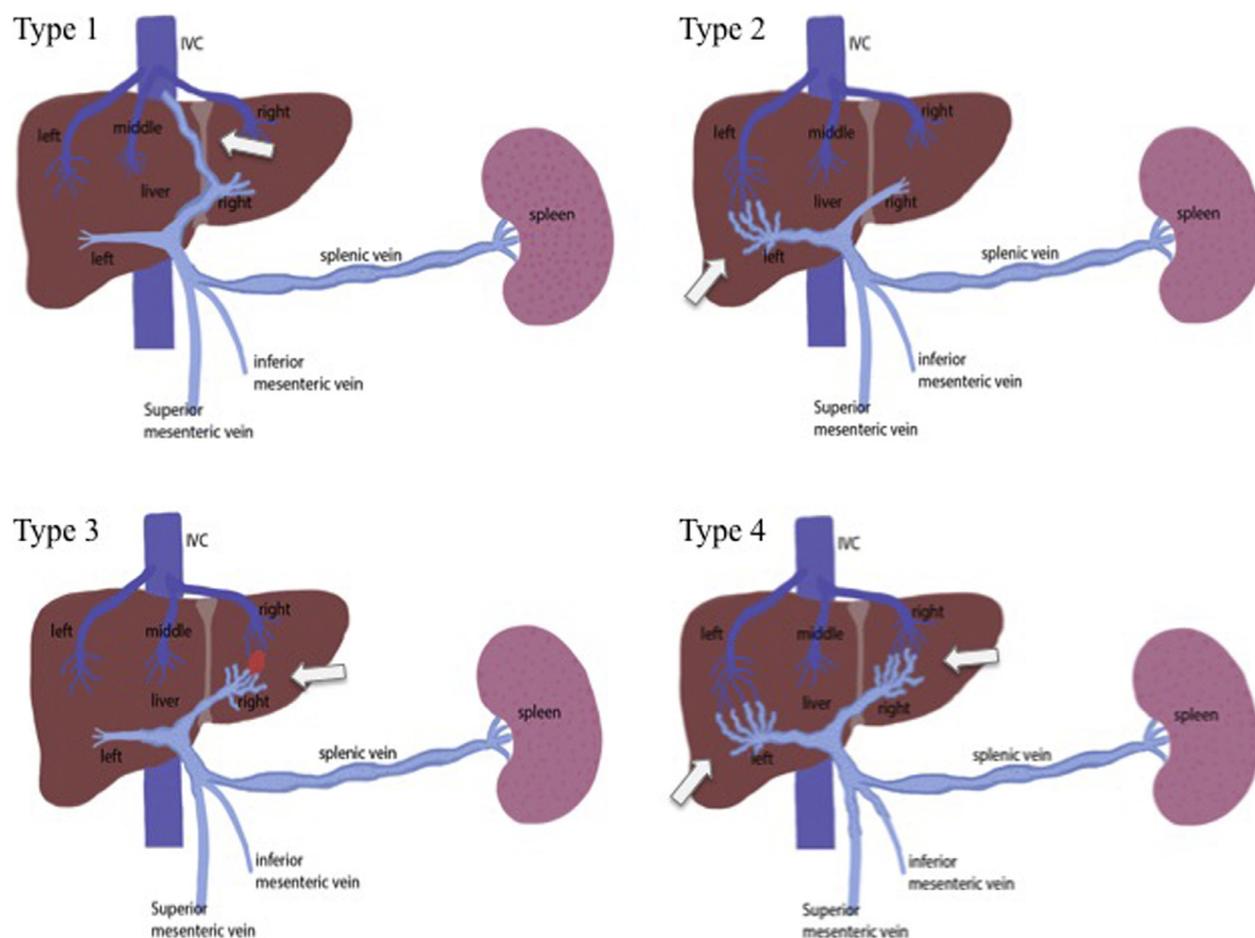
Fig. 9 – Schematic representation of 4 different variations of intrahepatic shunts. White arrows indicate the abnormality in the 4 types of intrahepatic shunts.

while performing the PVE, so, we theorized that the shunts were likely congenital in nature.

