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Case Report

Acute appendicitis in an adult patient with congenital intestinal malrotation

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ABSTRACT

Intestinal malrotation is a rare congenital disorder that consists of incomplete rotation or non-rotation of the mid-primitive intestine during embryonic development. It tends to manifest itself in the first months or years of life, and in adulthood its manifestation is rare. We present the clinical case of a 25-year-old woman with a history of acute abdominal pain predominantly in the right iliac fossa, vomiting and anorexia. Intraoperatively, the clinical suspicion of acute appendicitis was confirmed, however in an atypical location associated with intestinal malrotation, having led to the need to adapt the laparoscopic approach.

Keywords: Left-sided acute appendicitis, Intestinal malrotation, Appendectomy

INTRODUCTION

Acute appendicitis (AA) is often associated with a typical clinical presentation of abdominal pain that begins in the midline of the abdomen before locating in the right iliac fossa (RIF), which is the most common location of the cecal appendix.[1,2] In these cases, the diagnosis of AA is generally predicted by the clinical presentation, classically without the need to resort to an imaging study before requiring surgical intervention.[1] However, the location of abdominal pain may vary if the cecal appendix is located in another region of the peritoneal cavity, making an atypical clinical presentation more likely to occur.

Usually happens in patients with situs inversus totalis or intestinal malrotation.[2-4] Intestinal malrotation is a congenital abnormality that arises from incomplete rotation or non-rotation of the midgut about the axis of the superior mesenteric artery (SMA) during embryonic development.[2] The incidence of intestinal malrotation is 1 in 6,000 births and the diagnosis in adulthood is rare.[4-6] Most cases occur in the first weeks of life, with a clinical presentation of intestinal obstruction and the remaining cases generally are asymptomatic.[1,2,5] The probability of AA in patients with intestinal malrotation is the same as in the general population.[1] We present a case report of AA with a typical clinical presentation, presenting intraoperatively with intestinal malrotation and a need to readapt the surgical approach.

CASE REPORT

A 25-year-old female, with no relevant past medical history or previous surgeries or medication. The patient went to the emergency department presented with abdominal pain for six hours, with an insidious onset located in the epigastrium and subsequent migration to the RIF. The patient had also an episode of vomiting and developed anorexia. The patient denied fever, changes in bowel habits, urinary symptoms or leucorrhoea. The last menstrual period was 14 days ago and the patient reported unprotected sexual intercourse. On physical examination, the patient had no fever and was hemodynamically stable. The abdomen was painful on palpation of the suprapubic region and RIF with signs of

peritoneal irritation. The analyses showed leukocytosis (22.21×109/l) with neutrophilia (79%) and normal C-reactive protein. The urine analysis was normal and the serum beta HCG was negative. A computed tomography (CT) with contrast of the abdomen and pelvis was performed, which raised the suspicion of AA.

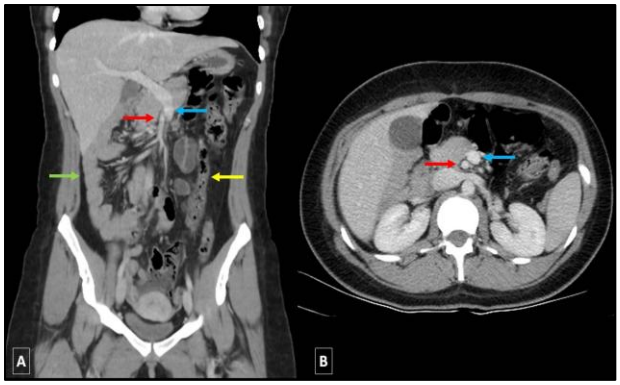

Figure 1: (A and B) CT scan showing the small bowel predominantly located on the right (green arrow) and the colon on the left of the peritoneal cavity (yellow arrow); Inversion of the relationship between SMA (red arrow) and SMV (blue arrow); coronal plane; and transverse plane.

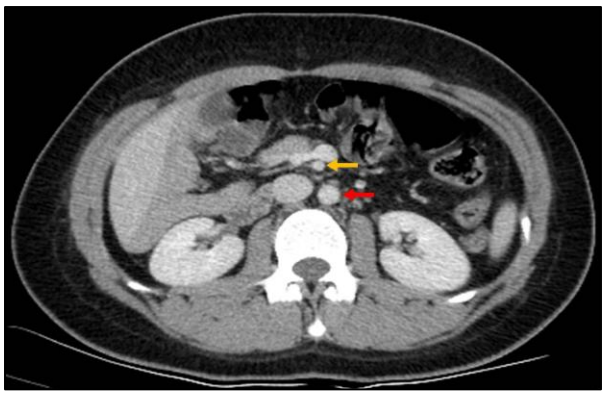

Figure 2: CT scan with absence of the third portion of the duodenum between the aorta (red arrow) and the SMA (orange arrow); transverse plane.

However, intestinal malrotation was not identified preoperatively despite the images showed the following findings: the colon was predominantly located on the left and the small bowel on the right of the midline, the relationship between the SMA and the superior mesenteric vein (SMV) was inverted with SMA on the right and the VMS on the left (Figure 1), the third portion of the duodenum was not located between the aorta and SMA (Figure 2) and, lastly, the cecal appendix was oriented from the caecum which was located on the left of the abdominal midline, with its apex ending in the suprapubic region, with a diameter of 9 mm.

