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**Name of Journal:** *World Journal of Gastrointestinal Oncology*

**Manuscript NO:** 76483

**Manuscript Type:** CASE REPORT

**Da Vinci robot-assisted pancreato-duodenectomy <sup>5</sup> in a patient with situs inversus totalis : A case report and literature review**

A **case** report and literature review

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## **Abstract**

### **BACKGROUND**

Situs inversus totalis (SIT) is an extremely rare congenital malformation characterized by mirror displacement of the thoracoabdominal organs such as the heart, liver, spleen, and stomach. Herein, we describe a patient with situs inversus totalis (SIT) complicated with cholangiocarcinoma who underwent successful pancreaticoduodenectomy with the assistance of a da Vinci robot.

### **CASE SUMMARY**

A 58-year-old female presented to the hospital with paroxysmal pain in her left upper abdomen, accompanied by jaundice and staining of the sclera as chief complaints. Imaging examination detected a mass at the distal end of the common bile duct, with inverted thoracic and abdominal organs. Endoscopic retrograde cholangiopancreatography (ERCP) forceps biopsy revealed the presence of a well-differentiated adenocarcinoma. The patient successfully underwent robotic-assisted pancreaticoduodenectomy; the operation lasted 300 minutes; intraoperative blood loss was 500 mL; there were no intraoperative and postoperative complications.

### **CONCLUSION**

Situs inversus totalis is not directly related to the formation of cholangiocarcinoma. Detailed preoperative imaging examination is conducive to disease diagnosis and also convenient for determining the feasibility of tumor resection. Robot-assisted pancreaticoduodenectomy for SIT complicated with cholangiocarcinoma provides a safe, feasible, minimally invasive, and complication-free alternative with adequate preoperative planning combined with meticulous intraoperative procedures.

**Key Words:** Situs inversus totalis; Cholangiocarcinoma; Da Vinci robot; Whipple; Surgery; Case report.

Li B, Lu S, He X, Lei B, Yao J, Feng S, Yu S. Da Vinci robot-assisted pancreatoduodenectomy in a patient with situs inversus totalis : A case report and literature review. *World J Gastrointest Oncol* 2022; In press

**Core Tip:** Situs inversus totalis (SIT) is an extremely rare congenital malformation characterized by a mirror image displacement of the thoracoabdominal organs such as the heart, liver, spleen and stomach. SIT combined with choledochal cancer is even rarer, and da Vinci robot-assisted pancreaticoduodenectomy in patients with SIT combined with choledochal cancer has not been reported. This case demonstrates that preoperative thorough planning, intraoperative precise anatomical knowledge, effective teamwork, meticulous treatment, and postoperative care are feasible with the aid of the da Vinci robot for pancreaticoduodenectomy in patients with SIT.

## **INTRODUCTION**

Situs inversus totalis (SIT) is characterized by complete mirror displacement of the anatomical positions of organs in the thoracoabdominal cavity. It has been reported that its incidence is 1/8000 ~ 1/25000 with a male to female ratio of 3:2 [1-3]. Although the exact cause of SIT remains unknown, it is generally accepted that it is caused by autosomal abnormalities [4]. In addition, some mutations, such as DNAH11 and Nme7, have been shown to be closely related to SIT [5, 6]. Patients with SIT typically exhibit no obvious symptoms, and their health and life expectancy appear to be unaffected [7]. Cholangiocarcinoma is a rare malignant tumor originating from bile duct epithelial cells, accounting for merely 3% of digestive tract malignant tumors [8]. It has an insidious onset, a high degree of malignancy, and a poor prognosis [9]. The Da Vinci surgical robot system appeared at the end of the 20th century, and the fourth generation of the robot was launched in 2014, with qualitative improvements in flexibility, accuracy, and imaging clarity. Notably, a remote observation and guidance system was developed in the second half of 2014 [10, 11]. According to reports, the da Vinci robot has evolved into its fifth generation [12]. To date, several cases of complete visceral

choledochal carcinoma have been reported [7, 13-18], but da Vinci robot-assisted pancreatoduodenectomy in a patient with SIT has not been reported so far. This is the case report of a patient with SIT complicated with cholangiocarcinoma who underwent pancreaticoduodenectomy assisted by a da Vinci surgical robot. We detailed the diagnosis and management of this case, as well as a thorough evaluation of the associated literature.

