Gastric cancer in a situs inversus totalis patient with multiple intestinal and vessel variations related to gastrectomy surgery

A case report and literature review

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Abstract
Rationale: Situs inversus totalis (SIT) is a rare congenital anomaly characterized by complete inversion of the abdominal and thoracic organs, and often involves multiple genetic mutations. The most suitable surgical technique for patients with multiple vessel and organ variations as well as SIT remains unclear. Furthermore, there has been insufficient clinical evidence that demonstrates which surgical techniques achieve the best outcomes. Finally, the standard of care has not yet been determined. We present the case of a 60-year-old man with SIT, who was diagnosed with moderately and poorly differentiated adenocarcinoma at the gastroesophageal junction. We further describe the advantage of using robotic-assisted laparoscopic surgery in patients with this anomaly.

Patient concerns: A 60-year-old man complained of pain in his upper abdomen for 3 months. Physical examination revealed an apex beat in the right fifth intercostal space, and vascular anomalies were noted on abdominal angiographic computed tomography.

Diagnoses: Moderately and poorly differentiated adenocarcinoma at the gastroesophageal junction with SIT.

Interventions: Robot-assisted total gastrectomy with D2 lymph node dissection and hand-sewn Roux-en-Y anastomosis was performed.

Outcomes: The postoperative course was uneventful, and the patient was discharged on the seventh postoperative day.

Lessons: Robotic surgery for gastric cancer is a safe and feasible alternative to laparoscopic surgery and it can be successfully used to treat gastric cancer in patients with SIT with multiple anatomic variations. As exemplified by our case, SIT might be accompanied by multiple anatomic variations. Detailed preoperative detailed imaging of the blood vessels and gastrointestinal tract is useful in these patients.

Abbreviations: ALHA = accessory left hepatic artery, GDA = gastroduodenal artery, HA = hepatic artery, LADG = laparoscopic-assisted distal gastrectomy, LGA = left gastric artery, SIT = situs inversus totalis, SMA = superior mesenteric artery, SPA = splenic artery, SPV = splenic vein.

Keywords: anatomic variation, D2 lymph node dissection, gastric cancer, robot surgery, situs inversus totalis, total gastrectomy

1. Introduction

The incidence of situs inversus totalis (SIT) in the general population has been discussed in several comprehensive studies. A review of the literature by Blegen[1] reported an incidence of 1:6600 based on chest radiography findings and 1:6200 based on autopsy findings. A review of Japanese studies showed incidences of 1:3834 for SIT, based on chest radiography (3 cases among 18,144 patients,[2] 15 cases among 49,690 patients,[3] and 10 cases among 39,526 patients[4]) and of 1:4563 based on barium upper gastrointestinal series (7 cases in 31,945 subjects[5]). Considering these data, an overall incidence from 1:4000 to 1:8000 seems plausible for SIT. Yaghan et al[6] said that in 1600, the first known case of SIT in a human was reported by Fabricius. In 1936, Allen[7] described a case of prepyloric carcinoma of the stomach in a 30-year-old man with SIT who died 3 weeks after gastrectomy. In 2003, the first case of laparoscopic surgery on a SIT patient with gastric cancer was reported.[8] In 2012, the first case of robot-assisted distal gastrectomy performed in a patient with SIT with gastric cancer was reported.[9] According to the literature, only 32 cases of gastric cancer have been reported in...
patients with SIT to date. Of these, only 1 patient underwent robot-assisted distal gastrectomy, 3 had vessel anomalies, and a few underwent laparoscopic-assisted distal gastrectomy (LADG). To our knowledge, there are no reports of robot-assisted total gastrectomy in the literature. Some journals cannot provide specific patient information, and we can only provide part of the patient’s information. Information on the cases in the literature is shown in Table 1. Here, we report the case of a patient with SIT with gastroesophageal junction cancer and discuss the vessel and organ variations relevant to total gastrectomy. Robot-assisted total gastrectomy with D2 lymph node dissection and hand-sewn Roux-en-Y anastomosis were performed.

