Midgut neuroendocrine tumour presenting with acute intestinal ischaemia in a regional centre

Joshua Sungho Hong, Elizabeth Lockie, Sam George

DESCRIPTION
A man in his early 80s presented to a regional hospital with 1 day of lower abdominal pain on a background of 3 months of diarrhoea and weight loss. He had no medical comorbidities nor surgical history, and was independent at home alone. Clinical examination revealed tachycardia (heart rate 115 beats/min) with otherwise normal vital signs, and localised peritonism of right central abdomen. Laboratory tests showed elevated C reactive protein (214 mg/L, normal range: 0–5 mg/L) with all other laboratory markers being normal including leucocytes and venous blood gas. Initial non-contrast CT scan showed large volume free fluid and pneumoperitoneum, congested mesentery and a solid mesenteric mass (2.3 × 4.5 cm) with internal calcification closely related to the third part of the duodenum, without definite cause for ruptured viscus (figure 1). Based on the clinical and radiological findings, the patient proceeded to emergency diagnostic laparoscopy with findings of four quadrant faecal peritonitis and ischaemic small bowel with perforation (figure 2), and thus decision was made to convert to a laparotomy. Careful inspection of the liver, peritoneum and the mesentery showed no metastatic features. The perforation was in the distal ileum approximately 30 cm proximal to the ileocecal valve, in a segment of bowel that was ischaemic likely secondary to the tumour in the root of mesentery obstructing the blood supply. The duodenojejunal flexure was dusky but viable. The ischaemic segment of small bowel was resected (195 cm) and side-to-side anastomosis was performed, followed by peritoneal washout and lavage. No somatostatin analogue was given intraoperatively as the patient did not present with abdominal carcinoid crisis as indicated by our clinical examination and a normal laboratory marker, and localised peritonism of right central abdomen.

Figure 1 CT of initial presentation in the emergency department.

Figure 2 Dusky appearing small bowel with faeculent peritonitis.

Figure 3 CT angiogram day 4 post operation.

Superior mesenteric vein (SMV) was also occluded with only an 11 mm portion of the terminal SMV remaining patent (figure 3). The small bowel resection contained a 6 mm well-differentiated neuroendocrine tumour, presumably the primary. The chromogranin A level sent preoperatively was within the normal range (74.0 µg/L, normal range <102.0). The patient progressed well post operatively and was discharged to a local healthcare facility for ongoing recuperation day 10 post operatively. The patient was scheduled for a 68Ga-Dotatate Positron Emission Tomography (PET)/CT scan for possible metastases and medically managed by an oncologist with somatostatin analogue (lanreotide 120 mg monthly). No other forms of adjuvant modalities were used for this patient.

Neuroendocrine tumours (NETs), previously known as carcinoids, are relatively rare but a tumour that is reported to be in increasing incidence. NETs can occur in any part of the body but most commonly presents in the gastrointestinal (GI) tract (60.9%) followed by in the bronchopulmonary system (27.4%). Within the GI tract, small intestine, rectum, colon, pancreas, stomach and appendix are the common locations in descending order. Intestinal obstruction and ischaemia are some of the well-known complications of midgut NETs which are due to the elastic sclerosis related to a desmoplastic reaction associated with the excessive hormonal release from the local tumour. However, the patient in this presented case did not present with abdominal carcinoid crisis as indicated by our clinical examination and a normal
Images in...

chromogranin A level. Surgery still remains as the mainstay of treatment for localised tumour, providing as the only curative option. However, no attempts to debulk the tumour was made intraoperatively for this patient as the tumour was encasing the distal superior mesenteric artery which could have led to unfavourable situation especially in a regional centre where there are no vascular or interventional radiology services on site. After careful discussion, debulking surgery of the primary tumour was not elected for this patient due to advanced age and comorbidities. However, alternative decision could be considered for patients with a better physiological reserve.

Patient’s perspective

Everything happened very quickly from the time of presentation to having the operation. Otherwise, I have good support network around me at home and feeling not too bad after what I have gone through.

Learning points

► Neuroendocrine tumour is relatively a rare tumour but it is increasing in incidence.
► Intestinal obstruction and ischaemia are some of well-known complications of midgut neuroendocrine tumours.
► Initial imaging investigation with intraoperative judgement is essential in determining the location of the tumour and surrounding vital structure. A damage control laparotomy for non-traumatic abdominal emergency can be performed then deciding on a definitive surgery.

In conclusion, the presented case highlights the initial presentation of a midgut NETs as a mesenteric ischaemia. Initial imaging investigation with intraoperative judgement is essential in a case like this where the tumour sits near a vital structure. Therefore, a damage control laparotomy for a non-traumatic abdominal emergency can first be performed then deciding on a definitive surgery.

Contributors JSH: conceptualisation; data curation; methodology; writing – original draft; writing – review and editing. EL: writing – review and editing. SG: conceptualisation; writing – review and editing.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Consent obtained directly from patient(s)

Provenance and peer review Not commissioned; externally peer reviewed.

Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

ORCID ID
Joshua Sungho Hong http://orcid.org/0000-0003-4610-297X

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