Unusual presentation of left sided acute appendicitis in elderly male with asymptomatic midgut malrotation

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A B S T R A C T

INTRODUCTION: Acute appendicitis in the setting of midgut malrotation is uncommon. Midgut malrotation commonly presents within the first month of life. A minority remain asymptomatic and may present with concomitant abdominal pathology making diagnosis difficult.

PRESENTATION OF CASE: This paper reports a rare case of a 73-year-old male diagnosed with acute appendicitis and asymptomatic MM. The patient underwent a laparoscopic appendectomy, but had an unplanned return to theatre for washout of post-operative intra-abdominal haematoma.

DISCUSSION: Midgut malrotation is commonly described by the stringer classification and type 1A is the most common in adults. There have only been a handful of documented cases of acute appendicitis with midgut malrotation occurring in the adult population. Previous delay in diagnosis has led to a delay in definitive management. Both laparoscopic and open surgery has been used in the past.

CONCLUSION: Acute appendicitis with malrotation should be considered in elderly patients presenting with atypical signs and symptoms. Imaging offers significant advantage for timely and definitive management.

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1. Introduction

Acute appendicitis is a commonly encountered surgical condition [1]. Presenting complaints and location of pain can mimic a variety of conditions including gastritis, gastroenteritis, cholecystitis, pyelonephritis and diverticulitis. Diagnosis is made even more difficult by the presence of concomitant anatomical abnormalities [2].

Midgut malrotation (MM) is associated with abnormal rotation of the primitive intestinal loop around the superior mesenteric artery axis during the first ten weeks of foetal life [1]. The majority (85%) of such cases will present in the first month of life with vague abdominal pain, distension, vomiting or obstruction. A minority progress to adulthood and remain asymptomatic, with incidence reported between 0.1–0.5% [3].

This paper reports a rare case of a 73-year-old male diagnosed with acute appendicitis and asymptomatic MM. There have been previous sporadic cases documented in the literature. We provide a review of these cases and discuss the importance of computed tomography (CT) for diagnosis and preoperative planning. Due to the abnormal anatomy, these patients often present with atypical signs/symptoms which can potentially delay timely diagnosis and management.

2. Case report

Mr. N is a 73 year old male who presented to emergency with a one week history of constipation and left-sided abdominal pain radiating diffusely across the lower abdomen. He denied any nausea or vomiting. He had been passing flatus but not opened his bowels for the past 3–4 days. He denied any other infective symptoms. His past medical history included bronchiectasis and ischaemic heart disease, aortic valve repair and previous coronary artery bypass graft (on aspirin). At presentation, he was febrile (38.5 °C) but otherwise haemodynamically stable—heart rate (92 beats/min), blood pressure (152/89 mmHg), respiratory rate (18 breaths/min) and saturations (99%). He had leucocytosis with left shift (white cell count 10.2 × 10⁹), neutrophils 7.5 × 10⁹, raised c-reactive protein (91 mmol/L), and elevated bilirubin (27 U/L). The remainder of his bloods were normal (Table 1).

At this time, provisional diagnosis was diverticulitis, with differentials including appendicitis, pyelonephritis, inflammatory bowel
disease and less likely, ischaemic bowel. CT abdomen and pelvis (with contrast) identified periappendiceal inflammatory changes consistent with appendicitis. He was also noted to have incidental CT findings consistent with type 1a MM, including right sided duodenojejunal (DJ) flexure, left sided caecal pole with midline appendix and inversion of superior mesenteric artery (SMA) and superior mesenteric vein (SMV) (Figs. 1 and 2). He underwent a laparoscopic appendectomy for suppurative perforated appendicitis with collection. A drain was inserted due to difficult haemostasis. He was monitored in HDU. Over the next day, he had approximately 200–300 ml of blood in his drain, and was given two units of packed red blood cells. As he remained haodynamically stable and was known to have been on aspirin pre-operatively (plus the intra-operative difficulty with haemostasis), this post-operative course was expected. On day four post-operatively, he acutely deteriorated becoming hypotensive and febrile with a repeat CT scan identifying the possibility of a collection at the operative site. He underwent a laparotomy and had washout of a large infected intra-abdominal haematoma. Intra-operatively, malrotation was confirmed on inspection of the DJ flexure and a left sided caecum. He made a complete recovery and was discharged home day thirteen post-operatively.

### Table 1

| Investigation | Result (normal reference range) |
|---------------|---------------------------------|
| Na            | 141 mmol/L (135–145)            |
| K             | 4.2 mmol/L (3.5–5.0)            |
| Cl            | 99 mmol/L (97–109)              |
| Creatinine    | 5.4 mmol/L (3.0–8.0)            |
| Bilirubin     | 27 micromol/L (<21)             |
| ALP           | 99 U/L                          |
| GGT           | 33 U/L                          |
| AST           | 22 U/L                          |
| ALT           | AST 26 U/L                      |
| Amylase       | 53 U/L (20–120)                 |
| Lipase        | 20 U/L (13–60)                  |
| WCC           | 10.2 × 10³/L (4.0–10.0 × 10³)   |
| Neutrophils   | 7.5 × 10⁹/L (2.0–7.0 × 10⁹)     |
| HB            | 146 g/L (130–170)               |
| Platelets     | 248 × 10⁹/L (150–400)           |
| C-Reactive protein | 91.0 mg/L (<5)                |

**Fig. 1.** Axial section of CT abdomen showing signs of malrotation: inversion of SMA/SMV relationship (long arrow), right-sided DJ flexure (short arrow).

