Laparoscopic Cholecystectomy and Appendectomy in Situs Inversus Totalis

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ABSTRACT

Situs inversus totalis is an uncommon anatomic anomaly that complicates diagnosis and management of acute abdominal pain. Expedient diagnosis of common intraperitoneal disease processes such as biliary colic, acute appendicitis and diverticulitis is often delayed as a result of seemingly incongruous physical findings. We present the case of a young woman with prior emergency room visits for complaints of a vague left upper quadrant abdominal pain. An ultrasound performed on her third presentation revealed visceral situs inversus with cholelithiasis and dilated intra- and extrahepatic biliary ducts. Standard laparoscopic cholecystectomy and cholangiography with a mirror-image surgical approach was performed successfully and without complication.

Key Words: Situs inversus totalis, Acute cholecystitis, Intraoperative cholangiography.

INTRODUCTION

Situs inversus totalis is a rare anatomic anomaly with an estimated incidence of 1:20,000 in the general population and an autosomal recessive mode of inheritance. Visceral situs inversus can occur with or without dextrocardia. One associated constellation of malformations includes Kartagener’s syndrome, first described in 1904 by Siewert in the Berliner Klinische Wochenschrift. He described a patient in 1901 with dextrocardia and situs inversus, who was also found to have bronchiectasis and sinusitis. Adams and Churchill reported a series of five patients with this same constellation of findings in 1937. Vazquez, et al noted that seven percent of babies with biliary atresia had situs inversus or cardiac defects.

While disease processes in patients with anatomic developmental anomalies such as gastrointestinal malrotation and nonrotation, visceral situs inversus and situs inversus totalis manifest faithfully their intra-abdominal locations, our standard clinical differential diagnoses often do not account for these variances. When evaluating a female with abdominal pain, ovulation, hemorrhagic ovarian cysts, pelvic inflammatory disease and endometriosis are all possible etiologies of acute pain, in addition to the gastrointestinal processes of appendicitis, cholecystitis and diverticulitis. Given the often overlapping clinical symptomatology of all of the above disease processes, a careful clinical history coupled with physical examination and an accurate localization of the offending organ becomes paramount.

CASE REPORT

A 20-year-old woman presented twice during the previous five months to the emergency room with complaints of post-prandial, vague left upper quadrant discomfort. She had no fever or leukocytosis at these evaluations and was discharged home on each occasion with a working diagnosis of gastroenteritis/gastritis, being treated with oral histamine-2 blockers. On the third presentation, she reported worsening pain with associated nausea and non-bloody emesis. On the third presentation, she reported worsening pain with associated nausea and non-bloody emesis. She also noted that food abstinence ameliorated her symptoms. Her past medical history was remarkable for two normal spontaneous vaginal deliveries—one five years prior to presentation and one only six months previously.
Physical examination showed the patient to be afebrile but in mild distress secondary to point tenderness on palpation of her left upper quadrant. She had no rebound or guarding but did exhibit a left-sided "Murphy's" sign. Laboratory examination revealed a leukocytosis of 12,000 cells/ml (normal range: 4.8 – 10.8), serum bilirubin = 2.0 mg/dl (0.2 – 1.2), alkaline phosphatase = 105 U/L (38 – 126), lactate dehydrogenase = 1000 U/L (313 – 618), serum glutamate-oxaloacetate transferase = 217 U/L (8 – 40), amylase 78 U/L (44 – 128) and lipase 146 U/L (23 – 208).

A chest radiograph demonstrated dextrocardia with an elevated (but normal in this case) left hemidiaphragm without other abnormalities (Figure 1). Abdominal ultrasonography showed a left-sided liver and gallbladder with cholelithiasis and dilated intra- and extrahepatic biliary ducts. The appendix was not visualized. Given these findings, a 12-lead electrocardiogram was performed, which demonstrated abnormal but characteristic wave morphologies for dextrocardia. A 2-D echocardiogram of the heart with M-mode Doppler yielded no intracardiac malformations.

