CASE REPORT

Gastrectomy for gastric carcinoma with situs inversus totalis: case report and literature review

Zhu H[1], Yang K[1,2], Hu JK[1,2]

[1]Department of Gastrointestinal Surgery
[2]Laboratory of Gastric cancer, State Key Laboratory of Biotherapy
West China Hospital, Sichuan University, Chengdu, China

Abstract

Background: Situs inversus totalis (SIT), a rare congenital anomaly, is characterized by a complete mirror image transposition of the thoracic and abdominal viscera. We report the case of a 66-year-old woman with SIT who was diagnosed with gastric antral carcinoma. Curative distal gastrectomy with Billroth-I anastomosis was performed.

Description of the case: A 66-year-old woman visited our outpatient department complaining of abdominal pain in the left upper quadrant for about one year. Physical examination revealed that the apex beat was in the right fifth intercostal space, just at the midclavicular line while a soft systolic murmur was audible at the upper right sternal border. The abdominal examination was unremarkable. The preoperative diagnosis was confirmed by gastroscopy and biopsy. Preoperative echocardiogram revealed the presence of dextrocardia and atrial septal defect. Preoperative contrasted computed tomography showed a complete right-left reversal of the thoracic and abdominal organs and thickened wall of gastric antrum without distant metastasis. Laparotomy through a midline incision confirmed the complete mirror-image transposition of the abdominal visceral organs and a 4-cm tumor with serosal involvement at the gastric antrum. Curative distal gastrectomy with D2 lymphadenectomy and Billroth-I anastomosis was performed. The patient had a rapid recovery and was discharged without any complications. The final staging of this case was pT4aN1M0, stage IIIa and she received chemotherapy with the SOX regimen for three cycles. Fifteen months after the operation, the patient is alive without any signs of recurrence.

Conclusions: The incidence of gastric cancer with SIT is very rare. Appropriate diagnostic modalities are very helpful for the diagnosis and preoperative planning. Gastrectomy with D2 lymphadenectomy in patients with SIT can be performed successfully with sufficient preoperative evaluation, comprehensive knowledge of anatomy, and meticulous surgical manipulation. Caution should be given to the possibility of coexisting cardiopulmonary malformations and synchronous cancers. Hippokratia 2015; 19 (4): 360-362.

Keywords: Gastric cancer, situs inversus totalis, operation, malformation

Corresponding authors: Kun Yang (e-mail: yangkun068@163.com) and Jian-Kun Hu (e-mail: hujkwch@126.com), Department of Gastrointestinal Surgery, and Laboratory of Gastric cancer, State Key Laboratory of Biotherapy, West China Hospital, Sichuan University, No. 37 Guo Xue Xiang Street, Chengdu 610041, Sichuan Province, China, tel: +862885422878, fax: +862885164047.

Introduction

Situs inversus totalis (SIT) is a congenital condition with complete mirror image transpositions of the thoracic and abdominal viscera. The overall incidence of SIT is reported from 1:4,000 to 1:8,000[1]. It seems that SIT does not influence normal health or life expectancy. Although there is no definite relationship between SIT and malignant tumors[2], several malignant neoplasms have been reported in association with SIT[1]. We report a case of gastric carcinoma in a patient with SIT. We describe this case because of the rarity of this anomaly in gastric cancer patients; furthermore, familiarity with this condition could help surgeons to recognize and avoid failures in diagnosis and treatment.

Case report

A 66-year-old woman visited our outpatient department complaining of abdominal pain in the left upper quadrant for about one year. Physical examination revealed a normal heart rate and blood pressure. However, the apex beat was in the right fifth intercostal space, just at the midclavicular line. Also, a soft systolic murmur was audible at the upper right sternal border. The respiratory system examination was unremarkable. Physical examination showed no abdominal mass, no enlargement of Virchow lymph nodes, and no abdominal tenderness. The liver and spleen were not palpable neither any other mass in the abdominal examination. The digital rectal examination did not find any abnormalities. Gastroscopy revealed a deep ulcer with an irregular raised margin located on the greater curvature of the gastric antrum. Histological examination of the biopsied specimen indicated poorly-differentiated adenocarcinoma. Preoperative echocardiogram revealed the presence of dextrocardia and atrial