2. Case presentation

2.1. Case report

A 60-year-old man presented with upper abdominal pain for 3 months. Gastroscopy revealed an ulcerated lesion on the gastric fundus, 1 cm beneath the esophagogastric junction, and a biopsy revealed adenocarcinoma. Physical examination revealed no Virchow lymph node, and the cardiac impulse was in the right fourth and fifth intercostal space at the midclavicular line. Percussion and auscultation of the heart were normal. The abdomen was soft with mild tenderness on the right upper quadrant and exhibited no rebound tenderness. The liver and spleen were not palpable. Chest radiography showed dextrocardia. Abdominal CT revealed situs with multiple spleens as well as celiac and hepatic vessel variations. (Fig. 1). The liver was rotated horizontally so that the larger right lobe was on the left side, and the right hepatic artery (HA) arose from the superior mesenteric artery (SMA) bridged by the gastroduodenal artery (GDA). There was no common HA, and the left hepatic lobe was supplied by an accessory left hepatic artery (ALHA) derived from the left gastric artery (LGA). The portal vein system was normal besides being horizontally rotated. Endoscopic ultrasonography showed a gastric ulcer below the cardia with concavity and yellowish mass around which the surface was uneven and swollen. Endoscopic ultrasonography revealed the gastric wall was thickened and hypoechoic at the lesion, which was 1.4 cm in diameter. The separation layer had completely disappeared, the local placenta percreta was not continuous, and there were no obvious enlarged lymph nodes near the lesion or in the mediastinum.

Informed consent was obtained from the patient. The authors have no funding and conflicts of interest to disclose.

2.2. Surgical procedure

Total gastrectomy with D2 lymph node dissection and hand-sewn Roux-en-Y jejunoesophageal anastomosis was performed robotically. The patient was under general anesthesia in a supine position with legs apart. The abdomen was explored laparoscopically to verify that no ascites or peritoneal implanting was present. The robot was docked and total gastrectomy with D2 lymphadenectomy was started. The adhesion between the omentum majus and the mesentery was detached and then peeled caudally to expose the duodenum. The transverse segment of the duodenum ran to the right side superficial to the superior mesenteric vessels (SMVs). After the gastroepiploic vessels below the pylorus were resected, an artery arch around the pancreatic head derived from the superior mesenteric artery (SMA) was found and could be traced into the hepatoduodenal ligament, where it became the proper HA. The duodenal bulb was transected after the lymphadenectomy in the superior pyloric area was completed. The portal vein was located on the inner side of and posterior to the proper HA, as normally found, while along the superior margin of the pancreas, the splenic vein (SPV) ran superficially to the artery and the root of the left gastric vein was easily seen and transected. The SPV was pressed caudally to expose the celiac trunk and its branches. Behind the right part of the SPV was the splenic artery (SPA), while on the left side, no common HA was found. The LGA was skeletonized and an
ALHA was found; therefore, the LGA was cut off distal to the bifurcation of the accessory left artery. By tracing the SPA behind the SPV, No. 11 group of lymphatic tissue was harvested. In the right upper quadrant 2 separate spleens were found. Because the carcinoma was located on the lesser curvature side, the splenic hila was not dissected. The postperitoneal fascia between the upper border of pancreatic tail and the cardia was dissected along a plane superior to the Gerota’s fascia. The esophagus was ligated at its lower end and then the pericardiac and periesophageal tissue in the esophageal hiatus was dissected, the 2 vagal trunks were transected, and the esophagus was transected 2 cm above the gastroesophageal junction. The lower segment of the esophagus was fixed on the crus to prevent the end of the esophagus from retreating into the mediastinum. Frozen section of the esophageal resection margin was pathologically examined. The en bloc specimen was collected into a bag and removed through a 4 cm upper middle abdomen incision. After establishing the pneumoperitoneum again, we explored along the duodenum and found the third part of the duodenum superficial to the mesenteric vessels from the left to the right side. The ligament of Treitz was unable to maintain the ascending duodenum in place. A short segment of the proximal jejunum was trapped behind the mesentery root by a ligament-like adhesion and, thereafter, the small intestine looped mainly on the right side of the abdomen. Only when the proximal jejunum was freed by lysis of the adhesion, was the jejunum able to be pulled upward to reach the esophageal end. A hand-sewn Roux-en-y esophagojejunostomy was made (Fig. 2).

