CASE REPORT

Subhepatic caecal tumour in an adult with intestinal malrotation

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SUMMARY

Intestinal malrotation is an abnormality that usually presents in infancy. Following correction, complications are rare in adults. We describe a case of a 64-year-old woman with a history of malrotation who presented with anaemia and weight loss. Colonic cancer was apparently excluded by colonoscopy. In fact, endoscopy had only been performed up to the hepatic flexure, which was unexpectedly positioned in the right iliac fossa. The patient then underwent a CT pneumocolon study, which demonstrated a caecal tumour, unusually located subhepatically. Repeat colonoscopy was performed to the true caecum and the lesion was biopsied. The patient underwent uncomplicated open right hemicolectomy and made a full recovery. To avoid diagnostic delay and, at worst, missing pathology entirely, clinicians must always thoroughly review a patient's history. Adults with malrotation may have atypical presentations of abdominal disease and so it is pertinent that surgeons, endoscopists and general practitioners remain alert and cognisant of their patients' history.

BACKGROUND

Colonic tumours and their associated sequelae compromise a significant part of general surgical on-calls and clinics, and indeed the day-to-day life of a general practitioner (GP). For this reason, it is our joint responsibility to develop our understanding of this common disease, including potentially unusual presentations.

Intestinal malrotation is rare, with an incidence of only 1 in 6000 live births;¹ however, bowel tumours are common with an incidence of over 1 000 000 new cases each year.² When the two are combined it can create a diagnostic conundrum that may delay diagnosis with subsequent inevitable disease progression.

Malrotation of the intestinal tract is believed to stem from incomplete rotation and fixation of the bowel during development. In normal embryogenesis, week 5 signifies a 270° clockwise rotation of the gut around the superior mesenteric artery following enlargement of the midgut portion. Then, at week 10, the tract normally attaches to the posterior aspect of the abdominal cavity. If this process is interrupted at any point, then intestinal malrotation will result.³ Malrotation typically presents in the first month of life—it is rare to find cases or complications from childhood disease in adults.⁴

An unwell, haemodynamically unstable child with malrotation and suspected volvulus represents

a paediatric surgical emergency. As definitive management of the unstable acute intestinal volvulus, the patient must be expedited to the operating theatre to undergo laparotomy and detorsion of the bowel. Surgical management of the malrotated bowel is done by Ladd's procedure in which torsion is reversed and congenital Ladd's bands are divided. Additionally, an incidental appendectomy is performed during a Ladd's procedure to minimise diagnostic confusion later in the patient's life.

In this case, our patient had a history of intestinal malrotation that then later in life presented with symptoms of a colonic tumour. The diagnosis of bowel tumour was delayed because of the unusual anatomy of her colon and the challenges that this presented to the endoscopist. Clearly, the aims of surgery in the paediatric setting are designed to treat and prevent acute complications from occurring, such as small bowel obstruction. Key facts such as a paediatric diagnosis of malrotation can fall far back into the medical history of patients—particularly if they are elderly. If these facts are not well documented, then future presentations may seem obscure.

This case teaches us that we should always review a patient's history rigorously and perform a thorough physical examination before performing any invasive investigation, and that we should remain vigilant to detect any history of intestinal malrotation when we are attending to patients with general surgical or gastroenterological pathologies.

CASE PRESENTATION

A 64-year-old Caucasian woman presented to her GP with a 4-month history of malaise, fatigue and abdominal pain. She had a medical history of small bowel obstruction that had been managed by midline laparotomy—at this operation she was found to have intestinal malrotation and volvulus. The intestinal volvulus was detorsed; however, a full Ladd's procedure was not performed. Additionally, she had a history of severe gastric ulceration treated with partial gastrectomy through a right-paramedian incision; and cataract disease, managed by phacoemulsification and intraocular lens implantation. A faecal occult blood test had returned as positive and routine full blood count and haematinics revealed iron-deficiency anaemia.

