

Intestinal Malrotation and Appendicitis in Adults

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Abstract

Background: Intestinal malrotation is a rare condition resulting from an incomplete midgut rotation and fixation; most of the cases were diagnosed in the newborn and the presentation at adulthood is traditionally considered as rare. In the same time appendicitis is a very common surgical condition with different clinical presentations and the diagnosis can be challenging especially when there is an alteration to the normal position of anatomical structures.

Case report: We present the case of 19 years old patient admitted for diffuse abdominal pain and dyspepsia. The physical exam revealed peri umbilical rebound tenderness. The biochemical tests revealed an inflammatory syndrome, with leucocytosis (WBC at 12,000/mm³) and elevated CRP (62 mg/L). The ultrasound exam was not contributive. A CT scan was performed and revealed the inflammation of ileo-caecal area and aspect of intestinal malrotation, the common mesentery. An exploratory laparoscopy was performed and confirmed the acute appendicitis and intestinal malrotation. Appendectomy was performed; the postoperative course was uneventful, and the patient was discharged at 3 postoperative days.

Conclusion: The intestinal malrotation in adults is a relatively rare condition and often asymptomatic. On the other hand, appendicitis is a common condition with various clinical presentations; in cases where peritoneal signs were located elsewhere other than the right iliac fossa, surgeons could bear in mind the possibility of underlying intestinal malrotation, as this could be the first presentation of this congenital condition. The laparoscopic approach is feasible and allows the diagnosis of the intestinal malrotation type and treatment of associated condition.

Keywords: Intestinal malrotation; Common mesentery; Appendicitis; Laparoscopy; Appendectomy

Introduction

Intestinal malrotation is a generic term and represents any abnormality in the rotation and fixation of the gastro-intestinal tract during its fetal development [1]. The typical 270° intestinal rotation during fetal development can occur at a wide range of locations and can lead to various acute and chronic presentations of the disease [1]. The malrotation abnormalities were reported in 1898 by Mall FP [2]. The Incidence is variable upon different studies: 1/6000 live births [3]; 0.2% in barium swallow studies in infants [3]; 1% of general population in autopsy studies [3,4]. Most of the cases were diagnosed in the first weeks of life and the presentation at adulthood is rare.

On the other hand, appendicitis is a common surgical condition, with various clinical presentations and could mimic other diseases as cholecystitis, diverticulitis, complicated ovarian cysts, and pelvic inflammatory disease. The diagnosis could be further obscured by underlying undiagnosed anatomical anomalies, for instance intestinal malrotation [5].

Case Presentation

A 19 years old, young woman, BMI at 19 kg/m², was admitted for diffuse abdominal pain and nonspecific dyspepsia. The physical exam revealed peri umbilical rebound tenderness. The biochemical tests revealed an inflammatory syndrome with leukocytosis (white blood cells count at 12,000/mm³) and elevated CRP (62 mg/L). The ultrasound exam was not relevant; however, excludes an adnexal pathology. The CT scan revealed inflammatory infiltration of the ileo-caecal area without intraperitoneal fluid collections and aspect of intestinal malrotation, the common mesentery type (Figure 1).

An exploratory laparoscopy was performed and confirmed acute appendicitis and the common mesentery. Appendectomy was performed in usual manner: trocars placement in umbilical region, suprapubic and

left iliac fossa; appendicular meso divided using bipolar and monopolar energy; appendicular base ligated by a Vicryl 1 endo-loop and then divided; the resected specimen was extracted through the umbilical trocar using an endo-bag (Figure 2; Video).

The postoperative course was uneventful, and the patient was discharged at 3 postoperative days. The pathological exam confirmed the acute catarrhal appendicitis.

Discussion

The intestinal tube, the aorta and the big vessels develop from the mesenchymal tissue [6,7]. In 3rd week the mesenchymal tissue fissures on both sides of the gastro-intestinal tube to form the coelomic cavity: future peritoneal cavity. [6,7] This cavity will progressively grow and its epithelium lines the digestive tract and vessels forming a dorsal meso (median and sagittal, between aorta and GI tube) and respectively a ventral meso, shorter, in front of the stomach and of the first duodenum, in which the liver buds [6,7].

