A Jejunal Gastrointestinal Stromal Tumour: an unusual cause of massive acute gastrointestinal haemorrhage with emphasis on pre intervention MDCT

Suchi Gaba^{1*}, Mansoor Aslam², Amjad Iqbal²

1. Department of Clinical Radiology, Nuffield Orthopaedic Centre NHS Trust, Oxford, UK

2. Department of Radiology, Pilgrim Hospital, United Lincolnshire Hospitals NHS Trust, Boston, UK

* Correspondence: Suchi Gaba, FRCR, MD, Department of Clinical Radiology, Nuffield Orthopaedic Centre NHS Trust, Oxford, OX3 7LD, UK (Segmail.com)

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ABSTRACT

Gastrointestinal stromal tumors (GIST) most commonly arise from the stomach followed by the small intestine and are common cause for an occult gastrointestinal (GI) bleeding. We present an unusual case of a jejunal GIST, which presented as an acute gastrointestinal haemorrhage. This case highlights the importance of an intravenous contrast enhanced abdominal CT with neutral oral contrast for the assessment of gastrointestinal bleeding where non-obstructive enhancing tumour, active extravasations and arteriovenous malformations (AVM) could potentially be obscured by the use of positive oral contrast. This case also emphasizes on the use of multiplanar image reconstructions that are extremely useful in determining the exact location and size of the pathology.

CASE REPORT

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A 67 years old female presented to accident and emergency department with history of a single episode of collapse and two episodes of melaena. Patient was treated for peptic ulcer disease 5 years ago and had no other significant past medical history. The haemoglobin on the day of the admission was 10.2 g/dL and this dropped to 6.3 g/dL on the day four as patient had two more episodes of melaena. A total of five units of blood transfusion were given to the patient.

An oesophagogastroduodenoscopy (OGD) showed a small sliding hiatus hernia and colonoscopy was unremarkable. The case was referred to our department for mesenteric angiogram with possibility of embolisation of a bleeding ulcer in the distribution of superior mesenteric artery. A pre intervention CT abdomen and pelvis was performed with intravenous contrast and oral water. The protocol in our department is to use a neutral/negative contrast in such situations.

Imaging technique

CT abdomen and pelvis with intravenous contrast was performed on a 4 slice helical Toshiba scanner. Positive oral contrast was not given. Patient had 750 ml of plain water and was not able to tolerate more neutral oral contrast. CT (Fig. 1a) with coronal (Fig. 1b) and sagittal multiplanar reformatted images (MPR) demonstrated 4.0 x 3.5 cms eccentric mass in the proximal jejunum. Mass had lobulated margins and revealed avid enhancement with intravenous contrast. No gross ulceration, necrotic change, fatty components or calcification were noted in the mass. No active extravasation of contrast was noted on the CT. There was minimal pelvic acsites. Rest of the abdominal viscera were unremarkable and no intrabdominal lymphadenopathy was demonstrated. The mass was thought to be submucosal rather than mural in origin due to its eccentric location and non-obstructive nature. Differential diagnosis of leiomyoma, leiomyosarcoma, GIST and gastrointestinal schwannomas were considered.

Laparotomy findings

Based on the CT findings and clinical condition with a presentation of acute haemorrhage an urgent laparotomy was performed. The laparotomy confirmed a hard, eccentric and nodular tumour in the antimesenteric border of the jejunum approximately 30 cm distal to the duodeno-jejunal flexure (Fig. 2). This part of the small bowel was kinked and adherent to the transverse mesocolon with vascular adhesions as well. This part of the bowel was resected with the tumour. Fresh blood was noted in the jejunal lumen with small amount of free fluid in the abdomen. Rest of the small and large bowel were unremarkable. There were irregularities on the mucosal surface covering the tumour but no frank ulceration was seen on the gross specimen examination.

Histopathology findings

Histopathology examination revealed that local excision was complete with clear margins and the edge of the neoplasm was smooth with no features of infiltration. There was central fibrosis and mucosal ulceration, which was covered by clot which accounted as a site for the blood loss (Fig. 3a). No necrosis was noted within the tumour, which correlated well with the CT findings. Tumour was composed of interwoven fascicles of spindle cells with mild nuclear anisocytosis and pleomorphism. There was no significant mitotic activity (Fig. 3b). The neoplastic cells illustrated wide spread positivity for CD117 (a proto-oncogene) and CD34 stains and were also focally positive for smooth muscle actin. Cells were found to be negative for desmin and S100 protein immunochemical stains.

The histological appearance and immunohistochemical profile of the mass confirmed it to be a low grade GIST and negativity for desmin and S100 suggested that mass was not of neurogenic origin. Patient had uneventful recovery. She was followed up after one and six-months without any untoward incident.

