Laparoscopic approach for the treatment of left-sided acute appendicitis associated with malrotation

Abstract

Introduction: Acute appendicitis is among the leading causes of general surgery emergencies and requires prompt diagnosis and treatment. However, the diagnosis can be especially challenging when the patient presents with left-sided abdominal pain. True left side originated appendix can occur in the setting of two conditions: situs inversus and intestinal malrotation.

Case Report: This article presents the case of a 31-year-old male with left lower quadrant (LLQ) abdominal pain, caused by left-sided acute appendicitis associated with intestinal malrotation. After confirmation of the diagnosis through radiologic studies, a laparoscopic appendectomy was performed, with adjustments to the position of the ports to allow for optimal access to the LLQ. There was no evidence of complications directly related to the malrotation, such as volvulus, small bowel obstruction or internal hernia. There were no complications in the intraoperative or postoperative period.

Discussion: The various forms of intestinal malrotations compose a spectrum of bowel positional anomalies, caused by nonrotation or incomplete rotation of the primitive bowel loop. Acute appendicitis poses a great risk to patients with malrotation due to the atypical presentation and the difficulty in the interpretation of radiologic studies, which may cause a grave delay in the diagnosis and surgical intervention, potentially resulting in higher morbimortality. This is aggravated by the very scant number of studies comparing different approaches and analyzing outcomes, which in turn is caused by the rarity of cases with the two conditions associated.

Conclusion: Although there is not enough evidence in the current literature to support the systematic use of laparoscopic approach in the treatment of appendicitis associated with intestinal malrotation, it is the opinion of the authors that the laparoscopic treatment, when performed by an experienced surgeon and with the proper adaptations arranged, can be feasible in those patients.

Keywords: left-sided appendicitis, intestinal malrotation, laparoscopic appendectomy, acute appendicitis in malrotation, situs inversus

Abbreviations: LLQ, left lower quadrant; RLQ, right lower quadrant; SMA, superior mesenteric artery LUQ, left upper quadrant

Introduction

Acute appendicitis represents one of the most common causes of abdominal pain and is among the leading causes of general surgery emergencies, affecting around 6 to 7% of western world countries’ population.1,2 Aiming at reducing its morbimortality, prompt diagnosis and surgical intervention should be employed, and therefore it is recommended that physicians adopt a low threshold for suspecting appendicitis, almost always including it in the differential diagnosis of abdominal pain.

In most cases, diagnosis can be made based on presenting signs and symptoms, especially when aided by a score system, such as the Alvarado score,1 which also takes into consideration laboratory parameters. The most frequent presentations include tenderness in the right lower quadrant (92.2%), leukocytosis (75.6%), neutrophilia (73.3%), rebound tenderness (72.2%), and migratory pain (62.2%).4 However, the point of maximal tenderness in acute appendicitis varies greatly due to the wide range of positions in which the appendix can be found, being in a radius of less than 3 centimeters of the McBurney’s point in only 40% of cases.3

Among all the possible sites of abdominal tenderness related to acute appendicitis, one of the most challenging presentation is left-sided abdominal pain. There are two possible explanations for such finding: either the patient has a long vermiform appendix, which originates in the right lower quadrant (RLQ) and extends to the left of the midline, since it can measure up to 20 centimeters;1 or the atypical presentation results from congenital defects in which the cecum is positioned in the left side of the abdomen, and thus the appendix truly originates from the left. Such anomalous anatomy can occur in the setting of two conditions: situs inversus and intestinal malrotation.

Objectives

To present a case of left-sided acute appendicitis associated with intestinal malrotation in an adult patient, review the literature regarding the subject, and discuss the feasibility of laparoscopy for the surgical intervention.
Case report

Patient is a 31-year-old male, admitted to the emergency department complaining of abdominal pain for 12 hours. The pain had been continuous, was localized in the left lower quadrant (LLQ) and periumbilical regions, graded 8 out of 10 in a pain scale and had no alleviating or aggravating factors. He also had nausea, vomiting and anorexia. He denied any episode of fever or change in bowel movements.

