CASE REPORT

Left-sided appendicitis in children with congenital gastrointestinal malrotation: a diagnostic pitfall in the emergency department

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SUMMARY

Acute appendicitis is one of the most common conditions requiring emergency surgery. However, acute appendicitis presenting with left lower quadrant abdominal pain is extremely rare. Imaging, particularly CT, plays an important role in establishing an accurate and prompt diagnosis, as delay in diagnosis may occur due to lack of uniformity in the clinical signs and symptoms. We report a rare case of a 10-year-old boy who presented with persistent left lower quadrant pain of several days duration, in which the CT scan of the abdomen and pelvis was essential in establishing the correct diagnosis. The malpositioned inflamed appendix was clearly identified in the left side of the abdomen, with the characteristic CT findings of uncomplicated intestinal malrotation. Left-sided acute appendicitis should be considered in the differential diagnosis of young patients presenting with left lower quadrant pain, in order to avoid delay in diagnosis and guide the surgical intervention.

BACKGROUND

The coexistence of intestinal malrotation and acute appendicitis often constitutes a diagnostic pitfall in the emergency department. This case report represents the 10th reported case, in the English literature, of acute appendicitis superimposed on asymptomatic bowel malrotation in the paediatric population. A thorough knowledge of the differential diagnoses of left-sided abdominal pain, as well as high clinical suspicion, is imperative to ensure an early correct diagnosis, to prevent further delay in diagnosis and to avoid complication and guide the management.

CASE PRESENTATION

A 10-year-old previously healthy boy presented to the emergency department with a 5-day history of persistent left lower abdominal pain, fever, nausea and vomiting. The patient was examined 3 days before presentation by a paediatrician and was diagnosed with gastroenteritis. On physical examination, the abdomen was rigid, with significant tenderness over the left lower quadrant.

INVESTIGATIONS

Laboratory tests revealed elevated white cell count of $11\,800/\text{mm}^3$, normal haematocrit and chemistry, and negative urine analysis. Owing to persistent symptoms for several days, contrast-enhanced CT scan of the abdomen and pelvis was performed

(figure 1A,B). A redundant floating ascending colon was discovered, with the majority of small bowels positioned in the right side of the abdomen and the caecum located in the left mid-abdomen, consistent with intestinal malrotation. No associated situs, caval or other congenital anomalies were present and no evidence of volvulus was identified. A dilated, tubular, blind-ending structure was seen arising from the caecum in the left mid-abdomen, measuring 1.6 cm in diameter with significant surrounding fat stranding and free fluid (figure 1A,B). The findings were consistent with acute appendicitis, with phlegmon formation.

TREATMENT

The patient underwent emergency open appendectomy through a left upper transverse incision since laparoscopy was deemed difficult in the presence of complicated appendicitis. At exploration, there was evidence of free flowing pus in the peritoneal cavity, and the appendix was noted to be associated with phlegmon with adherent bowels and thickened omentum. The malrotation was not corrected at that time, since the symptoms were not related to malrotation and because of the presence of severe mesenteric inflammation and adhesions. Pathology confirmed the diagnosis of acute appendicitis with perforation and serositis.

OUTCOME AND FOLLOW-UP

The patient had an uneventful recovery and was discharged home several days later in good condition. Follow-up upper gastrointestinal series (figure 1C,D) was performed one week after the procedure showing the duodenojejunal junction to the right of midline, below the level of the duodenal bulb, consistent with intestinal malrotation. One month later, the patient presented with symptoms of bowel obstruction. A CT scan was performed with intravenous and oral contrast, showing several dilated small bowel loops, with bowel faeces sign and a transitional zone in keeping with small bowel obstruction. There was no evidence of volvulus. The patient underwent a second operation that revealed simple adhesive small bowel obstruction which was released with a smooth and uneventful recovery thereafter.

DISCUSSION

The coexistence of intestinal malrotation and acute appendicitis often constitutes a diagnostic pitfall. A review of the English literature revealed 32

To cite: Taslakian B, Issa G, Hourani R, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009474

Figure 1 Axial (A) and coronal (B) CT images of the abdomen showing an enlarged fluid-filled thickened appendix, with enhancing wall, in the left side of the abdomen (white arrow). The small bowels are located on the right side of the abdomen (white open arrow). Radiograph from barium upper gastrointestinal series (C and D) shows contrast agent-filled duodenum and jejunal loops that remain right-sided. Duodenojejunal junction (black arrow) lies below the level of the duodenal bulb (asterix), without crossing the spine to left. Note the absence of colonic gas in right lower quadrant (black open arrow).

