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TRANSHIATAL DISTAL ESOPHAGECTOMY AND TOTAL GASTRECTOMY IN A CASE OF ESOPHAGOGASTRIC TUMOUR WITH SITUS INVERSUS TOTALIS

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ABSTRACT

Situs inversus totalis (SIT) is a rare congenital anomaly with a thoracic cavity, and abdominal structures are reversed. The most suitable approach and surgical technique in a patient with SIT and organ variation remain unclear. We report our experience with a case of situs inversus totalis diagnosed with the esophagogastric tumour. We report a 77 years old female diagnosed with locally advanced esophagogastric tumour classified as Siewert Type II. Preoperative CT scan shows a complete mirror image of the thoracic cavity and abdominal viscera and no vascular anomalies. She underwent distal esophagectomy and total gastrectomy with Roux-en-y anastomosis and D2 lymph node dissection. Distal esophagectomy was done via transhiatal approach for adequate proximal margin resection. There were no intra-operative problems, and the postoperative course was uneventful. Till present, we believe that this is the first case of Transhiatal distal esophagectomy and total gastrectomy in situs inversus totalis patient reported in Malaysia.

Keywords: Esophagectomy, Situs Inversus Totalis, Cancer

CASE REPORT

A 76-year old woman presented with a history of epigastric pain for four months was discovered to have Situs inverses totalis after diagnostic imaging was performed to investigate the cause of his symptoms. No other physical abnormalities were noted. The patient had not been diagnosed with SIT or had developed any specific disease until this point. Endoscopy was performed, and it showed a tumour measured 3.0 × 2.0 cm and classified as Siewert Type II. Histological examination of the biopsied specimen indicates moderately differentiated adenocarcinoma. Perioperative echocardiogram revealed the presence of dextrocardia with no septal and valvular defect. The thoracic and abdominal organs in a right-left reversal position (Figure 1).

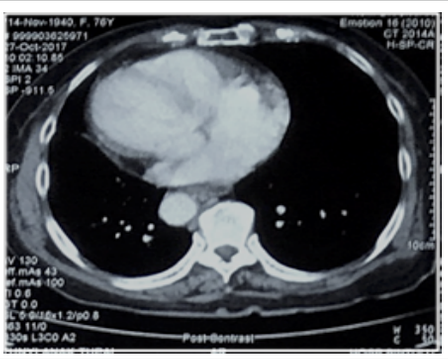

Figure 1: Computed tomography showing dextrocardia.

A computed tomography scan confirmed the 4.2 × 4.7 x 3.9 cm oesophageal lesion with enlarged perigastric nodes (Figure 2a, 2b).

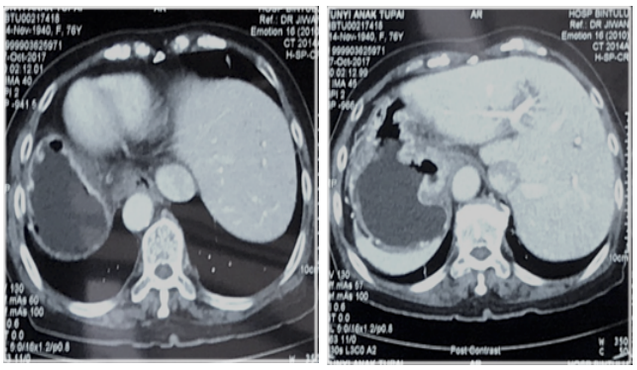

Figure 2: Computed tomography showing a) tumour at the esophagogastric junction (arrow), b) exophytic tumour with enlarged perigastric node.

There was no apparent hilar or mediastinal lymphadenopathy. There were no liver and pulmonary metastasis or visible invasion to the adjacent organs. No concomitant of abnormal vascularisation of the blood vessel nor malformations of the abdominal organs were observed on CT scan. The patient was elected for surgery and resection of the tumour was performed by Transhiatal esophagectomy and total gastrectomy with D2 dissection using an open approach following the Japanese Gastric Cancer Treatment Guidelines.

SURGICAL PROCEDURE

Distal esophagectomy, total gastrectomy with D2 dissection and Roux-en-Y jejunoesophageal anastomosis were performed in the open approach. The patient was under general anesthesia in a semi left-lateral position. The surgeon stands on the right side of the patient as the surgeon that operated is left-handed. The upper midline abdominal incision from the xiphoid process to the umbilicus was made, and abdomen was explored to verify that no metastatic disease and resectability of the tumour. The visceral organ arranges in complete mirror-image transposition. Thompson and Balfour's retractors were applied for better exposure of the hiatus and the abdominal cavity. The greater omentums were resected from the transverse colon. Right gastroepiploic was identified and ligated, and the infrapyloric lymph nodes basin was dissected. The location of the spleen was confirmed on the right side, and the node basin along the short gastric vessels and the left gastroepiploic vessels node basin were dissected. The left gastroepiploic vessels were ligated and divided. After kocherized the duodenum, the proximal portion of the second part of the duodenum divided using a linear stapling device.

