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CASE REPORT

Appendiceal atresia causing recurrent right lower quadrant pain without inflammation

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Abstract
An 11-year-old girl presented with recurrent right lower quadrant (RLQ) pain. There was no evidence of inflammation and appendiceal swelling except at the initial onset. The repeated presence of a small amount of ascites at the time of abdominal pain triggered the performance of exploratory laparoscopy. Intraoperative examination revealed a non-inflamed, unswollen appendix with a cord-like atretic segment at the middle part and an appendectomy was performed. At 46 months follow-up, she remained asymptomatic. In patients with recurrent RLQ pain of unknown cause, it is necessary to consider diagnostic laparoscopy while keeping appendiceal atresia in mind as a differential diagnosis.

Keywords Appendiceal atresia · Intestinal atresia · Appendicitis

Introduction

Right lower quadrant (RLQ) pain is a clinical condition commonly encountered in the setting of pediatric emergency care. RLQ has many causative diseases, including appendicitis, gastroenteritis, mesenteric lymphadenitis, testicular torsion, adnexal torsion, and pelvic peritonitis, which can be difficult to diagnose in patients without typical findings [1, 2]. Structural abnormalities of the appendix such as duplication, anomalous implantation, horseshoe appendix, and atresia may also cause RLQ pain and can result in a misdiagnosis of patients with acute abdomen [3–5]. We herein reported a case of appendiceal atresia causing recurrent RLQ pain diagnosed by exploratory laparoscopy.

Case report

An 11-year-old girl with RLQ pain was referred to Fujita Health University Hospital. She had no previous history of similar symptoms. On examination, tenderness was localized in the right lower abdomen, and a blood test revealed an elevated serum C-reactive protein (CRP) concentration of 4.18 mg/dL. Abdominal ultrasonography showed appendiceal swelling without ascites or abscesses (Fig. 1a). Blood flow within the wall of the swollen distal appendix was confirmed (Fig. 1b), but the continuity of the appendiceal lumen to the cecum was not evaluated. The patient was diagnosed with appendicitis catarrhalis and treated conservatively. Although her symptoms were initially relieved, she developed several recurrent episodes of RLQ pain without signs of infection during the next 2 months. Imaging studies at the time of the abdominal pain showed no swelling of the appendix but occasionally revealed right paracolic ascites.

At 13 years of age, the patient revisited our hospital because of persistent RLQ pain that had begun the previous day. On arrival, she was afebrile and had localized tenderness without rebound tenderness or guarding in the right lower abdomen. Laboratory tests revealed no significant abnormalities; her white blood cell count ($5.5 \times 10^3/\mu L$) and serum CRP concentration (< 0.01 mg/dL) were normal.

Abdominal ultrasonography showed a normally sized appendix and fluid collections on the dorsal side of the cecum. Abdominal contrast-enhanced computed tomography revealed ascites confined to the caudal side of the cecum (Fig. 2). However, we observed no appendiceal swelling, peri appendiceal fat stranding, or other abnormal findings that may indicate specific causes of the patient's abdominal pain, such as ovarian torsion, mesenteric lymphadenitis, or diverticulitis. Pelvic magnetic resonance imaging

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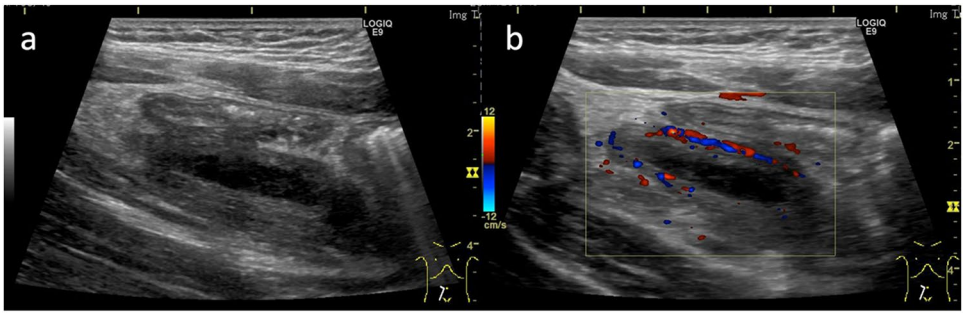

