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An Unusual Case of Intestinal Malrotation Causing Duodenal Obstruction by a Looped Appendix

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 30
Final Diagnosis: Duodenal obstruction caused by a looped appendix due to intestinal malrotation
Symptoms: Post postprandial vomiting
Medication: —
Clinical Procedure: Improved after unwinding of the looped appendix and subsequent appendectomy
Specialty: Surgery

Objective: Congenital defects/diseases

Background: Bowel obstruction is a mechanical or functional obstruction of the intestines which prevents the normal movement of the products of digestion. Intestinal malrotation is one of the rarest causes of mechanical bowel obstruction. In adults, the incidence rate is 0.2%, and 15% of all patients with confirmed diagnosis remain asymptomatic throughout life. Surgery is generally required when the patient is symptomatic.

Case Report: A 30-year-old man with multiple admissions for chronic intermittent colicky abdominal pain since childhood, was admitted for symptoms suggestive of proximal small bowel obstruction. Tomographic imaging identified a midgut malrotation and a duodenal obstruction by a non-diseased displaced appendix. Laparoscopic liberation of the duodenum and the terminal ileum was done successfully.

Conclusions: Intestinal malrotation is infrequently encountered in the adult population, but it should be kept in mind as a differential diagnosis whenever a case of acute intestinal obstruction in an adult presents without any significant past surgical history.

MeSH Keywords: Duodenal Obstruction • Intestinal Obstruction • Upper Gastrointestinal Tract

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Background

Bowel obstruction is a mechanical or functional obstruction of the intestines which prevents the normal movement of the products of digestion. Either the small or large bowel may be affected. Signs and symptoms include abdominal pain, vomiting, bloating, and non-passing of gas. Intestinal malrotation is one of the rarest causes of mechanical bowel obstruction. It is a congenital anomaly that was first reported by William Ladd in 1932. The source of this disease arises in the embryogenic stages when the bowels undergo a partial rotation or even non-rotation in the superior mesenteric artery axis [1]. Rotational anomaly of the midgut is uncommon in adults. The disease is usually symptomatic during the first couple of years, with some even claiming that 90% of patients will seek medical help during the first year of life [2]. However, certain cases are not symptomatic until much later in life, which may cause misdiagnosis [2]. Surgery is required generally when they are symptomatic. About 0.2% of the adult general population is affected, with 15% of diagnosed individuals remaining symptom-free [3]. The aim of this case report is to present a rare case of bowel obstruction consisting of duodenal obstruction caused by a displaced non-diseased appendix. This patient came to our hospital with a misdiagnosis and a delay in management, which might have increased the morbidity and mortality. We present this case to raise awareness of this rare diagnosis, thereby preventing delay of definitive diagnosis and management.

Case Report

A 30-year-old man who was a hepatitis B carrier with multiple hospital admissions for chronic recurrent post-prandial painless vomiting, was admitted to the hospital for investigations. A gastroscopy was done and was positive for a dilated fluid-filled stomach, without any signs of gastric outlet obstruction.

The patient was then discharged on medical treatment. A few weeks later the patient was admitted again for the same complaint but this time it was associated with a new onset of diffuse abdominal pain, colicky in nature, most severe in the epigastric area, radiating to the back. The pain increased in severity over the next 2 days and was associated with nausea and vomiting. During this admission, we (the general surgery team) were consulted for assessment.

His physical exam was unremarkable except for mild generalized abdominal tenderness and diffuse tympani on a soft abdomen. There were no signs of peritoneal irritation.

Laboratory tests were requested, including complete blood count, electrolytes, liver function tests, and pancreatic enzymes,

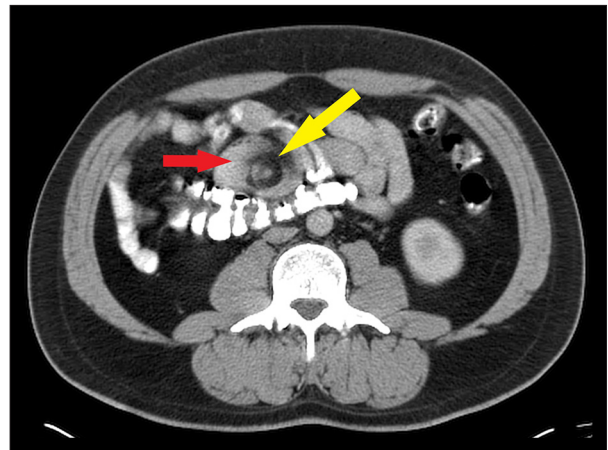


Figure 1. Abdo-pelvic CT scan with IV and PO contrast showing the whirlpool sign (yellow arrow) involving the jejunal loops (red arrow) at the level of the Treitz angle.