**Fig. 2.** Coronal section of CT abdomen showing left-sided caecal pole and acute appendicitis with peri-appendiceal inflammation (long arrow).

### 3. Discussion

Midgut malrotation is a group of congenital anomalies of the intestine related to non-rotation or malrotation of the intestine. In normal embryology, the midgut undergoes three stages of counterclockwise rotation to form the normal gastrointestinal anatomy [3,4]. Errors during this process can result in MM, classified by Stringer into Type 1–3 [4]. Mr. N presented with Type 1a malrotation which is a result of failure in the first stage of rotation. It is the most common malrotation reported in adults and is often asymptomatic [5].

Due to the variation in malrotation, the exact location of the appendix may vary from left side, midline, right upper or right lower quadrant. Imaging with computed tomography can help not only in pre-operative planning of such cases, but also minimises delay in diagnosis. Sensitivity and specificity of CT in setting of acute appendicitis in patients with midgut rotation has been reported between 90 and 97% and 94–100% respectively [6]. The most typically reported CT-findings in these patients include: right sided DJ flexure, left sided caecal pole, inversion of the SMA/SMV relationship and hypoplasia/aplasia of the pancreatic uncinate process [3,7–9]. In the case presented, all except uncinate process hypoplasia were present. Ely et al. reported a case series of 8 patients with acute appendicitis and incidental MM. All patients had abnormal uncinate process including one case with an absent pancreatic tail [3].

Acute appendicitis with MM has only previously been described in a handful of case report and small case series [1–3,5,6–10]. In a previous report, misdiagnosis as diverticulitis led to delay in definitive management, with the patient subsequently undergoing emergency laparotomy [2]. These cohort of patients are also at increased risk of volvulus and intestinal obstruction [11]. In most
reported cases, patients underwent laparotomy with attempted correction of the malrotation [1,3,5,6,9]. Other reports of laparoscopic appendectomy have also been documented [6]. Laparoscopy is useful not only in establishing differential diagnosis but for definitive surgery. The most common reasons in both groups for take-back laparotomies were due to collection, ileus or obstruction [6].

4. Conclusion

We present a case of acute appendicitis in an elderly patient with previously undiagnosed MM. Our case highlights the atypical presentation of these patients and importance of computed tomography in establishing a timely diagnosis. Initial laparoscopy may also be useful in ambiguous cases and for definitive surgery.

Conflict of interest

No conflicts of interest to declare.

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Ethical approval

Not Applicable

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. Patient consent was gained prior to writing and submission of this article.

Author’s contribution

All authors were involved in the study design, write up of the draft and final drafting of the paper with equal share of work.

Research registry

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Guarantor

Animesh Singla (corresponding author) will act as guarantor of submission.

References

[1] S. Akbulut, A. Ulku, A. Senol, M. Tas, Y. Yagmur, Left-sided appendicitis: review of 95 published cases and a case report, World J. Gastroenterol. 16 (4) (2010) 5598–5602.
[2] T. Hanna, J. Alob, Acute presentation of intestinal malrotation in adults: a report of two cases, Ann. R. Coll. Surg. Engl. 92 (2010) 1–4.
[3] A.B. Ely, N. Gorelik, Y. Cohen-Sivan, R. Zissin, L. Carpienta, A. Osadchy, et al., Appendicitis in adults with incidental midgut malrotation: CT findings, Clin. Radiol. 68 (2013) 1212–1219.
[4] D. Stringer, P. Babyn, Pediatric Gastrointestinal Imaging and Intervention, Toronto Press, Ontario, Canada, 2000.
[5] S. Israelit, O. Brook, B. Nira, L. Guralnik, D. Hershko, Left-sided perforated acute appendicitis in an adult with midgut malrotation: the role of computed tomography, Emerg. Radiol. 16 (2009) 217–218.
[6] A. Jones, D. Cassidy, Acute appendicitis presenting as acute gastritis in an adult patient with undiagnosed congenital gut malrotation: a case report, Emerg. Med. J. 44 (2) (2012) 153–155.
[7] R. Ratani, O. Haller, W. Wang, D. Yang, Role of CT in left-sided acute appendicitis: case report, Abdom. Imaging 27 (2001) 18–19.
[8] T. Sonomura, T. Royama, S. Ishii, T. Takeuchi, H. Sanda, K. Nakata, et al., Acute appendicitis with intestinal malrotation: the usefulness of coronal computed tomography, Intern. Med. 53 (2014) 1511–1513.
[9] R. Badea, N. Hajjar, V. Andreica, B. Procopet, C. Carazani, A. Tamas-Szora, Appendicitis associated with intestinal malrotation: imaging diagnosis features. Case report, Med. Ultrasonography 14 (2) (2012) 164–167.
[10] J. Moll, J. Marti, Left-sided appendicitis in a 47-year-old man with previously undiagnosed intestinal malrotation, Am. J. Emerg. Med. 31 (459) (2013) 5–6.
[11] C. Lin, C. Tiu, Y. Chou, J. Chen, W. Liang, C. Cheng, CT presentation of ruptured appendicitis in an adult with incomplete intestinal malrotation, Emerg. Radiol. 10 (2004) 210–212.

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