

## Case Report

# Situs inversus totalis with esophageal cancer: a case report

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### ABSTRACT

Situs inversus is a rare disorder and its association with esophageal cancer is seldom encountered in clinical practice. Its incidence is 1/8000 to 1/25000 live births. Only 12 cases have been reported in the literature so far. Here, we report a case of esophageal cancer with situs inversus totalis, which was successfully managed by esophago-gastrectomy via a high right thoracoabdominal approach and review the relevant literature. Although the surgery was technically challenging due to anatomic reversal of organs, it did not differ from the routine surgery. Moreover, it was observed that with careful recognition of mirror image anatomy preoperatively and during surgery, complications can be avoided.

**Keywords:** Situs inversus totalis, Lower esophageal cancer, Adenocarcinoma, Thoracoabdominal approach, Esophagectomy

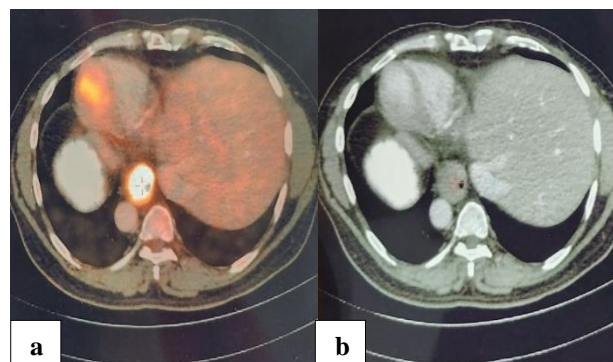
### INTRODUCTION

Situs inversus totalis (SIT) is a rare anomaly with complete transposition of thoracic and abdominal organs. Its incidence is 1/8000 to 1/25000 live births.<sup>1</sup> Only 12 cases of esophagectomies for esophageal cancer with SIT have been reported worldwide so far.<sup>2</sup> Mirror image reversal of organs poses a technical challenge to the operating surgeon. Hence, preoperative orientation of anatomic reversal is essential for one-lung anesthesia, incision and anastomosis. Herein, we present a rare case of lower esophageal and gastroesophageal junction (GEJ) adenocarcinoma who underwent esophagectomy. In this patient intraoperative and postoperative period was uneventful.

### CASE REPORT

A 62-year-old male, known case of situs inversus, presented with progressive dysphagia and weight loss (over 6 kg) of 3-months duration. He was evaluated with endoscopy showing circumferential growth in lower esophagus extending 35 to 40 cm from incisors. Computed

tomography (CT) scan confirmed 5.6 cm long lower esophageal-GEJ growth; liver and coeliac axis was normal; however, there were para-esophageal and peri-gastric nodes. Biopsy confirmed pathology as adenocarcinoma. PET-CT scan also ruled out any distant metastasis (Figure 1).

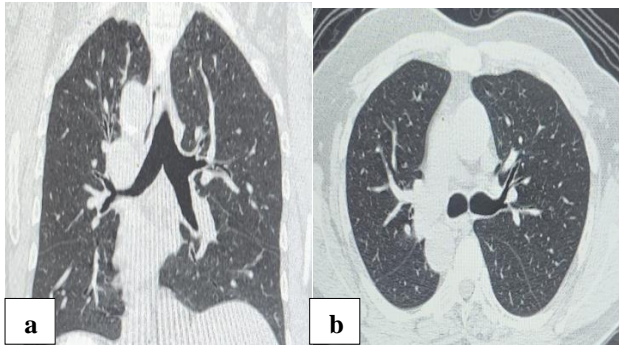


**Figure 1 (a and b): PET-CT scan showing lower esophageal and GEJ circumferential growth causing luminal narrowing with FDG uptake (SUVmax:18).**

The patient denied any neoadjuvant treatment and thus was taken up for esophago-gastrectomy. The anesthesia and operative challenges faced are as follows.

### One-lung anesthesia

In this patient the left side of lung had anatomical structure of right lung (Figure 2). We intended for collapse of right lung with one-lung anesthesia. So, we had put left double lumen endotracheal tube (DLET) size 32 Fr which failed to achieve one-lung anesthesia as left DLET did not match the bronchial structure of left lung.



