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FLIPPING THE SCRIPT: MANAGING CARCINOMA OF THE ESOPHAGUS IN THE PRESENCE OF SITUS INVERSUS TOTALIS - CASE REPORT AND LITERATURE REVIEW

Oncology

Mohammad Masoom Parwez M.Ch Resident, Department of Surgical Oncology, State Cancer Institute, Guwahati, Assam, India.

Naveen Saini M.Ch Resident, Department of Surgical Oncology, State Cancer Institute, Guwahati, Assam, India.

Zahid Zahiri M.Ch Resident, Department of Surgical Oncology, State Cancer Institute, Guwahati, Assam, India.

Rajiv Paul* Associate Professor, Department of Surgical Oncology, State Cancer Institute, Guwahati, Assam, India. *Corresponding Author

ABSTRACT

Introduction: Situs inversus totalis (SIT) refers to a mirror image reversal of the normal position of the internal organs. The incidence of situs inversus totalis is 1 in every 8,000 to 25,000 births, and the condition is most often diagnosed by radiographic examination. Only 12 cases of esophageal cancer with SIT have been reported worldwide. Here we report a case of left-sided hybrid VATS McKeown's Esophagectomy. **Case Presentation:** A 47-year-old male patient presented with progressive dysphagia to solids for 1 month. Endoscopy revealed infiltrating growth at 3-6 o'clock position starting 35cm. Punch biopsy demonstrated moderately differentiated squamous cell carcinoma. CECT scan revealed dextrocardia and situs inversus with liver, gall bladder, IVC on left side and aorta, spleen and sigmoid colon on the right and asymmetrical distal oesophageal wall thickening with luminal narrowing. After MTB approval, he underwent neoadjuvant concurrent chemoradiation as per CROSS protocol followed by left VATS McKeown's esophagectomy with standard two-field lymphadenectomy in the right lateral decubitus position. Postoperative period was uneventful with length of hospital stay being 10 days. Final histopathology revealed pathological complete response with margin free of tumor. Patient kept on close follow-up. **Conclusion:** In the realm of rare anatomical anomalies, the present case stands as a testament to the complexity and diversity of clinical challenges that medical professionals may encounter. The coexistence of esophageal cancer and SIT presents a formidable diagnostic and therapeutic puzzle. This report reiterates the need for constant adaptation, innovation, and multidisciplinary collaborations in the pursuit of enhancing patient outcomes.

KEYWORDS

Situs inversus, carcinoma esophagus, esophageal cancer, mckeowns esophagectomy, situs inversus totalis

INTRODUCTION

Situs inversus totalis refers to a mirror image reversal of the normal position of the internal organs [1]. The incidence of situs inversus totalis is 1 in every 8,000 to 25,000 births, and the condition is most often diagnosed by radiographic examination [2]. Only 12 cases of esophageal cancer with SIT have been reported worldwide [3]. Here we report a case of left sided VATS McKeown Esophagectomy with laparotomy for gastric conduit with thoracic part done in right lateral position.

CASE REPORT

A 47-year-old male patient presented with progressive dysphagia to solids for 1 month and weight loss with intact appetite. He is known smoker for last 10 years currently abstaining for last 1 month with no known co-morbidities and significant family history. Upper Gastro Intestinal Endoscopy revealed infiltrating growth at 3-6 o'clock position starting 35cm from upper incisor with near total luminal occlusion. Punch biopsy from the growth showed features of moderately differentiated squamous cell carcinoma. Contrast enhanced computed tomography scan revealed dextrocardia and situs inversus with liver, gall bladder, cecum, appendix on the left side. spleen and sigmoid colon on the right side and aorta seen to the right of the inferior vena cava (Fig- 1). Eccentric heterogenous wall thickening of the distal esophagus (length of 5-6 cm, maximum wall thickness of 15mm) with luminal narrowing and proximal esophageal dilatation. Loss of fat planes with left atrium. On clinical examination, ECOG-1, no palpable cervical lymphadenopathy, per abdomen examination within normal limits. He underwent nasogastric tube insertion on outpatient basis for feeding and nutritional rehabilitation.

Case was discussed in our Institutional multidisciplinary tumor board. Patient underwent neoadjuvant radiation (41.4Gy/23 fractions over 4-5 weeks) with concurrent chemotherapy (2 cycles of paclitaxel {50mg} and carboplatin{200mg}). Response evaluation was done after 6 weeks from completion of neoadjuvant treatment. Imaging revealed stable disease as per RECIST criteria and maintained fat planes with adjacent structures.

