A Rare Anatomical Variation Complicating A Diffuse Abdominal Pain Presentation: Colonic Perforation in Situs inversus totalis

Omar Elfanagely[1], Yousef Elfanagely[2], and Abimbol Pratt[3]

[1]Robert Wood Johnson University Hospital
[2]Brown University
[3]Hackensack Meridian Health

July 8, 2021

Abstract

Situs inversus totalis (SIT) is a rare anatomical variation of the thoracic and abdominal organs. We report a case of a 93-year-old woman who presented with pneumoperitoneum secondary to stercoral colonic perforation requiring emergent Hartman's procedure.

A Rare Anatomical Variation Complicating A Diffuse Abdominal Pain Presentation: Colonic Perforation in Situs inversus totalis

Authors: Omar Elfanagely MD[1], Yousef Elfanagely MD[2], Abimbola Pratt MD[3]

Affiliations:

[1]Department of Surgery, Robert Wood Johnson Medical School, New Brunswick, New Jersey, USA

[2]Department of Internal Medicine, Warren Alpert Medical School of Brown University, Providence, Rhode Island, USA

[3]Division of Trauma, Acute Care Surgery & Critical Care, Department of Surgery, Hackensack Meridian Health

ABSTRACT

Situs inversus totalis (SIT) is a rare anatomical variation of the thoracic and abdominal organs. We report a case of a 93-year-old woman who presented with pneumoperitoneum secondary to stercoral colonic perforation requiring emergent Hartman's procedure.

KEY CLINICAL MESSAGE

The anatomic variation seen in situs inversus totalis can complicate surgical interventions and clinical diagnoses. Proper planning, thorough imaging, and careful execution are necessary to ensure patient safety and care in patients with SIT.

INTRODUCTION

Situs inversus totalis (SIT) was first described in humans by Fabricius in 1600[1]. Since then, SIT remains a rare anatomic anomaly with an incidence of 1:10,000 to 1:20,000. Patients with SIT have a complete right-left inversion of the thoracic and abdominal viscera. The cause of SIT is not fully identified, but might be correlated with congenital and genetic factors. The diagnosis of SIT is often incidental and found on imaging.

As the diagnosis of SIT is often incidental, patients with SIT can present to the hospital with the manifestations of another process. However, anatomic variations seen in SIT can vary the expected presentation of typical medical conditions. Likewise, the mirror-image anatomy seen in SIT can complicate surgical interventions that are performed in a particular orientation. Here, we present a case of pneumoperitoneum incidentally found to have SIT.

CASE PRESENTATION

A 93-year-old woman with a past medical history of hypertension, hyperlipidemia, and atrial fibrillation (on anti-coagulation) presented to the emergency room with severe abdominal pain that started 4 days prior to presentation. Abdominal pain was non-radiating, diffuse, and persistent in nature. She also reported decrease appetite secondary to nausea and vomiting. She endorsed constipation for 3 to 4 days, but continued to pass flatus. She denied fevers, chills, and recent trauma. Her vital signs were notable for tachycardia (heart rate of 110 beats per minute), temperature of 98.7 degrees Fahrenheit, blood pressure of 119/58 millimeters of mercury, and respiratory rate of 20 breaths per minute.

On physical examination, she had diffuse abdominal tenderness and distension. There was also voluntary guarding, but no rebound tenderness or rigidity. The remainder of the physical exam was unremarkable, except for an irregular heart rate heard on the right side of the chest. Laboratory testing was notable for an elevated INR (INR 1.65), lactic acidosis (lactic acid 2.0 mEq/L), and acute kidney injury (creatinine 1.67 mg/dL). There was no evidence of leukocytosis (6.2 K/uL).

The patient underwent a computed tomography (CT) scan of the abdomen and pelvis (Figures A-B). The non-enhanced CT scan revealed multiple small foci of free intraperitoneal air, predominantly anteriorly and greater asymmetric to the right (Figure A, see red arrow). No definitive source was identified. Additionally, the liver was noted to be on the left (Figure B, see red star) and the stomach on the right (Figure B, see red O). A few borderline dilated small bowel loops were noted (Figure A).

