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Case Report

Left Sided Acute Appendicitis in Midgut Malrotation

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Abstract

Acute appendicitis is the most common acute surgical disease in adult, it affects about 7% of the Western population and has typical clinical presentations such as periumbilical pain radiating to the right lower quadrant of the abdomen with peritoneal reaction on palpation. But asymptomatic anatomic anomaly like midgut malrotation can lead to atypical clinical symptoms and result in misdiagnosis in the setting of relatively common intestinal disorders. Here, we report a case of a 48-year-old male diagnosed with acute appendicitis and asymptomatic midgut malrotation. The patient presented pain at the left lower abdomen associated with nausea and vomiting. Abdominal computed tomography revealed left sided appendicitis with non-rotational type midgut malrotation. Emergency exploratory laparoscopy revealed ruptured acute appendicitis with peritonitis. The complete intestinal malrotation was identified, and a perforated appendix was observed at the left of the abdomen. Laparoscopic appendectomy was performed. The postoperative course was uneventful, and the patient was discharged on the eighth postoperative day.

Anatomic anomaly in midgut malrotation affects the typical clinical presentations and results in delayed diagnosis of acute appendicitis. Surgeons should have the possibility of altered anatomy in mind and appropriate diagnosis and treatment based on imaging study should not be delayed.

Keywords: *Acute Appendicitis; Intestinal Malrotation; Laparoscopy; Appendectomy*

Abbreviations

CT: Computed Tomography; HS-CRP: High Sensitivity C Reactive Protein; ESR: Erythrocyte Sedimentation Rate

Introduction

Acute appendicitis is the most common acute surgical disease in adult and affects about 7% of the Western population, with an estimated mortality rate of about 0.2 to 0.4% [1]. Acute appendicitis usually has typical clinical presentations such as periumbilical pain radiating to the right lower quadrant of the abdomen with peritoneal reaction on palpation, fever, and anorexia. But asymptomatic anatomic anomaly can lead to atypical clinical symptoms and result in misdiagnosis in the setting of relatively common intestinal disorders. Midgut malrotation is associated with abnormal rotation and fixation of the primitive intestinal loop around the superior mesenteric artery axis during the first ten weeks of fetal life. Midgut malrotation is commonly diagnosed in neonatal period, while diagnosis in adults is rare and reported with an incidence of 0.2% [2]. We report a case of a 48-year-old male diagnosed with acute appendicitis and asymptomatic midgut malrotation.

Case Report

A 48-year-old man was admitted to the Emergency Department with an acute abdomen. He reported a one-week history of pain at the left lower abdomen associated with nausea and dyspepsia. He denied any other infective symptoms. He had visited his primary care

physician twice in one week, but no definite diagnosis was made. Vital signs were all within normal limits. On physical examination, the abdomen was mildly distended and tympanic on percussion. Physical examination revealed left lower quadrant tenderness and rebound tenderness, and audible hyperactive bowel sounds. There were no other remarkable findings. Laboratory tests showed normal white cell count (9.56 × 103/mL with left shift), high sensitivity C reactive protein (HS-CRP) level of 17.87 mg/dL (normal range < 0.75 mg/dL) and erythrocyte sedimentation rate (ESR) level of 17 mm/hr (normal range < 16 mm/hr). Provisional diagnosis was diverticulitis, with differentials including pyelonephritis, inflammatory bowel disease, and ischemic bowel disease. A computed tomography (CT) scan of the abdomen revealed left sided cecum and acute appendicitis with periappendiceal inflammation (Figure 1A and 1B), and findings of complete intestinal malrotation (type 1a, non-rotation) with entire colon shifted to the left and small bowel moved to the right (Figure 2A and 2B). Emergency exploratory laparoscopy revealed ruptured acute appendicitis with peritonitis. The complete intestinal malrotation was identified. Laparoscopic appendectomy was performed (Figure 3). The histopathologic finding of resected appendix revealed a suppurative stage of acute appendicitis. The postoperative course was uneventful. The patient was discharged on the eighth postoperative day.

