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Case report

A rare anatomical variation complicating a diffuse abdominal pain presentation: A case report of colonic perforation in situs inversus totalis

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ABSTRACT

Introduction: Situs inversus totalis (SIT) is a rare anatomical variation of the thoracic and abdominal organs. It is a congenital anomaly with an incidence of 1:10,000 to 1:20,000. Patients with SIT do not have a decreased survival rate as compared to patients without SIT because SIT generally does not have a pathophysiologic significance. However, the anatomical variations in SIT can cause some challenges when assessing intraabdominal and intrathoracic symptoms or performing operations.

Case presentation: We report a case of a 93-year-old woman with a past medical history of hypertension, hyperlipidemia, atrial fibrillation, and situs inversus totalis who presented with diffuse abdominal pain for 4 days. Abdominal exam was significant for diffuse tenderness. Computed tomography (CT) imaging was significant for pneumoperitoneum. She emergently underwent an exploratory laparotomy, descending hemicolectomy and left in discontinuity with an open abdomen. On postoperative day 2 she underwent a stamm feeding gastrostomy tube, incisional hernia repair, and maturation of end colostomy. Her remaining hospital course was complicated by a pelvic collection, which was managed by a percutaneous guided drain placement. She was ultimately discharge to rehab on hospital day 15.

Discussion: SITS can present a particularly challenging situation to clinical diagnoses and surgical procedures. However, when identified, these patients should warrant special considerations prior to proceeding with surgical intervention. This includes radiologic imaging and proper planning prior to the operating room, when possible.

Conclusion: We herein present a case of colonic perforation in a patient with situs inversus totalis. Proper planning, thorough imaging, and careful execution are necessary to ensure patient safety and care in patients with SIT. However, in the case of emergency this should not delay definitive management.

1. Introduction

Situs Inversus is a congenital anomaly characterized by the transposition of the thoracic and abdominal viscera. When associated with dextroardia, it is termed situs inversus totalis (SIT) which was first described in humans by in 1600 [1]. Since then, SIT remains a rare anatomic anomaly with an incidence of 1:10,000 to 1:20,000. The cause of SIT is not fully identified, but is believed to be partly due to the malrotation of the gastrointestinal system during the embryologic development [2]; as well as 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. This case report has been presented in line with the SCARE Criteria [3]. Below we describe the first ever-case report of perforated diverticulitis in a patient with SIT.

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2. Methods

2.1. Patient information

A 93-year-old woman with a past medical history of hypertension, hyperlipidemia, and atrial fibrillation (on anti-coagulation), situs inversus totalis, and uterine fibroids status post hysterectomy who presented to the emergency room of our academic hospital with severe abdominal pain that started 4 days prior to presentation. Of note, the patient has no toxic habits (smoking, drinking), allergies or contributory family history. She denied any known genetic abnormalities. 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 mm Hg, and respiratory rate of 20 breaths per minute.

On physical examination, she had diffused 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 (wbc 6.2 K/uL).

The patient underwent a computed tomography (CT) scan of the abdomen and pelvis (Figs. 1-2). The non-enhanced CT scan revealed multiple small foci of free intraperitoneal air, predominantly anteriorly and greater asymmetric to the right (Fig. 1, see red arrow). No definitive source was identified. Additionally, the liver was noted to be on the left (Fig. 2, see red star) and the stomach on the right (Fig. 2, see red O). A few borderline dilated small bowel loops were noted (Fig. 1).

Prior to the cross sectional and the blood work findings the differential diagnosis included nonsurgical conditions such as gastritis, enteritis, and constipation. Surgical pathologies included bowel obstruction (given her previous surgical history), ovarian torsion, appendicitis, volvulus, diverticulitis, colonic perforation and other hollow viscus perforation. However, with the return of the imaging and labs it was clear the patient was suffering from an acute surgical pathology. Our patient was then started on broad-spectrum antibiotics, cultures were drawn, received volume repletion, and taken emergently to the operating room by one of our trauma and critical care fellowship trained attending. In the operating room, the patient was underwent an exploratory laparotomy for which she was found to have perforation diverticulitis of her descending colon requiring a descending hemicolectomy. There was no associated malrotation identified. During the index procedure the patient developed increasing pressor requirements and coagulopathy for which the decision was made to leave her abdomen open with plan for resuscitation in the surgical intensive care unit (SICU). The patient was taken back to the OR on postoperative day 2 and underwent a reexploration, a stamm feeding gastrostomy tube, ventral incisional hernia repair (with inlay biologic mesh), and maturation of end colostomy. Pathology noted ischemic changes to the colon, acute serosal exudates and adhesions, and chronic diverticulitis. The patient was successfully extubated in the SICU and ultimately downgraded to the floors. Her postoperative course was complicated by a pelvic collection, which was amenable to interventional radiology guided drain placement and eventually removed prior to discharge. She was ultimately discharge to rehab on hospital day 15 on a dysphagia puree diet in satisfactory condition.