septal defect. Preoperative thoraco-abdomino-plevic general three-dimensional (3D) reconstructed contrasted computed tomography (CT) showed a complete right-left reversal of the thoracic and abdominal organs and thickened wall of gastric antrum (Figure 1A). There were no obviously enlarged lymph nodes in the CT images. There were no ascites, liver metastasis, pulmonary metastasis or visible invasion into adjacent organs. Also, no concomitant malformations of the abdominal organs nor any abnormal vascularization of the blood vessels were observed on CT scan. Laparotomy through a midline incision confirmed the complete mirror-image transposition of the abdominal visceral organs and a 4-cm tumor with serosal involvement at the gastric antrum. No peritoneal seeding and liver metastasis were found. No abnormality in vascular anatomy was found. Curative distal gastrectomy with D2 lymphadenectomy and Billroth-I anastomosis was performed (Figure 1B). Gross inspection of the specimen showed a malignant ulcerative appearance. The patient had a rapid recovery and was discharged without any complications. Finally, pathological examination confirmed the diagnosis of poorly differentiated gastric adenocarcinoma with serosa involvement. A total of 33 lymph nodes were harvested, of which two were metastatic (pT4aN1M0, stage IIIa). She received chemotherapy with the SOX regimen (oxaliplatin 150 mg/d day1 and S-1 75mg/d days 1-14, every three weeks) only for three cycles. Fifteen months after the operation, the patient is alive without any signs of recurrence.

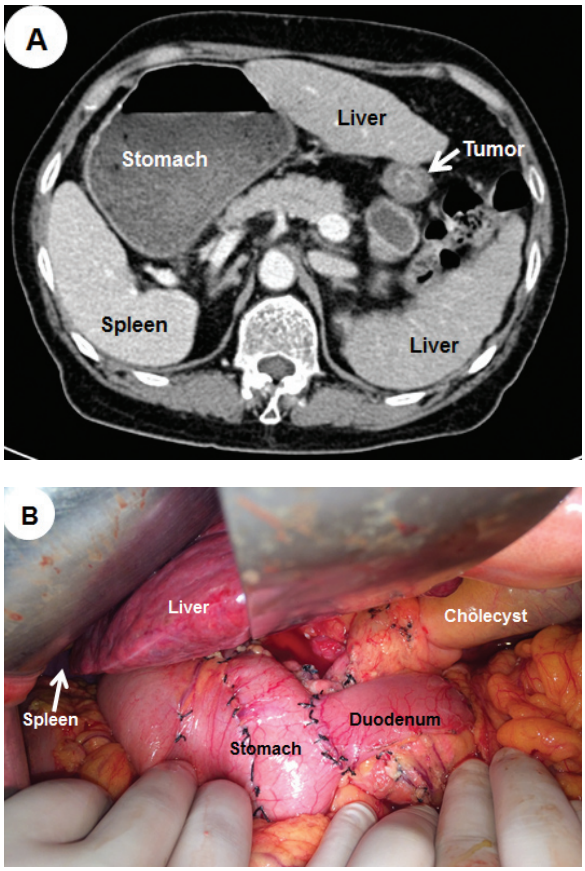

Figure 1: Abdominal computed tomography (CT) and laparotomy revealed situs inversus totalis. A) CT revealed a complete right-left reversal of the abdominal organs and thickened wall of gastric antrum without distant metastasis; B) Laparotomy confirmed the complete mirror-image transposition of the abdominal visceral organs and curative distal gastrectomy with Billroth-I anastomosis was carried out.

Discussion

We report this case due to the rarity of SIT. Therefore, the accumulation of more cases is important to understand better the characteristics and risks of this anomaly, which may facilitate patients' treatment and prevent the avoidable faults from surgery or other interventions, particularly in the emergency setting. Moreover, although there are several reports of gastrectomy in patients with SIT, most of them described the technical difficulties resulting from the unusual anatomy. Herein, we focused on the experience and tips learned from the reported case.

SIT is characterized by a left-to-right reversal of the abdominal viscera with dextrocardia, which is in contrast with situs inversus viscerum which means a complete mirror-image transposition of the abdominal visceral organs with normal orientation of the thoracic organs[2]. SIT could be accompanied by several abnormalities, such as cardiac malformation, long QT syndrome, bronchiectasis, rhinosinusitis, polysplenia, or some urologic anomalies[3]. In the reported case, the patient has an atrial septal defect.

