Splenopneumopexy as an Addition to the Sugiura Procedure for Refractory Life-Threatening Variceal Hemorrhage in a Patient with Noncirrhotic Complete Portomesenteric Thrombosis

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Background	Patients with acute variceal hemorrhage refractory to endoscopic intervention may be managed with esophageal devascularization; however, this does not provide long-term treatment of portal hypertension. In cases in which transjugular intrahepatic portosystemic shunts (TIPS) or other conventional surgical shunts cannot be performed due to lack of viable target outflow vessels, as in the case of this patient with complete porto-spleno-mesenteric (PSM) venous thrombosis, splenopneumopexy can be added to relieve portal hypertension.
Summary	A 54-year-old male presented with complete PSM thrombosis and esophageal variceal bleeding. Variceal banding and splenic artery embolization at an outside hospital were unsuccessful management strategies. Upon arriving at this institution, a Sengstaken-Blakemore tube was placed to temporize the acute bleeding. The patient underwent emergent esophageal devascularization and surgical splenopneumopexy to provide long-term portosystemic decompression. He is now four years post-surgery without further bleeding episodes. There is radiologic evidence of decreased portal hypertension.
Conclusion	Splenopneumopexy is a useful addition to the Sugiura procedure in cases of complete PSM thrombosis with associated upper gastrointestinal bleeding and should be considered by hepatobiliary surgeons dealing with complications of portal hypertension.
Keywords	Variceal bleeding, splanchnic venous thrombosis, modified Sugiura procedure, splenopneumopexy

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Case Description

Variceal bleeding has a mortality of 15 to 20 percent within six weeks of the bleeding episode in cirrhotic patients.[1] Much less is known about the risks and mortality of variceal bleeding in patients with noncirrhotic portomesenteric thrombosis.[2,3] Management focuses on control of bleeding via endoscopy and prevention of recurrence via pharmacologic, repeated endoscopic, and surgical interventions.[1,3,4] Esophageal devascularization is attempted when neither transjugular intrahepatic portosystemic shunt (TIPS) nor surgical shunts are possible, but the portomesenteric hypertension is not addressed by this procedure. We report a case of variceal bleeding secondary to porto-spleno-mesenteric (PSM) thrombosis refractory to pharmacologic and endovascular therapy that was treated with esophageal devascularization and splenopneumopexy to allow for spleno-pulmonary shunting and decompression of portomesenteric hypertension.

The patient is a 54-year-old male with significant medical history of a prothrombin gene mutation who presented to an outside hospital with grade IV esophageal variceal bleeding refractory to endoscopic banding. Abdominal computed tomography (CT) scan demonstrated complete portal, splenic, and superior mesenteric vein thrombosis with large venous collateral vessels. Transaminases and synthetic liver enzymes were normal. As an attempt to limit inflow to the mesenteric vessels, the splenic artery was embolized, but bleeding continued. Insufficient resuscitation resulted in cardiac arrest secondary to hemorrhagic shock. After successful resuscitation, he was transferred to this hospital for escalation of care.

The patient arrived in hemorrhagic shock, on multiple vasopressors. A Sengstaken-Blakemore tube was immediately placed to tamponade the variceal bleeding, which controlled the hemorrhage and stabilized the patient. The surgical plan needed to address the immediate, life-threatening variceal hemorrhage and long-term decompression of his portal system.

First, esophagogastric devascularization was performed, similar to the original Sugiura procedure, but without splenectomy, vagotomy, or pyloroplasty[5]. Instead, we performed a left thoracotomy at the level of the 8th intercostal space. The diaphragm was incised via the transthoracic approach from the esophageal hiatus anteriorly to the xiphoid process and the gastroesophageal junction was mobilized. Devascularization was performed by ligating the perforating vessels along the distal 8 cm of the esophagus and 5 cm of the proximal stomach. The Sengstaken-Blakemore tube was removed without noting further bleeding, and esophageal transection and anastomosis was performed 2 cm above the gastroesophageal junction via an anterior gastrotomy using one load of a 29 mm end-to-end anastomosis stapler.

