

Minimally invasive esophagectomy with intrathoracic anastomosis in a situs inversus totalis patient

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Case report

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Abstract

Background

Situs inversus totalis (SIT) is a rare congenital condition which is characterized by abnormal placement of the thoracic and abdominal organs. Laparoscopic and thoracoscopic esophagectomy with intrathoracic anastomosis is technically difficult and has rarely been reported in SIT patients with esophageal cancer.

Case presentation:

We report a SIT patient whose condition is complicated by middle and lower esophageal cancer. This 55-year-old patient underwent total laparoscopic and thoracoscopic esophagectomy with intrathoracic anastomosis. The overall operation time was 270 minutes. Estimated blood loss was 150 mL and the patient was discharged after 20 days.

Conclusions

Laparoscopic and thoracoscopic esophagectomy with intrathoracic anastomosis is technically feasible and secure in SIT patients with esophageal cancer.

Background

Situs inversus is a rare congenital disease characterized by the abnormal arrangement of organs in the thorax and abdomen. This condition can be divided into complete situs inversus, known as situs inversus totalis (SIT), and incomplete situs inversus (ISI) [1]. Arranged in a mirror image of their usual positions, the organs of SIT patients are completely transposed in a right-to-left reversal. SIT is rare, and SIT patients with esophageal cancer are rarer still [2]. To date, no more than 10 cases of video-assisted thoracoscopic esophagectomy for the treatment of SIT patients with esophageal cancer have been reported [3]. Moreover, intrathoracic anastomosis in conjunction with a minimally invasive esophagectomy of a SIT patient is technically demanding and has been reported only once [4]. Here, we discuss a SIT patient whose condition was complicated by middle and lower esophageal cancer. This patient underwent laparoscopic and thoracoscopic esophagectomy with intrathoracic anastomosis. We provide detailed description of these surgical procedures and valuable information as to the treatment of this rare condition.

Case Presentation

A 55-year-old man was admitted to hospital with progressive dysphagia. The patient was found to have mirror-image dextrocardia (Fig. 1) with an abnormally thickened esophageal wall (thoracic middle-lower

segment) observed by way of contrast-enhanced computed tomography (CT) of the chest (Fig. 2a). SIT was observed in the patient, with a right-sided position of the heart and aortic arch (Fig. 2b). Additionally, the spleen and stomach were observed on the right side and the liver on the left by means of CT of the upper abdomen (Fig. 2c). Further to this, an esophagogram indicated the presence of an irregular filling defect of approximately 6 cm in length on the middle-lower segment of the esophagus (Fig. 3).

Endoscopic ultrasonography of the upper digestive tract confirmed the presence of an esophageal tumor involving fibrous membrane 30 to 45 cm from the incisor teeth. No significant enlargement of lymph nodes was detected. A CT scan of the head, a bone scan, and abdominal ultrasonography revealed no distal metastases. A cardiopulmonary function test indicated that there were no apparent surgical contraindications. Based on the above preoperative examination, the patient was diagnosed with esophageal carcinoma, which was clinically staged as T3N0M0, stage IIA (Union for International Cancer Control [UICC] esophageal carcinoma pathological stage, 2017). Laparoscopic and thoracoscopic esophagectomy with intrathoracic anastomosis was determined to be the best treatment for the patient's middle-lower esophageal carcinoma.

Surgical Technique

The patient underwent intravenous-inhalation general anesthesia and double-lumen endotracheal intubation.

Throughout the laparoscopy-assisted abdominal surgery, the patient underwent two-lung ventilation and was placed in a supine position with five abdominal ports. The observation port was an 11-mm incision beside the umbilicus, and the other four ports were positioned at the incisions in the subcostal region of the left (5-mm port) and right midclavicular lines (5-mm port), and the incisions 2 cm superior to the umbilicus in the left (11-mm port) and right parasternal lines (5-mm port). The surgeon stood on the left side of the patient, which is the opposite of the standard position. Under the laparoscope, in accordance with preoperative imaging, the stomach was observed in the right hypochondriac region, the spleen to the right and rear of the stomach, and the liver on the left (Fig. 4a). The patient's stomach was routinely mobilized, the abdominal lymph node was dissected, and the anastomosis was circularly embedded with the pedicled omentum flap. An assistant abdominal incision was subsequently made below the xiphoid process, and a gastric conduit of approximately 3.5 cm in diameter was created using a linear stapler (the Endo-GIA 60 – 4.1; Johnson & Johnson) outside the abdominal cavity. A jejunostomy was performed approximately 30 cm distal to the ligament of Treitz.