The patient then underwent colonoscopy. Using a scope guide, the endoscopist visualised a clear lumen down to the right iliac fossa—the segment of bowel that most commonly represents the caecum (figure 1). The patient's symptoms

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Figure 1 Initial colonoscopic view (taken in right iliac fossa). The folds in the wall of the bowel superficially resemble, but are not, the ileocaecal valve. Additionally, no appendicular opening is seen. This is potentially a view of the hepatic flexure.

persisted for six more weeks before the GP arranged abdominal CT with oral contrast. This demonstrated aberrant colonic anatomy, with the caecum situated in a subhepatic position and the hepatic flexure in the right iliac fossa (figure 2A, B). A suspicious lesion with an intraluminal component was visualised in the right upper quadrant within the caecum. For further clarification, the patient then underwent a virtual colonoscopy (CT pneumocolon), which showed a large fungating caecal tumour near the ileocaecal valve (figure 3A). Finally, the patient underwent a repeat colonoscopy. This endoscopic investigation was challenging, as the tortuous and abnormal bowel was difficult to navigate. Once the true caecum was visualised, the fungating tumour was located and biopsied, confirming probable causation for the patient's symptomatology (figure 3B).

An elective open right hemicolectomy was performed. This was an uncomplicated operation in which a large polypoidal tumour

was resected along with the caecal specimen (figure 4A, B). A primary anastomosis was created and the patient made a swift recovery and was discharged.

INVESTIGATIONS

CT of the abdomen

With oral contrast, this demonstrated a malrotated large bowel and a subhepatic caecum (beneath the left lobe of the liver) with a suspicious lesion. With the addition of CT pneumocolon, this then showed a large fungating tumour adjacent to the ileocaecal valve. The patient was negative for metastatic disease on CT.

Colonoscopy

Initially, this showed nothing abnormal, as the scope was only inserted to the hepatic flexure. On repeat, colonoscopy demonstrated a fungating caecal tumour, from which biopsies were taken.

Contrast enema

The patient described here did not undergo contrast enema. However, contrast enemas do give good delineation of colonic anatomy. They are underutilised in contemporary surgical practice, however, if this patient had undergone a contrast enema, it is possible that the initial endoscopist would have been alerted to the patient's unusual anatomy at an earlier juncture—and as such may have progressed the initial colonoscopy past the right iliac fossa and up to the true caecum.

DIFFERENTIAL DIAGNOSIS

- Colonic tumour
- Iron malabsorption (following partial gastrectomy)
- Peptic ulcer disease

Figure 2 Abdominal CT with oral contrast (coronal reconstruction). (A) The large bowel is abnormally positioned as a result of intestinal malrotation. In the right iliac fossa, a kink can be seen. This feature could understandably mimic the caecum on colonoscopy—in reality, it was more probably the inverted hepatic flexure. (B) A section of large bowel with a wide lumen (likely the caecum) can be seen in a highly unusual subhepatic position.

Figure 3 (A) Three-dimensional reconstruction of virtual colonoscopy. The true caecum is visualised more successfully with CT pneumocolon. Three-dimensional reconstruction demonstrating a fungating tumour adjacent to the ileocaecal valve, which was missed on initial colonoscopy. (B) Repeat colonoscopic view (taken in right upper quadrant). When the colonoscope was passed further, the same fungating tumour, as visualised by the CT pneumocolon, was identified and biopsied.

TREATMENT

As definitive treatment, the patient underwent elective open right hemicolectomy. Open surgery was opted for over a laparoscopic approach as a challenging surgical environment was anticipated, with highly aberrant anatomy surrounded by numerous intra-abdominal adhesions, given the patient's surgical history. Indeed, both of these difficulties were encountered intraoperatively.

On the surface of her abdomen, the patient had two large scars. A right paramedian laparotomy scar from a previous partial gastrectomy and a large midline laparotomy scar from her childhood episode of bowel obstruction. Our incision was made down the same line as the midline laparotomy for maximal access with minimal cosmetic disruption.