Figure 1. Computed tomography showing transposition of the abdominal organs. ALHA=accessory left hepatic artery, HA=hepatic artery, L=liver, LGA=left gastric artery, SMA=superior mesenteric artery, SPA=splenic artery, SPV=splenic ven.
3. Outcome

Pathological examination revealed an ulcerative-type mass approximately 0.4 cm away from the esophagogastric junction line on the lesser curvature, and the tumor broke through the muscular layer. Microscopic examination revealed a moderately to poorly differentiated adenocarcinoma (Fig. 3) invading into the subserous layer (pT3), psychoneuroimmunology (+), and positivity for one of the 46 lymph nodes. The HER-2 result was + on immunohistochemistry, but the FISH result was negative. After the surgery, the patient recovered uneventfully. A liquid diet was started the morning after surgery. Flatus occurred 32 hours after surgery, and the patient was discharged on the eighth day after surgery. No postoperative complications occurred.

4. Discussion

SIT may be a genetic disease, but its clear genetic reasons are still unknown. Nonaka et al. suggested that immobility of nodal cilia inhibits the flow of extraembryonic fluid during embryogenesis leading to the development of SIT. The anatomical variants of patients with SIT are not simply related to the location. SIT with agnathia has been reported. Symptomatic abnormalities in the chest (septal defects, lungs being divided into lobelets, pulmonary arterial stenosis, tetralogy of Fallot, or transposition of the great vessels and total esophageal duplication) and problems inside the abdomen (symmetrical liver, absent spleen or multiple spleens, intestinal abnormalities, atresia or stenosis of the duodenum, and urologic anomalies) can also occur. Partial SIT with normocardia might also be associated with cardiac diseases, including, for example, familial long QT syndrome. In addition to the malformation, patients with SIT may have a higher risk of cancer due to malfunction of the KIF3 complex.

Although SIT is usually asymptomatic, it is often found in certain clinical situations. The inverse location of the viscera might lead to misdiagnosis of abdominal disease if SIT is not taken into consideration. In addition, some kinds of abnormalities, including organ location abnormalities and vessel abnormalities, might be an impediment to operation when surgery is needed.

The standard operation for advanced gastric cancer requires total or distal gastrectomy with D2 lymphadenectomy. The anatomic variation may make lymph node dissection inconvenient. Only 3 cases involving vascular malformation have been described in the literature to date; in first patient, the left HA branched from the SMA, in the second patient, the right gastric artery branched from the celiac trunk and the common HA branched from the SMA, and in the third patient, the left HA branched directly off the abdominal aorta. The abnormalities found in the third patient increased the difficulty of an operation. Meanwhile, in our case, abnormalities of the organs in the abdomen included multiple spleens and trapping of the proximal segment of the jejunum behind the mesenteric vessels due to...
intestinal malrotation, which restricts the manipulation of the jejunum during anastomosis. The free skirt of the major omentum adhered to the mesentery of the small intestine loops because there was no normal transverse colon; therefore, performing an omentectomy was markedly different than the usual procedure. In addition, vessel abnormalities in this patient included the lack of the common HA, so the proper HA derived from the SMA through the GDA. A branch of the LGA became an ALHA. In addition, the portal system was located superficially to the celiac branches. Therefore, the veins had to be retracted to expose the arteries during the lymphadenectomy. To the best of our knowledge this is the first report of vessel and organ variations during robot-assisted total gastrectomy.

Preoperative imaging was effective for detecting the vessel variants, which was helpful during the dissection. With the availability of high resolution CT data sets, a wide variety of 2-dimensional and 3-dimensional reconstructions can now be acquired. However, imaging did not show all the abnormalities in this patient, especially the intestinal malrotation. In our patient, the intestinal abnormality impeded the progression of the robotic surgery during reconstruction. During abdominal exploration, we found the malrotation, but did not notice the surgery during reconstruction. During abdominal exploration, we

5. Conclusion

The occurrence of gastric cancer in a patient with SIT is very rare. In few reports of SIT, variations in vessels and organs are mentioned, but details regarding how to deal with these blood vessels during surgery are not described. When operating on patients with SIT, it is important to be attentive to anatomic variations, particularly vascular anomalies. Detailed exploration might be more useful before surgery. Robotic surgery is an efficient method for treating gastric cancer in a patient with SIT with vessel and organ variations.

Acknowledgment

The authors sincerely thank the entire staff of our department for offering their assistance with the medical service and manuscript writing processes.

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