Before the two-port thoracoscopy-assisted thoracic surgery began, the patient was repositioned in a right prone position and underwent right single lung ventilation. The camera port was located at the eighth intercostal space on the posterior axillary line, and the operation port was positioned between the fourth and sixth intercostal space at the anterior axillary line. The surgeon stood on the abdominal side of the patient. Intraoperative observation revealed that the azygos vein and the three lobes lay in the left thoracic cavity (Fig. 4b). A tumor 5 cm in length with a maximum diameter of 3 cm was located in the middle and lower segment of the esophagus (from 2 cm above inferior pulmonary vein to 2 cm above the

diaphragm) (Fig. 4c). The esophagus was mobilized at a site 4 cm superior to the azygos vein, and the pleura surrounding the esophagus at the anastomotic stoma and thoracic lymph nodes were systematically dissected.

The remaining steps in this surgery resembled that of surgery performed on situs solitus patients with esophageal cancer and consisted of purse-string suturing, the insertion of the anvil of the stapler, distal esophagectomy, and intrathoracic anastomosis of the esophageal stump and the tubular stomach. After anastomosis, a thoracoscopy was performed by inserting an endoscope into the gastric conduit to ensure the integrity of the anastomotic stoma and that it was not bleeding.

The overall operation time was 270 minutes, comprising laparoscopic operation time (100 minutes) and thoracoscopic operation time (170 minutes). Estimated blood loss was 150 mL. Throughout the procedure, systematic lymphadenectomies were conducted at levels 2R (right recurrent laryngeal nerve), 8 (middle-lower segment of the esophagus), 17 (left gastric artery), and at the lesser curvature of the stomach and pylorus. Postoperative pathology confirmed that a well to moderately differentiated squamous cell carcinoma had invaded the entire esophageal wall, including the serosa, with metastasis to the lymph nodes, including the 2R (1/8) and the pylorus (1/3). Finally, the patient was diagnosed with well to moderately differentiated esophageal carcinoma which was pathologically staged as T3N1M0, stage IIIB (UICC esophageal carcinoma pathological stage, 2017) [5]. There were no postoperative complications such as trachyphonia or other symptoms related to recurrent laryngeal nerve injury. After 20 days, the patient was discharged and advised to undergo adjuvant therapy in the oncology department. A video demonstrating the operation accompanies this article (Additional file 1).

Discussion And Conclusions

A rare autosomal recessive disease, SIT was first described in 1600 by Fabricius [6], and its incidence ranges from 1/8000 to 1/25,000 in the general population [7]. Other than their anomalous organ arrangement, most SIT patients have relatively normal anatomy. However, 20–30% of SIT cases coexist with Kartagener syndrome, which is characterized by a triad of conditions: bronchiectasis, sinusitis or nasal polyps, and situs inversus. In clinical practice, we rarely encounter a case of esophageal cancer complicated by SIT. The surgical treatment of esophageal cancer have evolved from traditional open surgery to minimally invasive surgery. Yoshida et al. first reported a patient with SIT underwent simultaneous hand-assisted laparoscopic gastric mobilization and thoracoscopic esophagectomy in 2004[8]. Later, other case reports followed, with cervical anastomosis used in most of cases[2, 9–11]. Intrathoracic anastomosis was reported only once[4]. This 2014 study by Peel et al, however, did not focus on surgical techniques, but rather on the treatment of SIT in Kartagener syndrome. In our case, intrathoracic anastomosis was selected instead of cervical anastomosis to achieve radial resection of the nidus and decrease the incidence of severe complications such as recurrent laryngeal nerve injury and anastomotic leak. This surgical method is technically feasible and secure [12].

There are two predominant reasons why the presence of SIT can complicate surgeries. One is that the anomalous translocation of viscera that occurs in SIT requires a surgical team to be flexible and accommodate this reversal of organs by performing surgery in a mirror image of what it would be under normal circumstances. As described above, this was reflected in the positioning of the patient, respiratory management, endoscope incisions, mobilization of the organs, lymph node dissection, et cetera. Fortunately, the relative anatomical position of organs in most SIT patients is the same as that in situs solitus patients, which decreases the difficulty of thoracoscopic and laparoscopic surgery and ameliorates the risks involved. Another reason SIT increases the difficulty level of these surgeries is that SIT patients often have comorbid conditions, for example Kartagener syndrome. In our case, Kartagener syndrome wasn't found. Patients with Kartagener syndrome often have respiratory infections due to poor mucociliary clearance, especially in surgical conditions [13]. Complete preoperative examination, including necessary imaging and cardiopulmonary function assessment, and careful perioperative management of respiratory tract are therefore vital to avoiding severe postoperative complications such as sepsis and respiratory failure.