DISCUSSION

GISTs are rare with an incidence of 0.1-3% of all gastrointestinal (GI) tumours but are the most common mesenchymal tumours of the GI tract (1). 5% of GI haemorrhage is obscure in nature and GISTs have been described as one of the cause (2). An obscure gastrointestinal bleeding is difficult to treat and with patient dissatisfaction can disappoint the clinicians. GISTs predominantly occur in stomach followed by the small intestine, colon, rectum, and oesophagus (1). The most common clinical manifestation for symptomatic GISTs is occult gastrointestinal bleeding from mucosal ulceration (3).

Multidetector high-resolution CT (MDCT) images with multiplanar reformation can precisely silhouette the small bowel pathology (4). CT scan for our patient was performed on a 4 slice helical MDCT scanner however, now-a-days new generation multi slice scanners provide superior spatial resolution and aid in better characterisation of intestinal tumours. It also helps in delineating the bleeding source in patients with acute GI haemorrhage (5) and should be a part of the algorithm in the management of an acute gastrointestinal haemorrhage (6). In our case there was no active arterial bleeding seen on CT. It has been reported in a few studies that MDCT is more sensitive to digital subtraction angiography in revealing the gastrointestinal arterial bleeding in both animal (7) and vitro models (8). Recently it has been demonstrated that intravenous contrast enhanced MDCT has threshold of 0.35ml/min for the detection of an active arterial bleeding in comparison to DSA, which has 0.96ml/min (8).

The main differential diagnosis of benign or a small sized malignant GIST is gastrointestinal schwannomas (9). Gastric schwannomas are divided into two major subgroups as mesenchymal or neuroectodermal (9). It is crucial to differentiate GIST from GI schwannomas, as GI schwannomas are biologically benign tumours with an excellent prognosis (10) whereas GISTs carry a relatively higher malignant potential (11). 20-30% of the GISTs are malignant in nature but most (70-80%) are benign (1). On CT imaging, GI schwannomas have homogenous attenuation and can be readily differentiated from a large benign or malignant GIST, which demonstrates heterogeneous enhancement due to haemorrhage, necrosis and intra-lesional cystic changes (9). Gastrointestinal schwannomas are distinctive from conventional schwannomas, which originate from CNS or soft tissues (9).

Diagnosis on CT imaging becomes very difficult if the mass is small and lacks ulceration and necrosis. In such cases histopathology is often required for a confident diagnosis.

The density of the positive oral contrast is not uniform throughout the bowel and depends on multiple factors including the bowel motility, amount of fluid in the bowel lumen, obstructed, non obstructed and formulation of the oral contrast used. Our department protocol is to use 20 ml of 270mg Iodine/ ml diluted in 1000ml of water as positive oral contrast for CT, which can give an intra-luminal density ranging from 30 HU in segments with poor opacification to 200 HU in segments of bowel with good opacification. Lesions such as GIST, leiomyoma and leiomyosarcoma often show avid contrast enhancement with high Hounsfield value (114.8 HU in our case) which matches the density of the positive oral contrast in the segments of the bowel with good opacification. Hence, there is a possibility of an enhancing tumor, active extravasation and a submucosal AVM or angiodysplasia being obscured by the contrast in the bowel lumen. A neutral or negative oral contrast such as water or milk would be preferable if the patient can tolerate any oral intake. This will avoid potential pitfalls in the interpretation of the mesenteric angiograms, if a conventional angiogram were to be performed soon after the CT imaging.

TEACHING POINT

This case highlights the importance of post-intravenous contrast MDCT with neutral oral contrast to establish a cause for GI haemorrhage. The MDCT images should also be reviewed on a three dimensional workstation with multi-planar reformats.

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ABBREVIATIONS

GIST= Gastrointestinal stromal tumor GI = gastrointestinal CT= Computed Tomography AVM= Arteriovenous malformation OGD= oesophagogastroduodenoscopy MPR= multiplanar reformats MDCT =Multidetector high-resolution CT HU= Hounsfield Unit

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FIGURES

Figure 1: 67 year old female with GIST causing massive acute gastrointestinal haemorrhage. Axial (A) and coronal (B) contrast-enhanced CT images display an avid and heterogeneous enhancing mass (arrow) with average attenuation of 114 Hounsfield Unit, arising eccentrically from the proximal jejunum.



Figure 2: 67 year old female with GIST causing massive acute gastrointestinal haemorrhage. Photograph of the cut surface of the resected surgical specimen. A pale, lobulated mass with central scar and a small focus of ulceration.



Figure 3: 67 year old female with GIST causing massive acute gastrointestinal haemorrhage. A) Photomicrograph (H&E stain; original magnification x20) revealing a focus of mucosal ulceration covered by an intraluminal clot. B) Cytology (H&E stain; original magnification x200) demonstrating fascicles of spindle cells with mild nuclear anisocytosis and pleomorphism with no significant mitotic activity.

KEYWORDS

Gastrointestinal stromal tumors, GIST, CT, Gastrointestinal bleeding, MDCT, Multidetector Jejunum, Computed Tomography

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