Fig. 1 **a**, **b** Longitudinal ultrasound image of the tip of the swollen appendix with wall blood flow

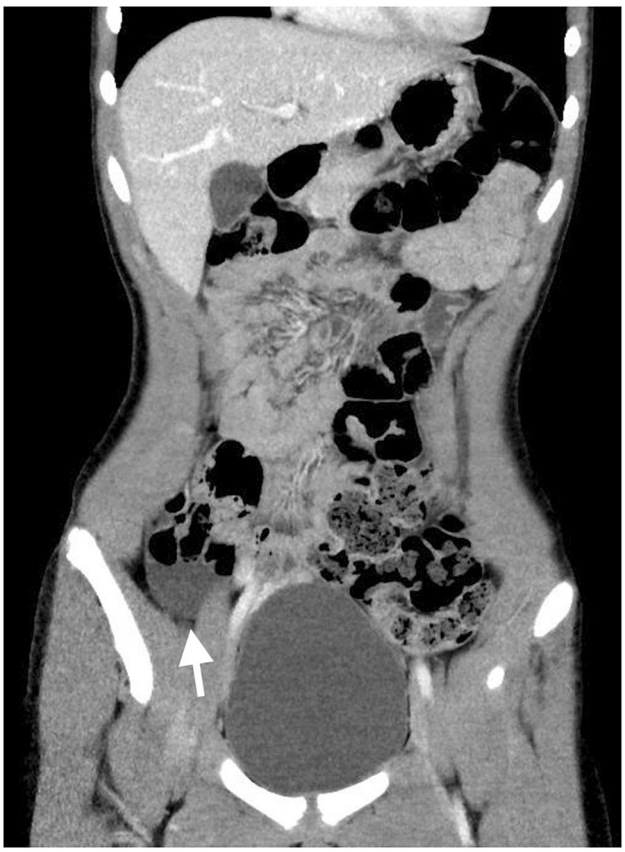

Fig. 2 Coronal view of abdominal computed tomography showing a small amount of fluid confined to the caudal side of the cecum (white arrow)

also showed no abnormalities except for ascites in the same region as that seen on computed tomography. Although these findings did not lead to the diagnosis, we planned to perform exploratory laparoscopy with the consent of the patient and her family.

With the patient under general anesthesia, a midline intraumbilical incision was made, and a Smart Retractor® (TOP Corporation, Tokyo, Japan) was inserted. A silicone cap (Free Access® XS; TOP Corporation) was attached for use as a multichannel port, and two 5-mm working ports were inserted through the cap. Intraoperative examination revealed a non-inflamed, unswollen appendix with cord-like narrowing at the middle part; an intact mesoappendix; and a small amount of serous ascites (Fig. 3a, b). There was no intraperitoneal adhesion and no abnormal findings of surrounding organs including the right ovary, ileum, and right-sided colon. The ileocecal area was pulled out through the umbilical wound, and an appendectomy was performed.

Histological examination of the appendix showed that the distal side of the cord was bilaterally blind-ended with mild chronic inflammation and fibrosis (Fig. 4a, b). Fibrous tissue was more prominent near the cordate than at the tip of the appendix. In the cordate region, the layered structure of the intestinal wall disappeared and was replaced by fibrous tissue, and no lumen was observed (Fig. 4c). The proximal aspect of the cord was dilated, with calcified fecal deposits in the lumen, but no evidence of acute inflammation. The findings around the peri choroidal area were the same as those of intestinal atresia type II, so the diagnosis of appendiceal atresia was made. Her postoperative course was uneventful, and the patient was discharged on postoperative day 3. She remained asymptomatic for 46 months postoperatively.