Preoperative cardiopulmonary assessment was done with pulmonary function testing and echocardiography which was within normal limits. A multidisciplinary meeting was called for prior to surgery involving patient and his caregivers, anesthetists, radiologist and surgical team. Surgical approach and expected difficulties and critical events were discussed. A detailed informed consent explaining the risks involved in vernacular language was obtained.

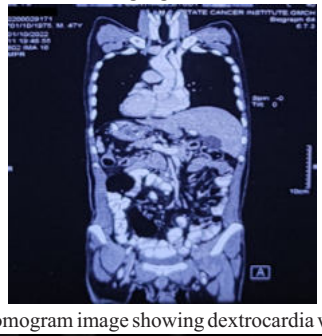

Fig 1 – CECT tomogram image showing dextrocardia with SIT

Fig 2 – Intraoperative image during thoracic esophagus mobilization showing mirror image of normal anatomy with azygous on the left side of esophagus.

Patient underwent left video assisted thoracoscopic McKeown's esophagectomy with standard two field lymphadenectomy in right lateral decubitus position with operating surgeon on left side of the patient. Intraoperatively the azygous vein and esophagus was found on the left side (Fig -2). During the abdominal part of the surgery spleen and greater curvature of the stomach was on the left and liver was on the right side of abdominal cavity (shown in Fig 3). The cervical part of the surgery was approached from the right side and stapled side to side esophago-gastric anastomosis was done using linear cutter GIA stapler. Feeding jejunostomy was done as a feeding procedure. Total operative time was 520 minutes and blood loss were 300ml. Patient was extubated on table and kept under observation intensive care unit. Patient was transfused two units of packed red blood cells. Patient ambulation and tube feeding was started gradually from 1st post-operative day. Chest drain was removed on 4th post-operative day. Nasogastric tube was removed on day 6 and oral trial feeds were started subsequently. Post-operative period was uneventful and the patient was discharged on 10th post-operative day.

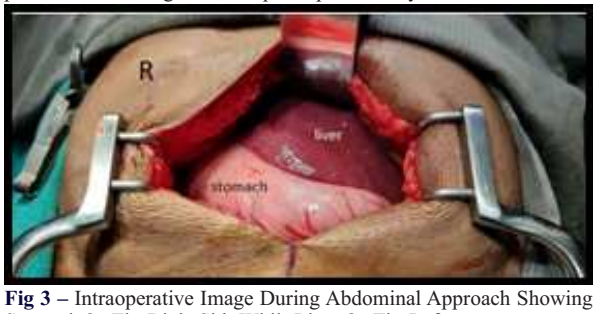

Fig 3 – Intraoperative Image During Abdominal Approach Showing Stomach On The Right Side While Liver On The Left.

Final histopathology report: No residual tumor seen, pathological complete response; TRG (Tumor Regression Grade) :0 (complete response); all margins free of tumor, regional LN (0/39); stage: ypT0N0Mx. Patient kept under routine follow-up every 3 monthly. Patient was seen in out-patient basis after 3 months; he was found to be recovering quite well (shown in Fig 5. a-c).

Fig 4 – Resected specimen showing esophagus and part of lesser curvature of stomach (fundus and cardia), rest of the stomach was fashioned into gastric conduit.

DISCUSSION

Fig 4 Left – anatomical position photograph of patient at 1st follow-up with post-operative incisions, Right – left lateral thoracoscopic port site positions with scars of intercostal tube insertion sites

Situs inversus totalis is a rare genetic condition in which the organs of the chest and abdomen are mirror images of their normal positions. The incidence of situs inversus totalis (SIT) is estimated to be around 1 in 10,000 people. This means that it is a relatively rare condition. However, situs inversus partialis (SIP), a related condition in which only some of the organs are reversed, is thought to be more common.

The exact cause of situs inversus totalis (SIT) is not yet fully understood, but it is believed to be a genetic condition that is inherited in an autosomal recessive pattern. An individual must inherit two copies of the mutated gene, one from each parent, to develop the condition. The mutated gene affects the normal process of organ development during embryonic development, leading to the mirror-image positioning of the organs.

Situs inversus totalis (SIT) can occur as an isolated finding or in association with other genetic conditions like Kartagener syndrome, Primary ciliary dyskinesia, Ivemark syndrome, Ellis-van Creveld syndrome, Heterotaxy syndrome.