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## **CASE PRESENTATION**

### ***Chief complaints***

A 58-year-old female patient presented to our hospital in November 2021 with jaundice, yellow sclera, paroxysmal pain in the left upper abdomen, itching, fatigue, tea-like urine, and weight loss of 1.5 kg.

### ***History of present illness***

The patient developed symptoms 2 mo ago and recurrent upper left abdominal pain.

### ***History of past illness***

The patient underwent surgery for cholecystolithiasis 10 years ago.

### ***Personal and family history***

The patient had no similar family history or genetic history of the disease.

### ***Physical examination***

Physical examination on admission revealed yellow discoloration of the skin and sclera, no chest deformity, and cardiac sound in the right chest. The abdomen was soft, while the upper abdomen was tender. There was no rebound tenderness or muscle tension. The liver and spleen were not palpable under the rib edge, and the activities of both lower limbs were unremarkable.

### *Laboratory examinations*

Preoperative auxiliary examinations were as follows: Total bilirubin (TBIL) 126.2 $\mu$ mol/L(normal range:3.4-20.5 $\mu$ mol/L);Direct bilirubin (DBIL) 97.2 $\mu$ mol/L(normal range: 0-6.8 $\mu$ mol/L); Indirect bilirubin (IBIL) 29.2 $\mu$ mol/L(normal range: 3.1-14.3 $\mu$ mol/L); Aspartate aminotransferase (AST) 19.0 U/L(normal range:13-40U/L);Alanine aminotransferase(ALT)28.0U/L(normal range: 7-45U/L);Alkaline phosphatase (ALP) 255 U/L(normal range: 50-135U/L); Abnormal prothrombin (DCP)76.60ng/mL(normal range: 0-40ng/mL);Carbohydrate antigen 19-9 (CA19-9) 197.96 U/mL(normal range: 0-37.00ng/m). Other blood biochemical indexes were within the normal range.

### *Imaging examinations*

The chest X-ray exposed that the right heart was mirrored (**Figure 1A**). Multi-slice spiral CT (MSCT) revealed complete transposition of the thoracic and abdominal viscera, as well as a tumor in the lower segment of the common bile duct, with minimal intestinal obstruction. The intrahepatic bile duct, extrahepatic bile duct, and pancreatic duct were dilated, and cholecystolithiasis with chronic cholecystitis was diagnosed (**Figure 1B, C**). Endoscopic retrograde cholangiopancreatography (ERCP) was performed, and the tissue was taken for pathological biopsy; the results showed a well-differentiated adenocarcinoma. Magnetic resonance imaging (MRI) showed intraluminal growth of cholangiocarcinoma in the lower part of the common bile duct with dilatation of the bile duct and pancreatic duct.

### *Further diagnostic work-up*

Postoperative histological results were as follows: 1. Well-differentiated adenocarcinoma of the common bile duct with invasion of the duodenal wall. No vascular tumor thrombus and nerve invasion were identified. 2. There were no signs of malignancy in the gastric, duodenal, and pancreatic margins. 3. Each of the 6 Lymph nodes manifested reactive hyperplasia. 4. Cholecystolithiasis with chronic

cholecystitis.<sup>5</sup> Immunohistochemical staining was positive for CDX-2, CK7 and CK19 (Figures 2A, B, C, D ,E).

### **Abbreviations**

SIT : Situs inversus totalis ;<sup>7</sup> ERCP: Endoscopic retrograde cholangiopancreatography;<sup>18</sup> TBIL: Total bilirubin; DBIL: Direct bilirubin; IBIL: Indirect bilirubin; AST: Aspartate aminotransferase; ALT: Alanine aminotransferase; ALP: Alkaline phosphatase; DCP: Abnormal prothrombin; CA19-9: Carbohydrate antigen 19-9; MSCT: Multi-slice spiral CT; MRI: Magnetic resonance imaging;<sup>10</sup> RGA: right gastric artery; LHA: left hepatic artery; RHA: right hepatic artery; EUS: endoscopic ultrasound;

### **FINAL DIAGNOSIS**

<sup>13</sup> According to the American Joint Committee on Cancer/International Alliance for Cancer Control (AJCC/UICC) TNM staging system, the patient's tumor stage was IIB stage<sup>[19]</sup>. The pathological stage of the patient was pT3N0M0.