3. Discussion

SIT is a rare anatomical anomaly in which there is complete right to left reversal of the thoracic and abdominal organs. Aristotle described the first historical description of SIT in animals, it was later reported in

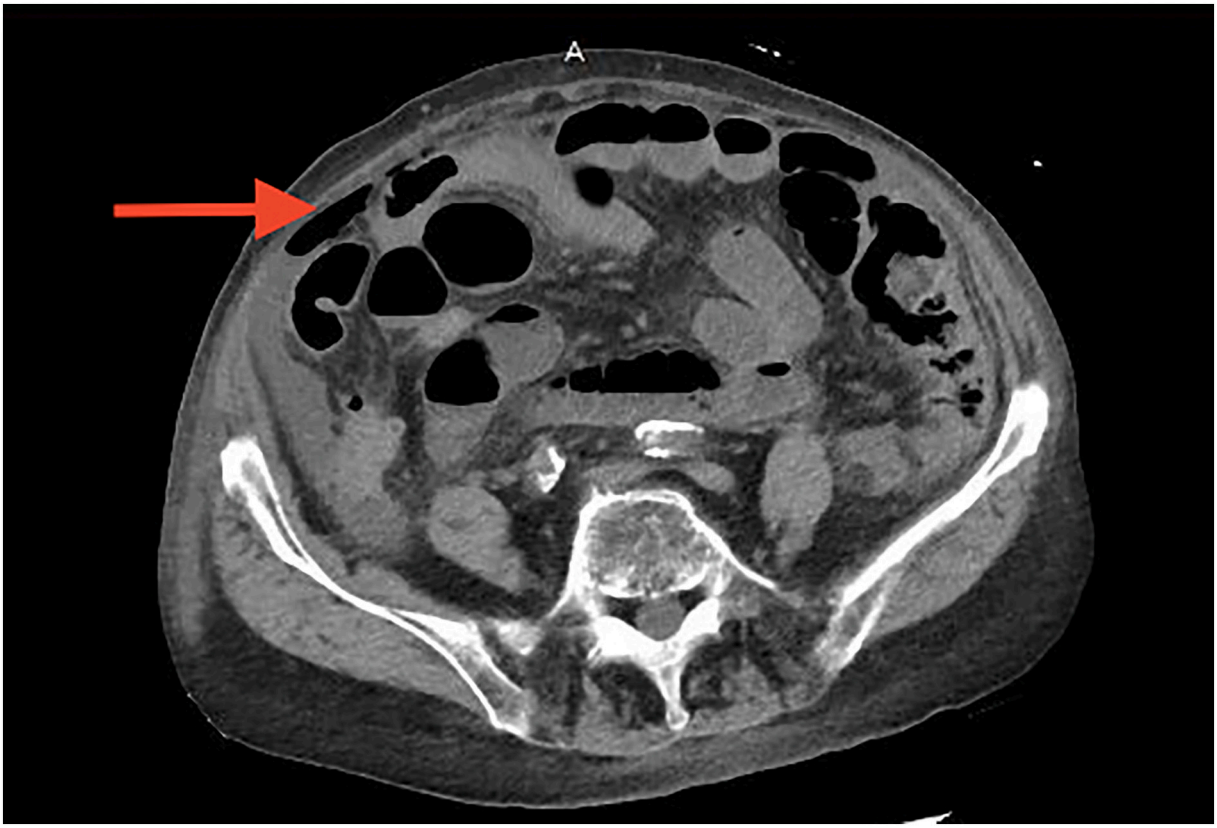

Fig. 1. Computed tomography (CT) scan of the abdomen and pelvis, axial view Arrow pointing to free air (pneumoperitoneum).