The patient underwent urgent exploratory laparoscopy. Initially, a 12 mm trocar was inserted at the supraumbilical level with identification of cloudy fluid in the pelvic cavity and RIF and with no visualization of the cecal appendix. Additionally, a 12 mm trocar was inserted in the suprapubic region and a 5 mm trocar in the left iliac fossa (LIF). Peritoneal cavity exploration identified that cecum was located in the LIF, the small bowel was on the right and acute edematous appendicitis was confirmed (Figures 3 and 4).

After these findings, it was necessary to place an additional 5 mm trocar in the RIF to allow triangulation and facilitate the ergonomics of the surgery. An appendectomy was then performed. The postoperative period was uneventful and the patient was discharged on the third postoperative day. The histopathological report confirmed the diagnosis of acute appendicitis.

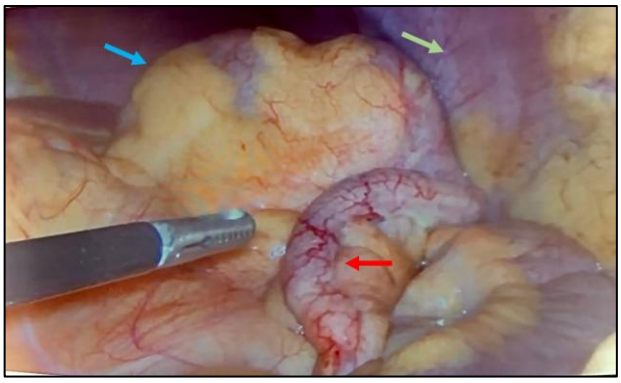

Figure 3: Location of the cecum (blue arrow) close to the left paracolic gutter (green arrow) and the cecal appendix with acute inflammation (red arrow).

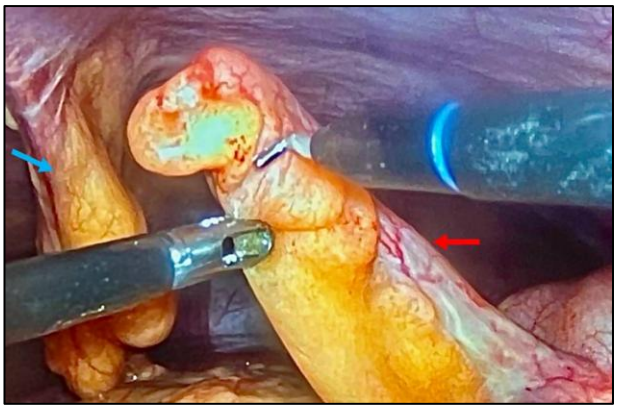

Figure 4: Cecal appendix (red arrow) located to the left of the falciform ligament (blue arrow).

DISCUSSION

After the fifth week of embryogenesis, the primitive intestine is made up of three divisions: anterior, middle and posterior primitive intestine. The midgut develops and gives rise to a large part of the adult intestine. This begins in the duodenum, at the level of the ampulla of Vater and extends to the junction of the proximal two thirds with the distal third of the transverse colon. During

embryonic development, the midgut begins a rapid stretching process, exceeding the capacity of the abdominal cavity to contain it, leading to a temporary physiological herniation through the umbilical cord at the sixth week of embryogenesis.

Coincidentally with the growth in extension of the midgut, there is a counterclockwise rotation of approximately 270 degrees around the axis of the SMA. In the tenth week of embryogenesis, there is a progressive reduction in physiological herniation, which is complete around the 11th week.[8] Specifically, at this stage there is the possibility to occur intestinal malrotation. Interruption in this process results in non-rotation or incomplete rotation of the intestine. Non-rotation occurs more frequently than incomplete rotation. When it happens, the small intestine is located to the right of the abdomen and the colon is in a contralateral position, a condition verified in this case report. Patients with non-rotation often remain asymptomatic and the majority are diagnosed incidentally through imaging studies or during surgical interventions motivated by other clinical conditions. Incomplete rotation occurs when there is a sudden stop in rotation, generally at 180 degrees.[9]

AA is one of the most common conditions requiring emergency surgery.[10] The typical clinical presentation consists of initially vague epigastric or periumbilical pain with subsequent migration to the RIF, associated with anorexia, nausea or vomiting and low-grade fever, with physical exam revealing peritoneal irritation with a positive Blumberg sign at McBurney's point. However, these typical findings occur in only 50 percent of patients.[2,3] When the clinical presentation is atypical with pain located in the LIF, there is an even greater probability of error or delay in diagnosis in around 24 percent of cases, which was not observed in this case.

CT scan raised the suspicion of acute appendicitis, however intestinal malrotation was only detected intraoperatively, leading to the need for adaptation by the surgical team with the placement of an additional trocar in the RIF.[3,11] The fact that it is a rare condition, diagnosing intestinal malrotation may pose a challenge for the Radiologist too. The identification of intestinal malrotation prior to surgery would allow the placement of the trocars/laparoscopic tower at the beginning of the surgery in a position that would allow the LIF approach early.

The most frequent imaging findings of acute appendicitis associated with intestinal malrotation correspond to the position of the duodenojejunal junction and the small bowel on the right, the colon on the left and the inversion of the position of the SMA and SMV, in association with the typical imaging findings of acute appendicitis.[2] The recommended treatment for acute appendicitis with intestinal malrotation consists of appendectomy, being the laparoscopy recommended as it allows the benefits of minimally invasive surgery, namely shorter recovery time, less pain in the postoperative period and better aesthetic results.[1,4]

CONCLUSION

Acute appendicitis in association with intestinal malrotation can lead to an atypical clinical presentation, delaying the diagnosis and corresponding treatment, raising the probability of complications occur. The diagnosis is generally challenging and CT scan is essential for the diagnosis to be established. Treatment consists of appendectomy, preferably laparoscopically.

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