Then the primitive bowel, initially in sagittal position, completes a 270° counter clockwise rotation around the superior mesenteric artery: 1) the first rotation 90° (during the week 6); 2) the second rotation 90° (during the week 7 to 11); 3) the third rotation 90° (during the week

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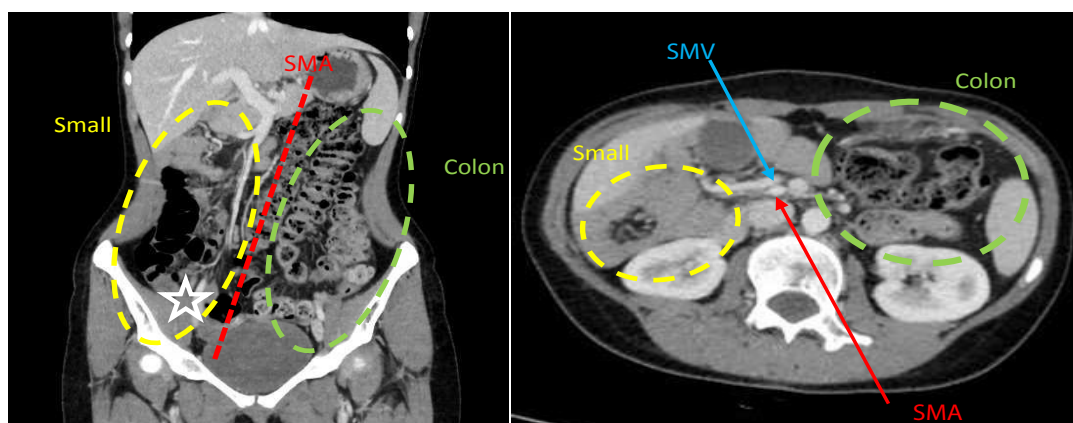


Figure 1: CT scan: coronal (left) and axial (right) sections. Aspect of common mesentery; small bowel is at the right side of the spine and of the superior mesenteric artery (SMA) axis and the colon is on the left side. The white star indicates the inflammation of ileo-caecal area.

