Hybrid Surgery for Portosystemic Encephalopathy in a Patient with Liver Cirrhosis: a case report

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ABSTRACT

Regarding the treatment for a portosystemic shunt, surgical or interventional radiological closure of the shunt was established. Interventional radiology including balloon-occluded retrograde transvenous obliteration can worsen portal hypertension and create a large thrombus close to the major venous system in the case of a huge portosystemic shunt. In contrast, it is also difficult to treat some cases through surgery alone when huge complicated shunts exist very deep in the body. Herein, we report a successful case of surgical shunt ligation for portosystemic encephalopathy in a hybrid operation room that enabled intraoperative angiography and computed tomography. A 62-year-old woman with chronic hepatitis C was referred to our hospital due to high levels of serum ammonia and hepatic encephalopathy. She had a massive, complicated portosystemic shunt from the inferior mesenteric vein to the left renal vein but did not have esophageal or gastric varices. It was difficult to occlude the portosystemic shunt by interventional radiologic techniques because the shunt had an extremely large amount of blood flow and many collateral routes. We performed the shunt ligation in the hybrid operation room. Intraoperative angiography provided detailed information about the portosystemic shunt, such as direction or volume of blood flow and collateral routes in real time. Her encephalopathy disappeared completely and she remains healthy with improved liver functional reserve to date. In conclusion, this is a successful case of a hybrid operation for an extremely large and complicated portosystemic shunt, providing for intraoperative angiography as a safe and reliable surgical treatment for portosystemic encephalopathy in patients with liver cirrhosis.

Key words: Intraoperative angiography, Hybrid operation room, Hepatic encephalopathy

Hepatic encephalopathy is the loss of brain function that occurs when the liver is unable to remove toxins from the blood. The development of hepatic encephalopathy negatively impacts patient survival. There are two types of encephalopathy related to liver cirrhosis: portosystemic encephalopathy and end-stage hepatic encephalopathy in severe liver dysfunction. The portosystemic shunt involves blood flow mainly from the supra mesenteric vein (SMV) to the systemic vein, and results in high systemic blood ammonia levels. Regarding the treatment for portosystemic encephalopathy, endovascular shunt obliteration including balloon-occluded retrograde transvenous obliteration (B-RTO) results in good outcomes. However, B-RTO can worsen portal hypertension and create a large thrombus close to the major venous system such as the inferior vena cava and portal trunk when the shunt is very large.

Intraoperative angiography in the hybrid operation room has been previously described in neurosurgery for cerebral arteriovenous malformations and aneurysms and in cardiovascular surgery. However, this procedure is rare in gastroenterological surgery. Herein, we describe a successful case of surgical shunt ligation for portosystemic encephalopathy in a hybrid operation room. Hybrid operation rooms offer high-end imaging with angiography devices for complex and delicate interventions. The described operating room is

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equipped with an angiography device (Artis zee biplane system®, SIEMENS) and a computed tomography (CT) scan machine (SOMATOM Definition AS Open - RT Pro edition®, SIEMENS). The Artis zee biplane system features excellent performance for imaging and position flexibility. SOMATOM Definition AS Open - RT Pro edition® can become a useful tool for delivering images to evaluate the result of intervention during an operation.

CASE PRESENTATION

A 62-year-old woman (height, 158 cm; weight, 89 kg) with chronic hepatitis C was referred to our hospital for the treatment of severe hepatic encephalopathy that interfered with everyday life and was resistant to medication treatment. She had a history of hysterectomy and hepatectomy (partial resection of S4) and a high body mass index (BMI) of 35.7. Laboratory tests revealed neither jaundice nor hepatic insufficiency and scored Child-Pugh B (8) as follows: total bilirubin level, 0.8 mg/dl; prothrombin time and international normalized ratio, 62% and 1.27; creatinine level, 0.91 mg/dl; platelet count, 96000/μl; albumin, 3.6 g/dl, ammonia, 96 μmol/liter, and indocyanine green retention rate at 15 min(ICG-R15), 42.5%. Enhanced CT showed a massive portosystemic shunt from the inferior mesenteric vein (IMV) via the left ovarian vein to the left renal vein, and splenomegaly (568 ml) (Fig. 1a). There was no ascites. Angiography also showed portal hypoperfusion and portal flow steal via a huge portosystemic shunt (Fig. 1b,1c,1d). Portal vein pressure (PVP) measured by the wedge hepatic venous pressure was 20 mmHg. Gastroscopy revealed no hemorrhagic disease, such as tumors, ulcers, or varices, in the upper gastrointestinal tract. Echocardiography revealed no heart or fluid decompensation due to heart or renal failure.

