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Case Report

Arterial-portal fistula treated with hepatic arterial embolization and portal venous aneurysm stent-graft exclusion complicated by type 2 endoleak

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ABSTRACT

Intrahepatic arterioportal fistulas may be complicated by portal hypertension. An associated portal venous aneurysm (PVA) may impinge upon adjacent structures or rupture. We present a 65-year-old man with an intrahepatic arterioportal fistula and 6.4 × 5.8 cm right portal vein aneurysm extending within 0.4 cm of the hepatic margin, associated with pain concerning for impending rupture. The PVA was refractory to transarterial embolization due to recruitment of arterial collaterals. Therefore, it was additionally excluded from the portal vein with a 12 mm × 9.5 cm venous stent graft. Although endovascular therapy thrombosed the aneurysm and improved symptoms, it was complicated by a type 2 endoleak into the PVA.

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Introduction

Intrahepatic arterioportal fistulas (APFs) may be congenital or acquired from cirrhosis, trauma, surgery, or percutaneous hepatic intervention. APFs are often inconsequential but

can be complicated by symptoms of portal hypertension. Progressive enlargement and development of a portal venous aneurysm (PVA) may lead to compression of biliary ducts and other adjacent structures, or rupture [1]. Transarterial embolization (TAE) has been described in treatment of APF but may be ineffective due to recruitment of collateral arterial

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perfusion [2]. We describe a case of an APF complicated by a large PVA refractory to TAE alone and ultimately treated with combined TAE and portal venous stent-graft exclusion. Endovascular therapy thrombosed the aneurysm, ameliorated symptoms, and reduced aneurysm size, but was complicated by type 2 endoleak. This case highlights the limitations of arterial coil embolization alone for the treatment of complex vascular lesions.

Case report

Our institutional review board does not require approval for single retrospective case reports. A 65-year-old man with a history of hepatitis C virus cirrhosis and orthotopic liver transplantation (OLT) 16 years prior developed right upper quadrant abdominal pain. He underwent contrast-enhanced magnetic resonance imaging at an outside hospital to further evaluate a PVA initially detected on ultrasound (US). MRI demonstrated arterial-phase enhancement of the right portal vein and peripheral parenchymal enhancement suggestive of APF, with an associated 6.4×5.8 cm PVA arising from the origin of the right portal vein and involving the anterior and posterior branches (Fig. 1A). The aneurysm extended to within 0.4 cm of the hepatic margin. Review of previous imaging demonstrated a small, peripheral APF on a CT scan performed in 2003 with enlargement to 3.7×3.5 cm on a CT scan performed in 2007; the fistula was at the site of prior percutaneous liver biopsy.

The enlargement of the aneurysm, proximity to the liver capsule, and associated pain raised concern for impending rupture. Multidisciplinary review including Hepatology, Surgery, and Interventional Radiology concluded that right hepatectomy was undesirable due to scarring from previous OLT and small future liver remnant volume; therefore, the decision was made to perform arteriography with intention of TAE to close the APF, with concurrent listing for second OLT due to concern for precipitating hepatic decompensation and biliary ischemia. At arteriography, a hypertrophied right hepatic artery (RHA) was identified with contributors arising from all segmental arterial branches of the RHA to the PVA. Embolization of posterior subsegmental branches with 0.018-inch Nester coils (Cook Medical, Bloomington, IN) resulted in decreased but persistent perfusion from additional branches of the RHA (Fig. 1B).

A subsequent embolization procedure targeted additional RHA subsegmental branches in the same fashion; however, new perfusion to the PVA developed from branches of the LHA (Fig. 1C). Given the failure of durable occlusion with coil embolization, additional embolization was performed of subsegmental arterial branches of the segments II, III, and IV arteries with N-butyl cyanoacrylate (NBCA, Trufill, Cordis Neurovascular Inc., Miami Lakes, FL) in a ratio of 1:3 with ethiodized oil (Lipiodol; Guerbet LLC, Princeton, NJ) and resulted in complete occlusion of the arterial supply to the PVA on angiography.