The exact etiology of SIT remains unclear. It is thought that SIT has a genetic predisposition to autosomal recessive inheritance abnormalities which could lead to inhibition of extra-embryonic fluid flow by immobility of nodal cilia during the embryonic period and consequently cause the development of situs inversus[4]. Recently, it was proved that the KIF3 complex (an intracellular motor protein) and cell adhesion factors (including N-cadherin and β-catenin) were involved in the development of situs inversus, and the growth as well as the progression of synchronous cancer[5].

In general, SIT is detected or diagnosed incidentally during thoracic and abdominal imaging tests, such as CT, magnetic resonance imaging or barium meals, etc. SIT is frequently associated with concomitant malformations of abdominal viscera (eg. polysplenia, or some urologic anomalies) and often with abnormal vascularization of the blood vessels. Therefore, careful preoperative planning and preoperative anatomic assessment of vascularization are crucial for patients undergoing surgery[6], which would prevent misidentifying anatomy and avoid unexpected injury of important vessels. Advanced diagnostic imaging techniques, such as 3D-CT or angiography might be helpful to demonstrate the structures of blood vessels and organs preoperatively[7]. It has been reported that 3D reconstruction of abdominal CT angiography was an optimal non-invasive modality to maximize the precise assessment of vascular anatomy[8]. Although the 3D reconstruction of CT angiography was not performed in the present case, the images of general

3D reconstructed CT scan were sufficient for evaluation by the experienced radiologist and surgeons. In this case, the preoperative contrast CT revealed no malformations among the abdominal organs and no abnormal vascularization. Also, the findings of the operative exploration confirmed this preoperative evaluation. Since general 3D reconstructed CT scan is both accurate and easily available even in small district hospitals, it could be the preferred modality for preoperative planning and anatomic assessment compared with other imaging modalities. We should take note that in this case, the pain of the antral lesion was located in the left upper quadrant, opposite the usual location. Therefore, we should keep in mind that the symptoms of diseased organs may be manifested at opposite locations in patients diagnosed with SIT. In addition, more attention needs to be paid in emergency conditions. For instance, the pain of acute appendicitis appears at left lower quadrant[9].

Surgical intervention for patients with SIT is technically more complex, due to the anatomical abnormalities. Some surgeons recommend exchanging the positions of operator and assistant during surgery[6], which was believed to facilitate the surgical team's adaptation to the mirror image of the standard procedure and help avoid intraoperative complications. However, we performed the operation with our usual setup and positioning. With detailed preoperative assessment and careful manipulation, abdominal organs and vessels were detectable without difficulty, and an uneventful operation without major bleeding was performed in our case. In fact, there were no anatomical problems when we performed gastrectomy for this patient. There was also no intraoperative complications encountered. Therefore, we infer that sufficient preoperative evaluation, comprehensive knowledge of anatomy, and meticulous surgical manipulation would minimize the obstacles associated with SIT. Although more difficult than ordinary cases, laparoscopic or robotic gastrectomy for patients with SIT were also reported to be safe and feasible approaches[6,10-12]. Because of the high possibility of accompanying cardiopulmonary malformation, strict postoperative monitoring and management are mandatory.

Although the incidence of SIT is low and it is not considered to be premalignant, more than 60 cases of malignancies with SIT have already been reported. Therefore, we consider it necessary to perform tumor and organic malformation screenings simultaneously in patients with SIT. Synchronous cancers in patients with SIT were also reported[1,13]. We should be mindful of the possibility of a concurrent tumor in patients with SIT who have been diagnosed with one kind of cancer disease.

Conclusion

The incidence of gastric cancer with SIT is very rare. Appropriate diagnostic modalities are very helpful for diagnosis and preoperative planning. Gastrectomy with D2 lymphadenectomy in patients with SIT can be performed successfully with sufficient preoperative evaluation, comprehensive knowledge of anatomy, and meticulous surgical manipulation. Caution should be given to the possibility of coexisting cardiopulmonary malformations and synchronous cancers.

Conflicts of interest

The authors declared no conflicts of interest.

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