After an exhaustive assessment, we made a decision to select surgical treatment for hepatic encephalopathy instead of B-RTO, because B-RTO could increase the risk of severe complications for this patient. The shunt had an extremely large amount of blood flow and several drainage vessels. It came from the IMV to the left renal vein, by way of the left ovarian vein, which was resected at the distal end due to a past history of total hysterectomy.

The operation strategy is shown in Fig. 2a. First, we performed splenectomy and confirmed the condition of the portosystemic shunt by intraoperative angiography. The PVP was measured through the catheter in the SMV trunk in Fig. 2b. The portal flow was also measured at main trunk of right PV by Doppler ultrasonography in Fig. 2b. After splenectomy, the PVP decreased (29 to 15 mmHg) but the hepatofugal flow still continued in the portal vein by angiography (Fig. 3a). Three collateral veins were confirmed under direct vision. Angiography also revealed the collateral veins from the IMV to the left renal vein. Next, we tried to clamp the route of the portosystemic shunt. After clamping three collateral veins, the PVP mildly increased from 15 to 20 mmHg, and the portal flow slightly increased to 2.5 cm/sec. Angiography revealed that the hepatofugal flow still persisted (Fig. 3b). Furthermore, the additional occlusion of the IMV raised the portal flow to 15.2 cm/sec and the PVP increased up to 25 mmHg immediately after occlusion of the IMV. After a while, the PVP became stable at around 20 mmHg with restored hepatopetal flow. All clamping veins including the collateral veins and IMV were finally ligated and there was no blood flow from SMV to the systemic vein in the SMA angiography (Fig. 3c).

The operation dramatically reversed the patient’s encephalopathy and she gradually recovered from high systemic blood ammonia levels. The postoperative course was uneventful except

Fig. 1. (a) A massive portosystemic shunt from the IMV via the left ovarian vein to the left renal vein, and splenomegaly on contrast computed tomography (CT). (b) Blood flow to the left ovarian vein on splenic artery arteriogram. (c) Blood flow to the left ovarian vein in the SMA arteriogram. (d) Blood flow to the left ovarian vein in the IMA arteriogram. The arrows indicate the direction of blood flow. SV: splenic vein, SMV: superior mesenteric vein, IMV: inferior mesenteric vein, PV: portal vein, IVC: inferior vena cava, LRV: left renal vein

Children's digital health care system equipped with an angiography device (Artis zee biplane system®, SIEMENS) and a computed tomography (CT) scan machine (SOMATOM Definition AS Open - RT Pro edition®, SIEMENS). The Artis zee biplane system features excellent performance for imaging and position flexibility. SOMATOM Definition AS Open - RT Pro edition® can become a useful tool for delivering images to evaluate the result of intervention during an operation.

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Hybrid Surgery for Hepatic Encephalopathy

Drainage vessels into the systemic circulation. The hybrid operation room consists of intraoperative angiography and computed tomography devices. These enable the evaluation of blood flow in multiple porto-systemic shunts in real time, so that the patient's encephalopathy can be reduced dramatically.

Our patient required treatment for chronic encephalopathy because of impaired ability to perform daily life activities, and non-responsiveness to medication. The treatment for portosystemic shunt includes surgical treatment, interventional radiological treatment, and conservative therapy. Interventional radiology including B-RTO is the first choice of treatment for portosystemic encephalopathy. B-RTO has been shown to be a suitable continuous ascites, which was controlled by medication. She was discharged with no complications related to the shunt ligation on postoperative day 21. As of now, after a follow-up of 6 months, the patient has showed no recurrence of hepatic encephalopathy or gastrointestinal bleeding.

DISCUSSION

We describe a successful surgical treatment for the extremely large and complicated portosystemic shunt in the hybrid operation room. It was difficult to occlude the portosystemic shunt by interventional radiology technique alone because the shunt had an extremely large amount of blood flow and several drainage vessels into the systemic circulation. The hybrid operation room consists of intraoperative angiography and computed tomography devices. These enable the evaluation of blood flow in multiple porto-systemic shunts in real time, so that the patient's encephalopathy can be reduced dramatically.