Second, a splenopneumopexy was performed to facilitate a long-term decompression of splanchnic venous blood flow. The superior pole of the spleen was brought through the defect in the diaphragm, the diaphragm was sewn to the splenic capsule with interrupted 0-chromic sutures, and the superior pole of the spleen transected to expose a 3 cm in diameter bare area of splenic parenchyma. Minimal bleeding from the surface was easily controlled with electrocautery. Next, a size-matched area of the left lower lobe of the lung was denuded off its visceral pleura. The spleen and lung parenchyma were then sewn together using interrupted 2-0 Chromic sutures (Figure 1). The chest was closed in layers and a chest tube was place.

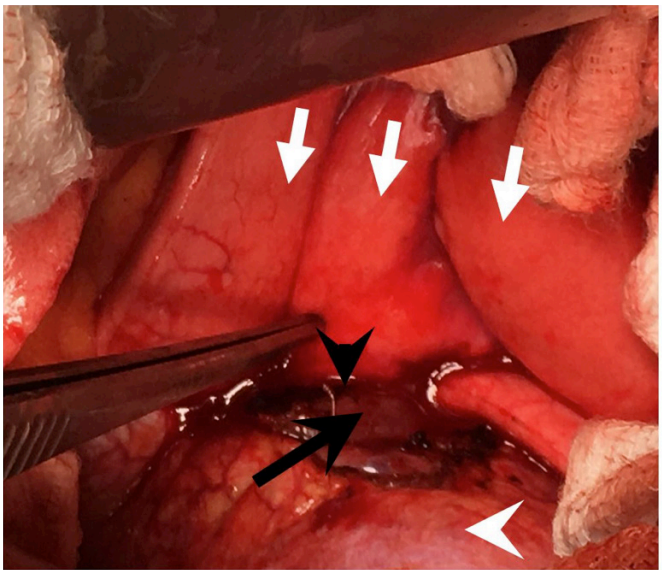

Figure 1. Intraoperative photograph of the splenopneumopexy (black arrowhead), spleen (black arrow), diaphragm (white arrowhead), and lung (white arrows)

The chest tube was removed on postoperative day (POD) three. The patient recovered well, tolerated a general diet, and was discharged home on POD 10. He has been followed in clinic for four years without any new episodes of gastrointestinal bleeding. A CT scan of the chest after four years showed venous vascular signal that extended from the spleen into the left lung vasculature (Figure 2). The spleen decreased in size from 1045 cm^3 to 305 cm.^{3} After surgery, the patient was started on therapeutic warfarin to manage his hypercoagulable state that he continues to date.

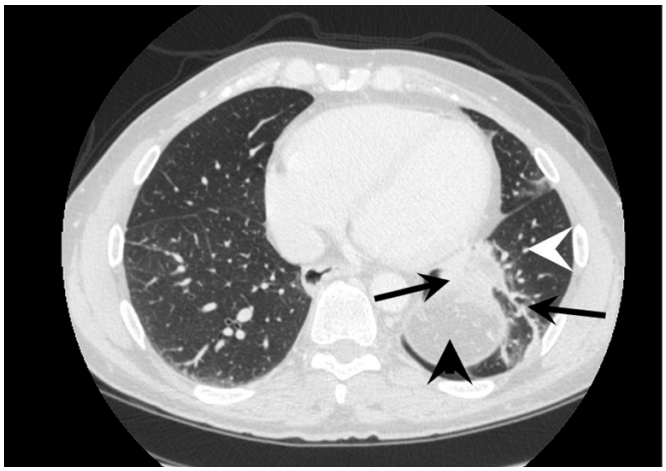

Figure 2. Postoperative (four-year) CT scan in the porto-venous phase demonstrating collateral circulation (black arrows) between the spleen (black arrowhead) and lung (white arrowhead)

