Emergency thoracic endovascular aortic repair with celiac artery coverage in hereditary hemorrhagic telangiectasia

Yuji Kawano, MD,[a] Yuji Kanaoka, MD,[b] Nobuhiko Hiraiwa, MD,[a] Daisuke Nakatsuka, MD,[a] *and* Minoru Tabata, MD, MPH,[a] *Chiba and Tokyo, Japan*

Celiac artery (CA) coverage during thoracic endovascular aortic repair has been demonstrated to be a feasible and effective strategy for selected cases. However, there is a potential risk of ischemic complications due to CA coverage in patients with certain types of hereditary hemorrhagic telangiectasia (HHT). Herein, we report a case of thoracoabdominal aortic rupture in a patient with HHT that was successfully treated with emergency thoracic endovascular aortic repair covering the CA preceded by hepatic artery bypass. We also review the hepatic circulatory derangements and unique considerations in the surgical management of HHT. (J Vasc Surg Cases and Innovative Techniques 2017;3:57-9.)

CASE REPORT

A 64-year-old man was transferred to us from another hospital with a diagnosis of ruptured thoracoabdominal aortic aneurysm. On admission, his vital signs were stable. He had abdominal distention and mild epigastric tenderness. Although his body temperature was 36.8°C, his white blood cell count and serum C-reactive protein level were elevated to 20,200/μL and 24.45 mg/dL, respectively. His hemoglobin level was 8.1 g/dL.

The patient had a history of hereditary hemorrhagic telangiectasia (HHT) and recurrent epistaxis for 30 years, and he also had a long history of hypertension. A week before emergency transfer, he was brought to the referring hospital with mild abdominal distention and constipation. Computed tomography (CT) demonstrated normal-size thoracoabdominal aorta with a diameter of 25 mm and no abnormal findings except for slight thickening of the retroperitoneum. The patient was admitted for further examination of suspected retroperitoneal fibrosis. His symptoms had been unchanged during hospitalization; however, follow-up contrast-enhanced CT that was performed on the day of transfer showed a saccular aneurysm of the thoracoabdominal aorta and retroperitoneal hematoma. He was then immediately transferred to our hospital for emergency repair of ruptured thoracoabdominal aortic aneurysm. Considering the rapid progression of the aneurysm and elevated white blood cell count, we made a diagnosis of ruptured infected aortic aneurysm. Blood culture specimens were obtained, followed by empirical antibiotic therapy. The last CT scan showed that the lower end of the aneurysm was located 10 mm away from the celiac artery (CA) and 24 mm away from the superior mesenteric artery (SMA; Fig 1, *A*). It also detected simultaneous opacification of arterial and portal vessels, indicating arterioportal shunts (Fig 1, *B*), which is one of the characteristic findings in HHT. We decided to perform emergency thoracic endovascular aortic repair (TEVAR) followed by intensive antibiotic therapy instead of open surgical repair of the thoracoabdominal aorta because it seemed to carry a high risk of hemorrhagic complications in this HHT patient. To minimize the risk of liver ischemia after CA coverage, we performed the left external iliac to proper hepatic artery bypass with a 6-mm Teflon graft (Gelsoft Plus; Vascutek Ltd, Glasgow, UK) before TEVAR. The graft was first anastomosed to the left external iliac artery, pulled into the peritoneal space through the sigmoid mesocolon, routed anteriorly to the pancreas through the transverse mesocolon, and then anastomosed to the proper hepatic artery. The patient underwent TEVAR using a 34 × 100-mm TAG thoracic stent graft (W. L. Gore & Associates, Flagstaff, Ariz) with CA coverage to obtain adequate distal sealing. The distal edge of the endograft was fixed just proximal to the SMA. Intraoperative arteriography showed no sign of endoleak and excellent blood flow to the hepatic artery through the bypass. The retroperitoneal hematoma did not expand or rupture during surgery. However, because the bowel was heavily distended, we left the abdomen open to avoid the abdominal compartment syndrome. The abdomen was successfully closed on postoperative day 6. Preoperative blood cultures were positive for methicillin-sensitive *Staphylococcus aureus*. After 4 weeks of intravenous antibiotic therapy, life-time oral suppressive antibiotic therapy was continued. The patient recovered without any visceral ischemic complications, and he was discharged home on postoperative day 44. Postoperative three-dimensional CT demonstrated the stent graft situated just proximal to the SMA with no endoleak and patent bypass graft (Fig 2). The follow-up CT scan taken 6 months after the operation revealed no aneurysm and no fluid collection or endoleak (Fig 3). There has been no sign of recurrent infection for a year since surgery. The patient gave his full consent for the information described in this report and its publication.