We proceed with lymphadenectomy of the group along the lower mediastinum, supra pyloric lymph nodes, common hepatic, coeliac trunk and left gastric artery. Then, the node basins along the proximal splenic artery, right pericardial lymph nodes, left pericardial lymph node, and along the lesser curvature were dissected thoroughly.

The triangular and coronary ligament of the liver was divided. Segment 2 and 3 of the liver retracted to upward and to the left for better exposure of the distal esophagus. The distal esophagus and all periesophageal tissue are then dissect carefully, and the dissection was extended proximally in a circumferential manner. The gastrophrenic ligament is divided, and the dissection carried over the anterior portion of the phrenoesophageal ligament. The crura were incised to provide better exposure of the mediastinum. Before dividing the esophagus, lateral stay suture was placed to secure the esophagus and prevent retraction into the mediastinum. The distal esophagus was divided sharply with 5cm clear proximal margin. A Roux-en-y esophagojejunostomy was performed by stapled anastomosis (CDH). Operating time was 270 min, and estimated blood loss was 200 mL.

OUTCOMES

The final pathology showed a poorly differentiated adenocarcinoma with invasion limited to the subserosal connective tissue (pT3). Three out of 35 lymph nodes were positive for metastatic tumour deposits (pN3). The proximal and distal margins were free from tumour involvement. The final stage was pT3N2M0, stage IIIB according to the American Joint committee on Cancer (AJCC)/ Union for International Cancer Control (UICC) 8th edition. There were no immediate postoperative complications, and the patient was discharged ten days after the operation.

DISCUSSION

Situs inversus totalis (SIT) is a rare congenital anomaly characterised by asymmetrical 'mirror-image' orientation of all organs in relation to the midline. The incidence of SIT is one in 10,000 to 50,000 live births (Fujikawa, 2013). It was suggested that immobility of nodal cilia inhibits the flow of extraembryonic fluid during embryogenesis leading to the development of SIT. Situs inversus can include both the thoracic and abdominal cavities (totalis), or only one cavity (partial). About 60% of patients with SIT have other congenital anomalies of the gastrointestinal tract, including rotational anomalies, biliary atresia, splenic agenesis, small bowel atresia, duplication, and colon aganglionosis (Lee, 2006). It has been suggested that no direct relationship between SIT and gastric cancer (Gündeş, 2018).

Patient with SIT is usually asymptomatic, and it is often found in specific clinical situations. The inverse location of the viscera might lead to misdiagnosis of disease if the SIT is not taken into consideration. Before performing surgery in a patient with SIT, the surgeon must be aware of the presence of vessel variance, and organ location abnormalities because it might be an encumbrance during operation. Preoperative evaluation of cardiopulmonary status is essential to detect any potential severe abnormalities before preceding any major surgery. Preoperative CT angiography is useful for detection of any vascular anomalies in which can be a guide during surgery. With vascular mapping, the surgeon may avoid unnecessary catastrophes intra-operatively.

In total, there are about 4 cases of esophagogastric

junction cancer with SIT reported in English literature but none from Malaysia (Iwamura *et al*. 2001; Morimoto *et al*. 2015, Cao *et al.* 2017 & Singh et al. 2016). All of the patients received curative surgery with two of having open total gastrectomy with D1 dissection. One of the patients received Robot-assisted total gastrectomy with D2 dissection (Cao, 2017) and another patient having minimally invasive Ivor Lewis procedure using left video-assisted thoracoscopic surgery approach (Singh, 2016). All patient was treated successfully, and no significant complication postoperatively was reported. None of the cases that reported have mentioned about distal esophagectomy via transhiatal approach in a patient with SIT diagnosed with the esophagogastric tumour.

In our case, it was an advanced esophagogastric tumour and classified as Siewert type II. We decided to performed transhiatal distal esophagectomy, total gastrectomy with D2 dissection. As our patient was an elderly patient, we want to avoid thoracotomy incision, which thereby minimised pain and subsequent postoperative pulmonary complication. By performing the transhiatal approach also, we able to achieve adequate proximal margin and circumferential resection margin. During lymphadenectomy, extra precaution and advanced skill were needed to perform it precisely as the vascular anatomy in mirror image. However, there was no vascular variant and organ malformation in our patient.

The critical factor in performing surgery in a patient with SIT is knowing the anatomy, mainly anatomic variation and vascular anomalies. Preliminary discussion between the surgeon, first assistant and operating team is necessary to execute a successful operation.

CONCLUSION

Transhiatal distal esophagectomy, total gastrectomy can be performed safely in a patient with SIT diagnosed with esophagogastric junction tumour. These may be the first case reported to date. Radiologic assessment is mandatory to asses any vascular variance and organ anomalies. The surgeon needs to imagine the mirror image during the surgery and must be aware of the anatomy anomalies to accomplish the surgery.

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