The right hemicolectomy continued without complication following adhesiolysis and mobilisation of the ascending colon from the right side of the abdomen, and the caecum from its subhepatic position. A stapled primary ileocolic anastomosis was

created, the abdomen was closed and the patient returned to the ward.

OUTCOME AND FOLLOW-UP

Postoperatively, the patient made a rapid and successful recovery.

A modern colorectal enhanced recovery programme was followed and the patient was discharged within 3 days postoperatively. The total length of her hospital stay was 4 days: the first day was the day of the elective surgery and then, postoperatively, she remained under our care for three further days until she returned to eating/drinking and opening her bowels normally, after which she was discharged home.

The patient was seen in the postoperative colorectal clinic 6 weeks after her right hemicolectomy. She was well and happy at the clinic appointment, with resolution of her anaemic symptoms. Her midline laparotomy scar had healed without problem.

Figure 4 (A) Intraoperative photography, taken during open right hemicolectomy. The left side of the photo is cranial and the right is caudal. The twisted, subhepatic caecum is very obviously seen in the abdominal wound, located at the top of the abdomen. (B) Pathological specimen of caecum and tumour. For the benefit of the reader, we have inverted the specimen. On the right side of the photograph the polypoid tumour is seen to be contained completely within the specimen, with decent margins achieved proximally and distally.

Clearly, given the extent of her intra-abdominal adhesions, it is pertinent that the patient and the GP remain mindful of the possibility of another episode of adhesional small bowel obstruction in the future.

DISCUSSION

A colorectal tumour against a background of intestinal malrotation is a rare clinical entity that poses specific challenges to diagnosis and management. We identified seven cases of colorectal tumours in the presence of intestinal malrotation in the extant literature; these cases are discussed below.

Ren and Lu⁵ reported a case of a man with altered bowel habits, who underwent laparotomy, demonstrating an ascending colon tumour located posteriorly to the superior mesenteric vessels, on a background of undiagnosed malrotation. Similarly, Michalopoulos *et al*⁶ presented a case of a patient who was found to have incidental malrotation and ascending colon carcinoma at laparotomy. Brilliantino *et al*⁷ also resorted to laparotomy for a caecal carcinoma located in the left abdomen in a patient with malrotation associated with situs inversus. Donaire *et al*⁴ performed laparoscopy and laparotomy, revealing intestinal malrotation with left upper quadrant caecal carcinoma in a man with unexplained anaemia. Hirano *et al*,⁸ Morimoto *et al*⁹ and Hirano *et al*¹⁰ utilised laparoscopy in patients with malrotation, to resect colonic tumours located in the ascending colon (located in the left abdomen), in the caecum (located posterior to the superior mesenteric artery) and in the transverse colon, respectively.

These seven existing case reports show that intestinal malrotation can go undiagnosed for many years, and that the process may result in several unusual colonic tumour positions. They also show that a minimally invasive management may be appropriate in select patient groups.

In adulthood, intestinal malrotation has an incidence of 0.2% and typically these patients will have had no congenital anomalies associated with childhood malrotation. The actual clinical diagnosis can often be difficult, thus explaining the late presentation in a number of patients.¹¹ Patients will frequently present acutely with bowel obstruction with or without intestinal ischaemia. However, chronic presentations are usually non-specific with patients reporting vague abdominal cramp. As such a significant proportion of adult malrotation is picked up radiologically using several modalities such as barium studies, CT and angiography, and also incidentally at laparotomy.¹²

The patient described in our case report had a good clinical outcome following definitive surgical management. However, her initial negative colonoscopy did jeopardise this. Having analysed the case, we feel that there are at least three themes that warrant discussion.

