Laparoscopic Cholecystectomy in Situs Inversus Totalis with Post-ERCP Pancreatitis & Masri-Larkin Syndrome: A Novel Diagnosis

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Introduction

Situs viscerum inversus (SVI) is a rare autosomal recessive condition in which the organs are transposed from their normal location to the opposite side of the body. It is found in one of every 10,000 to 20,000 births. There are two types of SVI, which include situs inversus totalis (SIT), where both thoracic and abdominal organs are reversed, and the more rare type, situs inversus partialis (SIP), where the reversal is only partial. Retroperitoneal dysplasia, biliary atresia, congenital heart disease, and pancreatic fibrosis are some of the congenital anomalies that can be associated. If SIT is associated with bronchitis, chronic sinusitis, and deficient tracheobronchial cilia, it is known as Kartagener’s Syndrome.

Laparoscopic Cholecystectomy (LC) has become the gold standard for surgical therapy in gallbladder disease and has been rarely documented to be performed in patients with SIT, with only 50 known cases in the literature. We present the only known case in the literature of a SIT patient with choledocholithiasis and post-ERCP pancreatitis who subsequently had a Laparoscopic Cholecystectomy performed. We also submit the first known presentation of a prominent syphon of the hepatic artery, just posterior to the gallbladder, and propose it to be named Masri-Larkin Syndrome.

Case

A 79 year old female presented to the hospital with two days of nausea and vomiting. Her history included SIT. Physical exam was significant for LUQ tenderness to palpation. An abdominal ultrasound confirmed that she had SIT and was notable for cholelithiasis and choledocholithiasis. She had an initial CT A/P and was admitted to the hospital and subsequently underwent an MRCP followed by an ERCP, which confirmed the diagnosis, and a sphincterotomy was performed and the CBD stones were removed.

Following this, the patient developed post-ERCP pancreatitis and was monitored in the hospital until her LFT’s normalized and she was optimized for surgery. She was taken to the operating room at Larkin Community Hospital for Laparoscopic Cholecystectomy with Intraoperative Cholangiogram and Umbilical Herniorraphy.

Operative Technique

Operative monitors were positioned bilaterally over the patient’s shoulders. The surgeon and camera assistant were positioned on the patient’s right side and the first assistant on the patient’s left side. This patient also had an umbilical hernia present, so a supraventricular transverse incision was made and carried down to the fascia. The sac of the hernia was identified and separated from the hernia and the sac was excised and sent for pathology. A 10mm Hasson port was introduced into the peritoneal cavity and secured in place with two stay sutures of 0 Vicryl. The abdomen was insufflated with CO2 to 15 mmHg. A 10mm 0° scope was introduced, and the patient was placed in reverse Trendelenburg and rotated to the right. The gallbladder was thought to be distended and surrounded with dense adhesions but the liver looked normal. Two 5mm trocars were placed on the left subcostal margin, at the midclavicular and anterior axillary line and one 10mm port was placed just below the xiphoid and to the left of midline. Through the anterior axillary port, the first assistant used a grasper to retract the fundus superiorly and medially. The surgeon used another grasper through the subxiphoid port to retract the infundibulum medially, and with a Maryland dissector through the midclavicular port, peeled the peritoneum off the gallbladder starting laterally at first, and then moved anteriorly and medially.

Through careful dissection, the cystic duct and artery were clearly identified. It is noteworthy to mention that a prominent syphon of the hepatic artery was seen, which was close to the gallbladder and was significantly pulsating. The right hepatic artery was carefully separated from the gallbladder inferiorly. The cystic artery and the cystic duct were clearly identified at Calot’s triangle and the cystic artery was then quadri-clipped and divided. Next, a clip was placed on the cystic duct at the junction with the gallbladder. A partial ductotomy was then performed with laparoscopic metzenbaum scissors, through the midclavicular port, and the cystic duct was partially clipped and divided. The cystic duct was then clipped and divided. The cystic duct and artery posterior branch of the artery were clipped and divided. The entire gallbladder was removed from its fossa, using a spatula with electro cautery, placed in a pouch and delivered through the epigastric port. There were stones as well, which were all retrieved and irrigation was done. Once hemostasis was secured, a size-10 Jackson Pratt was placed in Morrison’s pouch and pulled through the anterior axillary 5 mm port and secured to the skin with 3-0 Prolene.

The cholangiogram was done under fluoroscopic guidance and showed no evidence of any common bile duct stones. The proximal biliary system was normal and there was contrast in the duodenum. The clip holding the catheter was removed, as well as the catheter, and then three clips were placed around the cystic duct ensuring both sides were seen. The cystic duct and artery and the posterior branch of the artery were triply clipped and cut. The entire gallbladder was removed from its fossa, using a spatula with electro cautery, placed in a pouch and delivered through the epigastric port. There were stones as well, which were all retrieved and irrigation was done. Once hemostasis was secured, a size-10 Jackson Pratt was placed in Morrison’s pouch and pulled through the anterior axillary 5 mm port and secured to the skin with 3-0 Prolene.

Through the epigastric port, a general laparoscopic exploration showed a normal bowel with no evidence of any vascular or bowel injuries. Of note, the exploration showed the appendix to be on the left side with sigmoid colon on the right side. The laparoscope and ports were removed and the umbilical hernia was repaired using a Mayo-type hernia repair. Operative time was sixty-nine minutes for both the Laparoscopic Cholecystectomy and intraoperative cholangiogram and Umbilical Herniorraphy. The patient had an uneventful postoperative recovery and after monitoring the LFT’s for a few days, the drain was removed and the patient was discharged from the hospital. After a postop visit in the clinic, the patient was doing well and had no complaints.

Discussion

Laparoscopic cholecystectomy in a patient with Situs Inversus was first described by Campos and Sipes in 1991, since then only a few dozen surgeons have had the privilege of saying they have performed this unique and challenging surgery. The contralateral orientation of the viscera provides an opportunity to perform a familiar surgery in an unfamiliar environment and requires rethinking every maneuver of the operation from beginning to end. The contralateral orientation of the viscera provides an opportunity to perform a challenging surgery.

In this case, the four-port mirror-image approach was used with 10mm ports in the epigastric and supraumbilical sites. The surgeon alternated between right and left hands for the dissection. The midclavicular port was used to place the cholangiocather. Stapling was done through the epigastric port with the left hand. The procedure took sixty-nine minutes.

Although there have been four cases of choledocho-liathiasis in a patient with SIT, all of whom underwent ERCP, to our knowledge this is the first case of post-ERCP pancreatitis in a patient with SIT that subsequently underwent LC with ICG. A notable anatomic anomaly found during surgery was the prominent syphon of the hepatic artery, located just posterior to the gallbladder and Calot’s Triangle, that we propose be named Masri-Larkin Syndrome.

References