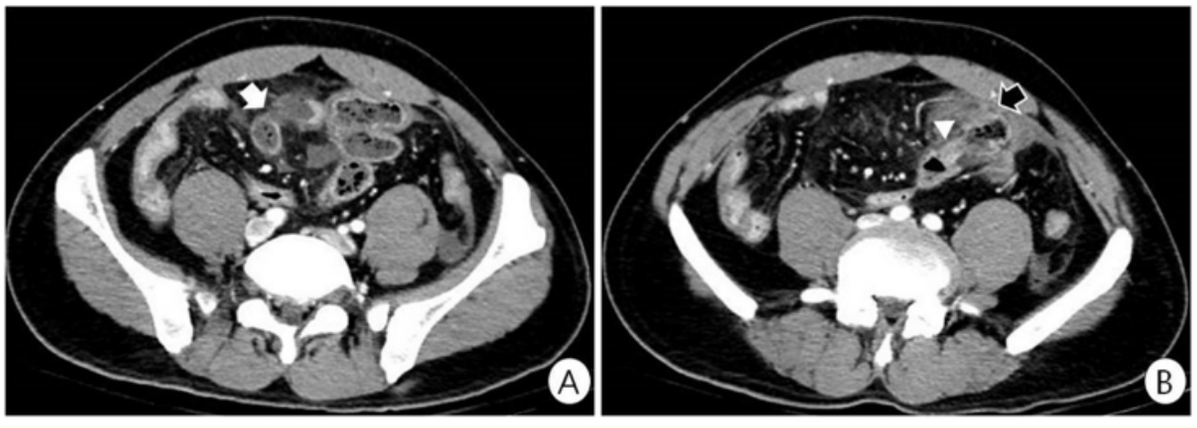

Figure 1: *CT scan (axial image at the level of sacral promontory) shows (A) dilated left sided appendix (arrow) with wall enhancement, fat stranding and peritoneal thickening, (B) cecum (arrow) and ileocecal valve (arrowhead).*

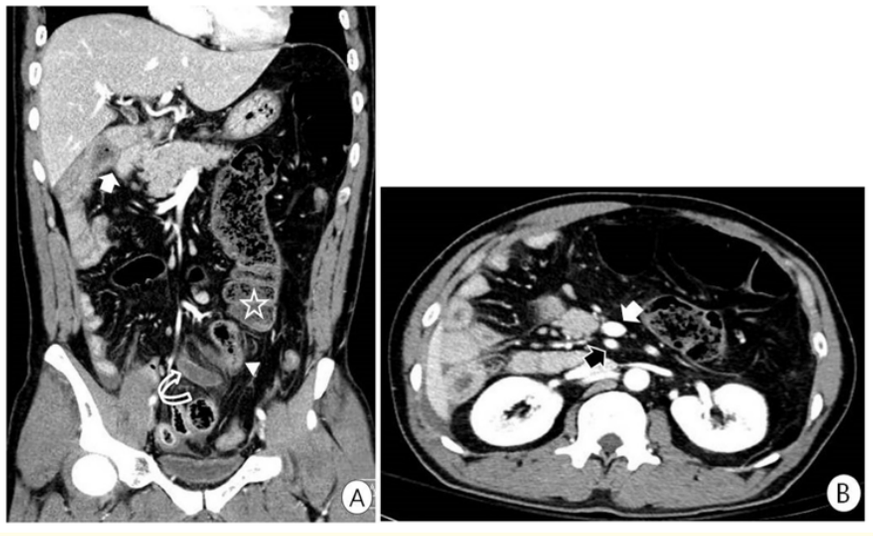

Figure 2: *CT scan shows type 1a (non-rotation) midgut malrotation, (A) right sided duodenojejunal junction (arrow), left sided cecum (star) and terminal ileum (arrowhead) with midline appendix (curved arrow) and (B) inversion of superior mesenteric artery (black arrow) and superior mesenteric vein (white arrow).*