With occlusion of the arterial supply to the PVA achieved, the decision was made to pursue exclusion of the PVA to achieve thrombosis and reduction in size. A multidisciplinary review was held and the decision was made to pursue stent-graft exclusion of the PVA. Diagnostic mapping of the PVA and

portal vein was initially performed. The left portal vein was accessed under US guidance with a 21-gauge needle and an Accustick (Boston Scientific, Marlborough, MA) was utilized to accommodate a 0.035-inch guidewire. A 6-French sheath was then placed, and a 5-French pigtail marking catheter advanced into the main portal vein. Portal venous pressure measured 15 mm Hg. On portal venography the PVA involved the entire RPV, with 2.2 cm diameter at its origin and enlarging over a short segment to 6.8 cm (Fig. 1D). The left portal vein landing zone measured 1.5 cm in diameter and the main portal vein landing zone measured 1.8 cm. Given the large caliber of the left portal vein, placement of a standard stent graft was not feasible, and as such an iliac limb endoprosthesis was selected for use.

The 6-French sheath was exchanged for a 12-French GORE DrySeal Flex Introducer Sheath (GORE Medical, Newark, DE). A 20 mm \times 9.5 cm GORE Excluder Contralateral Leg Endoprosthesis (PLC 201000, GORE Medical, Newark, DE) was reverse-loaded [3] into the sheath to deploy the 20 mm aspect in the main portal vein and the 16 mm end in the left portal vein. The stent graft was apposed to the main and left portal veins with a compliant Q50 Plus Stent Graft Balloon Catheter (GORE Medical, Newark, DE). Following stent-graft placement, portal venography demonstrated no enhancement of the excluded aneurysm (Fig. 1E), and immediate thrombosis was observed of the PVA on US. Portal venous pressure measured 17 mm Hg. Microfibrillar collagen paste (Avitene; CR Bard, Murray Hill, NJ) was prepared by mixing the dry flour with 3 mL of iodinated contrast material (Omnipaque-300, GE Healthcare, Chicago, IL) and 3 mL of normal saline. The sheath was withdrawn into the hepatic parenchymal tract and an 8-French sheath was advanced into the 12-French sheath; the track was embolized with microfibrillar collagen paste through the 8-French sheath under fluoroscopic guidance [4]. US 1 week later demonstrated echogenic thrombus and no Doppler signal within the PVA; the stent graft and LPV remained patent. Abdominal pain had resolved, and liver function tests remained normal.

A CT performed 3 months later showed that the PVA had decreased in size to 4.5×4.4 cm. However, peripheral enhancement within the PVA in the arterial phase suggested new arterial-portal shunts. Five months after stent-graft exclusion, the patient developed atrial fibrillation associated with a new pericardial effusion attributed to a viral infection, and anticoagulation was initiated with warfarin. On CT follow-up at 6 months, the PVA remained 4.5×4.4 cm in diameter but with increased peripheral enhancement (Fig. 1F). Angiography confirmed a type 2 endoleak supplied by new branches of the segment 4 hepatic artery, reconstituting branches of the RHA distal to coils (Fig. 1G).

Given the extensive network of perfusing arteries refractory to prior embolization, decreased size of the aneurysm, and resolution of symptoms, further endovascular therapy was withheld. CT follow-up at 11 months demonstrated stable size of the PVA with further increase in peripheral enhancement. The patient was listed for OLT due to exhaustion of all endovascular treatment options for the PVA. A donor liver was procured and retransplantation was performed 14 months after stent-graft exclusion. During transplant, the portal vein was transected and the stent graft was extracted prior to creation of an end-to-end donor-recipient portal vein

