Laparoscopic Resection of Osler-Weber-Rendu Lesion

John Park, MD, Bryan Ellis, DO, Christopher Juergens, MD

ABSTRACT

Background and Objectives: Osler-Weber-Rendu is a hereditary disease characterized by telangiectasias, arteriovenous malformations, and aneurysms involving the cutaneous, gastrointestinal, pulmonary, and central nervous systems. This report describes a combinatorial approach using laparoscopic and intraoperative endoscopy to perform a partial gastric resection of bleeding arteriovenous malformations.

Methods: A 70-year-old female with a history of bleeding from Osler-Weber-Rendu disease presented to the emergency department complaining of hematemesis. Her vital signs were unstable, and she was immediately resuscitated with intravenous fluids and transfusions. A combined laparoscopic and intraoperative endoscopic approach to a partial gastric resection was planned. Intraoperatively, a single lesion was identified along the posterior aspect of the greater curvature of the stomach by using endoscopy. The blood supply was taken down with a Ligasure and gastric resection with a 60-mm Echelon stapler.

Results: The pathology report confirmed the complete resection of the arteriovenous malformations. The patient recovered well and was discharged home several days later without any pain complaints.

Conclusions: Little has been written about the medical treatment of Osler-Weber-Rendu arteriovenous malformations, let alone surgical treatment. A combinatorial laparoscopic and intraoperative endoscopic approach to gastric resection allows both minimization of the gastric resection and the complete identification and removal of the arteriovenous malformations.

Key Words: Osler-Weber-Rendu, Arteriovenous malformations.

INTRODUCTION

Osler-Weber-Rendu (OWR) or hereditary hemorrhagic telangiectasia (HHT) is a hereditary disease of vascular malformation transmitted as an autosomal dominant trait affecting 1 in 5000 to 8000 men and women.1 The signs and symptoms involve telangiectasias, arteriovenous malformations (AVMs), and aneurysms involving the cutaneous, gastrointestinal (GI), pulmonary, and central nervous systems (CNS). The skin areas most commonly affected are the mucosa of the mouth and GI tract, which are characteristically described as small, pulsating, macular and papular, and usually punctate telangiectasias. The most common initial symptom is recurrent epistaxis, which develops in most cases by puberty and eventually occurs in 80% of cases. The main risk is GI, pulmonary, and CNS hemorrhage.

The literature is limited regarding the medical or surgical therapeutic options for OWR malformations. Medical treatment options for the cutaneous telangiectasia of OWR include no treatment for mild cases, or electrosurgery and pulse-dye laser to destroy cutaneous and accessible mucosal lesions.

Surgical approaches to epistaxis and arteriovenous malformations have been reported in the literature.2-5 However, no standard surgical approach to recurrent bleeding from Osler-Weber-Rendu malformations has been established. In this case report, we describe the intraoperative endoscopic and laparoscopic approach to partial gastric resection of an Osler-Weber-Rendu AVM.

METHODS

A 70-year-old Caucasian female with a history of Osler-Weber-Rendu and a long history of multiple GI bleeds presented to the emergency department with complaints of vomiting bright red blood. She complained of weakness but no loss of consciousness. Vital signs showed a systolic blood pressure of 60 mm Hg to 70 mm Hg. Her
last episode and hospitalization was 5 months earlier when she was treated with gastric electrocautery and approximately one month earlier when she had gastric vascular ectasias electrocauterized. She had received a total of 7 units of blood the year before this hospitalization. Laboratory work revealed her hemoglobin and hematocrit (H&H) levels decreased from 14.1/42% to 10.8/29%. Her family history revealed that her father and sister both have Osler-Weber-Rendu, though her disease is more aggressive.

The patient was aggressively rehydrated and given 2 units of packed red blood cells with stabilization of serial H&H and vital signs. The gastroenterology department was consulted and an esophagogastroduodenoscopy (EGD) was performed, which revealed a large amount of clot in the patient’s stomach, and ulceration of her gastric wall with a 1-cm AVM located along the greater curvature. Due to her previous hospitalizations and blood transfusions for her disease, a long discussion was had with the patient and all services involved about partial gastric resection of the bleeding AVM. Everyone was in agreement that surgical resection was indicated, and she was prepared for surgery. Current recommendations for surgery include a bleed greater than 6 units, a 1500-mL to 2000-mL first-episode bleed, or a blood loss greater than 1000mL in the first 24 hours. A bleed of more than 5 units and an ulcer larger than 1 cm in diameter are risk factors for death from ulcer.

The operation was planned as a laparoscopic partial gastric resection with intraoperative EGD. A supraumbilical 5-mm port, 2 right ports (10 mm and 5 mm), and one left lower lateral 5-mm port were placed. After all ports were in place, another surgeon placed an Olympus GIF-160 endoscope into the stomach. As soon as the stomach was entered with the endoscope, the AVM was identified on the posterior aspect of the greater curvature of the stomach, while the rest of the stomach and duodenum were normal (Figure 1). The location of this lesion was ideal for a laparoscopic wedge resection as planned. In efforts to minimize the gastric resection while including the AVM in our resection, we used endoscopic guidance to aid our laparoscopic placement of metallic clips on the stomach, both proximally and distally to the lesion (Figure 2).

Next, we used the Ligasure (ValleyLab, Mansfield, MA) to enter the lesser sac and take down the short gastric vessels. After we had created a window just large enough for a 60-mm Echelon stapler (Ethicon, Cincinnati, OH), we completely excised the lesion with 3 reloads. Endoscopy was then repeated to confirm both the removal of the AVM and to test our staple line. The specimen was opened on the back table, and it was clear that we had included the lesion in the specimen (Figure 3). The pathology report also confirmed that the AVM was contained completely within our specimen, and that there were no malignant characteristics. The patient recovered well postoperatively and was discharged to home several days later.
DISCUSSION

Laparoscopic treatment of Osler-Weber-Rendu disease is not well cited in the literature, let alone in combination with intraoperative endoscopy. The major advantage with this combination procedure is minimization of the gastric resection. We were able to precisely locate and remove just the AVM without performing a hemigastrectomy or resection of a large amount of gastric tissue. This benefits the patient by avoiding the morbidity and mortality that is associated with a hemigastrectomy.

We should state that the location of the AVM on the greater curvature was ideal for a wedge resection. The argument could be made that this lesion could have been removed without endoscopy. However, using this combination technique we have both minimized the gastric resection while confirming removal of the lesion. The combination technique would actually have been more useful if the lesion had been in a more difficult location, such as the cardia or near the gastroesophageal junction, due to the ability to minimize the gastric resection.

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