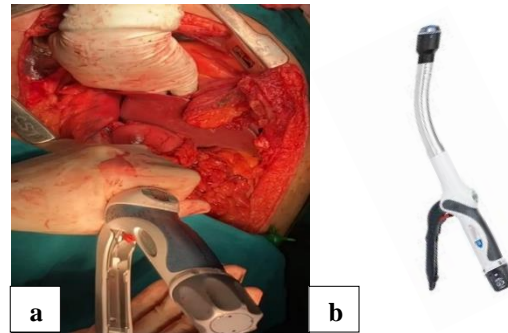
**Figure 2 (a and b): HRCT image where left lung is showing anatomical structure of right lung.**

### Operative procedure

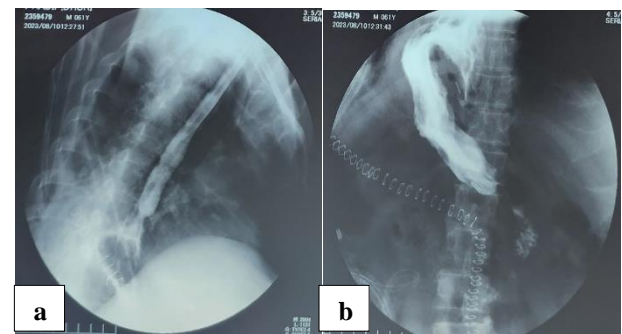
We took a high right thoraco-abdominal approach along the 7th intercostal space (ICS) for adequate exposure. Phrenic nerve saving peripheral diaphragmatic incision was taken. Cuff of diaphragm excised near cardio-esophageal junction for tumor free margins. The stomach was mobilized. The left para-esophageal (PE) lymph node was felt just below carina, so we thought bilateral thoracotomy may be required to get nodal clearance and tumor free margin. However, we mobilized esophagus much beyond the right inferior pulmonary vein (IPV) across the midline in front of the vertebra and after vagotomy, the left PE node could be dissected down completely. Left gastric lymph node clearance was also done with excision of entire lesser curvature of stomach. The lower esophagus-GEJ was resected with adequate margins followed by esophago-gastric end to end anastomosis using circular stapler (Figure 3). Later, pyloromyotomy and feeding jejunostomy was also done.

In the postoperative period, the patient recovered well. On postoperative day nine, oral contrast study showed no anastomotic leak and good pyloric emptying (Figure 4). The patient was started on liquid diet and later soft diet by end of fifteen days.

The final histopathology report was suggestive of moderately differentiated adenocarcinoma involving gastroesophageal junction invading muscularis propria with lympho-vascular and perineural invasion and ten metastatic lymph nodes (pT2 N3 M0).



**Figure 3 (a and b): Operative image showing circular stapler being fired across midline; a hand lifting right IPV and pericardium upwards for safe stapler application.**



**Figure 4 (a and b): Oral contrast study showing no anastomotic leak and good pyloric emptying.**

### DISCUSSION

Situs inversus is a rare condition. Aristotle first described this condition in animals, whereas Fabricius reported the first known human case in 1600.<sup>3,4</sup>

In clinical practice, we rarely encounter cases of esophageal cancer associated with SIT. Its incidence is between 1/8000 to 1/25000 live births.<sup>1</sup> Surgical procedure in SIT patients are challenging owing to their mirrored anatomy. Therefore, recognition of their variations is very important to avoid intraoperative accidental injuries. The surgical treatment of esophageal cancer has evolved from traditional open surgery to minimally invasive surgery (Table 1).

Yoshida et al first reported a patient with SIT underwent simultaneous hand-assisted laparoscopic gastric mobilization and thoracoscopic esophagectomy in 2004.<sup>5</sup> In this study, although esophagectomy was performed only thoracoscopically, it was a little difficult to recognize the anatomical relationship of the pleural contents in the thoracoscopic view and because of the absence of the surgeon's tactile sensation. Hence, they suggested that introduction of hand-assisted thoracoscopic esophagectomy might be one of the ways to achieve a safer procedure.

