Laparoscopic Cholecystectomy in Situs Inversus Totalis: A Case Report

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Introduction

Situs inversus totalis (SIT) is a rare congenital abnormality, where there is transposition of both the thoracic and the abdominal organs, thus forming a left-right asymmetrical picture of the normal anatomical placement of the organs.[1] It is kept under an umbrella term, heterotaxia, which encompasses all congenital disorders associated with malposition about the left-right axis.[2] An important distinction is from simply situs inversus, where only some of the major abdominal and thoracic organs are reversed from their normal anatomical positions.[3] SIT is even more of a rare condition, with an incidence of 1:10,000, and males being affected more than females (1.5:1).[1] Due to this anatomical asymmetry, it may pose difficulties during diagnostic and therapeutic procedures.[1,3]

Situs inversus totalis appears to be familial and laterality is established early on in the developmental process, during gastrulation. A protein, namely "Sonic Hedgehog" (Shh) is deemed to be responsible for the expression of two growth factors, Nodal and Lefty. If these proteins are released on the right side, the rotation of the heart occurs to the left side. However, if they are released on the left side, rotation occurs to the right. A gene named PITX2 has been identified, which controls the secretion of Shh and Nodal.[4] The exact mechanism behind this is yet unknown, however, it has been speculated that the mutation in the PITX2 and Nodal gene is responsible and is also associated with other conditions like primary ciliary dyskinesia (PCD) and Kartagener syndrome.[1,3,4,5]

Due to the unique transposition of the anatomical structures in SIT, the usual causes of abdominal pain may be a misdiagnosis. Similarly, these patients with cholelithiasis may have a presentation, with left upper quadrant pain, instead of the usual right, likely posing a diagnostic dilemma.[6,7] Operative management in SIT patients also presents with technical and ergonomic challenges, especially for right-handed surgeons.[8] The mirror image of the anatomy causes difficulty in the dissection of the Calot's triangle, thus requiring appropriate tweaks to the normal preoperative and intraoperative setting.[8] This report describes a case of a SIT patient with symptomatic cholelithiasis, managed by laparoscopic cholecystectomy.

Case History/Examination

A 30-year-old female, presented with colicky pain abdomen, located over the epigastric and the left hypochondrium regions for the last 4 months. The pain was intermittent in nature, non-radiating, and mild in severity. She complained of an increase in pain after the consumption of fatty food. She gave history of occasional nausea without vomiting. There was no history of fever, jaundice, alteration in bowel habits, and recent significant weight loss. She was diagnosed with SIT, whilst a medical check that was required for a job application. She underwent lower section cesarean section 3 years back. There was no other significant medical or surgical history. Her family history was also non-significant. On examination, her general condition was

fair, afebrile, and with no icterus. Examination of the abdomen revealed mild tenderness on palpation over the left with no guarding. Normal heart sounds were heard over the right side of the thorax.

Methods (Differential Diagnosis, Investigations and Treatment)

With the possible differential diagnosis of gastritis and biliary colic, the following investigations were done. Her laboratory investigations showed normal white blood cell counts of 9800 cells per cubic millimeters and liver function test was also found to be normal. Chest X-Ray (Figure 1) revealed dextrocardia with gastric bubble on the right side and liver on the left. Ultrasonography of the abdomen showed multiple echogenic foci of 3 to 7 mm in size in the gallbladder and common bile duct (CBD) of 6 millimeters. She was diagnosed with symptomatic cholelithiasis and planned for laparoscopic cholecystectomy. Routine preoperative workup was done and was found to be normal

The operating theatre was prepared such that the surgeon and camera assistant were in right side, and, assistant surgeon and the laparoscopic tower in left side of the patient (Figure 2A). The surgeon in this case was right-handed. The patient was kept in supine position with left side slightly elevated. Four ports were created. The first port of 10mm positioned supraumbilically was created, followed by the creation of pneumoperitoneum using carbon dioxide gas. After which, a 10 mm port in epigastric region and two 5 mm ports were positioned in the left mid-clavicular line and left anterior axillary line respectively (Figure 2B).