The patient’s past medical and surgical history, family history and social history were unremarkable. On physical examination, the patient was alert and fully oriented. His temperature was 36°C, the blood pressure 150/98 mmHg, the pulse 73 beats per minute, and the oxygen saturation 99% while breathing ambient air.

Skin was warm and moist, without pallor or icterus, no rashes. There was no lymphadenomegaly. Lungs were clear to auscultation. Heart examination revealed no abnormalities, and the point of maximal impulse was felt on the left midclavicular line at the fifth intercostal space. Abdomen was soft and non-distended, tender to palpation of the LLQ, with rebound tenderness, without guarding, no masses or hepatosplenomegaly. The remainder of the examination was normal.

Even though the presentation was highly atypical for acute appendicitis, it was considered as one of the differentials for the case. To clarify the diagnosis, a workup including laboratory exams and computed tomography of the abdomen was ordered. Complete blood count showed leukocytosis of 13500 per mm$^3$ and neutrophil left shift, with 10% bands. Chest CT confirmed situs solitus (Figure 1A). Abdominal CT showed signs of intestinal malrotation, with abnormal relative position of the superior mesenteric vein and artery (Figure 1B), small bowel redundancy to the right of the midline, and colon and cecum to the left of the midline (Figure 1B–1F). The appendix was long and anterior. It originated from the cecum in the left iliac region (Figure 1D) (Figure 1E) and extended to the hypogastrium (Figure 1F). The appearance of the appendix showed significant dilation and a thickened and enhancing wall, with the presence of periappendiceal inflammation demonstrated by stranding of the adjacent fat (Figure 1D–1F). There was also a conglomerate of small bowel loops near the appendix tip due to the inflammation induced by the appendix (Figure 1D) (Figure 1E). Video of the abdominal CT is available as a supplementary material.

In face of the findings, a diagnosis of acute appendicitis associated with intestinal malrotation was made, and emergency surgical intervention was planned. After careful consideration, the surgical team opted for a laparoscopic approach to perform the appendectomy. The positioning of the ports, however, had to be adjusted to allow for optimal access to the LLQ. The camera port was normally positioned in the umbilicus, while the two working ports were in the suprapubic and RLQ positions (Figure 2).

The laparoscopy confirmed the radiologic findings, as the small bowel was to the right of the midline and the cecum to the left. There was absence of the Treitz ligament. Those findings correspond to intestinal malrotation type 3A according to Stringer classification (Table 1). There was no evidence of complications directly related to the malrotation, such as volvulus, small bowel obstruction or internal hernia. The appearance of the appendix suggested uncomplicated acute appendicitis, as was later confirmed by the histopathological analysis. After bipolar cauterezation of the mesoappendix, appendectomy was performed with the use of a linear stapler at the base of the appendix. There were no complications in the intraoperative or postoperative period. Follow up showed a healthy patient, without any early morbidities related to the surgery.

