CASE REPORT

A jejunal GIST presenting with obscure gastrointestinal bleeding and small bowel obstruction secondary to intussusception

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Accepted 15 October 2015

SUMMARY

A 68-year-old man with episodes of overt obscure gastrointestinal (GI) bleeding was investigated with multiple upper and lower GI endoscopies, CT enterography and capsule endoscopy, but no cause was found. He then presented acutely with small bowel obstruction. A laparotomy revealed complete small bowel obstruction secondary to jejunal intussusception over a 4 cm intraluminal polyp. Following resection and primary anastomosis, histology revealed that the polyp was a GI stromal tumour (GIST). This is an exceptionally uncommon presentation of a rare tumour. It is surprising that this tumour was not detected by CT enterography and not seen on capsule endoscopy. Immunohistochemistry and mutation analysis of the GIST suggested that it had a low risk of metastatic disease, but a high risk of recurrence. Staging CT scans did not reveal evidence of distal spread. The patient is currently receiving 3 years of chemotherapy with imatinib.

BACKGROUND

The diagnostic work up and final diagnosis illustrated in this case are both remarkable. This case report illustrates the challenges of investigating obscure gastrointestinal (GI) bleeding (OGIB), where the relative inaccessibility of the small bowel means that more standard endoscopic techniques tend to be preceded by less invasive, but less interventional techniques. It is still surprising that neither capsule endoscopy (CE) nor CT enterography (CTE) detected a 4 cm intraluminal small bowel tumour (SBT). Finally, the patient's journey through successive negative investigations culminated in a more dramatic manner with emergency surgery and a diagnosis of an uncommon tumour presenting in an exceptionally rare manner—a jejunal GI stromal tumour (GIST) causing bowel obstruction due to intussusception.

CASE PRESENTATION

We present a case of a 68-year-old man with ischaemic heart disease, hypertension and insulin-dependent type 2 diabetes mellitus. In 2009, he was being investigated by his general practitioner for mild normocytic anaemia (haemoglobin 12.8 g/dL, mean cell volume (MCV) 81.2 fL). Two colonoscopies revealed sigmoid diverticulosis, colonic polyps and telangiectasia in the ascending colon. An oesophagogastrroduodenoscopy (OGD) revealed a hiatus hernia, but no bleeding source. By early 2013, the patient developed a microcytic anaemia

(haemoglobin 10.5 g/dL, MCV 74.2 fL) and intermittent diarrhoea.

In June 2013, the patient presented with an acute episode of melaena and an associated drop in haemoglobin (from 13.9 to 8.4 g/dL). An OGD showed Barrett's oesophagus, mild gastritis and duodenitis, but no active bleeding. A *Campylobacter*-like organism test and duodenal biopsies were negative. In September 2013, the patient had a similar presentation, but with a negative OGD. He had multiple subsequent outpatient investigations. A colonoscopy revealed diverticulosis, and three polyps in the ascending colon. Histology of these revealed low-grade dysplastic tubulovillous adenomas. CTE was performed in August 2013. Oral negative contrast (Klean-Prep) and single-phase intravenous iodine-based contrast (Omnipaque) were used. The consultant gastro-intestinal radiologist reported that the distal small bowel was well distended with no small bowel wall thickening or mass seen. An outpatient CE was performed in November 2013. The bowel preparation was a 10 h fast following 12 h of a clear fluid diet. The images were read by a consultant gastroenterologist. Small bowel passage time was reported to be 5 h and 45 min. The reporter stated that the capsule probably remained in the small intestine at the end of the study, and that the luminal view was mainly obscured by bowel contents (figure 1). Although no blood or likely bleeding source was seen, the study was considered incomplete, and it was recommended by the author, if symptoms persisted, to repeat CE and include one sachet of Picolax in the bowel preparation.

One week following CE, the patient presented with abdominal pain and distension, vomiting and constipation. The patient did not recall passing the capsule prior to this episode. A contrast-enhanced multidetector abdominal CT scan revealed dilated small bowel loops with a transition point in the pelvis, but no identifiable cause. No capsule was detected. Two days later, the patient underwent a laparotomy for non-resolving small bowel obstruction. The intraoperative findings were complete small bowel obstruction with dilated proximal small bowel and collapsed distal small bowel. The transition point was a 45 cm jejunal segment intussuscepted over a large intraluminal polyp. No capsule was found at surgery. Ten centimetres of small bowel with the polyp were excised and sent for histology followed by primary anastomosis. The postoperative period was uncomplicated.

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To cite: Sadeghi P, Lanzon-Miller S. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-207650

Figure 1 The jejunum obscured by bowel contents.

Macroscopically, the polyp was fleshy and white, measuring 40×40×20 mm (figure 2). Microscopically, there were spindle cells, epithelioid cells, ulceration and a mitotic index of 0–1/5 mm². There was no evidence of distal spread, and the resection margins were clear. Immunohistochemistry was positive for CD117 (KIT), SMA and DOG1, and negative for Desmin, CD34 and S100. The findings were consistent with a small bowel GIST. Based on the tumour size, site and mitotic index, it was considered to be at low risk (4.3%) of metastatic disease, but mutation analysis revealed a deletion at 556–558 of exon 11, indicating a poor prognosis with a recurrence rate of 40–60%.

OUTCOME AND FOLLOW-UP

Following discharge, a staging CT and positron emission tomography CT did not reveal any evidence of metastasis. In May 2014, the patient started imatinib 200 mg once daily.