Diagnostic laparoscopy was performed and the laterality was visualized (Fig 3). The fundus of the gallbladder was then retracted towards the right shoulder and the retraction of the Hartmann's pouch was done to the left by the assistant. The posterior dissection was then performed for the clearance of the cystic duct and artery. This manoeuvre prevented the crossing of hands of the surgeon. Thus, the Calot's triangle was identified and dissection of the fibrofatty tissue was done, to achieve the critical view of safety, such that only the cystic artery and duct was seen to be entering the gallbladder. The gallbladder was then slightly separated from its attachment to the liver to expose a part of the cystic plate, following which the cystic artery and the duct were clipped and divided. The gallbladder was then dissected off its liver bed and removed through the umbilical port. On opening of the retrieved gallbladder, two calculi were found inside, with its walls mildly inflamed. This was then then sent for histopathological examination. The 10mm ports were closed using polygalactin 1-0. The histopathological report concluded the diagnosis to be chronic calculous cholecystitis.

Conclusion and Results

SIT is a rare condition, but for symptomatic cholelithiasis in this subset of patients, laparoscopic cholecystectomy is a rather standard procedure. However, special care must be taken while positioning the ports and placement of the surgeon and the assistants, due to ergonomic and technical challenges surgeons face, especially right-handed ones. We presented a case where a right-handed surgeon was situated in the right side of the patient, with the assistant retracting the gallbladder while the primary surgeon dissected. This prevented the crossing of the hands of the surgeon. Her post-operative period was uneventful and she was discharged on the second post-operative day. Follow up on the seventh day showed a healthy surgical site. She was doing well at one year follow up, with no recurrence of biliary symptoms or signs of infection.

Discussion

SIT is a rare condition characterized by the complete transposition of both the thoracic and the abdominal viscera along the left-right axis, thus forming a mirror image of the normal anatomical picture.[1] SIT alone is asymptomatic, except for the accompaniment of cardiac anomalies associated with dextrocardia, such as atrial situs solitus, discordant atrioventricular (AV) connection, and to a lesser extent congenitally corrected transposition of great arteries (TGA).[6,9] Situs inversus is closely associated with a condition known as PCD. Fifty percent of the patients with PCD are associated with situs inversus, and together is known as Kartagener's Syndrome. [10,11] However, there isn't enough evidence that establish the association between SIT and gallbladder disease. [6,12]

While the diagnosis of symptomatic cholelithiasis in patients with SIT is challenging, since our patient was diagnosed with it on routine health checkup prior to the development of illness, no particular challenge

was faced in this case. The first successful laparoscopic cholecystectomy in a patient with situs inversus was performed in 1991 by Campos and Sipes.[13] In a similar report, Arya et al. performed the procedure by placing the ports and the instruments in a mirror-image fashion to the conventional laparoscopic cholecystectomy. The primary surgeon performed the dissection, while the first assistant retracted the Hartmann's pouch throughout the surgery, thus preventing the crossing of the hands of the right-handed surgeon.[14] However, Alam and Santra reported conduction of the procedure with the first assistant holding the camera through the infraumbilical port and the second assistant retracting the Hartmann's pouch.[8]

Other procedures have also been reported with several modifications. One of them being adopting the Llyod-Davis Position, with the primary surgeon standing in between the abducted legs of the patient.[14,15] Cases of Single incision Laparoscopic Surgery (SILS) have also been reported, with the aim of reducing invasiveness, improving cosmesis, and avoidance of the crossing of the instruments.[16,17]

Key Clinical Message

In patients with situs inversus totalis, the main challenge lies in the diagnosis itself. The surgical treatment for cholelithiasis in these patients can be a standard procedure, if it is well planned.

Ethics Statement

This case report involves one patient, so IRC approval is not required.

Conflict of Interest

The authors declare no conflict of interest.

Author Contributions

P.L: Wrote the whole manuscript and reviewed the literature; M.S: was involved in the management of the patient and was responsible for data curation; R.S: was involved in the management of the patient and reviewed the manuscript; P.J.L: was involved in the management of the patient, supervised the process of manuscript writing and reviewed the final manuscript.

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Figure Legends

1. Figure 1: Chest X-ray showing dextrocardia, right-sided gastric bubble, and shadow of the liver on the left.
2. Figure 2A: The operative setup demonstrating the primary surgeon present on the right side of the patient.
3. Figure 2B: Incision sites for trocar placement.
4. Figure 3: Laparoscopic picture showing the gallbladder placed towards the left side of the falciform ligament.