### **TREATMENT**

Prior to surgery, the anesthesiologist assessed the patient's perioperative risk score.<sup>8</sup> According to the American Society of Anesthesiologists Physical Status classification system (ASA PS)<sup>[20]</sup>, the ASA score of the patient was P2.

The detailed procedure of the operation is as follows:

1. The surgery was prepared in accordance with the specifications of the Da Vinci System. The layout of the operation window and the instrument arm are illustrated in **Figure 3**.

2. Upon entering the abdominal cavity, it was observed that the abdominal organs were fully inverted (**Figure 4A**).

3. The gallbladder was anterogradely dissected while the cystic duct was retained. Then, the common hepatic artery was exposed (**Figure 4B**). Following this, the<sup>12</sup> right gastric artery (RGA), the left hepatic artery (LHA), and the right hepatic artery (RHA)



were exposed. Next, the suprapyloric lymph node (NO.5), anterosuperior lymph node of common hepatic artery(NO.8a) and hepatoduodenal ligament lymph nodes (along the hepatic artery) (NO. 12a) were dissected, and the portal vein and its left and right branches were exposed. Afterward, hepatoduodenal ligament lymph nodes (along the portal vein) (NO. 12p) were dissected, the common hepatic duct and common bile duct were exposed, and hepatoduodenal ligament lymph nodes (along the bile duct) (NO. 12b) were dissected (**Figure 4C**). Finally, the hepatic portal vein was skeletonized, and the common bile duct was excised.

4. The gastroduodenal and hepatogastric ligaments were dissected. Thereafter, the right blood vessels of the gastric reticulum were clamped and cut off, and the subpyloric lymph nodes (NO.6) were dissected. A Kocher incision was made to expose the inferior vena cava (**Figure 4D**). Most of the pancreatic head covered the descending and horizontal part of the duodenum, making it difficult to expose the superior mesenteric artery (**Figure 4E**). The superior mesenteric vein was subsequently exposed along the lower margin of the pancreas, following which the posterior space of the pancreas was separated. Next, the pancreas was cut off, and the location of the pancreatic duct was determined (**Figure 4F**). The superior mesenteric artery was protected under direct vision. The uncinate process was subsequently cut off along the right side of the superior mesenteric vein. Lastly, the pancreaticoduodenum was resected (**Figure 4G**).

5. Pancreaticojejunostomy and choledochojejunostomy were performed. The specimens were removed from the abdominal cavity *via* a median 8 cm incision in the upper abdomen, and gastrojejunostomy was executed through this incision.

6. A total of 3 drainage tubes were placed in Wen's foramen, behind the pancreaticojejunostomy, and in the pelvic cavity. The operation lasted 300 min, and the intraoperative bleeding was only 500 mL.

### **OUTCOME AND FOLLOW-UP**

There were no severe complications during or after the operation. The patient stayed in the hospital for a total of 30 days and was discharged 22 days following surgery. At the



last follow-up (March 15, 2022), there was no evidence of recurrence, and tumor markers had fallen within the normal range. After treatment, he lived completely independently and is still alive.

## **DISCUSSION**

At present, it is generally acknowledged that SIT with malignancies is a rare coincidence, although SIT with gastric cancer, rectal cancer, pancreatic cancer, liver cancer, and other tumors have been reported [21-24]. Indeed, only 8 cases of SIT complicated with choledochal carcinoma have been reported so far [7, 13-18]. The clinicopathological features of the 8 previously reported cases and our case are summarized in Table 1. The cohort consisted of 6 men and 3 women, ranging from 33 to 74 years, including 3 from Japan, 2 from China, 2 from the United States, 1 from Italy, and 1 from Morocco. 8 out of the 9 patients underwent the Whipple procedure, and 1 patient underwent choledochectomy and Roux-en-Y hepaticojejunostomy. Histopathological results determined that there were 4 cases of well-differentiated adenocarcinoma, 1 case of medium-differentiated adenocarcinoma, 1 case of poorly-differentiated adenocarcinoma, and 1 case of high-grade intraepithelial neoplasia.