**Table 1: Summary of 12 reported cases and our case of esophagectomy for esophageal cancer with situs inversus totalis.**

Case	Age (years)	Sex	Comorbidity	Operative procedure	Position in thoracic surgery	Operative time (min)	Blood loss (ml)	Perioperative complications	Hospitalization days
Yoshida et al <sup>5</sup>	57	M	None	Thoracoscopic + hand-assisted laparoscopic surgery	Right decubitus	540	340	None	Not described
Mimae et al <sup>6</sup>	57	M	None	Thoracotomy + laparotomy	Right decubitus	512	585	None	16
Aoki et al <sup>7</sup>	53	M	None	Thoracotomy + laparotomy	Right decubitus	463	762	None	18
Yagi et al <sup>8</sup>	73	M	None	Thoracoscopic + hand-assisted laparoscopic surgery	Right decubitus	390	130	None	Not described
Peel and Darling <sup>9</sup>	67	M	Kartagen er syndrome	Thoracoscopic + laparoscopic surgery	Right decubitus	Not described	Not described	Not described	Not described
Ujiie et al <sup>10</sup>	63	M	None	Thoracoscopic + hand-assisted laparoscopic surgery	Right decubitus	621	310	None	17
Chinusa-my et al <sup>11</sup>	62	M	None	Thoracoscopic + laparoscopic surgery	Prone	286	Not described	None	11
Hosoda et al <sup>12</sup>	78	M	None	Thoracoscopic surgery + laparotomy	Right semiprone	861	978	Right recurrent laryngeal nerve palsy	30
Nakano et al <sup>13</sup>	82	M	None	Thoracoscopic + hand-assisted laparoscopic surgery	Prone	661	157	None	34
Nakano et al <sup>13</sup>	66	M	Intestinal malrotation; polysplenia	Thoracoscopic surgery + laparotomy	Prone	637	210	None	17
Feng et al <sup>14</sup>	54	M	None	Thoracotomy + laparotomy	Right decubitus	Not described	Not described	None	10
Xie et al <sup>2</sup>	66	M	None	Thoracoscopic + laparoscopic surgery	Right lateral-prone	480	80	N	
Our case	64	M	None	Thoracoabdominal approach	Left lateral decubitus	540	100	None	10

Later in 2008 Mimae et al reported a case of middle 1/3<sup>rd</sup> esophageal carcinoma where they said it was easier to perform esophagectomy through a left thoracotomy for the patient with situs inversus totalis, because the organs are arranged in a mirror image of the normal positions and curative surgery can only be achieved through a left thoracotomy.<sup>6</sup>

However, the location of important organs such as the recurrent laryngeal nerves and bronchus was unclear and unfamiliar at the beginning of the operation where they tried to always imagine the mirror-imaged anatomy during surgery and dissected the lymph nodes very carefully so as not to injure the important organs.

In our case, certain challenges were faced. For one lung anesthesia, we should have put right DLET on left side in view of reversed bronchial anatomy of right and left lung. Because of which we probably failed to achieve one-lung anesthesia. Also, our initial preoperative plan was bilateral thoracotomy to remove the left PE node. However, mobilization of esophagus beyond IPV, across midline on opposite side up to subcarinal area and after vagotomy, left PE node could be approached for nodal clearance. Following this esophago-gastric anastomosis was done using circular stapler across midline, thus avoiding opposite side thoracotomy.

There are few studies showing minimal invasive approaches. Xie et al in 2021 reported a case of esophageal cancer with SIT successfully treated by total MIE, with a right lateral-prone position adopted during the thoracic procedure that had combined advantages of both the lateral decubitus position (allowing quick conversion to open surgery) and prone position (providing a well-exposed operative field for esophagus).<sup>2</sup> They faced mainly two challenges, first was the fact that the surgeon found it difficult to localize and identify the recurrent laryngeal nerves because the left-side recurrent laryngeal nerve looped under the left-side subclavian artery while the right-side recurrent laryngeal nerve looped under the right aortic arch.

The second came from the fact that the right-handed surgeon felt more impairment when dissecting with his left hand for some procedures, such as mobilizing and dissecting the tissues in the outlet of the thoracic cavity. But they used preoperative three-dimensional (3D) image reconstruction which was a helpful adjunct as it provided the most graphic representation of the orientation of organs, especially with SIT.

## CONCLUSION

Situs inversus appears as a mirror image in imaging system. However, technical execution of surgical management is a task. A perfectly changed anatomical orientation is necessary for suitable anesthesia and a suitable approach.

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