DISCUSSION

Approximately 5% of GI bleeding cases are caused by small bowel lesions.¹ Primary SBTs account for only 5% of all primary GI tract tumours.² The diagnosis of SBTs is often delayed because symptoms and signs are often absent or non-specific.³ For example, OGIB secondary to an SBT may be overt

Figure 2 Surgically resected portion of the jejunum containing an intraluminal gastrointestinal stromal tumour.

(presenting as haematemesis and/or melaena) or occult (presenting as iron-deficiency anaemia).

Following negative upper and lower GI endoscopies, our patient was appropriately investigated with CTE and CE for overt OGIB.⁴ Enteral contrast for CT examinations may be administered orally (CTE) or via a nasojejunal tube (CT enteroclysis). The combination of small bowel distension with negative contrast and mural enhancement with intravenous contrast allows detailed assessment of the small bowel wall.⁵ A recent meta-analysis (n=696) reported that CT enteroclysis had a per-patient pooled sensitivity of 92.8% for SBTs.⁶ There is, however, a relative paucity of studies looking at CTE detection of SBTs. In a retrospective study comparing CE and CTE in 41 patients with SBT by Hakim *et al*,⁷ CTE had a sensitivity of 92.7% for SBT. Similarly, a prospective study comparing CE and CTE in 59 patients with OGIB by Huprich *et al*⁸ reported that CTE had 100% sensitivity for SBT, although there were only nine patients in this subgroup. Given the reported high sensitivity of CTE for SBT, it is surprising that the patient's 40×40×20 mm SBT was not seen on CTE—specifically, in the case of a GIST, an intraluminal enhancing soft tissue density mass with adjacent wall thickening.⁹

CE can potentially visualise the entire mucosal surface of the GI tract. In a retrospective study (n=145), CE had a sensitivity of 83.3%, a specificity of 100%, a positive predictive value of 100% and a negative predictive value of 97.6% for SBT.¹⁰ However, CE lacks the interventional capabilities of standard endoscopy; it only visualises the mucosal surface of the bowel and thus cannot detect submucosal or serosal abnormalities, and lesion localisation is not precise. The patient's poor bowel preparation may have been a contributing factor to the SBT being missed. In a meta-analysis of bowel preparation protocols in CE (n=401) by Niv,¹¹ sodium phosphate or polyethylene glycol preparations improved bowel wall visualisation compared to standard fasting guidelines. Indeed, Zagorowicz *et al*¹⁰ report that the three patients in whom SBTs were missed by CE had inadequate bowel preparation. Therefore, had the patient not presented acutely with small bowel obstruction, it would have been appropriate, as per the reporting Consultant's recommendation, to repeat CE with an osmotic laxative included in the bowel preparation protocol. It is interesting to note that the capsule appears to have been temporarily retained in this case, which, in retrospect, may have been due to the partially obstructing intraluminal GIST. Although the patient did not recall passing the capsule before his final acute presentation, no capsule was detected on CT or found at surgery; therefore it is assumed that the capsule had been passed. Finally, if a high clinical suspicion remains for a small bowel lesion, but CE is negative, some authors report that balloon-assisted enteroscopy (including possible total enteroscopy) should then be considered.^{12 13}

GISTs are found throughout the GI tract but are most commonly in the stomach (60%), followed by the jejunum and ileum (30%). The existence of primary extraintestinal GISTs is controversial.¹⁴ In a Swedish population-based study,¹⁵ approximately 70% of GISTs were associated with clinical symptoms, 20% were not (detected incidentally during endoscopy, radiological scans, or surgery) and 10% were only detected at autopsy. The likelihood of a GIST being symptomatic was proportional to its size. GISTs most commonly present with dyspepsia and GI bleeding, caused by pressure necrosis and ulceration of the overlying mucosa.¹⁴ Rarely, they may present with bowel obstruction¹⁶ or tumour rupture with haemoperitoneum.¹⁴ Bowel obstruction due to intussusception in this

context is extremely rare since GISTs tend to grow exophytically,¹⁷ not intraluminally, as in this case. A literature search revealed only a handful of case reports of a GIST presenting with pure jejunal^{18–21} or jejunoileal²² intussusception.

This interesting case illustrates the challenges of investigating OGIB. The relative inaccessibility of the small bowel means that more standard endoscopic techniques allowing real-time visualisation of, and intervention to, the GI tract (eg, OGD, colonoscopy) tend to be preceded by less invasive techniques such as CE and CTE. It is indeed surprising, however, that neither CE nor CTE, with reported sensitivities of 83.3% and 92.7–100% for SBTs, respectively, detected a 40×40×20 mm intraluminal SBT. Where there is a high index of suspicion for a small bowel bleeding source despite multiple negative investigations, we would advocate considering repeating investigations in an optimised form, for example, using osmotic laxatives to improve bowel wall visualisation during CE. It may even be appropriate to consider beginning the diagnostic work up again.

In summary, it is not only the diagnosis, but also the diagnostic work up illustrated in this case that makes it remarkable. The patient's unfruitful journey through successive negative investigations for OGIB culminated in a more dramatic manner with emergency surgery and a diagnosis of a rare tumour—a jejunal GIST—presenting in an exceptionally rare manner—bowel obstruction due to intussusception.

Learning points

- ▶ The relative inaccessibility of the small bowel means that less invasive though less interventional techniques, such as capsule endoscopy and CT enterography, tend to be first line in the investigation of obscure gastrointestinal (GI) bleeding.
- ▶ If multiple investigations for obscure GI bleeding are negative, it may be appropriate to consider optimising and repeating certain investigations.
- ▶ Gastrointestinal stromal tumours typically present with dyspepsia and GI bleeding, but rarely may present with obstruction due to intussusception.

Contributors PS researched and wrote the article. SL-M reviewed and edited it.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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