Bleeding Jejunal GIST: An uncommon cause of Gastrointestinal Bleeding

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SUMMARY

Gastrointestinal stromal tumours (GIST) are rare gastrointestinal tumours and are one of the causes of obscure gastrointestinal bleeding. We report a case of massive gastrointestinal bleeding secondary to bleeding jejunal GIST in a 43 years old gentleman. Endoscopic intervention failed to identify the source of bleeding and CT Angiography (CTA) showed a jejunal mass and patient underwent laparotomy and resection of the bleeding jejunal GIST. This article highlights the rare cause of the massive GI bleeding and also emphasise the role of CTA in obscure GI bleeding

INTRODUCTION

Gastrointestinal stromal tumours (GIST) are rare with an incidence of 0.1-3% of all gastrointestinal (GI) tumours but are the most common mesenchymal tumours of GI tract. [1, 3, 4] 5% of GI haemorrhage is obscure in nature and GIST has been described as one of the causes.[1] Obscure GI bleeding has always been a challenge for clinicians to diagnose them and advocate appropriate treatment.

This article reports a case of massive GI bleeding secondary to Jejunal GIST and also emphasise the role of CTA in an obscure GI bleeding.

CASE REPORT

A 43 years old gentleman with underlying diabetes mellitus and ishaemic heart disease, presented with 1 day history of passing out malenic stools prior to admission. On admission to hospital, he was hypotensive and haemoglobin level was 5.6 g/dl. Upon admission to the ward patient still had ongoing bouts of malena. He required 5 pints of packed cell transfusion. OGDS revealed no evidence of upper gastrointestinal bleeding and colonoscopy showed blood throughout the length of the colon and there was blood in the terminal ileum. We proceeded with a CTA which showed an exophytic, well defined enhancing mass, which was arising from the proximal jejunum, and a provisional diagnosis of bleeding jejuna GIST was made. It measured 4.4cmx3.4x5cm and within this mass, there was a small hypodensity area, representing central necrosis. Branches from the jejunal arteries were seen coursing through this mass with large draining vein from the mass at the lateral aspect of the jejuna wall and into the superior mesenteric vein. Patient underwent laparotomy and Intra operative findings were proximal jejunal GIST, 10 cm from the D-J flexure. Patient underwent resection of tumour with primary anastomosis. Patient recovered well from the operation and did not have any further episodes of GI bleeding.

The HPE results showed a submucosal tumour with well demarcated edges expanding to serosa. The tumour composed of fascicles of spindly cells with elongated oesinophilic cytoplasm. Mitotic figures were occasionally seen, average count 3/50 high power fields. The immunohistochemical stain: vimentin, CK117 were positive, meanwhile EMA, S100, Desmin and myogenin were negative. It was classified as a GIST with low risk.

DISCUSSION

The term GIST was first coined by Mazur and Clark to describe a heterogenous group of gastrointestinal non – epithelial neoplasms. It was further defined by Hirota and co – works in 1993 and finally GISTs are defined as pleomorphic mesenchymal tumours of the GI tract that express the KIT protein CD 117 and also CD 34 on immunohistochemistry.[5]

GISTs predominantly occur in stomach followed by the small intestine, colon, rectum and esophagus. The most common clinical manifestation for symptomatic GISTs is occult GI bleeding from mucosal ulceration. There are few factors that may contribute to the haemorrhage of jejunal GIST, firstly the location at the small bowel is associated with the highest incidence of bleeding. 64% of small bowel GISTs present with bleeding, whereas gastric, colonic and rectal GISTs have been associated with <50% incidence of bleeding. 2 Second, although extra luminal in origin, GIST may ulcerate through the overlying mucosa, causing intraluminal bleeding. [2] Third, stromal collagen is minimal in most GISTs, but delicate, thin walled vessels may be prominent, making stromal haemorrhage a common feature of these tumours. [2]

The diagnosis in small bowel GIST is difficult as they are inaccessible to endoscopy and the standard angiography has low specificity especially in light or intermittent bleeding. [2] These patients usually end up with exploratory laparotomy to identify the source of the bleeding.