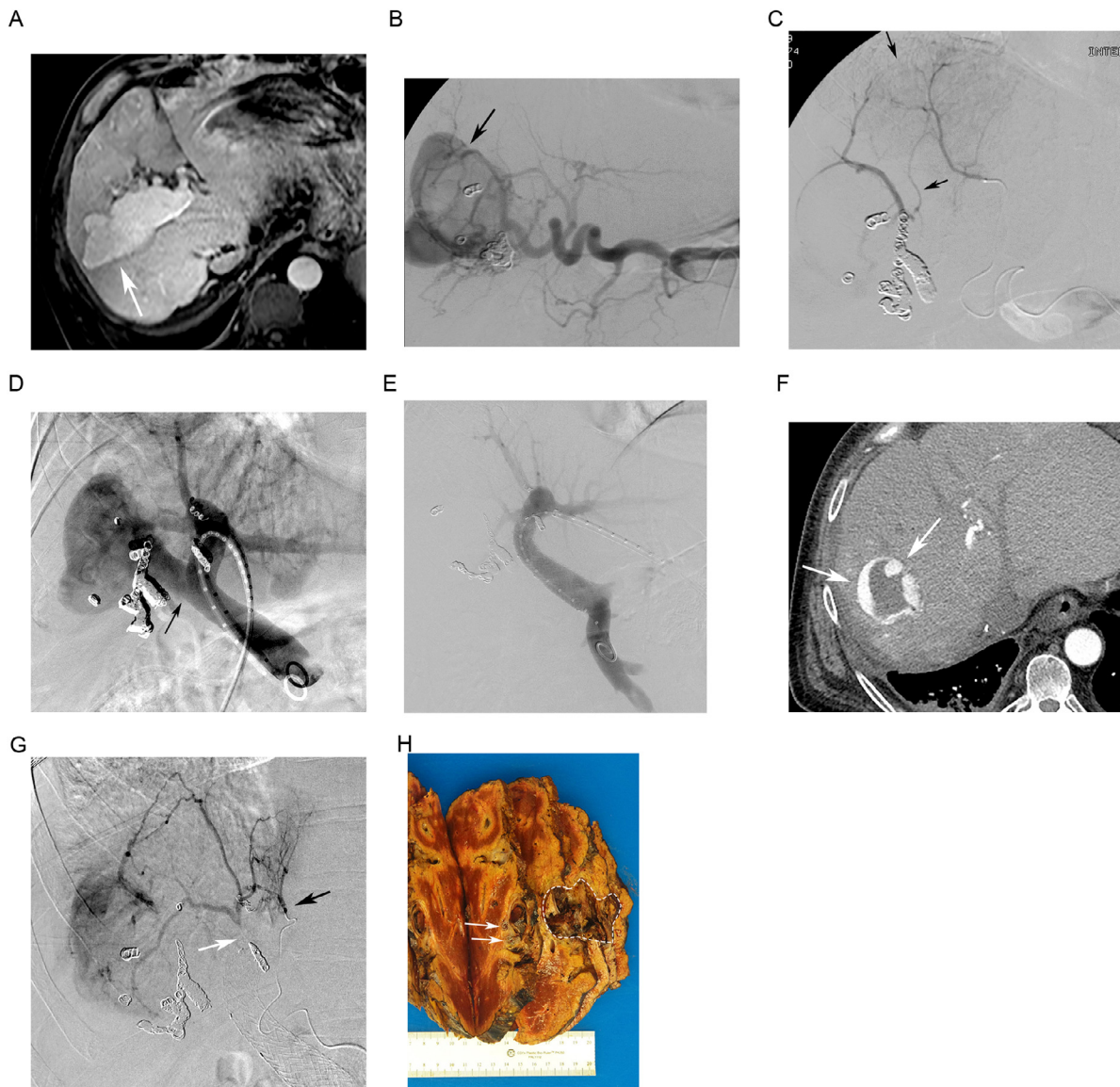


Fig. 1 – Images of a 65-year-old man status post-OLT 16 years prior with abdominal pain. (A) Axial contrast-enhanced MR image demonstrates arterial-phase enhancement of the right portal vein with an associated 6.4 × 5.8 cm PVA (arrow). (B) Celiac angiography following coil embolization of multiple subsegmental RHA contributors shows persistent perfusion of the PVA (arrow) via additional branches of the RHA. (C) On angiography 1 month later, collateral reconstitution (arrows) of RHA branches have developed from the LHA. (D) Portal venography 4 months after initial angiography opacifies the right PVA with origin (arrow) measuring 2.2 cm in diameter. (E) Following stent-graft placement portal venography confirms exclusion of the PVA with preserved flow within the superior mesenteric vein and left portal vein. (F) Axial contrast-enhanced CT image 6 months after stent-graft exclusion demonstrates arterial-phase peripheral enhancement of the PVA (arrows). (G) Subsequent angiography confirmed a type 2 endoleak supplied by branches of the segment 4 hepatic artery (black arrow) reconstituting branches of the RHA distal to coils (white arrow). (H) Representative gross pathologic section of the PVA (dashed outline) with multiple adjacent embolized feeding vessels (arrows) containing coils.

anastomosis. Gross pathology of the explanted liver demonstrated cirrhotic morphology and a PVA containing organizing thrombus with multiple vessels adjacent to the PVA, some containing coils from prior embolization (Fig. 1H). The patient is currently alive 6 months post-transplant.

Discussion

This case demonstrates the difficulty of endovascular treatment of a complex APF with associated PVA, and the

limitations of treatment by TAE alone due to recruitment of perfusion by arterial collaterals.

