CASE REPORT

Small bowel carcinoid: a rare cause of bowel obstruction

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SUMMARY

Carcinoid tumours though commonly affect the appendix, are a rare cause of small bowel obstruction, causing a diagnostic dilemma. We presented a 70-year-old man with small bowel obstruction, not responding to conservative management, which required an emergency laparotomy and was found to have a mass encasing the mid-jejunal loops and mesentery that was resected and reported to be a carcinoid tumour.

BACKGROUND

Carcinoid tumours are a group of low-grade, slow-growing, well-differentiated tumours originating from the endochromaffin cells of Kulchitsky, that is, neural crest cells situated at the base of the crypts of Lieberkuhn in the gastrointestinal tract (GIT) and can produce several biologically active substances (hormones) and hence are classified into 'functioning' and 'non-functioning' types.¹ Though the appendix is commonly affected, they can rarely occur in any part of the small and large bowel and usually present with abdominal pain, bowel obstruction and metastases, which might require surgical management.² Diagnosis of carcinoid tumours is made on the basis of clinical manifestations, peptide and amine secretion and radiological/nuclear imaging but confirmation is made by histological analysis.³ Medical treatment is mainly with somatostatin analogues, which are effective in controlling most symptoms. Surgery is the cornerstone of treatment for intestinal carcinoids.⁴

CASE PRESENTATION

A 70-year-old man with no comorbidities was referred with generalised abdominal pain, distension and obstipation of 2 days duration. He had nausea but no vomiting or fever with normal urine output. There were no similar or other significant systemic symptoms. General examination was unremarkable and he was neither pale nor dehydrated. Vital signs were normal. Abdominal examination revealed generalised gaseous distension with no tenderness, guarding, rigidity, mass or free fluid. On per-rectal examination, there was no tenderness and the rectum was empty.

INVESTIGATIONS

All blood and biochemical investigations were normal. An erect abdomen X-ray performed showed multiple air-fluid levels suggesting small bowel obstruction (figure 1). Owing to financial constraints, an abdominal CT scan was deferred.

Figure 1 Plain erect X-ray abdomen showing multiple air-fluid levels indicating small bowel obstruction.

TREATMENT

As the conservative management for 48 h showed no improvement in symptoms, an emergency exploratory laparotomy was performed. Peroperatively, there was a hard mass in the mid-jejunum encasing the jejunal loops and mesentery. The proximal and distal small bowel was densely adherent to this mass and could not be separated. Hence a resection of the mass along with the involved small bowel was performed (figure 2). Rest of the bowel and abdominal organs were normal.

OUTCOME AND FOLLOW-UP

Postoperatively, the patient made an uneventful recovery and was started on orals on day 4. The final histopathology revealed a tumour consisting of uniform cells with regular central nuclei, faint eosinophilic cytoplasm with mitosis, a vascular

Figure 2 Excised specimen consisting of the mid-jejunum and mesentery.

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stroma with hyaline change and dense fibrosis, consistent with small intestinal carcinoid (figure 3). The patient has been kept under close follow-up for last 1 year, is asymptomatic and is doing well.

DISCUSSION

Carcinoid tumours were originally termed 'karzinoide' or 'cancer-like' by Oberndorfer in 1907 due to their slower growth in comparison with adenocarcinomas but the current nomenclature for tumours of epithelial origin occurring in the GIT with neuroendocrine differentiation is gastroenteropancreatic neuroendocrine tumours or 'GEP-NETs' and are specifically found in the ileum and appendix. They can also be found in the rectum, caecum, jejunum, pancreas, biliary duct, oesophagus, liver and lung. The incidence rates of carcinoid tumours is between 2 and 5/100 000.⁵

