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Rare disease

Gastrointestinal stromal tumour presenting as gastroduodenal intussusception

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Summary

Gastroduodenal intussusception secondary to gastrointestinal stromal tumour is a very rare cause for intestinal obstruction. The diagnosis of this condition can be challenging, as symptoms are often non-specific and intermittent. This article reports a case where the diagnosis was made preoperatively with abdominal imaging and was treated by a combination of endoscopic reduction and laparoscopic resection.

BACKGROUND

Intussusception can be defined as the invagination of one part of the gastrointestinal (GI) tract into another and most commonly involves the invagination of a proximal segment of bowel (intussusceptum) into an adjacent distal segment (intussuscipiens). Overall, adult intussusception represents approximately 5% of all intussusceptions[1] and accounts for approximately 1% of all bowel obstructions.[2] In contrast to children, however, 90% of adult intussusceptions have an associated anatomical abnormality, usually a benign or malignant neoplasm.

Intussusception can occur at any site in the gastrointestinal tract. In children, the typical site for intussusception is ileo-colic but in adults, the majority involves either ileo-ileal or colo-colic segments. Intussusception involving the proximal gastrointestinal tract is exceedingly rare.

CASE PRESENTATION

A 78-year-old woman was referred with a 1-week history of persistent upper abdominal discomfort, vomiting and anorexia. She had similar, but milder, episodes intermittently over the preceding 2 months. Gastroscopy performed at the referring hospital reported what appeared to be invagination of the distal stomach into the duodenum. Her history included hypertension, osteoporosis and previous laparoscopic cholecystectomy. Her sister was recently diagnosed with bowel adenocarcinoma.

INVESTIGATIONS

The only abnormal finding on haematological investigations was a mildly elevated urea level, which was consistent with her dehydrated state secondary to vomiting. She underwent a contrast-enhanced abdominal CT scan and this demonstrated a dilated stomach with intussusception of the distal stomach extending as far as the second part of the duodenum. The intussusception lead point appeared to be a polypoid gastric mass.

DIFFERENTIAL DIAGNOSIS

Distal gastric tumour, foreign body, tumour of the duodenum, ectopic pancreatic tumours and head of the pancreas tumours.

TREATMENT

Intraoperative gastroscopy revealed a distended stomach with a polypoid mass intussuscepted into the first part of the duodenum. Endoscopic reduction was carried out and the polyp appeared to originate from the distal body and antrum of the stomach.

Laparoscopy did not reveal any evidence of metastases and laparoscopic exvagination of the gastric mass was performed via anterior gastrotomy. A laparoscopic linear stapler (Endo GIA Ultra Universal Stapler; Covidien Surgical, Mansfield, MA, USA) was used to take a wedge resection of the stomach encompassing the lesion. Several applications of the stapler were required to complete the resection. The mass was placed in the endoscopic retrieval bag and removed through the umbilical port. Air insufflation of the stomach was carried out with a gastroscope and the no evidence of air leakage was seen under direct laparoscopic visualisation. The patient was returned to the postanaesthetic care unit and the specimen was sent for histopathological analysis.

OUTCOME AND FOLLOW-UP

Postoperatively the patient was allowed clear fluids the following morning and was started on a soft diet on day 3. She was discharged home on postoperative day 4. On follow-up, she developed a minor superficial wound infection at the umbilical port wound. This was treated with oral antibiotics and it resolved quickly with an otherwise uneventful recovery.

The surgical specimen was 45×33×34 mm in size. Histopathological analysis of the polypoid mass showed an admixture of spindle and epithelioid cells and areas of myxoid change with the presence of vacuolated cells. Immunohistochemistry with c-kit antibody confirmed the diagnosis of gastrointestinal stromal tumour.

DISCUSSION

Gastroduodenal intussusception is a rare cause of intestinal obstruction. When it occurs, the most common aetiologies are intussusception of the gastric remnant through a gasrtojejunal anastomosis or of gastric polyp through

the pylorus into the duodenum.[3] It presents with a variety of symptoms that can be acute, intermittent or chronic. An epigastric mass may be palpated on physical examination. The widespread availability of abdominal imaging has made preoperative diagnosis of gastroduodenal intussusception far more common.[4 5] CT is especially useful and can provide excellent anatomic detail of the intussusception morphology.

Gastroduodenal intussusception secondary to gastrointestinal stomal tumours (GIST) appears to be a very rare cause for intestinal obstruction. It is caused by prolapse of the tumour and subsequent invagination of a portion of full thickness gastric wall into the duodenum.

GISTs are mesenchymal neoplasms and are thought to develop from the interstitial cells of Cajal. These tumours have been reported ranging in size from smaller than 1 cm to as large as 40 cm in diameter. The annual incidence and prevalence of GIST has been reported to be 14.5 per million population and 129 per million population, respectively.[6] They can occur at any point of the GI tract but occur most commonly in the stomach (47–60% of cases). Most (50–80%) GISTs arise because of a mutation in a gene called c-kit. The c-*kit* proto-oncogene encodes KIT, which is a transmembrane tyrosine kinase receptor. All GIST tumours should be considered to have malignant potential and no GIST tumour can be correctly classified as 'benign'. Predictors of behaviour include size, mitotic rate, location, complete surgical resection and tumour rupture. Tumours <2 cm in size with a mitotic rate of <5/50 high-power field have been shown to have lower risk of recurrence.

Surgery offers the only treatment option that can definitively cure the disease and the standard initial treatment for non-metastatic disease is en-bloc resection. Lymphadenectomy not usually indicated. Five-year survival rates are between 48% and 65% after complete resection. GISTs are notoriously resistant to chemotherapeutic agents. The only effective, specific, non-surgical therapy for GISTs is imatinib (Gleevec; Novartis, East Hanover, NJ, USA). This is a selective tyrosine kinase inhibitor with action against mutant c-kit as it occurs in association with GISTs.

Imatinib has a 40–70% response rate in metastatic or inoperable cases. The 2-year survival of patients with advanced disease has risen to 75–80% following treatment with imatinib.[7 8] It is considered a significant breakthrough in antineoplastic drug therapy because it is targeted against a specific molecular derangement.

Learning points

- Gastrointestinal intussusception due to gastrointestinal stromal tumour is an exceedingly rare cause for intestinal obstruction.
- Preoperative diagnosis can be challenging as symptoms are often vague, intermittent and long standing.
- CT is very useful in the diagnosis and in demonstrating the morphology of the intussusception.
- A combination of endoscopic reduction and laparoscopic resection is a safe approach to treatment.

Competing interests None.

Patient consent Obtained.

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