CT angiography can overcome the shortage of standard angiography; multi detector row CT has an accuracy of 100% for localisation of acute GI Bleeding. Multi detector row CT (MDCT) features strikingly increased image resolution and markedly decreased scanning time. These attributes enable acquisition of accurate arterial phase images and thus

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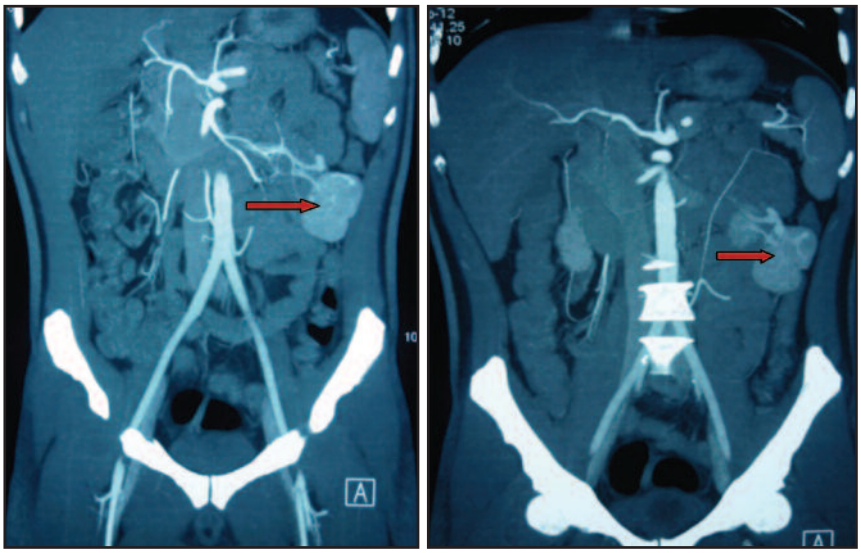

Fig. 1: The Pre-operative CTA showing the exophytic, enhancing mass (red arrows) which represents jejunal GIST.

identification of extravasations of contrast material into the intestinal lumen, a finding diagnostic of acute GI bleeding. 2 Intravenous contrast enhanced MDCT has threshold of 0.35ml/min for detection of an active arterial bleeding in comparison to DSA which is 0.96ml/min. [1]

Characteristics of GIST in MDCT are large (>5cm), well circumscribed, lobulated, free of calcifications and with spontaneous central area of necrotic or haemorrhagic density and have an exophytic growth. [2] These findings correspond to the CTA features in our case.

The main differential diagnoses of benign or small sized malignant GISTs are gastrointestinal schwannomas. On CT imaging, GI schwannomas have homogenous attenuation and can be readily differentiated from large benign or malignant GIST, which demonstrates heterogenous enhancement due to haemorrhage, necrosis and intra-lesional cystic changes. [1]

The mainstay of management for jejunal GIST is a complete surgical excision. The surgical imperative is a complete gross resection with an intact pseudo-capsule and negative microscopic margins. Adjacent organs adherent to the mass should be resected en bloc with the tumour, in order to avoid capsule rupture and intra- abdominal spillage. Lymphadenectomy is usually not required because these tumours do not show lymph – node metastases. [2]

The roles for laparoscopy in the resection of GISTs continue to expand. The same principles of complete macroscopic resection and avoidance of tumour rupture observed durind laparotomy apply to laparoscopy. Generally, Gastric GISTs 5cm in size or less may be removed by a laparoscopic wedge resection. Those GISTs greater than 5cm in size may benefit from a laparoscopic assisted technique with a hand port. Data on laparoscopic resection of GISTs at other sites (including small bowel) are limited. [4]

Favourable prognosis for GIST are tumour size <10 cm, the lesion without metastases, low mitotic index and complete resection of the tumour with negative microscopic margin and with no intraperitoneal dissemination. However, regardless of the mitotic index, the location of the tumour in the small intestine and its size more than 5 cm predict an intermediate malignant potential. [2] Based on published series of GISTs, the 2 most important prognostic features of a primary tumour are its size and mitotic index. [4] The prognostic significance of mutations in the KIT and PDGFRA genes has been examined in GISTs from the pre-imatinib era and tumours with KIT exon 11 mutation are associated with a worse outcome than tumour with other KIT or PDGFRA mutant isoforms or with no detectable mutation. [4]

In Patients who have undergone surgical resection of the GISTs, CT is performed for surveillance of metastatic or recurrent disease. CT abdomen and pelvis should be obtained every 3-6 months. For ver low risk GISTs, less frequent follow up is appropriate. CT is also used to monitor systemic therapy. CT (with or without PET) is recommended within 3 months of initiating thyrosine kinase inhibitor (TKI) therapy in patients with definitively unresectable or metastatic disease. [4]

CONCLUSION

Bleeding jejunal GIST is rare cause of GI bleeding and can be extremely difficult to diagnose due to inaccessibility of endoscopy. CTA has been shown to be an important tool to identify the site of GI bleeding and facilitate the diagnosis of GIST.

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