Discussion

Intestinal atresia, which can occur at any point in the gastrointestinal tract, is a major cause of neonatal surgical disease, and its incidence ranges from 1.3 to 3.5 per 10,000 live births [6, 7]. The duodenum is affected in approximately 50% of cases, followed by the jejunoileal region. Appendiceal atresia is extremely rare, with only three cases reported to date; these are briefly described as follows.

The first case of appendiceal atresia involved a 4-day-old boy weighing 2500 g who underwent emergency laparotomy based on a preoperative diagnosis of ileojejunal atresia. Intraoperative findings revealed multiple atresias of the proximal jejunum and jejunal diverticulum. Additionally, a swollen appendix of 16 mm in length with a 3-mm fibrous atretic segment close to the base was detected. The mesoappendix was absent [8].

The second case involved a 59-year-old man who had typical symptoms of appendicitis, including significant

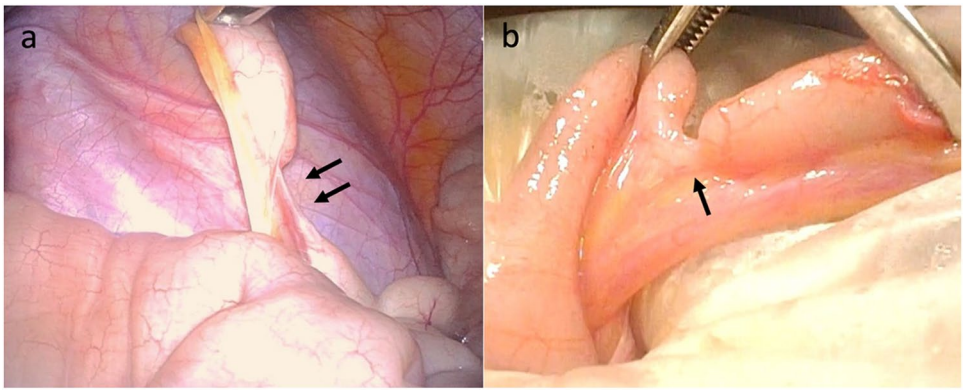

Fig. 3 **a**, **b** Intraoperative view showing a non-inflamed, unswollen appendix with cord-like narrowing at the middle part (black arrows) and an intact mesoappendix

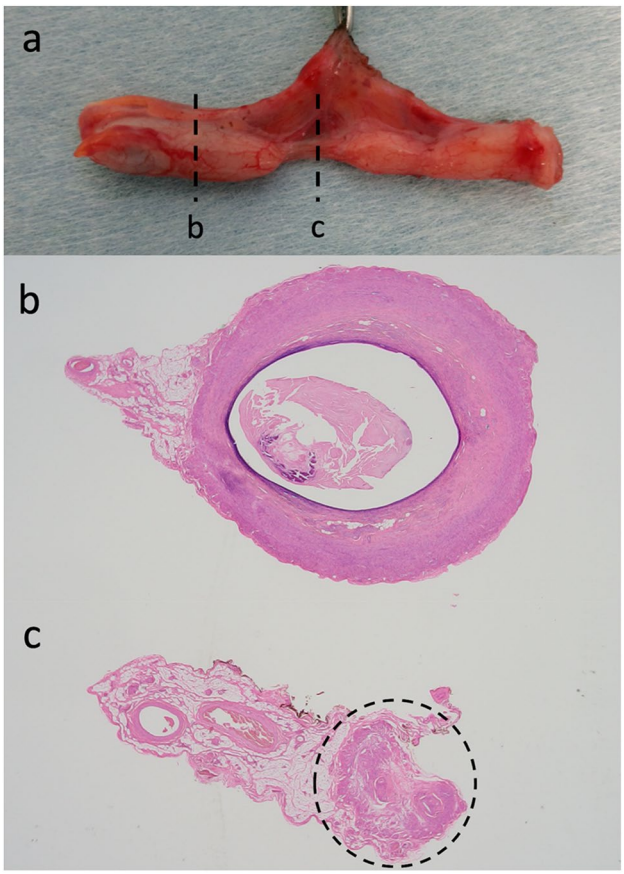

Fig. 4 **a** Resected appendix. **b** Cross-sectional slice of the appendix distal to the cordate region demonstrating prominent fibrotic tissue with mild chronic inflammation. Hematoxylin and eosin stain; original magnification, × 12.5. **c** The lumen and wall structure are not identified in the cross-sectional slice of the cordate region (dotted circle). Hematoxylin and eosin stain; original magnification, × 12.5