From the Department of Cardiovascular Surgery, Tokyo Bay Urayasu Ichikawa Medical Center, Chiba[a]; and the Department of Surgery, The Jikei University School of Medicine, Tokyo.[b]

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Correspondence: Minoru Tabata, MD, MPH, 3-4-32, Todaijima, Urayasu-shi, Chiba 279-0001, Japan (e-mail: mtabata@post.harvard.edu).

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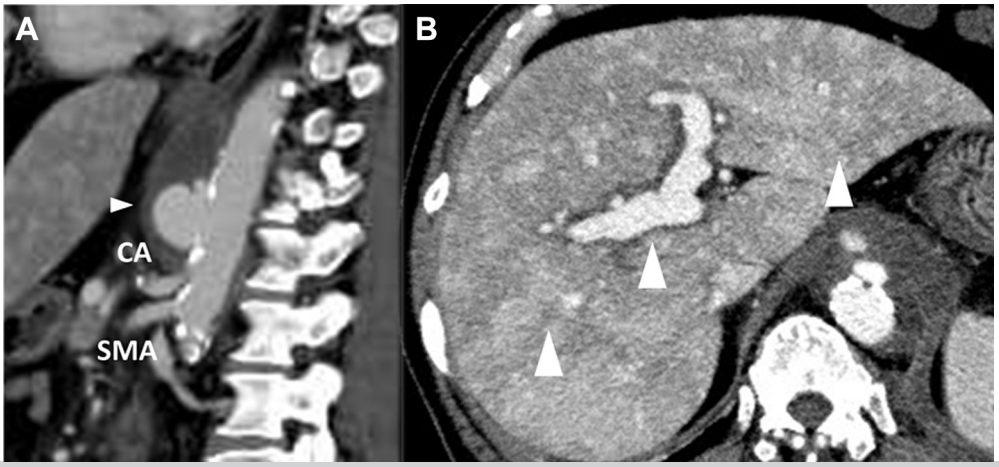

Fig 1. Preoperative contrast-enhanced computed tomography (CT) scan. **A,** Aortic rupture site is located adjacent to the celiac artery (*CA*) ostium (*arrowhead*). **B,** Simultaneous opacification of portal vein at arterial phase indicates arterioportal shunts (*arrowheads*). *SMA,* Superior mesenteric artery.

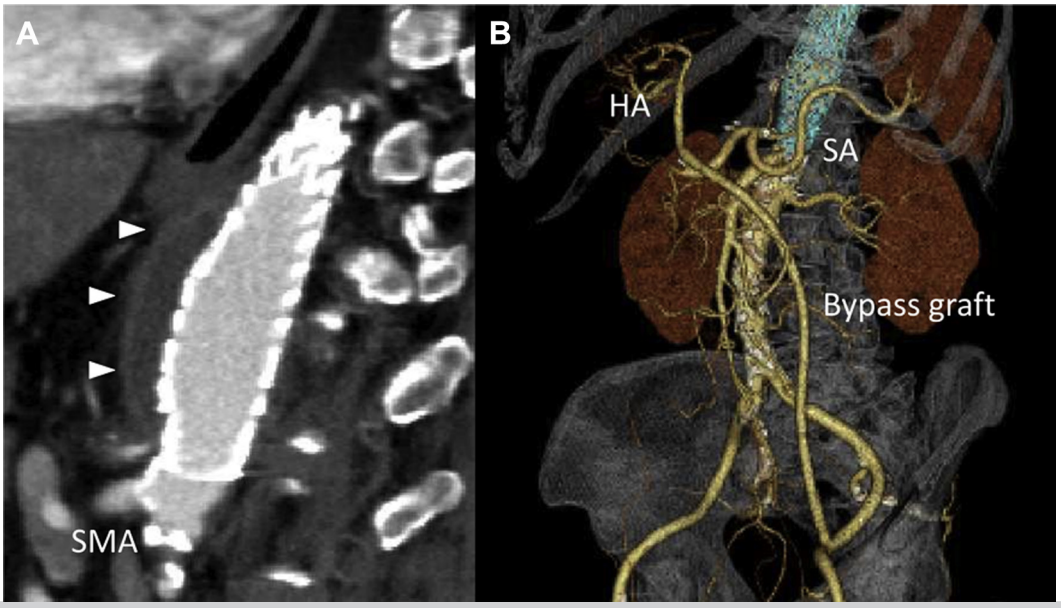

Fig 2. Postoperative contrast-enhanced computed tomography (CT) scan. **A,** There is no sign of endoleak at ruptured site (*arrowheads*). **B,** Three-dimensional image demonstrates patent graft and celiac artery (CA) branches. *HA,* Hepatic artery; *SA,* splenic artery; *SMA,* superior mesenteric artery.