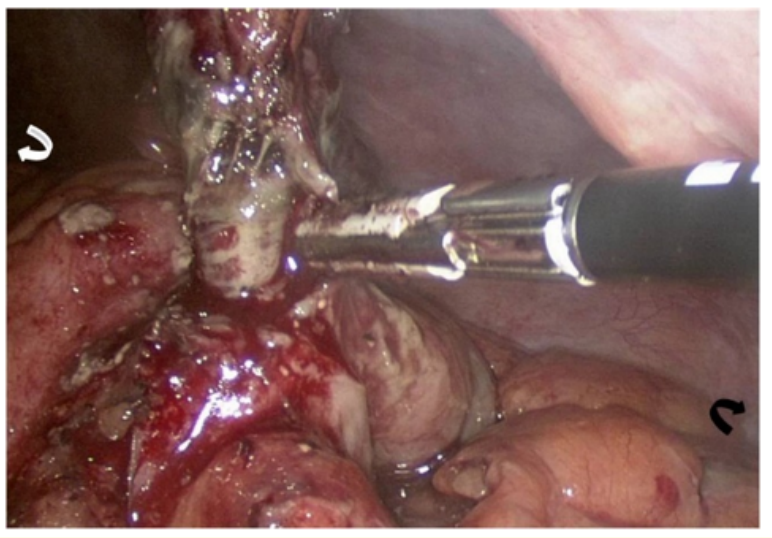

Figure 3: *Finding of an exploratory laparoscopy: Gangrenous and perforated appendix was observed at the left of the abdomen (white curved arrow: cranial, black curved arrow: caudal).*

Discussion

Midgut malrotation is a rare congenital anomaly resulting from incomplete or failure of midgut rotation and fixation during embryonic development. Normal midgut rotation consists of the change from a short straight gut at about the 4th week of gestation having rotated 270° counter clockwise at about the 12th week. The first stage is the physiologic herniation of the midgut between weeks 6 and 8 of development, the second stage (tenth week of gestation) is its return to the abdomen, and at 12th week the third stage is the fixation of the midgut. Abnormalities in this normal process lead to various malformations. It has been classified into three major types, including non-rotation (type I), duodenal malrotation (type II), and combined duodenal and cecal malrotation (type III) [3,4]. This presented case is in the non-rotation type (type I), which is characterized by right-sided duodenojejunal junction, predominantly right-sided small bowel, left-sided colon with an unusual position of the cecum, transposition of the superior mesenteric artery and vein, and hypoplasia of the uncinate process of pancreas. Type I (non-rotating) is estimated to be an incidental finding in 0.2% of adults and is expected to be the most frequent [5]. Acute appendicitis is one of the most common abdominal surgical diseases [1]. Although right lower abdominal pain is a typical symptom, several cases of acute left-sided appendicitis with midgut malrotation have been reported with an estimated incidence of 0.04% [6]. The diagnosis of this combination is challenging because of atypical symptom and sign. Left-sided appendicitis generally presents with left lower abdominal pain and is confused with diverticulitis or gynecological diseases. Altered anatomy in acute appendicitis with midgut malrotation affects the clinical presentation and results in delayed diagnosis and possibly also in an increased incidence of complications such as abscess and perforation, resulting in severe consequences or increased morbidity and mortality rates. Midgut malrotation in adults is diagnosed incidentally when detected in imaging studies or during evaluation of another intra-abdominal pathology.

Conclusion

Anatomic anomaly in midgut malrotation affects the typical clinical presentations and results in delayed diagnosis of acute appendicitis. Surgeons should have the possibility of altered anatomy in mind and appropriate diagnosis and treatment based on imaging study should not be delayed.

Conflict of Interest

The author has no conflict of interest.

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