After 24 hours of intravenous antibiotics, the patient was taken to the operating room for laparoscopic exploration. General endotracheal anesthesia was administered, and an infraumbilical port was placed using the open technique for access to the abdominal cavity. Exploratory laparoscopy confirmed the ultrasonographic finding of visceral situs inversus totalis. A standard laparoscopic cholecystectomy was performed, including an intraoperative cholangiogram. The latter showed no evidence of choledocholithiasis and free efflux of contrast into the duodenum with a mirror-image configuration of the biliary tree (Figure 2). The appendix was visualized in the left lower quadrant, isolated from the mesoappendix, and resected with an endoscopic stapling device.

Final pathology showed a normal vermiform appendix and gallbladder with chronic cholecystitis and cholelithiasis, containing 11 yellow-green calculi, the largest of which was 5 mm in diameter. As the bile ducts were free of stones on cholangiography and the alkaline phosphatase level was normal, we believe the ductal dilation noted was secondary to hepatic irritation from the inflammatory process in the gallbladder. The patient did well postoperatively, with repeat laboratory examination on postoperative day two showing a normal white blood cell count (7.5 K), serum bilirubin = 1.5 mg/dl, alkaline phosphatase = 111 U/L, lactate dehydrogenase = 492 U/L and serum glutamate-oxaloacetate transferase = 97 U/L. The patient was discharged home on the second postoperative day.

DISCUSSION

The value of laparoscopy in evaluation of patients with atypical abdominal pain has been well established. A small, but real, subset of the general population has situs inversus totalis or some other type of previously unestablished anomaly of bowel rotation. While chance favors the prepared mind, we are frequently unprepared to evaluate usual disorders set in unusual circumstances. Other investigators have reported the utility of laparoscopy, not only in establishing the diagnosis of rotational bowel abnormalities but also in treating dis-
Dextrocardia, or right looping of the embryological truncus arteriosus and bulbus cordis over the ventricle and atrium, is frequently accompanied by visceral situs inversus. The incidence of concomitant intracardiac structural defects in this situation is low. Isolated dextrocardia, by contrast, however, is associated with a high incidence of cardiac defects. Patients with indeterminate visceral situs frequently have asplenia or polysplenia as well as multiple complex cardiac anomalies (eg, atrial and ventricular septal defects, endocardial cushion defects, double-outlet right ventricle). We believe the presence of dextrocardia mandates further investigation of the status of visceral situs for two reasons. First, dextrocardia is a predictor of the relative frequency of congenital cardiac defects. Second, it alerts clinicians and, more importantly, the patient of a condition whose repercussions can bring to bear surgical consequences.

Sporadic reports of successful laparoscopic cholecystectomy in visceral situs inversus have appeared in the worldwide literature since 1992. All stress the feasibility of the laparoscopic approach to treatment with few technical modifications. After entering the peritoneal cavity and insufflation with carbon dioxide, exploration of the abdominal cavity is undertaken, and the diagnosis of visceral situs inversus, if unknown preoperatively, is discovered. If suspected preoperatively, it is confirmed by direct inspection. A standard subxiphoid port is placed, while lateral abdominal ports are placed through the left, rather than right, side of the abdominal wall. The operative surgeon is positioned to the patient’s right rather than left side and the operation proceeds as usual. These minor modifications readily facilitate operative exposure of the gallbladder and biliary tree for cholecystectomy and intraoperative cholangiography. Having discussed the issue with our patient preoperatively and in order to preclude any future diagnostic confusion, the appendix was sought, visualized in the left lower quadrant, and excised with an endoscopic stapling device.

Minimal modification to the standard laparoscopic cholecystectomy protocol is required to treat patients with visceral situs inversus totalis or bowel malrotation anomalies. Given the low, albeit real, incidence of associated gastrointestinal conditions, we strongly favor intraoperative cholangiography to clearly delineate the course of the biliary tree and ensure the absence of intraluminal strictures or filling defects. These considerations, when paid appropriate heed, make laparoscopy not only safe but also expedient and ensure the optimal treatment of patients who have these special instances of common disease processes.

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