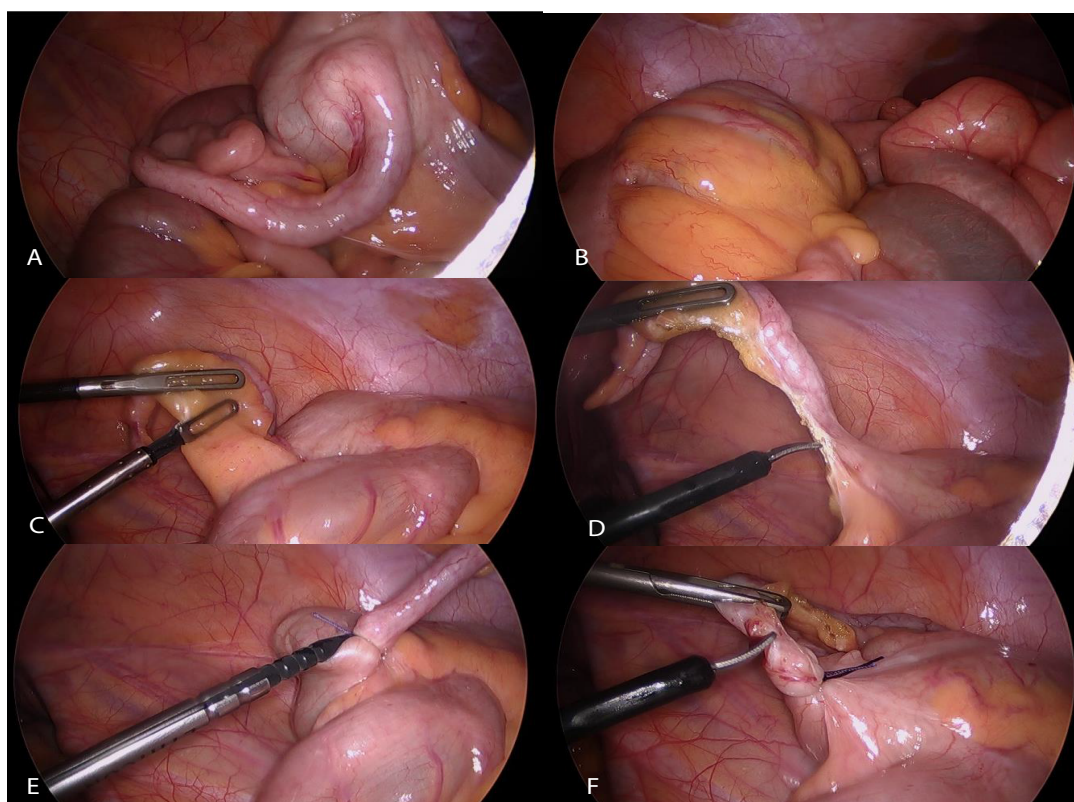


Figure 2: Laparoscopic appendectomy. A) Confirmation of acute appendicitis; B) Confirmation of common mesentery; the right colon is mobile and the small bowel is located on the right side of midline; C) Division of mesoappendix; D) Completing the mesoappendix division; E) Appendicular base ligation using Endoloop; F) Division of appendicular base.

12) [6-8]. The next step is the fixation of the ascending and descending colon forming Treitz and Toldt fascias with the final arrangement of the digestive tube. So, after a complete rotation and fixation, the duodeno-jejunal flexure is located at the left side of the spine (first lumbar vertebra) and ileo-caecal junction is in the right iliac fossa; the mesenteric root with superior mesenteric artery (and vein) lies between these two landmarks [6-8].

The rotation of the primitive intestine can stop at different moments resulting different forms of malrotation [1-4]. The common mesentery is the complete form, a total non-rotation of the intestine [1-4]; the small bowel, including the duodeno-jejunal flexure is located on the right side of the spine, like in our case. In the incomplete forms, the caecum can be situated at various levels

of the abdomen and can be attached to posterior peritoneum by fibrous tissue, the Ladd's bands (named after American pediatrician William Edwards Ladd (1880-1967)) [9]. The intestinal malrotation can lead to volvulus with intestinal obstruction and intestinal ischemia. Ladd's bands can pass over the duodenum causing extrinsic compression and obstruction. In these cases "Ladd procedure" is indicated to divide the bands, revealing the cause of obstruction [3].

The etiology of the intestinal malrotation is not known; several studies revealed a possible genetic etiology especially in syndromic form (in association with other malformation): in this way it was described mutations in the forkhead box transcription factor FOXF1, and mutations in genes controlling L-R patterning [10].

The intestinal malrotation is traditionally considered to be diagnosed in new-borns and infants [1]. However, a recent study on about 200 cases found that 31% were infants, 21% were aged 1 to 18 years, and the remaining 48% were adults [11].

Unfortunately for the new-born and infants the first sign is often the intestinal obstruction secondary to volvulus. The mortality remains elevated due to extensive intestinal ischemia [1]. In adults the diagnosis of intestinal malrotation is often an incidental finding in barium studies, angiographies, and computer tomography scans as found in literature and in our case [1,11-13].

The laparoscopic approach allows the exploration of entire abdominal cavity and diagnosis of the malrotation and of the type of malrotation, and the division of Ladd's bands and the treatment of the associated disorder (appendectomy, as in our case, colic, or bariatric surgery) [14-17].

Conclusion

The intestinal malrotation in adults is a relatively rare condition and often asymptomatic. On the other hand, appendicitis is a common condition with various clinical presentations; in cases where peritoneal signs were located elsewhere other than the right iliac fossa, surgeons could bear in mind the possibility of underlying intestinal malrotation, as this could be the first presentation of this congenital condition. The laparoscopic approach is feasible and allows the diagnosis of the intestinal malrotation type and treatment of associated disease.

Conflict of Interest

Authors have no conflict of interest to disclose.

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