Based on our observations, we can conclude that the key to successful treatment of SIT patients with esophageal cancer through laparoscopic and thoracoscopic esophagectomy with intrathoracic anastomosis is the ability of a skilled surgical team to adjust to this reversal of organs and invert the positioning of the operation. Additionally, staff need to carefully assess whether patients have other anatomical abnormalities that may cause severe complications and attend to them during the perioperative period.

Abbreviations

SIT: Situs inversus totalis; ISI: incomplete situs inversus; CT: Computed tomography

Declarations

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Availability of data and materials

All data generated or analyzed during this study are included in this manuscript.

Authors' contributions

YL was the main surgeon of the operation and conceived of the study. KL and ZZ were the assisted surgeons of the operation. YM, ZZ and KL collected the clinical data. YM, HX and WQ drafted the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The study design was approved by the appropriate ethics review boards.

Consent for publication

Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images

Competing interests

The authors have no conflicts of interest to declare.

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Figures



Figure 1

Chest X-ray showing dextrocardia

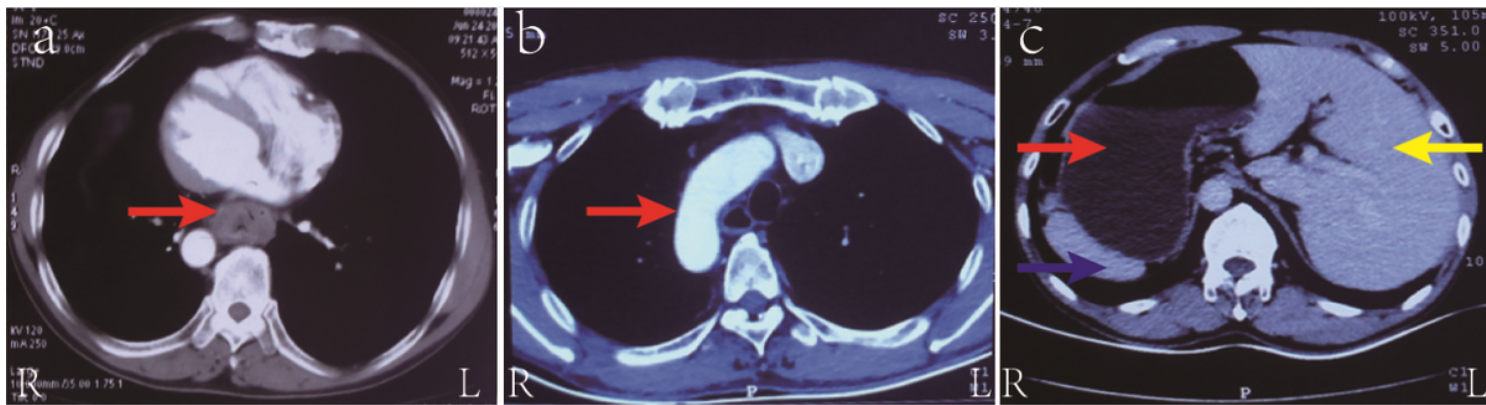


Figure 2

Contrast-enhanced computed tomography of the chest and upper abdomen showing (a) the abnormally thickened esophageal wall of the thoracic middle-lower segment (red arrow); (b) the aortic arch as arranged in SIT (red arrow); and (c) the liver (yellow arrow), stomach (red arrow), and spleen (blue arrow) as arranged in SIT.

