

Acute Appendicitis in a Patient With Situs Inversus Totalis and Malrotation

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ABSTRACT

Left-sided acute appendicitis is a rare condition that occurs most frequently in patients with congenital abnormalities. It can be a surgical emergency and a diagnostic challenge for clinicians. We present a case of acute appendicitis in a patient with situs inversus totalis and intestinal malrotation.

Key Words: left-sided appendicitis, malrotation, situs inversus.

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INTRODUCTION

Acute appendicitis is a relatively common disease process with an annual incidence of 1:1000.¹ It is one of the most common presentations of acute abdomen requiring surgical consultation. Left-sided acute appendicitis (LSAA) is a rare condition that occurs most frequently in patients with congenital abnormalities, most commonly situs inversus totalis, followed by malrotation.^{2,3} In this case report, we describe a patient with known situs inversus and LSAA in the setting of intestinal malrotation who was diagnosed with the use of preoperative computed tomography (CT) scanning and treated with laparoscopic appendectomy.

CASE REPORT

The patient was a 34-year-old man with known past medical history of situs inversus diagnosed on previous chest radiogram. He presented to the emergency department (ED) with a 2-day history of right lower quadrant pain with associated anorexia, fever, and watery diarrhea. He did not report previous periumbilical pain. Physical examination demonstrated right lower quadrant and supra-

pubic tenderness without signs of diffuse peritonitis. He had right lower quadrant pain with palpation of the left lower quadrant but did not have exacerbation of his pain with extension of the right or left hip. Laboratory examination revealed leukocytosis (white blood cell count [WBC] 14,900/ μ L). The CT scan of the abdomen/pelvis showed known situs inversus totalis and findings consistent with intestinal malrotation. The small bowel was located in the left abdomen, and the colon was noted to be on the right. The duodenum did not cross the midline, and the superior mesenteric vein (SMV) was to the right of the superior mesenteric artery (SMA) (**Figure 1**). The cecum and appendix were located in the mid to left pelvis. Radiological findings confirmed appendicitis (mucosal enhancement and mild adjacent fat stranding) (**Figure 2**).

The patient underwent laparoscopic appendectomy. The open Hasson technique was used to enter the abdomen. Diagnostic laparoscopy revealed a right-sided stomach and left-sided liver. Additional 5-mm trocars were placed in the suprapubic region and the right lower quadrant. The small bowel was retracted caudad, and the appendix was identified in the left pelvis. It was inflamed and



Figure 1. CT scan (coronal cuts) showing the superior mesenteric vein (SMV, red arrow) lying to the right of the superior mesenteric artery (SMA).

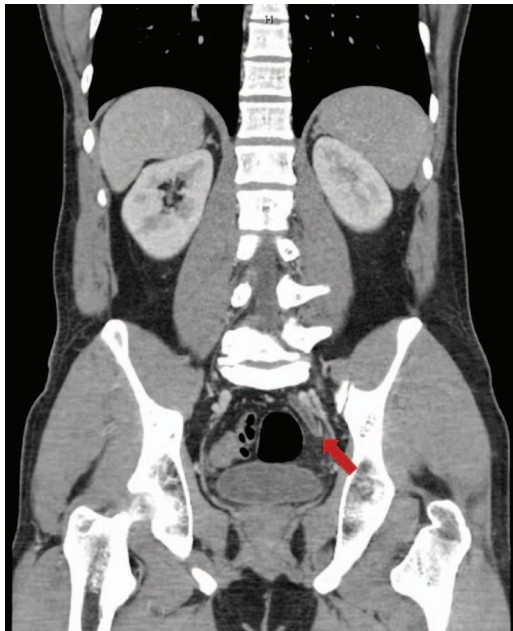


Figure 2. CT scan (coronal cuts) showing the appendix (red arrow) lying in the left pelvis. It is distended with mucosal enhancement.

suppurative, consistent with acute appendicitis. The mesoappendix was secured with a LigaSure™ Retractable L-Hook device (Medtronic, Minneapolis, MN), and the base of the appendix was transected with a 45-mm Tri-Staple™ purple load (Covidien). Surgical pathology was consistent with acute appendicitis. The patient recovered well and was discharged home on the first postoperative day.

DISCUSSION

Acute appendicitis is one of the most common presentations of acute abdomen requiring surgical evaluation.^{1,4,5} The classic presentation of acute appendicitis is vague abdominal pain that starts at the umbilicus and then localizes to the right lower quadrant, near McBurney's point. It is associated with anorexia and fever, and CT scanning of the abdomen/pelvis is the most sensitive and specific diagnostic modality.^{1,4,6} However, the classic clinical presentation occurs in only about 60% of patients,⁴ and approximately one-third of patients with acute appendicitis have pain localized outside of the right lower quadrant.² Classic treatment for acute appendicitis is appendectomy (open or laparoscopic).⁷ Recently, nonoperative pathways with antibiotics as definitive treatment of acute appendicitis have shown success rates of up to 75%.^{5,8} However, standard treatment for uncomplicated acute appendicitis remains appendectomy in the United States.