These findings were consistent with the diagnosis of pneumoperitoneum in a patient with situs inversus totalis. There was additional concern for an early small bowel obstruction given borderline small bowel dilatation. Our patient was started on broad-spectrum antibiotics, received volume repletion, and taken emergently to the operating room. In the operating room, the patient was found to have stercoral colonic perforation of her descending colon requiring Hartmann's procedure (colectomy with end colostomy).

DISCUSSION

SIT is a rare anatomical anomaly in which there is complete right to left reversal of the thoracic and abdominal organs. The first historical description was in animals by Aristotle[1]. In 1600, the first known case of SIT in humans was reported by Fabricius[1]. The incidence is thought to be 1:10,000 to 1:20,000[2]. Although the etiology has not been fully clarified, most cases are thought to be related with sporadic genetic mutations during embryonic development[3, 4]. SIT is diagnosed with diagnostic imaging. Patients with SIT are at increased risk of heart, spleen, and hepatobiliary malformations[5]. However, survival rate of patients with SIT is not different from that of patients without SIT[6].

Given the complete mirror-image anatomy, clinical diagnoses and performing procedures can be particularly challenging. Patients with SIT who develop appendicitis, for example, can be confused with other intraperitoneal processes such as diverticulitis[7]. Surgeons and proceduralists operating on patients with SIT have to aware of the anatomic variants present and risk of organ malformation.

Patients with SIT should undergo thorough radiological examination before undergoing invasive procedures such as surgery, percutaneous endoscopic gastrostomy, or hemostasis of gastrointestinal hemorrhage because of the increased risk of malformations in anatomical structures and reversal of organ location. Additional considerations such as positioning of patient and/or proceduralists are important. Patients with SIT who require Endoscopic retrograde cholangiopancreatopgraphy (ERCP) or endoscopy are placed in the right lateral decubitus position to account for the organ reversal[6]. Surgeons performing a laparoscopic procedure

on a patient with SIT may does so in the lithotomy position to improve orientation to the reversed anatomy[8]. Overall, patients with SIT require proper planning prior to surgical intervention to ensure patient safety.

CONCLUSION

We report a case of pneumoperitoneum found in a patient to have situs inversus totalis, which is rare anatomical anomaly. The anatomic variation seen in situs inversus totalis can complicate surgical interventions and clinical diagnoses. Proper planning, thorough imaging, and careful execution are necessary to ensure patient safety and care in patients with SIT.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING STATEMENT

No funding sources to declare

REFERENCES

1. Pathak K, Khanna R, Khanna N. Situs inversus with cholelithiasis. Journal of postgraduate medicine 1995;41:45.

2. Feng Q, Yao J, Lu S. Gastrointestinal: Situs inversus totalis with esophageal cancer. Journal of gastroenterology and hepatology 2020;35:709-709.

3. Benhammane H, Kharmoum S, Terraz S, et al. Common bile duct adenocarcinoma in a patient with situs inversus totalis: report of a rare case. BMC research notes 2012;5:1-5.

4. Casey B. Genetics of human situs abnormalities. American journal of medical genetics 2001;101:356-358.

5. Fonkalsrud EW, Tompkins R, Clatworthy W. Abdominal manifestations of situs inversus in infants and children. Archives of Surgery 1966;92:791-795.

6. Lee HK, Cho KB, Kim ES, et al. Gastrostomy in a patient with situs inversus totalis. Clinical endoscopy 2013;46:662.

7. Navarro V, Oshiro H, Peña B, et al. Situs inversus totalis. Revista de Gastroenterología del Perú 2013;33:345-347.

8. Rosen H, Petrosyan M, Mason RJ. Cholecystitis in situs inversus totalis. Radiology case reports 2008;3:226.

FIGURES

Figure A . Computed tomography (CT) scan of the abdomen and pelvis, axial view

Arrow pointing to free air (pneumoperitoneum)

Figure B . Computed tomography (CT) scan of the abdomen and pelvis, coronal view

Star is left-sided liver. Circle is right-sided stomach. Rectangle is a right-sided heart.