<table>
<thead>
<tr>
<th>Type</th>
<th>Fetal development stage in which the error occurred</th>
<th>Resulting defect</th>
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<tbody>
<tr>
<td>Type 1: nonrotation (less than 90 degrees rotation).</td>
<td>Stage 1 (before 6 week): during physiologic herniation.</td>
<td>The duodenum remains to the right of the midline, and the colon to the left. Absence of the ligament of Treitz.</td>
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<td>Type 2: malrotation primarily affecting the duodenum.</td>
<td>Stage 2 (6 to 10 weeks): normally the duodenal loop rotates, while the colon stays relatively fixed, therefore the colon is less affected by a defect in this stage.</td>
<td>Type 2A: Interrupted duodenal rotation with normal colonic position. Ladd bands formation</td>
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<td>Type 3: malrotation primarily affecting colonic rotation or fixation.</td>
<td>Stage 3 (after 10 weeks): normally is when the bowel loops return to the abdominal cavity, initiated by the duodenojejunal segment, followed by the colon and cecum.</td>
<td>Type 2B: Reverse rotation of the duodenum and colon. Duodenum is anterior to the SMA, while the transverse colon is posterior to it. Cecum may be normally located in the RLQ.</td>
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<td>Type 2C: Reverse rotation of the duodenum with normal colon rotation. Small bowels are surrounded by mesentery, with the risk of internal herniation. Cecum may be normally located in the RLQ or shifted to the left by the small bowel loops.</td>
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<td>Type 3A: Duodenum is to the right of the midline and the cecum is at a high position. The mesenteric vascular pedicle is very narrow, predisposing to volvulus. This is the most typical pattern of malrotation.</td>
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<td>Type 3B: Duodenum is to the right of the midline. Cecum may be in the RLQ, but the colon does not become fixed to the hepatic flexure, predisposing to Ladd bands formation.</td>
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<td>Type 3C: Incomplete fixation of the cecal mesocolon, leading to a mobile cecum. Duodenum is usually in its normal position, with a wide vascular pedicle. Patients are usually asymptomatic.</td>
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<td>Type 3D: Abnormal fixation of the duodenum and jejunum, leading to paraduodenal hernias. This type does not represent a true intestinal malrotation, instead it is a defect of intestinal fixation.</td>
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</table>

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Figure 1: Chest and abdominal CT images. A) chest CT showing situs solitus; B) abdominal CT with abnormal relation between the superior mesenteric vein and artery, small bowel loops on the right side of the abdomen, and both the ascending and the descending colon on the left side, the latter being more posterior and more to the left; C) abdominal CT showing the position of the cecum in the left iliac region; D) abdominal CT showing the appendix origin from the cecum in the LLQ, and extending anteriorly and to the right; E) abdominal CT that shows the conglomerate of small bowel loops induced by the inflammatory process; F) abdominal CT with the tip of the appendix ending in the hypogastric region. A, aorta; IVC, inferior vena cava; SMA, superior mesenteric artery; SMV, superior mesenteric vein; LRV, left renal vein; AC, ascending colon; DC, descending colon; SBL, small bowel loops; C, cecum; I, ileum; AB, appendix base; MA, mid appendix; AT, appendix tip; SC, sigmoid colon; * conglomerate of small bowel loops.

Figure 2: Immediate postoperative image of the abdomen, showing the position of A) the camera port; B) one working port in the suprapubic region and C) another working port in the RLQ.

Discussion

The various forms of intestinal malrotations compose a spectrum of bowel positional abnormalities, caused by nonrotation or incomplete rotation of the primitive bowel loop. During normal fetal development, the midgut herniates through the umbilical ring into the vitelline duct in the fourth week. In the tenth week of development, the intestine initiates its return to the abdominal cavity, completing a 270-degree rotation in the counterclockwise direction around the axis of the superior mesenteric artery (SMA). The duodenojejunal segment returns first to the cavity and becomes fixed to the left upper quadrant (LUQ) through the ligament of Treitz. Afterwards, the cecum returns and becomes fixed to the RLQ. By the twelfth week, the rotation is complete, and the colon becomes fixed to the retroperitoneum.1,7 When something goes wrong during this process, depending on the stage in which the error occurs, different types of malrotation result.

