

Upside-Down Appendix: Report of a Rare Case

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Abstract

Background: Intestinal malrotation is a rare disorder that is usually diagnosed in infancy or childhood. In adults, it can manifest with a broad range of symptoms, ranging from acute abdomen to vague abdominal complaints, or it may be discovered incidentally during investigation or laparotomy for unrelated disease. Among types of intestinal malrotation, it is rare for the appendix to be rotated into the subhepatic space “upside-down appendix”.

Case Report: An 83-year-old woman underwent laparoscopic cholecystectomy for gallstones. During intraperitoneal lavage, the appendix was found to be in the subhepatic space. Review of preoperative abdominal computed tomography (CT) showed that the cecum and appendix had rotated into the subhepatic space. Postoperative barium enema examination confirmed that the appendix was located in the right upper quadrant of the abdomen.

Conclusion: With advances in imaging and surgical techniques, asymptomatic malrotation is being detected with increasing frequency. An atypically located appendix can manifest with atypical presentation, which may result in misdiagnosis. Clinicians should be aware that the appendix can have an atypical location.

Key words: *Subhepatic appendix; intestinal malrotation; adults, laparoscopy; appendicitis*

Introduction

Intestinal malrotation is a congenital anomaly of midgut rotation that usually presents in childhood. In older children and adults, malrotation can manifest with a broad range of symptoms, ranging from acute abdomen to vague and intermittent abdominal complaints, or it may be an incidental finding [1]. Among the types of intestinal malrotation, the subhepatic location of an undescended cecum and appendix “upside-down appendix” is rare.

Case Report

An 83-year-old woman was admitted to hospital with gallstones which had been diagnosed 20 years earlier. She had experienced no abdominal symptoms for many years, but had developed subcostal pain over the previous few months. Abdominal ultrasonogram (U/S) and computed tomography (CT) revealed a gallbladder filled with gallstones. She underwent uncomplicated laparoscopic cholecystectomy, during which her appendix was identified in the subhepatic space (Figure 1). Review of the preoperative abdominal CT showed intestinal malrotation of the

cecum and appendix into the subhepatic space (Figure 2). No morphological abnormality was found in the other segments of the small intestine and colon. The positional relationship of the superior mesenteric vein and superior mesenteric artery was also normal. Postoperative barium enema examination performed with the patient's consent revealed that the terminal ileum ascended toward the subhepatic portion in the retroperitoneum and was connected to the cecum (Figure 3). The ascending colon descended and passed across the transverse colon at the umbilical level. Her postoperative course was uneventful, and she has been symptom-free for years.



Figure 1. Subhepatic appendix in an 83-year-old female. Intraoperative image of the appendix identified within the subhepatic space during intraperitoneal lavage during laparoscopic cholecystectomy.

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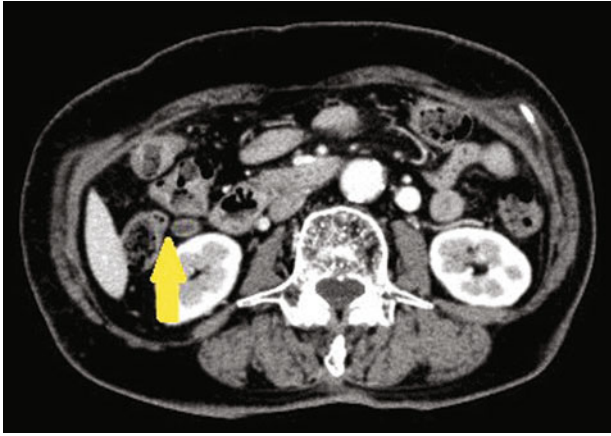


Figure 2. Subhepatic appendix in an 83-year-old female. Preoperative abdominal computed tomography (CT) showing the cecum and appendix rotated into the subhepatic space.

Discussion

The incidence of intestinal malrotation is 1/200 to 1/500 births, but it is estimated to be symptomatic in only 1/6,000 live births [2]. It is usually diagnosed in infancy or childhood. In adults, the diagnosis of midgut malrotation is very rare, with an incidence of 0.001%–0.19% [3], although, according to Rock and colleagues, about half (48%) of patients of all age diagnosed with intestinal malrotation were aged > 18 years, while 21% were aged 1–18 years, leaving only 31% diagnosed in the first year of life. Many cases of intestinal malrotation, therefore, remain asymptomatic in adults.

Among the various types of intestinal malrotation, it is rare for the appendix to occupy the subhepatic space. Development of the midgut occurs in three stages, namely umbilical herniation and rotation of the midgut loop, followed by further rotation and reduction into the abdominal cavity, and ending with fixation to the posterior abdominal wall [4]. In the 10th gestational week, a herniated midgut loop retracts into the abdominal cavity with counterclockwise rotation of 270° along the axis of the superior mesenteric artery. The first part to re-enter the abdominal cavity is the proximal jejunum, while the last to enter is the cecal bud. Just after retraction, the cecum and appendix occupy the subhepatic space. In the 11th gestational week, the cecum descends into the right iliac fossa [5]. As in the case presented here, failure of this step can result in a subhepatically located cecum and appendix.

Subhepatic appendix is usually identified at the onset of acute appendicitis. Although often found during surgery for other diseases, no reports have confirmed the existence of malrotation on postoperative radiology. This is a report of radiological demonstration of the presence of a subhepatic appendix in an adult.



Figure 3. Subhepatic appendix in an 83-year-old female. Barium enema examination showing the cecum and appendix located in the upper right abdomen (arrow).

Clinicians in daily clinical practice should be aware that the appendix can have an abnormal location due to intestinal malrotation.

Conclusion

We reported here the rare case of an appendix located in the subhepatic space, which was found incidentally during laparoscopic cholecystectomy. With advances in imaging and surgical techniques, asymptomatic malrotation of the gut is being detected with increasing frequency. Clinicians should pay close attention to slight differences from normal findings in daily practice.

Conflict of Interest: The authors declare that they have no conflict of interest.

Ethical Approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent: Informed consent was obtained from all individual participants included in the study.

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