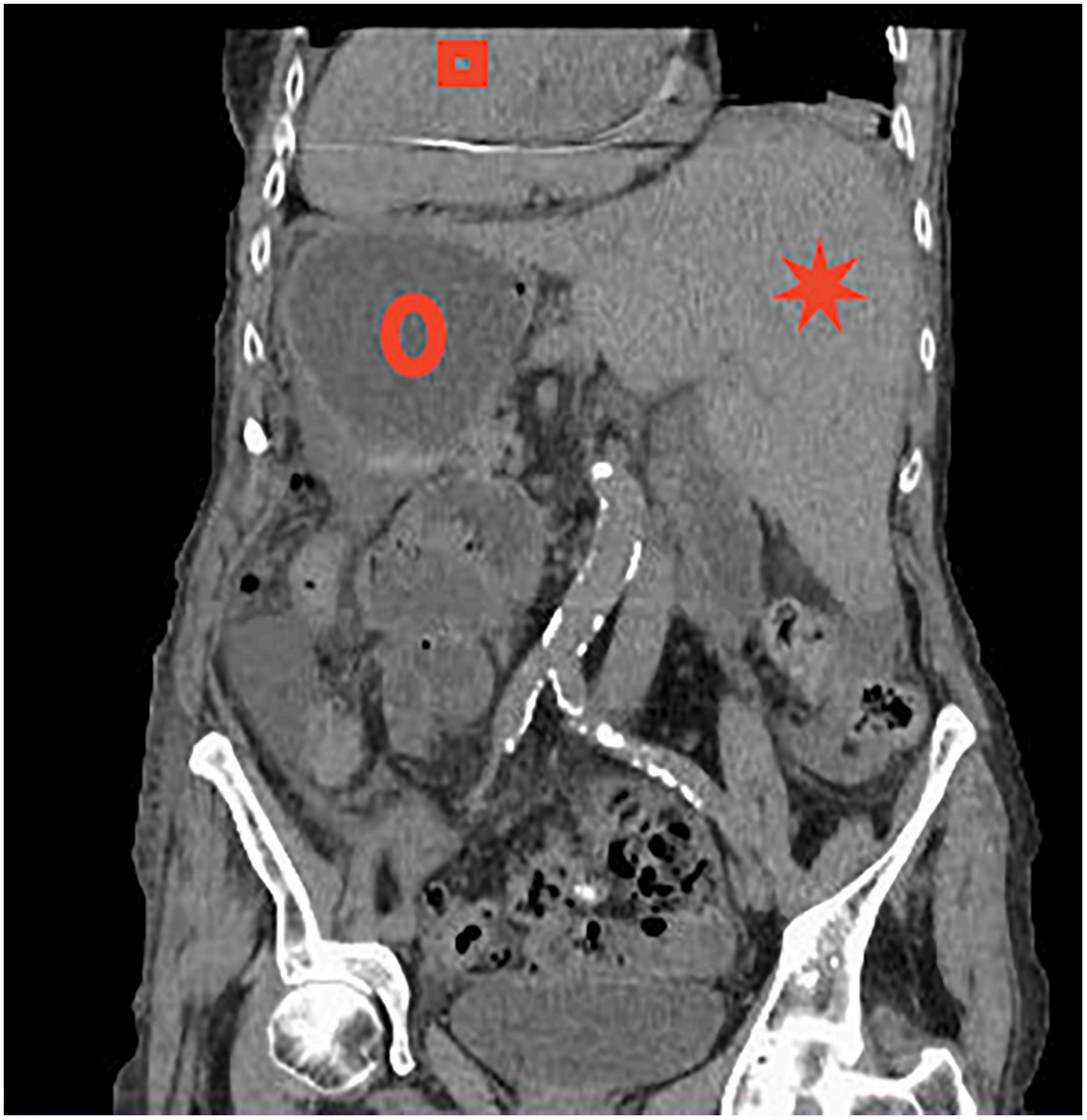

Fig. 2. Computed tomography (CT) scan of the abdomen and pelvis, coronal view Star is left-sided liver. Circle is right-sided stomach. Rectangle is a right-sided heart.

human in 1600 by Fabricius [1]. The incidence is thought to be 1:10,000 to 1:20,000 [4]. Although the etiology has not been fully clarified, most cases are thought to be related with sporadic genetic mutations during embryonic development [5]. SIT is diagnosed with diagnostic imaging. Patients with SIT are at increased risk of heart, spleen, and hepatobiliary malformations [6]. However, survival rate of patients with SIT is not different from that of patients without SIT [7].

Perforation peritonitis is one of the most common causes of surgical emergencies worldwide [8]. Etiology of intestinal perforation can be categorized due to ischemia, infection, erosion, and mechanical disruption [9]. Perforation peritonitis in situs inversus is extremely rare with limited case reports in the literature. To date, there are descriptions of perforation due to gallbladder [10], appendicitis [11], duodenal ulcer [12,13], and gastric [14]. To the best of our knowledge, this is the first case report of a patient with SIT presenting with perforated diverticulitis.

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 [15]. Additionally, females presenting with lower or right-sided abdominal pain may undergo a work up for genitourinary pathology and thus have a delay in diagnosis and appropriate treatment. Surgeons and proceduralists operating on patients with SIT have to be 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 [7]. Surgeons performing a laparoscopic procedure on a patient with SIT may do so in the lithotomy position to improve orientation to the reversed anatomy [16]. Therefore to choose the proper surgical incision site for abdominal exploration preoperative recognition of this condition is important. Overall, patients with SIT require proper planning prior to surgical intervention to ensure patient safety.

4. Conclusion

We report a case of colonic perforation 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.

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Ethical approval

Ethical approval not required.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request

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Declaration of competing interest

None to declare.

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