The pathogenesis of situs inversus totalis (SIT) is related to abnormal development of the organs during embryonic development. Normally, during embryonic development, the organs form and position themselves in a specific pattern. This pattern is established by the movement of tiny hair-like structures called cilia. The cilia move in a coordinated way, generating fluid flow that helps guide the organs into their proper positions.

The current view among researchers is that organ rotation and migration are the result of a chain of signals. Secretion of a protein named "Sonic hedgehog" (Shh) influences the expression of two transforming growth factors referred to as Nodal and Lefty. When these proteins are secreted on the left side of the embryo, the heart loops to the right, resulting in situs solitus. If the Shh protein is secreted on the right side, the heart loops to the left, resulting in situs inversus. If the Shh protein is secreted on both sides, the signal is unclear; 50 percent of such cases will result in situs solitus and 50 percent will result in situs inversus. However, it remains uncertain what causes the production of the Shh protein [4].

The diagnosis of situs inversus totalis (SIT) usually begins with a physical examination and medical history, as well as imaging tests to confirm the reversed orientation of the organs. During a physical exam, a healthcare provider may check for liver or spleen enlargement, which can also be located on the opposite side in individuals with SIT. A medical history can provide important information about any family history of SIT or related conditions. Imaging tests such as X-rays, CT scans, or MRI scans can provide a visual confirmation of the reversed orientation of the organs. In some cases, genetic testing may also be recommended to confirm a diagnosis of SIT and identify any potential underlying genetic mutations.

There is no established association between situs inversus totalis (SIT) and carcinoma of the esophagus. However, both conditions have their own incidence rates. As previously mentioned, SIT is a rare genetic condition with an incidence of around 1 in 10,000 people. On the other hand, the incidence of carcinoma of the esophagus varies depending on factors such as age, gender, lifestyle, and geographic location. In general, it is more common in men than women and is more frequently diagnosed in older individuals. In some parts of the world, such as China and parts of Africa, esophageal cancer is more common due to factors such as dietary habits and exposure to environmental toxins. The incidence of carcinoma of the esophagus in individuals with SIT is not well established due to the rarity of both conditions.

There have been several case reports of individuals with SIT who have been diagnosed with carcinoma of the esophagus. One such case report described a 54-year-old man with SIT who presented with dysphagia and weight loss. He underwent imaging tests which revealed a mass in the esophagus. A biopsy confirmed the diagnosis of squamous cell carcinoma of the esophagus. Due to the unique anatomy of the patient, careful surgical planning was required to ensure the correct approach and orientation for the surgery [5].

With the advances in surgical techniques, minimal invasive surgeries have been at par with the open surgery in terms of oncological outcomes and survival. Many centers are opting for either Hybrid or total minimal invasive surgeries for carcinoma esophagus in recent

times. Positioning of the patient depends on the surgeon's discretion with some favoring prone over lateral decubitus position.

Here we report a case of carcinoma esophagus with situs inversus totalis successfully managed with hybrid minimal invasive esophagectomy in lateral decubitus position. The surgery was challenging owing to the unfamiliar anatomy of the patient and mirror image of the normal anatomical organs. Delineating the vascular anatomy, recurrent laryngeal nerves, construction and positioning of the neo-esophagus (gastric conduit) was an arduous task. As such, the operative time was a bit longer than usual despite the surgical team rapidly adapting to the abnormal anatomy. The cross-sectional imaging films were an important adjunct in clinical decision making.

CONCLUSION

In the realm of rare anatomical anomalies, the present case of esophageal carcinoma occurring in conjunction with situs inversus totalis stands as a testament to the complexity and diversity of clinical challenges that medical professionals may encounter. With a worldwide reported incidence of only 12 cases, the coexistence of esophageal cancer and situs inversus totalis presents a formidable diagnostic and therapeutic puzzle. Despite the inherent technical intricacies associated, the multidisciplinary approach, meticulous planning, and surgical expertise culminated in a favorable outcome, reaffirming the significance of personalized patient care in challenging scenarios. As medical knowledge continues to expand, sharing experiences and insights from such exceptional cases contributes not only to the collective understanding of rare conditions but also serves as a beacon of hope for patients and medical practitioners alike.

Abbreviations –

SIT – Situs Inversus Totalis, VATS – Video Assisted Thoracoscopic Surgery, ECOG – Eastern Cooperative Oncology Group, RECIST – Response Evaluation Criteria in Solid Tumors, GIA – Gastro-intestinal Anastomosis, TRG – Tumor Regression Grade, LN – Lymph Node, CT – Computed tomography, MRI – Magnetic Resonance Imaging

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