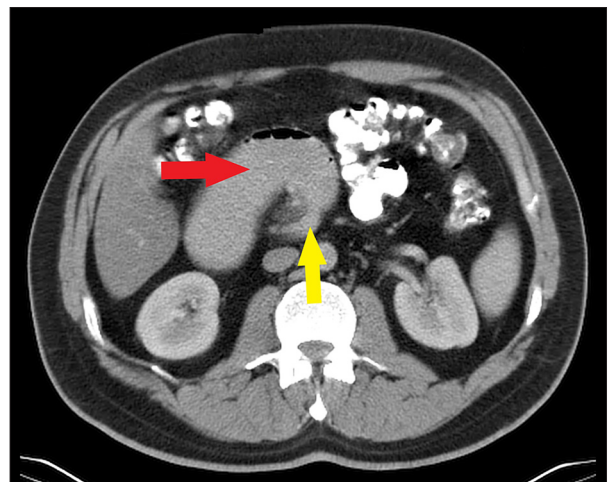


Figure 2. Abdo-pelvic CT scan with IV and PO contrast showing a dilated duodenal loop (red arrow) with a deflated bowel transition zone (yellow arrow).

and all results were within normal range. Computed tomography (CT) imaging was done and revealed a dilated fluid-filled stomach and duodenum (Figure 1). Along the axis of the superior mesenteric artery, intestinal malrotation was apparent, with a whirlpool sign at the level of the proximal jejunum (Figure 2). The patient was prepared for laparoscopic repair.

After establishing pneumoperitoneum and entering the abdominal cavity, our exploration showed that almost the entire large bowel was displaced to the left side and the small bowels to the right. The caecum was located to the right upper quadrant next to the dilated duodenum passing beneath it (Figure 3).

The jejunum was also found to be in a defect through the terminal ileum mesentery, without any signs of obstruction (Figure 4). Further dissection showed a band-like structure that

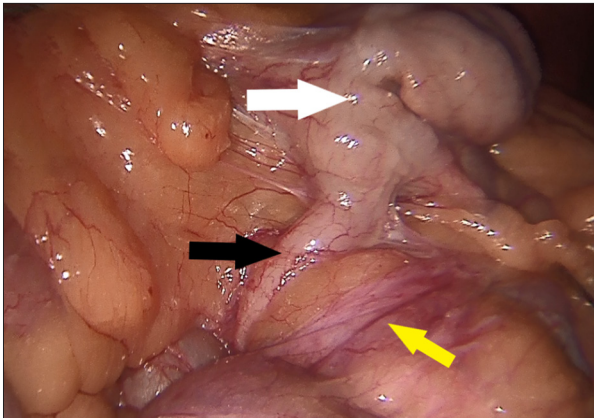


Figure 3. An intraoperative capture during laparoscopic exploration showing the highly positioned caecum (white arrow) and the base of the appendix (black arrow) covering the duodenum (yellow arrow).

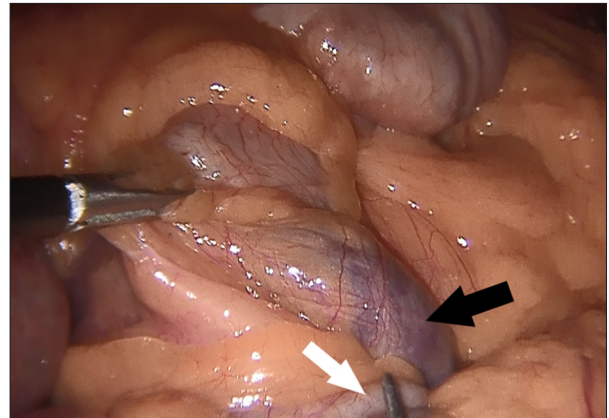


Figure 5. An intraoperative capture during laparoscopic exploration showing the appendix (white arrow) wrapped around the dilated duodenum (black arrow).

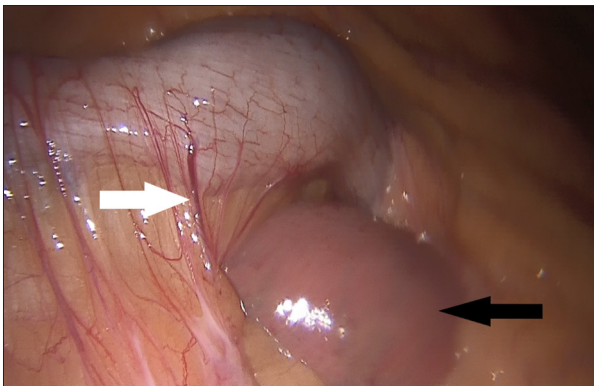


Figure 4. An intraoperative capture during laparoscopic exploration showing the jejunum (black arrow) passing through the defect in the terminal ileum mesentery (white arrow).