Carcinoid tumours can produce several biologically active substances, such as serotonin, histamine, dopamine, gastrin, kinins, kallikrein, adrenocorticotrophic hormone, growth hormone, substance P, calcitonin and prostaglandins. Hence, they can be classified into 'functioning' and 'non-functioning' types on the basis of the hormone production.^{6–7} Non-functioning carcinoids are usually asymptomatic or may show symptoms of mass effect, as in our patient. They are found incidentally during surgery, with their carcinoid nature being ascertained only following histological analysis.⁸ Functioning carcinoids have the potential to cause carcinoid syndrome, resulting from excessive activation of bioactive mediators throughout systemic circulation and manifests with a plethora of symptoms including nausea, vomiting, watery diarrhoea, facial flushing, wheezing, weight loss, abdominal pain, GI blood loss and heart failure. This is most likely when the carcinoid tumour is situated in the small intestine (90%).^{9–10} In some rare cases, tumour manipulation, anaesthetic induction or other invasive therapeutic procedures such as embolisation or radiofrequency ablation (RFA) may induce a sudden release of large amounts of vasoactive peptides into the systemic circulation resulting in carcinoid crisis, which consists of severe flushes, diarrhoea, dehydration, hypotension and arrhythmias. Octreotide prophylaxis is recommended during invasive surgical procedures involving carcinoid tumours.¹¹

The majority of patients with small intestinal carcinoids present with abdominal pain, bowel obstruction and metastases. Metastases is most common to liver (60–80%) and lymph nodes.¹² Another complication of small intestinal carcinoids is their tendency to cause mesenteric fibrosis, which could lead to kinking of the bowel, ischaemia, necrosis and perforation.¹³

Figure 3 Photomicrograph showing uniform cells with regular central nuclei, mitosis, vascular stroma with dense fibrosis (H&E $\times 40$).

The diagnosis of carcinoid tumours is made on the basis of clinical manifestations, peptide and amine secretion, radiological and nuclear imaging. Confirmation is by histological analysis. The commonly found biological markers in small intestinal carcinoids are chromogranin A, pancreatic polypeptide, neurokinin A and 5-hydroxyindoleacetic acid.³ CT scans are the most common imaging modality for small intestinal carcinoids, while endoscopic ultrasounds and intraoperative ultrasounds (IOUS) are also routinely used.¹⁴ Somatostatin receptor scintigraphy is the most sensitive modality to identify carcinoid tumours and their hepatic metastases. It utilises the affinity of radioactive-labelled somatostatin analogues to G protein-coupled glycoproteins, which are expressed in 70–90% of carcinoid tumours.¹⁵ The combined use with CT scans improves accuracy of localisation of tumours.

Surgery has been the cornerstone of treatment of small intestinal carcinoids. It remains as the chief modality of cure in patients with limited disease, especially when margin-negative resection is achieved.⁴ However, liver metastases develop in 50–75% of cases and curative liver resection is possible only in about 10% of cases. Therefore, surgery can also be employed for cytoreductive or palliative purposes in patients with extensive disease or metastases. Irrespective of the presence or absence of liver metastasis, resection of the primary tumour and extensive mesenteric lymphadenectomy is indicated to cure or delay progression of the disease. Mesenteric and retroperitoneal fibrosis with associated venous engorgement can lead to excessive bleeding intraoperatively. Clearance of metastatic lymph nodes affords symptomatic relief in addition to improving long-term survival.^{16–17}

Patients presenting with small bowel obstruction or other GI emergencies may require resections to treat the presentation, in addition to further radical resections following definitive histopathology. Radical resections are generally advocated for all tumours over 2 cm in diameter. For smaller tumours, presence of vascular or lymphatic invasion is an indication for further radical surgery.¹⁷ Postoperatively, patients must be followed up for 10 years. The 5-year survival rates following surgical resection of primary and metastatic small intestinal carcinoid tumours range between 60% and 80%.¹⁶

Medical treatment is chiefly with somatostatin analogues, which are very effective in control of most symptoms of carcinoid syndrome. They may also have an antiproliferative effect on the tumour.⁴ Other therapeutic modalities include RFA, cryoablation, targeted radionuclide therapy, chemoembolisation and chemotherapy with streptozotocin and 5-fluorouracil. Newer medications for advanced and inoperable tumours include sunitinib and everolimus.^{11–18}

Learning points

- ▶ Carcinoid tumours are a group of low-grade, slow-growing, well-differentiated tumours.
- ▶ They can produce several hormones.
- ▶ Appendix is commonly affected.
- ▶ Though the diagnosis is made on the basis of clinical manifestations, laboratory investigations and radiological/nuclear imaging, confirmation is always by histological analysis.
- ▶ Medical treatment is mainly with somatostatin analogues.
- ▶ Surgery is the cornerstone of treatment of intestinal carcinoids.

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Competing interests None.

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