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able therapeutic option for patients with portosystemic encephalopathy due to various types of extrahepatic portosystemic shunt\textsuperscript{13,18}. It has been shown to be effective in treating hepatic encephalopathy and restoring hepatopetal portal blood flow\textsuperscript{9,10}. However, this technique is not very effective when there are many collateral routes and an extremely large amount of blood flow\textsuperscript{9}. Moreover, B-RTO alone increases portal pressure further if the patient already has portal hypertension. Therefore, simultaneous splenectomy would be better in such cases. Furthermore, pulmonary embolus after B-RTO is also a theoretical concern, given the possibility of systemic sclerosing leakage, but was only seen with a pooled rate of 0.13\%. Overall, the pooled rate of major complications was 2.6\%, and a total of two procedure-related deaths out of 1,016 total cases were seen (0.2\%\textsuperscript{14}). Additionally, six cases of either inferior vena cava (IVC) or right iliac vein thrombus were seen and all noted in a single study\textsuperscript{21}. Pulmonary glue emboli may occur when the shunt drains directly into a large vein such as the left renal vein or IVC and lacks drainage into intervening small vessels\textsuperscript{17}. In our case, three collateral veins drained into the left renal vein separately, and hence, our patient was considered to be at risk for developing pulmonary embolism.

Surgical treatment for hepatic encephalopathy has been reported earlier\textsuperscript{4,7,8,16}. However, it was difficult to treat this case through surgery alone. First, the patient had an extremely high BMI. Second, the portosystemic shunt was present very deep in the surgical field, and was very complicated, extending from the IMV via some collateral veins to the left renal vein. Third, the patient had poly-surgery. A superior navigator is needed to provide accurate information regarding the portosystemic shunt during the operation. Therefore, hybrid operation was selected as the treatment in this case.

Hybrid surgery provides much information to improve the shunt ligation during the operation. First, CT angiography reveals the accurate location of the shunt, thus facilitating access during surgery. Second, radiologists perform a selective contrast study from the celiac axis, the SMA, and the IMA to confirm whether shunt ligation would work well. Third, digital subtraction angiography provides a high quality of imaging for the detection of small shunts. Our patient could have been operated upon using C-arm in a common operation room, however, the hybrid surgery makes the shunt operation safe and easy.

Some problems may arise from this kind of treatment. First, the shunt ligation leads to increase in the PVP, which may worsen esophageal gastric varices and result in the formation of new portosystemic shunts\textsuperscript{21}. Hence, close monitoring of the PVP is needed during the surgery. Futagawa et al suggested that risk factors for developing liver failure are severe cirrhosis and a 60\% or higher increase in baseline PVP after surgical occlusion of portosystemic shunts\textsuperscript{6}. Jiao et al\textsuperscript{10} reported that portal blood flow could be increased by up to 50\% leading to increased blood flow in the hepatic microcirculation without adverse events after surgical occlusion of portosystemic shunts. The subsequent increase in the PVP is associated with high mortality\textsuperscript{20}. The significance and the purpose of the splenectomy is to reduce the portal pressure after the shunt ligation\textsuperscript{20}. Intraoperative portal angiography under laparotomy is also useful for evaluation of the blood flow direction. Unfortunately, intraoperative portal angiography was not performed in this case. It is suggested that intraoperative portal angiography should be introduced in the next hybrid surgery. Second, patients with liver dysfunction classified as Child B or C also have a higher risk of surgical complications. Hence, appropriate modifications of the surgical procedures are needed for such patients.

CONCLUSIONS

This was a successful case of a hybrid operation for the extremely large and complicated portosystemic shunt. The hybrid operation room enables the evaluation of blood flow in multiple portosystemic shunts in real time, as a superior navigator would. Hence, this strategy is likely to provide a safe and reliable surgical treatment for portosystemic encephalopathy in patients with liver cirrhosis.

Consent: Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images.

Competing interests: The authors declare that they have no competing interests.

Author contributions: Y.I. and M.O. performed the research/study, analyzed the data and wrote the manuscript. S.K., H.T., K.I., K.I., and T.K. performed the research/study, analyzed the data and wrote the manuscript. Y.I. and M.O. performed the research/study, analyzed the data and wrote the manuscript. S.K., H.T., K.I., K.I., and T.K. performed the research/study, analyzed the data and wrote the manuscript. H.O. designed the study and interpreted the results.

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