LSAA is extremely rare and can occur with congenital abnormalities such as malrotation, situs inversus, or a right-sided but very long appendix projecting into the left lower quadrant.^{2,3} In 2010, Akbulut et al² published a case report and literature review of LSAA. Almost 97% of cases of LSAA were associated with a congenital abnormality: 69.4% with situs inversus and 24.2% with malrotation. However, our patient's presentation of LSAA with both situs inversus totalis and intestinal malrotation is even more rare.

Situs inversus is an uncommon condition caused by a single autosomal recessive gene with incomplete penetrance. It occurs in 1:5000 to 1:10,000 live births and with variable penetrance. Situs inversus totalis is the complete reversal of all organs, but there is also partial situs inversus, where only one of either the thoracic or abdominal cavity is reversed.² In addition to situs inversus totalis, there is situs ambiguous, or heterotaxy, which is a symmetric arrangement of organs that are typically asymmetric (eg, duplicate and symmetric spleen or liver). Situs ambiguous is frequently associated with malrotation (40% to 90%).⁹ In a retrospective review conducted by Choi et al⁹ in 2016, malrotation was seen in 16 of 33 patients with

a situs anomaly; however, among the 16 patients with situs inversus, only 1 patient had malrotation (6.25%). Among the 17 patients with situs ambiguous, 15 patients exhibited malrotation (88.2%), and situs ambiguous was significantly associated with intestinal malrotation compared with the patients with situs inversus.⁹

The incidence of malrotation varies from 0.03% to 0.5% of live births and is a spectrum of congenital positional abnormalities of the intestine caused by noncomplete or incomplete rotation of the bowel around the axis of the SMA during fetal life. Approximately 80% of all cases are diagnosed in patients younger than 1 month, although it can be diagnosed at any age.^{2,3} It can be a life-threatening condition if midgut volvulus occurs, which can lead to intestinal ischemia, short gut, and even death.¹⁰ All symptomatic patients with malrotation require Ladd's surgery; however, for those patients who remain asymptomatic beyond age 2 years, there is little evidence to guide treatment. At this time, prophylactic Ladd's procedure is generally not recommended in truly asymptomatic patients older than 2 years.¹⁰

Although this patient presented with classic right-sided pain, the diagnosis of LSAA can be a clinical challenge.^{4,6,11} The pain caused by LSAA has been reported in the right lower quadrant in 18.4% to 31% of patients with situs inversus or malrotation but more often presents with left lower quadrant pain.^{2,3} The traditional differential diagnosis for left lower quadrant pain in adults is diverticulitis, renal colic, ovarian cyst rupture, Meckel's diverticulitis, epididymitis, incarcerated or strangulated hernia, bowel obstruction, gastroenteritis, psoas abscess, and appendicitis.³ Our patient did not present with the classic diffuse periumbilical pain but instead had localized right lower quadrant pain, although he did have a Rovsing's sign.

There are 3 main imaging modalities for diagnosing acute appendicitis in adults and children. Intravenous contrast agent enhanced CT is the most sensitive and specific method used in adults and children.¹² However, iodinated contrast medium is nephrotoxic and CT requires exposure to ionizing radiation. Ultrasound is recommended when feasible in children and young adults up to age 30¹³ and has been shown to have sensitivity and specificity of 88% and 94% in pediatric patients and 83% and 93% in adult patients, respectively.¹² Risk factors for a nondiagnostic ultrasound include increasing age and being overweight.¹⁴ Increasingly, magnetic resonance imaging has been used as an alternative imaging modality that does not require ionizing radiation. Newer ultrafast protocols lasting less than 10 minutes have been shown to be fea-

sible in pediatric patients at select centers, requiring neither sedation nor intravenous contrast agents, with a reported sensitivity and specificity of 100% and 99%, respectively.¹⁵ CT, however, will likely remain a reasonable option for the evaluation of abdominal pain in the non-pregnant adult without clear pathology from history, physical, and laboratory evaluation.

The advent of laparoscopy has introduced a new dimension of diagnostic abilities. Although open appendectomy with an incision at McBurney's point is still performed in the community, a 2014 updated Cochrane review and meta-analysis showed that laparoscopy in women with acute lower abdominal pain, nonspecific lower abdominal pain, or suspected appendicitis led to a higher rate of specific diagnoses being made and a lower rate of removal of a normal appendix compared with open appendectomy alone. Additionally, patients returned to work faster, and their hospital stay was shorter.¹⁶

Our patient had a unique presentation of situs inversus totalis and malrotation demonstrated by lack of the duodenal C-loop, cecum in the midline pelvis, and an SMV located in what would be the normal position in a normal adult (to the right of the SMA) but denotes malrotation in a person with situs inversus totalis. Indeed, in malrotation, the small bowel is usually found entirely on the right side of the abdomen with the colon on the left side,¹⁷ but our patient presented with his small bowel on the left and large bowel on the right.

We present a case of acute appendicitis in a patient with both situs inversus totalis and intestinal malrotation. LSAA remains a diagnostic challenge, and prompt diagnosis can help prevent sepsis and further complications from acute appendicitis.

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