DISCUSSION

CA coverage during TEVAR has been established as a valid procedure when it is necessary to facilitate adequate distal stent graft fixation. However, the anatomic suitability for this procedure should be carefully evaluated before surgery. It carries a potential risk of ischemic complications because occlusion of the CA leads to decreased blood flow through its branches. Two recent studies have shown that operative mortality rates of TEVAR with CA coverage were 6% in both series and that the incidence rates of visceral ischemic complications were 6% and 11% despite the presence of collaterals between the CA and the SMA.[1,2] In emergency situations, the risk of ischemic complications may be even higher because it is often difficult to make a full assessment of functional or anatomic distribution of blood flow related to the CA branches before surgery.

HHT is an autosomal dominant vascular disease characterized by various systemic angiodysplastic structures. When the hepatic vessel system is involved, there are three types of intrahepatic shunts: arterioportal, arteriovenous, and portovenous (PV) shunts.[3] In our case, preoperative contrast-enhanced CT showed arterioportal shunts. Memeo et al analyzed CT images of hepatic lesions in patients with HHT. According to their report,

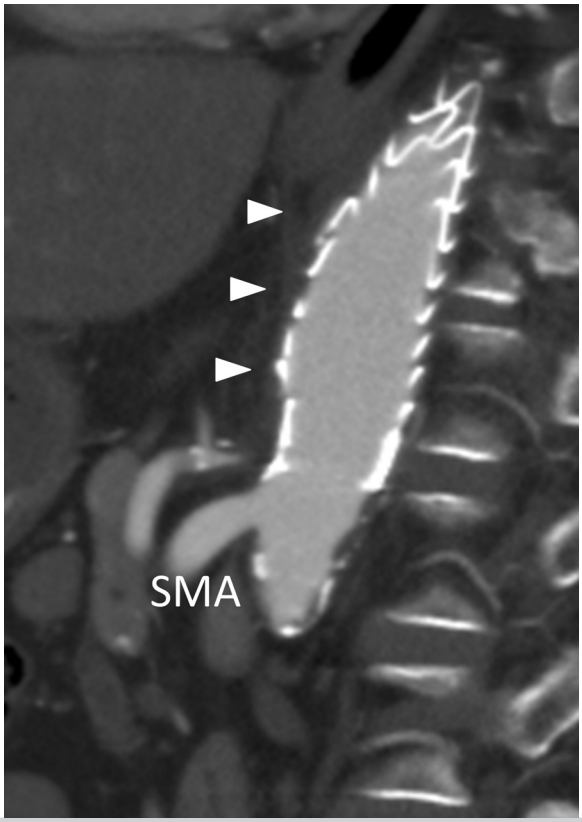

Fig 3. Contrast-enhanced computed tomography (CT) scan (6 months after the operation). The aneurysm has completely disappeared. No endoleak was found (*arrowheads*). *SMA*, Superior mesenteric artery.

arterioportal and arteriovenous shunts can be identified by CT, but PV shunts are difficult to detect because it is impossible to clearly separate a "portal" from a "venous" phase even with a multiphase dynamic CT study.[3] A biopsy and histologic study are useful to make a diagnosis of PV shunts in HHT.[4] In the presence of PV shunts, blood supply to the liver is largely dependent on the hepatic artery, and any procedure decreasing hepatic arterial flow can cause severe liver ischemia. Several studies have shown that embolization or ligation of the hepatic artery could cause hepatic or biliary necrosis in HHT patients. They claim that those interventions should not be done without ruling out PV shunts.[5,6] In our case, we diagnosed hepatic HHT from CT findings, but we could not rule out the presence of PV shunts. Thus, we decided to perform hepatic artery bypass before TEVAR with CA coverage. A saphenous vein graft might have been ideal in this infective case. However, we selected the prosthetic graft because we wanted to save time for vein harvesting in the emergency case. We kept the prosthetic graft away from the retroperitoneal hematoma to minimize the risk of prosthetic infection.

Endovascular treatment of infected aortic aneurysm has demonstrated excellent short-term outcomes. A multicenter study reviewing 123 cases retrospectively has shown that 30-day survival was 91%. Infection-related complications occurred in 27% of the cases, mostly in the first postoperative year.[7] Emergency open surgical repair would have been another option. Previous studies have shown that operative mortality rates of emergency open repair of the thoracoabdominal aorta were considerably high, 17% and 43.1%.[8,9] Operative risk is expected to be much higher in patients with HHT and infection. We chose emergency TEVAR rather than open repair, considering short-term benefits of endovascular treatment in this high-risk patient. Fortunately, the patient has been free from late visceral ischemic complications and recurrent infection in the first postoperative year.

CONCLUSIONS

Emergency TEVAR with CA coverage preceded by hepatic artery bypass was successfully performed for ruptured infected aneurysm of the thoracoabdominal aorta in a patient with HHT. Because full preoperative assessment of intrahepatic shunts is difficult in many HHT cases, securing blood flow to the hepatic artery should always be considered before TEVAR with CA coverage in patients with HHT.

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