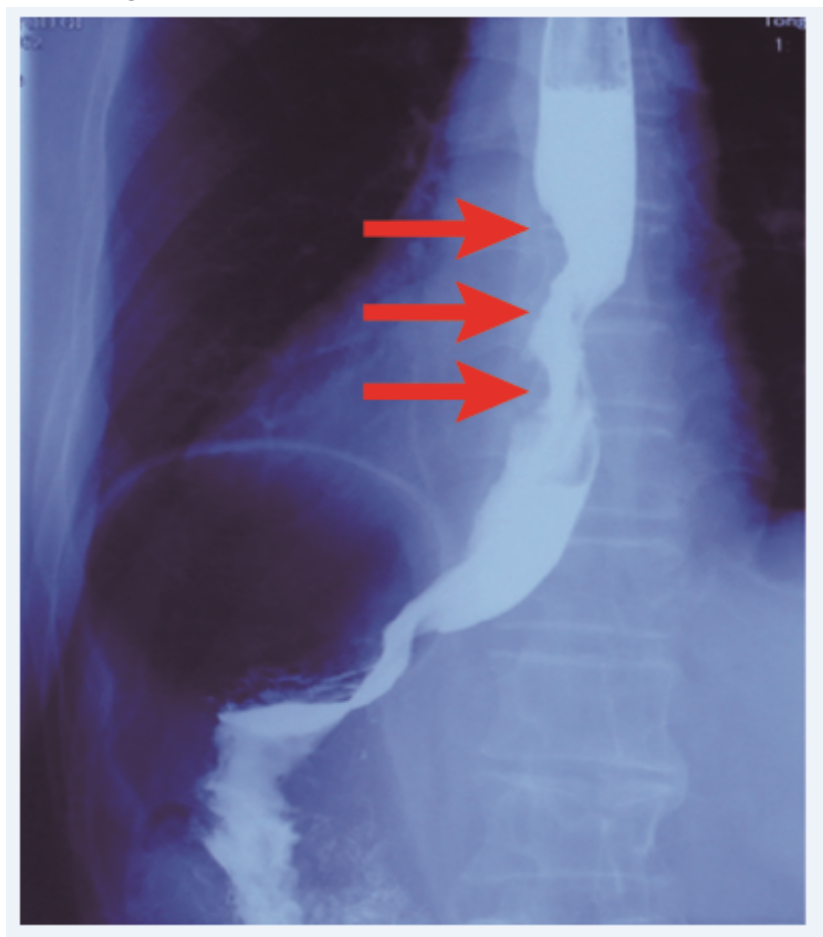


Figure 3

Esophagogram indicating the presence of an irregular filling defect approximately 6 cm in length on the middle-lower segment of the esophagus (red arrows).

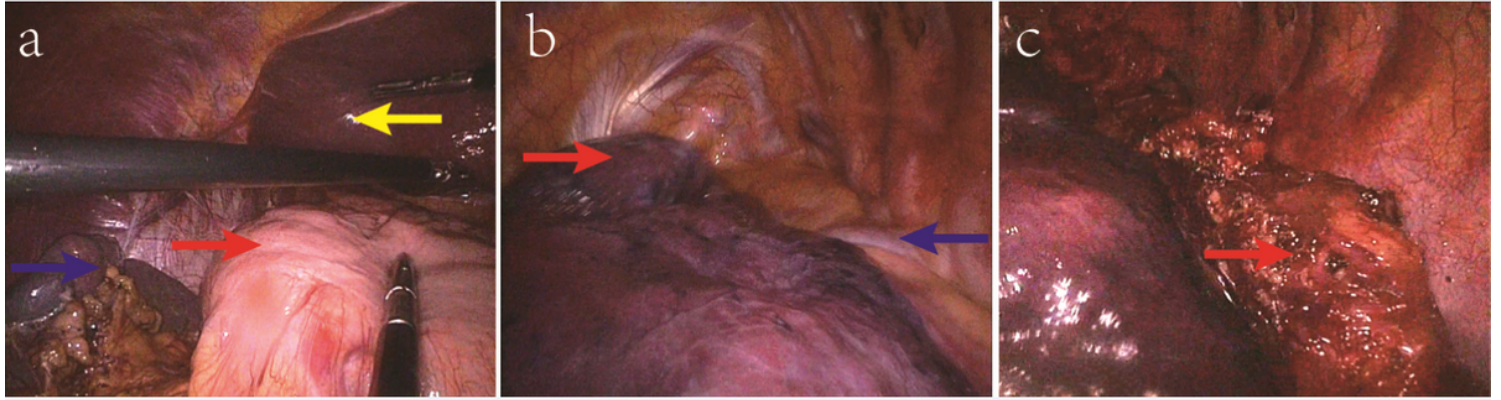


Figure 4

Intraoperative observation demonstrating (a) the stomach (red arrow) in the right hypochondriac region, the spleen (blue arrow) on the right rear of the stomach, and the liver (yellow arrow) on the left; (b) the azygos vein (blue arrow) and the three lobes (red arrow) in the left thoracic cavity; and (c) the esophageal carcinoma (red arrow).

Supplementary Files

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