One of the first proposed classification for intestinal malrotation was published by Long et al.4 The study described the anatomical variations found in 69 children with malrotation of the gut, based on radiographic studies and surgical records. Seven patterns of duodenal malrotation were identified: a) nonrotation of the duodenum, in which the duodenum and the jejunum remain to the right of the midline and the ligament of Treitz is absent (30%); b) malrotation with a corkscrew duodenum and jejenum (29%); c) partial rotation of the duodenum, in which the duodenum rotates behind or anterior to the SMA but does not reach the LUQ, so the jejunum remains in the right upper quadrant, and the ligament of Treitz may be absent or abnormally positioned (22%); d) partial rotation of the duodenum with the duodenojejunal junction over the right vertebral pedicle, and the ligament of Treitz may be absent or abnormally positioned (4%); e) malrotation with low position of the duodenojejunal junction (3%); f) malrotation with redundant duodenum to the right of the midline, and with the jejunum in the LUQ (4%); g) malrotation with a Z-shaped duodenum and jejenum (4%) (Figure 3). Those patterns of duodenal malrotation were combined with three patterns of colonic rotation: a) normal rotation of the colon; b) partial rotation of the colon, in which the cecum is in the right or left upper quadrant; c) nonrotation of the colon, in which the cecum is in the LLQ (Figure 4). There was only one case in the series that had isolate malrotation of the colon, without duodenal malrotation.

A few years later, in 2000, Stringer et al. proposed his homonymous classification for intestinal malrotation, based mainly on the embryologic stage in which the rotation or fixation error would have occurred6 (Table 1). Although it is hard to define the actual incidence of intestinal malrotation, it is estimated to be around 1:6000 live births.1 Contrary to the common belief that malrotation is an infancy problem rarely seen in adults, a study that analyzed data from 170 malrotation patients showed that 48% of the cases were diagnosed in adulthood.9 Only 31% were diagnosed under one year of age and 21% were between one and eighteen-year-old patients.9 Another study showed that among 24 cases of malrotation, 42% were adults10. Both studies also showed that symptomatic adults tend to have a more chronic course of signs and symptoms, and diagnosis is usually delayed in that age group compared to the pediatric population.9,10

Complications related to malrotation of the gut can be acute or chronic, the latter being more common in adults.9,10 in some cases presenting symptoms of malrotation for nearly 20 years.11

Complications are generally caused by the anomalous anatomy commonly found in those patients, including narrowed superior mesenteric vascular pedicle, increasing the risk of volvulus; presence of Ladd bands or reverse rotation of the duodenum, both of which may cause small bowel obstruction; and bowel or mesentery fixation defect, potentially leading to the formation of internal hernias.

Although acute appendicitis is probably not directly affected by the abnormal anatomy as are the aforementioned complications, it also poses a great risk to patients with malrotation. If we can assume that the high incidence of acute appendicitis in the general population can be extrapolated into the population with malrotation, we conclude that it is a very common problem among malrotation patients. However, the atypical presentation and the difficulty in the interpretation of radiologic studies in that group may cause a grave delay in the diagnosis and surgical intervention, potentially resulting in higher morbimortality.

Another factor that contributes to the higher susceptibility in those patients is the very scant number of studies comparing different approaches and analyzing outcomes. A study by Akbulut et al. showed there were only 26 cases of left-sided appendicitis associated with intestinal malrotation published in the English language literature from 1893 to 2010. Other 69 cases of left-sided appendicitis were analyzed in the same study, but they were due to situs inversus. Of the total 95 cases, only eleven (12%) underwent laparoscopic appendectomy. Laparoscopy was also performed in two other patients, but in those cases there was conversion to open surgery due to technical reasons. There aren’t any comparisons of outcomes and complications related to the technique chosen, since the cases are so sparse.

![Figure 3 Patterns of duodenal malrotation according to Long et al.](Image)

![Figure 4 Patterns of colonic malrotation according to Long et al.](Image)

**Conclusion**

Although there is not enough evidence in the current literature to support the systematic use of the laparoscopy approach in the treatment of appendicitis associated with intestinal malrotation, it is the opinion of the authors of this paper that the laparoscopic treatment, when performed by an experienced surgeon and with the proper adaptations arranged, is feasible in those patients. Besides the possible advantage of decreased need for postoperative analgesia, more rapid return to full activities, shorter hospital stays and less wound infections, laparoscopy also allows for a minimally invasive thorough evaluation of the abnormal anatomy, providing a more precise understanding and inventory of the anomalies found. Therefore facilitating a more effective assessment and planning of additional surgical procedures when needed, including possible correction of the malrotation and its associated complications.

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