Discussion

This case illustrates that variceal hemorrhage not amenable to endoscopic treatment can be controlled with a modified Sugiura procedure. Additionally, long-term decompression of the portosplenic system may be achieved by creating a surgical shunt between the parenchyma of the spleen and left lung. Ten percent of portal hypertension is caused by non-cirrhotic diseases.[6] These can be hereditary, including hypercoagulable disorders like Factor V Leiden mutation, Protein C or S deficiency, or Prothrombin gene mutation, or acquired, secondary to malignancy, abdominal surgery, or blunt abdominal trauma among others.[2,6] Chronic mesenteric thrombosis may lead to portal hypertension when it involves the portal and splenic veins, and may present with variceal bleeding.[3,4] In this case, the patient had a prothrombin gene mutation, complete PSM thrombosis with portal hypertension and variceal bleeding. Typical management of variceal hemorrhage aims to control variceal bleeding and prevent re-bleeding. First-line therapy includes pharmacologic and endoscopic intervention, with emergency surgical shunting as a last resort.[2,3,6,7] In therapy refractory hemorrhage, initial mechanical compression of gastroesophageal varices should be performed.[1,8]

In this case, first-line therapy failed. A Sengstaken-Blakemore tube was placed to temporarily stabilize the patient. Since the thrombosis affected all splanchnic veins, emergency TIPS or common surgical shunt procedures were not an option; therefore, a modified Sugiura procedure was performed to stop bleeding. A similar modified Sugiura procedure described by Ginsberg et al. consisting of esophageal devascularization, vagotomy, and Nissen fundoplication had a two-year rebleed rate of 25 percent when done in cirrhotic patients.[9] In addition, a splenopneumopexy was completed to relieve the PSM hypertension and to induce new collateral formation between the spleen and left lung.[10] Splenopneumopexy was initially described in a case series of 15 patients with Budd-Chiari syndrome. The series demonstrated reduced splenic pulp pressures after surgery (from a mean of 24.8 mm Hg to a mean of 19 mm Hg), and collateral circulation through the spleen and lung was shown, via splenoportography with ^{85}Kr tracing, to carry 47.5 percent of total portal flow within one month after the procedure. Moreover, all but one patient had resolution of ascites, and the two-year survival rate was 73 percent.[10] Although Budd-Chiari is routinely managed currently by portosystemic shunting, this original series suggested the effectiveness of long-term splanchnic decompression by shunting blood via the spleen to the lungs. In this case, we did not assess initial and repeated splenic pulp pressures like the above-mentioned study. However, a dynamic CT scan was performed four years after treatment and the extent of varices and collaterals was investigated. The CT demonstrated collateral circulation through the splenic-pulmonary interface, reduction in spleen size, and resolution of ascites. Our patient is now four years postsurgery without further bleeding episodes.

This case illustrates that splenopneumopexy may be beneficial when added to the Sugiura procedure in cases of extensive PSM thrombosis that is not manageable by repeated banding of gastroesophageal varices alone. It combines two surgical procedures for short-term (modified Sugiura procedure) and long-term (splenopneumopexy) alleviation of portal hypertension and variceal bleeding, and it may result in excellent clinical outcomes.

Conclusions

Splenopneumopexy in addition to the Sugiura procedure allows for the development of portosystemic collaterals in patients with no other portosystemic shunt options. It should be considered by surgeons who deal with difficult-to-treat portal hypertension where the PSM system is occluded.

Lessons Learned

Emergent esophageal bleeding is not always caused by cirrhosis and may be secondary to complete mesenteric thrombosis. When TIPS and conventional surgical shunts are not an option to relieve pressure, esophageal devascularization can stop active hemorrhage and splenopneumopexy can potentially provide long-term venous decompression via the development of splenopulmonary collaterals.

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