reported cases of acute appendicitis associated with malrotation, nine of which only have been reported in the paediatric age group (table 1). In the previously reported cases, most of the patients presented with left-sided abdominal pain, usually of several days duration. Correct diagnosis was delayed, leading to progression of the disease and significant morbidity because of perforation and peritonitis.^{1–3}

The differential diagnosis of left lower quadrant abdominal pain includes diverticulitis, renal colic, ruptured ovarian cyst, epididymitis, incarcerated or strangulated hernia, bowel obstruction, regional enteritis, psoas muscle abscess and right-sided and left-sided appendicitis.⁴ Acute appendicitis presenting with left lower quadrant pain occurs with congenital abnormalities that include true left-sided appendix.⁵ It may also represent an atypical presentation of long right-sided appendix, projecting

into the left lower quadrant⁵ or of a redundant and loosely attached caecum with exceptional mobility.^{6–7} Left-sided acute appendicitis can develop in association with two rare anatomical abnormalities, situs inversus and less commonly midgut malrotation.⁴

Imaging plays an important role in the evaluation and management of patients presenting with acute abdominal pain. CT scan has been increasingly employed in establishing accurate diagnosis in patients with non-traumatic acute abdomen and distinguishing surgical from non-surgical conditions. Contrast-enhanced CT has a reported positive predictive value of 95% for the diagnosis of appendicitis.⁸ The most specific sign of acute appendicitis on CT examination is a dilated, fluid-filled tubular structure that measures more than 6 mm in diameter, with a thickened enhancing wall in the right lower quadrant (RLQ), the expected location of the appendix. The presence of calcified appendicoliths and periappendiceal fat stranding are useful secondary imaging findings.⁸

Malrotation, a congenital abnormal position of the bowel within the peritoneal cavity, occurs approximately 1 in 500 births.⁹ It usually involves the small and the large bowel and is accompanied by abnormal bowel fixation by Ladd's bands or absence of fixation of portions of the bowel. Most cases of midgut malrotation are diagnosed in infancy and early childhood where, with up to 75% of symptomatic cases occurring in newborns, and up to 90% cases within the first year of life.⁹ It may present with an acute duodenal obstruction, chronic abdominal pain, intermittent vomiting, chronic diarrhoea, malabsorption and failure to thrive.¹ However, some cases have been reported in older children and adults and were typically asymptomatic or manifested as non-specific symptoms.¹⁰ They were often discovered during the investigation of other conditions.

In malrotation, the small bowel is located predominantly on the right side within the peritoneal cavity and the colon is located on the left side.¹¹ Various degrees of malrotation of the small or large bowel may occur, and the positions of the duodenojejunal junction (and, by implication, the ligament of Treitz) and colon depend on the developmental stage at which normal embryologic rotation failed. Because the second and third stages of rotation differ for the small versus the large bowel, the rotation of one may be normal while that of the other is abnormal. Broadly, intestinal malrotation can be categorised into incomplete (partial rotation) and non-rotation, with the latter being the most common type in the older population.¹²

Conventional radiography is neither sensitive nor specific for malrotation, although right-sided jejunal markings and the absence of a stool-filled colon in the RLQ may be suggestive of

Table 1 Reported cases of acute appendicitis associated with intestinal malrotation in children

Author (year)	Age (years)	Sex	Presentation	Duration	References
1. Tawk <i>et al</i> (2012)	15	Female	Left upper quadrant pain, nausea, low-grade fever and vomiting	3 days	<i>Int J Surg Case Rep</i> 2012;3:399–401
2. Kamiyama <i>et al</i> (2005)	14	Male	Not available	Not available	<i>Radiat Med</i> 2005;23:125–7
3. Tsumura <i>et al</i> (2003)	15	Female	Umbilical and left lower quadrant pain	Not available	<i>Surg Endosc</i> 2003;17:657–8
4. Hollander <i>et al</i> (2003)	9	Male	Left-sided abdominal pain and diarrhoea	1 week	<i>Pediatr Radiol</i> 2003;33:70–1
5. Ratani <i>et al</i> (2001)	8	Female	Lower abdominal pain, vomiting, and diarrhoea	2 days	<i>Abdom Imaging</i> 2002;27:18–19
6. Garg <i>et al</i> (1991)	17	Male	Right lower quadrant pain and vomiting	24 h	<i>Indian J Gastroenterol</i> 1991;10:103–4
7. Shapiro <i>et al</i> (1963)	15	Male	Lower abdominal pain, fever, diarrhoea and vomiting	2 days	<i>Calif Med</i> 1963;98:158–9
8. Haddon <i>et al</i> (1945)	19	Male	Right lower quadrant pain, vomiting	36 h	<i>BMJ</i> 1945;2:569
9. Crook <i>et al</i> (1918)	14	Not available	Left iliac pain, nausea and vomiting	Few hours	<i>Southern Med J</i> 1918;6:318