PVE aims to redirect the blood flow from the diseased part(s) of the liver to the FLR. Embolization will stimulate an adaptive mechanism to adjust for the changes in portal pressure with subsequent release of hormonal messengers and chemical signaling cascades leading to enlargement of the FLR to compensate for loss of hepatic function from the embolized liver. The FLR undergoes hyperplasia under the influence of a variety of intrahepatic (tumor necrosis factor-α, transforming growth factor-α, and extrahepatic growth factors [1]. The portal vein plays a crucial role as indicated by decreased proliferation of hepatocytes between periportal zone and hepatic vein. The pressure gradient in the portal system leads to the release of nitrous oxide and several cytokines and growth factors. Nitrous oxide induces local vasodilation and expression of several growth factors such as hepatocyte growth factor. Hepatocyte growth factor leads to hepatic hyperplasia directly by upregulating hepatocyte replication and indirectly through the release of other growth factors such as Interleukin-6 and tumor necrosis factor-α [1]. Proliferation of hepatocytes leads to enlargement of the FLR and eventual atrophy of the embolized liver. The increase in functional vol-

ume of the FLR renders patients with unresectable disease into surgical candidates with a lowered risk of liver insufficiency postoperatively.

Multiple factors such as age, baseline liver function, the extent of required liver resection, and the stage of disease are considered before selecting a patient for PVE. Patients with advanced cirrhosis are not usually good candidates for PVE. General guidelines recommend the FLR to be at least 20% of the normal liver, 30% of the diseased volume, and 40% in patients with underlying cirrhosis. In other words, if the liver occupies a volume of 10 cm^3 with 60% of it undergone cirrhosis, the FLR needs to be the bigger number between 2 cm^3 (20% of the normal liver) and 2.4 cm^3 (40% of the diseased 6 cm^3) [22]. Since the 40% option usually provides the bigger number, lack of "un-diseased" liver is one possible reason that excludes patients with advanced cirrhosis as possible PVE candidates. Note that the numbers used here were for discussion purposes only.

PVE generally utilizes 2 main approaches—contralateral and ipsilateral. The contralateral approach punctures the transhepatic portal vein within the FLR using ultrasound guidance and hence confers advantages through ease of catheter manipulation in the diseased liver along with a relatively low

risk of embolic material dislodgement in last stages of PVE. However, this method punctures the FLR and renders cannulation of Segment IV branches difficult during embolization [22]. The ipsilateral approach is the exact opposite of contralateral approach in that the portal vein is punctured within the diseased liver. Thereby, the FLR is unscathed during this process. However, this process is rendered difficult due to the challenges in catheter manipulation and higher risk of nontarget embolization of the FLR.

Several embolic materials may be used alone or in combination during portal vein embolization to block the blood flow to the diseased hepatic lobe and stimulate hypertrophy of the contralateral lobe. A thorough knowledge of hepatic vasculature is a prerequisite for a durable and complication free PVE. Several materials such as N-butyl cyanoacrylate, dehydrated ethanol, fibrin glue, spherical microparticles, or vascular plugs have been used. However, each of these materials also comes with their own disadvantages. For example, microparticles, coils, and ethanol while offer durable portal vein occlusion; they often require large quantities. In addition, ethanol has been shown to cause periportal necrosis and fibrosis.

N-butyl cyanoacrylate is a newer liquid embolic with an initial steep learning curve and has been associated with peribiliary fibrosis, periportal inflammation and may not be used in patients with reduced hepatopetal flow [23]. The material used for occlusion is often on a case-by-case basis or operator dependent, and currently, there is no consensus for one material over the other in the available literature [24,25]. Embolic materials should be used with extra caution in the vicinity of intrahepatic shunts. Immediate recognition of these intrahepatic portal-venous shunts is paramount to avoid catastrophic intraprocedural complications. Initial targeted embolization of the intrahepatic shunt should be performed before proceeding with PVE.

To our knowledge, there are no reports on the finding of intrahepatic portal-venous shunts while performing a PVE, which is rather surprising given that these intrahepatic shunts are not uncommon. The identification of these shunts preprocedurally on cross-sectional imaging would be extremely valuable for adequate preprocedure planning. In our case, the 2 intrahepatic portal-venous shunts in a noncirrhotic patient were not identified on prior cross-sectional imaging and prompt identification of them intraprocedurally allowed for corrective action to be taken to safely continue with a standard PVE.

R E F E R E N C E S

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