A proposed classification of arterial portal fistulae by Guzman et al categorized APFs into 3 types based on etiology [5]. The current case describes an example of a Type II fistula, defined as an acquired large central fistula, resulting in an expanding PVA. APFs are not uncommon in liver transplant recipients; in post-transplant patients undergoing hepatic angiography, the incidence of APFs has been reported as high as 5.4% [6]. However, in a retrospective audit of 1992 cadaveric and living donor liver transplant recipients performed over a 13-year period, only 0.2% were found to have hemodynamically significant APFs resulting in liver function test abnormalities, suggesting that the majority of APFs do not cause adverse sequelae [7].

If small and peripherally located, an APF is likely to resolve spontaneously within 1-3 months [5,8]. When large and more centrally located, as in the current case, APFs are unlikely to resolve and are likely to become symptomatic [5,9]. While management in prior decades has been surgical, involving ligation of the involved hepatic artery, resection, or retransplantation [10], hepatic artery embolization has become the accepted initial treatment of choice for APFs with a reported clinical success rate of 83% (15 of 18 patients) in prior case series [2,9,11–13]. Previously described arterial embolization interventions for APFs include Amplatzer plugs, coils, detachable balloons, NBCA, and sclerotherapy, with agent selection tailored to the anatomy and complexity of the shunt [9,11].

Hepatic arterial embolization may fail to durably occlude an APF, particularly if supplied by multiple arterial branches. Wada et al described a small PVA supplied by multiple feeding arteries, with persistent APF despite 2 TAE procedures with NBCA and ethiodized oil [2]. The fistula was ultimately treated successfully with retrograde transvenous obliteration with ethanol and ethiodized oil, with a balloon-mounted catheter inflated within the segment VIII portal vein outflow of the APF; concurrently a balloon-mounted catheter was inflated within the RHA. In the current case, coil embolization prompted recruitment of collateral circulation to the APF/PVA complex. Failure was attributed in part to the inability to position coil packs sufficiently adjacent to the fistulous connection, as evidenced by reperfusion of the fistula distal to coils on subsequent angiography. Complete closure was achieved after embolization with NBCA and ethiodized oil. These findings suggest that preferential consideration should be given to liquid embolics over metallic coils in APF treatment, particularly if coils cannot be placed directly adjacent to the fistulous connection.

Embolization of the PVA, or balloon-occluded retrograde transvenous obliteration of the PVA and APF as described by Wada et al, was anatomically infeasible due to the size and configuration of the PVA. Stent-graft exclusion was therefore performed after embolizing the hepatic arterial inflow. This procedure achieved clinical goals of reduction of the PVA size and resolution of symptoms, and initially achieved thrombosis of the APF and PVA. However, complete APF occlusion was transient with development of a type 2 endoleak 3 months after stent-graft exclusion of the PVA. Warfarin therapy has been associated with reduced rates of spontaneous thrombosis of type 2 endoleaks [14] and may be a contributing factor to

persistence of the endoleak in the presented case. Although occlusion of RHA branches and the portal vein may elevate risk for hepatic decompensation or biliary ischemia, these sequelae did not occur, likely because of collateral hepatic artery recruitment visualized on imaging follow-up after stent-graft placement.

The literature regarding the management of type 2 endoleaks is largely confined to complications following endovascular aortic repair [15,16], with treatment generally reserved for patients with growth of the excluded aneurysm sac. With no reported literature regarding an intrahepatic type 2 endoleak, the risk of late rupture relative to an aortic type 2 endoleak is unknown but may be catastrophic as the liver is surrounded predominantly by the peritoneal space. Conversely the likelihood of success of additional percutaneous intervention, such as direct sac puncture and embolization [16] and risk of resulting hepatic infarction and failure, are unknown. Given reduction of sac size from baseline and resolution of symptoms following stent graft exclusion, additional endovascular treatment was deferred in favor of active clinical and imaging surveillance and listing for a second OLT. The patient remained clinically asymptomatic until the time of OLT at 14 months following stent-graft exclusion of the PVA, although follow-up CT examinations had confirmed recruitment of new contributing vessels.

Conclusion

Although a complex hepatic APF with associated PVA is rarely encountered, the principles and techniques used for treatment in this case can be translated to similar lesions in different clinical scenarios. Importantly, the limitations of coil TAE alone are made apparent by the rapid recruitment of perfusion by arterial collaterals, and preferential consideration should be given for glue embolization in analogous lesions. Additionally, stent grafts may be successfully employed for aneurysm exclusion, even using endoprosthesis designed for other applications.

Declaration of Competing Interest

The authors have declared that no competing interests exist.

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