this finding. The upper gastrointestinal series remains the imaging standard for the diagnosis of malrotation, with a reported sensitivity of 95%.⁹ The normal position of the duodenojejunal junction is to the left of the left-sided pedicles of the vertebral body at the level of the duodenal bulb on frontal views and posterior (retroperitoneal) on lateral views.¹³ A duodenojejunal junction that fails to cross the midline and lies below the level of the duodenal bulb on upper GI series, with small bowels present in the right side and colon in the left side, is of diagnostic value in malrotation. Contrast enema examination is helpful in the diagnosis in urgent situations, and shows malposition of the right colon, but the caecum may assume a normal location in up to 20% of patients. This normal location may cause malrotation to be missed on this type of study.

Many cases of malrotation in older children and adults are currently being detected incidentally on cross-sectional imaging performed for various unrelated reasons. CT scan is very useful in detecting the rotational anomalies and the related complications. In addition, CT scan can show associated extraintestinal findings not evident on conventional examinations, which are useful indicators of malrotation.¹² This includes deviation from the normal relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV), and underdevelopment or absence of the uncinate process.¹ Most patients with malrotation will show a vertical relationship between SMA and SMV or left-right inversion.¹² However, these findings are not entirely diagnostic. Some patients with malrotation will have a normal relationship, and a vertical or inverted relationship can also be seen in patients without malrotation.¹² Therefore, isolated detection of such an abnormality is not sufficient for diagnosis; but should warrant closer examination of the bowel. Other associated anomalies that should be looked up on imaging are visceral situs anomalies, inferior vena cava anomalies, polysplenia and preduodenal portal vein.¹² Our patient had normal relationship between the SMA and SMV, and showed no evidence of other associated anomalies.

Imaging signs of acute appendicitis in cases of intestinal malrotation are similar to those of normally located appendix; however, due to the delayed diagnosis, they are often more severe with findings of abscess, phlegmon and free intra-abdominal gas indicating perforation. Ultrasonography (US) is also helpful in establishing the diagnosis. Colour Doppler can visualise the inflammatory hyperaemia at the level of the appendix and surrounding bowels, in the left lower quadrant.³ Contrast-enhanced US demonstrates enhancement of the affected bowel wall, and is particularly useful in the assessment of intestinal ischaemia.¹⁴

Surgical correction (Ladd's procedure) is usually the standard treatment for intestinal malrotation. However, the recommendations for treatment are still controversial. In order to prevent the feared complication of volvulus, some advocate the surgical intervention whenever radiographic evidence of intestinal malrotation is present, regardless of the type. Others resort to surgery only when the patients have symptoms related to malrotation and recommend that observation alone is an acceptable approach for atypical or incomplete malrotation.¹⁵ Our patient had complicated appendicitis with phlegmon formation. Appendectomy was performed, without Ladd's procedure since the symptoms were not related to malrotation. Laparoscopic appendectomy has been the gold standard technique for the treatment of acute appendicitis.¹⁶ However, the patient underwent an open appendectomy, through a mini-laparotomy incision, to avoid poor visualisation due to malrotation and the phlegmon formation identified on the CT scan. Although he later presented with intestinal obstruction, this time also his

symptoms did not result from the malrotation but rather he had an adhesive obstruction that was managed with simple surgical release of the adhesions.

Learning points

- ▶ Acute appendicitis is one of the most common conditions requiring emergency surgery.
- ▶ Coexistence of intestinal malrotation and acute appendicitis is rare and constitutes a diagnostic pitfall. Paediatricians should be aware of this condition.
- ▶ Acute appendicitis presenting with left lower quadrant pain occurs with congenital abnormalities, as an atypical presentation of long right-sided appendix or of a redundant caecum.
- ▶ A thorough knowledge of the differential diagnoses of left-sided abdominal pain, as well as high clinical suspicion, is imperative to ensure an early correct diagnosis, to prevent further delay in diagnosis and to avoid complication.
- ▶ Imaging plays an important role in establishing an accurate and prompt diagnosis, as delay in diagnosis may occur due to lack of uniformity in the clinical signs and symptoms.

Contributors All authors have significantly contributed in writing the manuscript. All authors have read and approved the final draft before submission.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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