CASE REPORT

Midgut malrotation presenting with left-sided acute appendicitis and CT inversion sign

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SUMMARY

In patients presenting with abdominal pain, appendicitis is the most common surgical disorder. Appendicitis causing pain in the left lower quadrant is extremely rare and can occur with congenital abnormalities that include true left-sided appendix or as an atypical presentation of right-sided long appendix, which projects into the left lower quadrant. We report a case of a 69-year-old man showing midgut malrotation with acute appendicitis presenting as left lower quadrant abdominal pain.

BACKGROUND

In patients presenting with abdominal pain, appendicitis is the most common surgical disorder. Appendicitis causing pain in the left lower quadrant is extremely rare and can occur with congenital abnormalities that include true left-sided appendix or as an atypical presentation of right-sided, but unusually long appendix, which projects into the left lower quadrant.¹ Left-sided acute appendicitis develops in association with two types of congenital anomalies: situs inversus totalis and midgut malrotation.^{1 2} Intestinal malrotation is a developmental anomaly of the midgut in which the normal fetal rotation of intestines around the superior mesenteric artery (SMA) and their fixation in the peritoneal cavity fail. Eighty-five per cent of such cases have been estimated to present in the first 2 weeks of life. Presentation is rare at adulthood. The clinical diagnosis of left-sided acute appendicitis is difficult in adults because the presentation is usually non-specific and malrotation is rarely considered.³

CASE PRESENTATION

A 69-year-old man presenting with a 1-day history of left lower abdominal pain was admitted to our emergency department. He did not have any significant medical history or treatment. Abdominal examination revealed tenderness in the left lower quadrant. Laboratory tests showed a raised white cell count ($12.2 \times 10^9/L$) and a raised C reactive protein concentration (110 mg/L). Abdominal X-ray was unremarkable. Based on his worsening of symptoms and the clinical findings, an abdominal CT scan was performed. Contrast-enhanced CT showed evidence of intestinal malrotation including right-sided duodenojejunal junction, left-sided hepatic flexure, superior mesenteric vein (SMV) inversion sign (figure 1), dilated appendix with wall-thickening positioned in the left side of the midline, and asymmetrical enlargement in the left abdomen (figure 2). These findings were diagnostic of

Figure 1 Superior mesenteric vein (SMV) inversion sign (thick open arrow). Inversion is shown as left SMV relating to artery.

malrotation with appendicitis. Appendectomy was performed, and pathology revealed acute appendicitis.

OUTCOME AND FOLLOW-UP

After appendectomy, 6 months later, there was no abnormality in transabdominal ultrasonography.

DISCUSSION

Intestinal malrotation can be broadly defined as any deviation from the normal 270° counterclockwise rotation of the midgut during embryological development. Midgut malrotation has been estimated to occur in approximately 1 in 500 live-births.^{4 5} In 85% of the patients with congenital malrotation, it

Figure 2 A contrast-enhanced abdominal CT scan shows a left side of the midline positioned dilated appendix with wall thickening at umbilical region (open arrow).

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presents in the first 2 weeks of life, usually with intestinal obstruction. In adults, malrotation is rarely suspected and is generally first visible on imaging studies as an incidental imaging finding as a rare cause of acute abdominal symptoms. In patients with abdominal abnormalities unrelated to coexisting malrotation, the altered anatomy may result in an atypical clinical presentation: appendicitis is a prime example, which may present with symptoms that are more left-sided. Careful review of the CT images, however, should yield the correct diagnosis, including the unsuspected malrotation. CT not only shows the intestinal malpositioning but also depicts associated extra intestinal findings not evident on conventional examinations. For example, deviation from the normal relationship between the SMA and SMV is a useful indicator of malrotation.^{5 6} In most patients with quiescent malrotation, the SMA and SMV will assume a vertical relationship or show left–right inversion as in the presenting case.³ Surgical treatment is the only real safe-

guard against appendicitis. CT clarifies unusual location of appendicitis, and is also useful in detecting associated rotational anomalies and related complications. Our patient is unusual in two respects. First, acute appendicitis is less common in the elderly. Second, even more unlikely is a diagnosis of ‘left-sided appendicitis’ in a patient not previously known to have intestinal malrotation. The location of his pain and his age minimally delayed the correct clinical diagnosis. Minimum delay led to no serious effect but it could lead to a serious outcome in other patients. Malrotation is rarely seen in the adult population. It is difficult to diagnose adult patients who have malrotation with appendicitis. Malrotation should be considered in the differential diagnosis of an adult patient’s left-sided acute appendicitis presenting with left lower quadrant pain.

Contributors EÇ and RT diagnosed the patient and prepared the case report. SK and BA helped to prepare the case report.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Left-sided acute appendicitis develops in association with two types of congenital anomalies: situs inversus totalis and midgut malrotation.
- ▶ The clinical diagnosis of left-sided acute appendicitis is difficult in adults because the presentation is usually non-specific and malrotation is rarely considered.
- ▶ CT clarifies unusual location of appendicitis, and is also useful in detecting associated rotational anomalies and related complications.

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