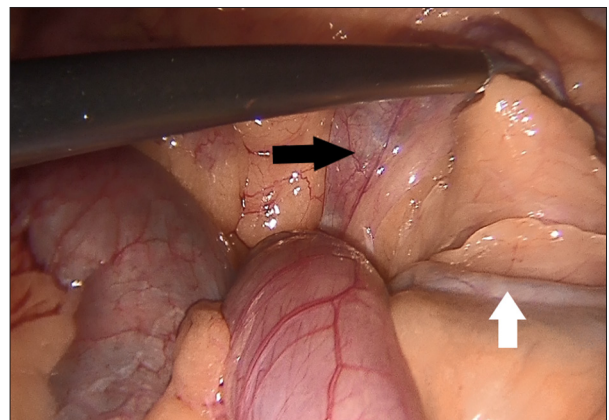


Figure 6. An intraoperative capture during laparoscopic exploration showing the appendix (white arrow) wrapped around duodenum (black arrow).

was encircling the duodenum, causing its narrowing, which was later found to be the appendix (Figures 5, 6).

After careful dissection, the anatomy was difficult to assess, so the decision was made to convert to laparotomy. After reentering the abdomen, the appendix was found to be encircling the duodenum two and a half turns, narrowing it and causing its obstruction, so an appendectomy was done followed by rotating the small bowels anticlockwise back to near-normal anatomy after liberation of the duodenum and terminal ileum. The post-operative course was smooth and unremarkable, with the patient passing flatus and stools on post-operative day 3, and he was then discharged home the following day.

Discussion

Intestinal malrotation is a developmental anomaly of the midgut in which there is failure in rotation of the bowels around the

superior mesenteric artery and in their fixation to the peritoneal cavity [1]. Rotational anomalies of the midgut are rare in adults, and, according to medical history, most cases are asymptomatic and go undiagnosed [2,4]. Such patients who become symptomatic will have an either acute or chronic clinical scenario: acutely, they present with bowel obstruction and ischemia, but chronically, they present with vague abdominal pain and recurrent vomiting [2,4].

In our case, changes were acute on top of a chronic congenital condition. The intestinal volvulus encountered was caused by the incorrect placement of the small bowels narrowing the pedicle of superior mesenteric vessels. These chronic vague symptoms increase the likelihood of misdiagnosis, and, more likely than not, the patient will seek medical help from several doctors, who will eventually attribute the complaints to intestinal motility or psychiatric illnesses [1,3].

Ultrasound can detect the reversal of the normal anatomic relationship between the superior mesenteric vessels, the whirl-pool sign of the midgut volvulus, and the bird-beak appearance of the duodenal obstruction, but with a false-positive rate of up to 21%, but contrast-enhanced CT of the abdomen shows intestinal malrotation with the midgut volvulus and anatomic location of small bowels and colon with features of obstruction and gangrene with very high sensitivity and specificity [3]. However, in acute and complicated cases, the diagnosis must be confirmed by surgical exploration [3].

Regarding treatment, there is no standard of care. Surgical correction of asymptomatic or incidentally discovered disease is controversial. Many clinical scenarios and complications exist because the intestinal malrotation is not a single congenital entity, but rather is a spectrum of rotational anomalies [5]. Elective Ladd procedure is becoming more commonly used for patients with a chronic clinical presentation, and emergent laparotomy is still the mainstay for patients with an acute and complicated presentation [6]. There is much debate surrounding asymptomatic patients. Some authors argue that early surgical intervention for an incidentally discovered malrotation is warranted because currently there are no modalities able to predict which asymptomatic patients will become symptomatic. This will avoid more catastrophic and probably

fatal complications such as midgut volvulus [6]. Other authors support watchful waiting for asymptomatic patients [7].

Conventional Ladd procedure remains the most common and effective method to alleviate intestinal malrotation. Currently, the growing number of laparoscopic Ladd procedures performed is encouraged by its good results, low morbidity, and short hospital stay [8].

Conclusions

Intestinal malrotation is infrequently encountered in the adult population; however, it should be considered as a differential diagnosis whenever a case of acute intestinal obstruction in an adult presents without any significant past history.

Department and Institution where work was done

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Conflicts of interest

None.

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