First, the issue of negative colonoscopy. Colonoscopy is generally regarded as the gold standard in screening for colorectal cancer—this is because, unlike other similar modalities, for example, flexible sigmoidoscopy, the entire large colon can be visualised. Nevertheless, there have been cases in which colorectal malignancy has been subsequently diagnosed despite recent negative colonoscopy. In a North American study, 52% of late-presenting colorectal cancers were attributed to being missed at colonoscopy (or incompletely removed at recent colonoscopy).¹³ To reduce the chance of missing pathology, colonoscopists strive to identify anatomical markers that signify a comprehensive investigation. Complete colonoscopy visualises the entire large bowel

up to and including the ileocaecal junction. Anatomically, three landmarks can aid the endoscopist: the most reliable of which is the readily identifiable ileocaecal valve; the appendicular orifice may also be useful, however, one would not strive to isolate this in patients who have undergone appendectomy; and, finally, the trans-illumination of the scope light through the right iliac fossa skin.¹⁴ The experience of the clinician in locating the mentioned landmarks is significant as a positive predictor of caecal intubation; this is reflected in the literature in which successful intubation rates are seen to rise accordingly with the number of years of training a clinician has. Factors associated with failed caecal intubation were poor bowel preparation and history of stomach surgery.¹⁵ We strongly recommend that these landmarks are used to determine the successful completion of all colonoscopic studies.

The second issue is the shortfalls that surround colonoscopic scope guidance. Guidance systems are a modern solution to the problem of not knowing the orientation and location of the scope. They relay extra information to the endoscopist, in addition to that from the standard scope camera. With a scope guide the endoscopist can more accurately estimate how far they have twisted the scope and the approximate position of the scope tip. Unfortunately, in patients with unusual or bizarre anatomy, this information can be misleading. In this case, the initial endoscopist was led to believe that he had fully visualised the large bowel lumen up to the caecum by the endoscopic guidance system. This would have been corroborated by a kink in the bowel thus mimicking the junction between caecum and terminal ileum. If patients have a known history of malrotation, scope guidance systems should not be used; instead, practitioners should rely on their own acumen and the aforementioned features to ascertain total completion of colonoscopy.

Last, the paramount importance of the clinical history. All patients undergoing any endoscopic procedure must have their history meticulously reviewed. In doing this, clinicians arm themselves for difficult situations. In our case, if the initial endoscopist had been aware of the history of malrotation, he may have proceeded further down the bowel lumen, thus discovering the tumour sooner. We appreciate that there are factors that may work against this: in the emergency setting, such as with the haemodynamically unstable patient with an upper gastrointestinal bleed, it might be difficult to review archived clinic letters before addressing the bleed. Indeed, in busy endoscopy departments there is also continual pressure from endoscopy sisters to complete cases as rapidly as possible. We urge endoscopists to pause and resist, where possible, to review the clinical history; this action should reduce long-term costs by identifying and minimising morbidity as early as possible.

Patient's perspective

The patient was pleased and keen to share her story with BMJ Case Reports. Reflecting on her childhood experience of intestinal obstruction and malrotation and its surgical management, she remembered how her mother had described to her about how her 'duodenum had become all twisted and that her mesentery was the wrong way round!' We thank the patient for her willingness and for her good humour.

Learning points

- ▶ Accurate history taking and full physical examination is absolutely essential before all endoscopic procedures. Detailed background information can inform the endoscopist of any potential anatomical abnormalities that may be encountered.
- ▶ Endoscopic adjuncts or aids, such as scope guides must be used with caution and ideally avoided in those with known intestinal malrotation.
- ▶ Remember, patients with bowel malrotation are just as susceptible to colorectal pathology, including tumours, obstruction and diverticulitis.
- ▶ Aberrant visceral anatomy can make operating on patients with intestinal malrotation difficult (whether surgically managed previously or not) and so the involvement of senior surgeons early on is crucial.

Contributors JMN performed the initial drafting of the case report and coordinated all subsequent tasks in preparation for publication. DO was responsible primarily for the literature review cited in the discussion section. AAA co-wrote the manuscript and was involved with the acquisition of some of the figures. KR was the senior consultant who managed the patient clinically, performed the right hemicolectomy, and provided the important learning and discussion points for the case report. He also selected the most pertinent figures for inclusion.

Competing interests None.

Patient consent Obtained.

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