tenderness and voluntary guarding at McBurney's point with leukocytosis and an elevated serum CRP concentration. He had a medical and surgical history of coronary heart disease. At laparotomy, the origin of the appendix was absent and an inflamed mass with a free tip was found 10 cm proximal to the ileocecal valve. This case was considered type III a atresia of the appendix because the feeding vessels of the mass were derived from the ileocecal artery [9].

The last case was a 17-year-old female who was treated conservatively with typical findings of appendicitis with low-grade fever, rebound tenderness in the right lower abdomen, and appendicular lump. Six months later, an interval appendectomy was performed, which revealed an apical and basal appendix with a blind-end separation and both connected by a fibrous strand, accompanied by a normal mesoappendix [10].

Intestinal atresia is generally caused by a vascular accident to sections of the bowel during fetal development, and the etiology of appendiceal atresia is thought to be the same as that of other sites of intestinal atresia. Other conditions that result in an obstruction of the appendix include fibrous obliteration. Fibrous obliteration of the appendix is considered as a part of the aging process that results in loss of the normal appendiceal mucosa, which eventually replaces the mucosa and submucosa with fibrotic tissues. It can be differentiated from appendiceal atresia by the preservation of the muscular and serous layer structure at the site of obstruction. On the basis of the timing of diagnosis in the first case and the intraoperative findings and the lack of history of RLQ pain in the second case, the atresia was deemed to have formed congenitally in both patients. The findings of the appendix in the third case and the present case were similar, and it is unclear what led to the reduced blood supply of the middle part of the appendix because the mesoappendix was intact. Although the inflammation was mild and blood flow in the tip of the appendix was at least confirmed by ultrasound, it is possible that appendicitis at the initial presentation may have played a role in the impaired blood flow. The continuity of the appendiceal lumen to the cecum was not fully evaluated by ultrasound at the initial visit and preoperatively since the diagnosis of appendiceal atresia was not in mind.

Possible causes of RLQ pain in our patient include infection distal or proximal to the site of atresia, or increased

internal pressure without infection at the double-blind appendiceal tip. There were no findings suggestive of appendicitis on blood tests or imaging studies except at the initial presentation, and pathology of the resected specimen showed only mild chronic inflammation distal to the atresia and no evidence of acute inflammation proximally, although fecaliths were present in the proximal lumen. Therefore, the increased internal pressure at the tip of the appendix may have contributed to the abdominal pain except at the initial onset. The repeated presence of a small amount of ascites at the time of abdominal pain triggered the performance of laparoscopic surgery, although appendiceal swelling could not be detected. Some patients with recurrent RLQ pain but without remarkable abnormalities in various imaging tests are followed up for a long period with a diagnosis of functional abdominal pain. In patients with recurrent RLQ pain of unknown cause, it is necessary to consider diagnostic laparoscopy while keeping appendiceal atresia in mind as a differential diagnosis.

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Author contributions Conception and design of the study: MM, MI, and TY. Acquisition of data: MM and AN. Drafting of the article: MM. Critical revision of the manuscript for important intellectual content: MM, MI, AN, TY, and TS. Final approval: TS.

Data availability The data that support the findings of this study are available from the corresponding author upon reasonable request.

Compliance with ethical standards

Conflicts of interest The authors declare that they have no conflicts of interest.

Ethical approval All procedures followed have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its subsequent amendments.

Informed consent Informed consent was obtained from the guardian of the patient whose case details appear in this manuscript.

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