Most of the early clinical manifestations of cholangiocarcinoma are atypical, and patients seek medical treatment owing to epigastric discomfort, abdominal pain, abdominal distension, loss of appetite, and other symptoms [25]. A minority of patients with cholangiocarcinoma caused by the tumor may develop symptoms of typical obstructive jaundice with severe colic [26]. However, SIT generally lacks specific clinical manifestations, considering it is frequently accompanied by other diseases or discovered during physical examination [27]. By reviewing the literature, it was noted that the reported clinical manifestations of SIT complicated with cholangiocarcinoma include jaundice, abdominal pain, weight loss, and other symptoms. Among the cohort, 5 cases developed jaundice, as was the case in our patient. Thus, jaundice can be considered the primary clinical manifestation of SIT complicated with cholangiocarcinoma. Earlier studies have reported that multi-slice spiral

CT(MSCT),MRI, endoscopic ultrasound (EUS), and endoscopic retrograde cholangiopancreatography (ERCP) are of tremendous value in the diagnosis and preoperative evaluation of SIT complicated with cholangiocarcinoma<sup>[7, 18]</sup>.

The following points need to be considered when performing robot-assisted pancreaticoduodenectomy in SIT patients. Compared to healthy individuals, the gall bladder, bile duct, and gastrointestinal tract form a symmetric structure in SIT patients. Therefore, surgeons must be accustomed to preoperative images of SIT patients to avoid prolonged operation time due to unfamiliarity with the anatomy of SIT patients. Additionally, we uncovered for the first time that the thoracic and abdominal organs of SIT patients with cholangiocarcinoma have not only left and right reverse positions but also anterior and posterior reverse positions. During the operation, the pancreatic head of the patient covered the descending and horizontal parts of the duodenum, which was inverted from its normal anatomical structure. Considering the atypical anatomy of this patient, there was no reference for the preoperative procedure. In addition to the conventional operation with three mechanical arms, two auxiliary operation holes were used to improve operability and safety. Due to the atypical anatomy of SIT, routine thinking should be avoided during the operation, and the operation should be initiated with familiarity and a clear anatomical position to avoid accidental injury. During the operation, with the help of da Vinci robot 3D and highly clear magnified view, using the method of small steps to dissect and separate, for the bleeding or blurred parts of the visual field, different from normal anatomy, try to avoid blind anatomical separation. For example, when the KOCHER technique is used to dissociate the descending part of the duodenum and other parts, the anatomy is not forced to follow the conventional technique, but in the da Vinci robot high-definition field of vision, from shallow to deep, rapid, initial resection of tiny tissue, abnormal bleeding and other cases in a timely and effective manner. The humanoid wrist function of the Da Vinci robot also highlights its superiority to laparoscopic surgeries in narrow spaces and unconventional suture operation, as well as the effect of open surgery. The patient recovered well following surgery, without developing complications such as biliary

fistula, pancreatic leakage, gastric emptying dysfunction, pseudocolicostheresis deficits and airway obstruction [28-31]. Furthermore, incision healing was also significantly better than open surgery.

## **CONCLUSION**

Situs inversus totalis (SIT) is a rare genetic disease and da Vinci robotic surgery is the current poster child for minimally invasive surgery. We believe that with thorough preoperative planning, precise intraoperative anatomical knowledge, effective teamwork, meticulous treatment, and postoperative care, da Vinci robotic-assisted pancreaticoduodenectomy in patients with SIT is feasible and developmental.

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## **ACKNOWLEDGEMENTS**

I would like to express my gratitude to all those who helped me during the writing of this thesis and their efforts in the management of this patient.

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