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Case Report

A Rare Case of Life-Threatening Bleeding Caused by a Jejunal GIST

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ABSTRACT

Gastrointestinal Stromal Tumours (GISTs) are rare mesenchymal tumours that are specific to GI tract. GISTs usually associated with advanced age and have a slight male preponderance. GISTs are commonly found in stomach. Jejunal GISTs are the rarest and account for about 0.1-3% of all GI tumours [1]. The most common clinical manifestation of symptomatic GISTs includes intermittent bleeding due to mucosal ulceration. Massive, life threatening gastrointestinal (GI) bleeding is a rare occurrence. We report a rare case of bleeding Jejunal GIST in a 32-year-old female who presented with haemorrhagic shock that required resuscitative laparotomy. Histopathology and immunohistochemistry confirmed the tumour to have features of GIST with clear margins and post-operative cross-sectional imaging excluded any metastatic deposits.

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Introduction

GISTs are rare neoplasms of GI tract (account for 1% of all GI tumours) and are usually found in advanced age (Median age 67 years). They are mesenchymal tumours specific to GI tract and are commonly found in stomach. However about 20% of them occur in small intestine. Jejunal GISTs are the rarest of all and account for only 0.1-3% of all GI tumours [1]. These tumours can cause intermittent GI bleeding (commonly occult) as it grows in size and result in ulceration of the overlying mucosa. Massive life-threatening GI bleeding, however, is a rare occurrence. We report a rare case of life-threatening GI bleeding caused by a jejunal GIST in a young woman.

Case Report

A 32-year-old woman was admitted to A&E having collapsed at home after massive rectal bleeding of 6 hours. On admission she was paper pale and sweaty. Her extremities were cold and clammy. Pulse was thready and systolic blood pressure was un-recordable. Abdomen was soft and the rectal examination revealed fresh blood and clots. She was resuscitated with un-crossmatched blood and FFP. Her initial Hb was 60 g/l, platelets and clotting were normal. She was too unstable for cross

sectional imaging (In fact CT angiogram was unavailable in out of hours in this setting) and a resuscitative laparotomy was performed. On table gastroscopy excluded any varices or peptic ulcers. At laparotomy a large mass with areas of haemorrhage and necrosis was found protruding the ante-mesenteric border of mid jejunum (Figure 1). The mass was excised with clear macroscopic jejunal margin with intact capsule and side to side jejuno-jejunal anastomosis was created using linear staplers (Barcelona technique). Patient recovered from laparotomy without any complications and was discharged home on post-operative day 6. Macroscopic assessment of the specimen revealed 6.5 x 5.5 x 4 cm size mass with patchy areas of necrosis and haemorrhage attached to the ante-mesenteric border of the jejunum with mucosal ulceration. Both surgical margins and mesentery were free from tumour. Microscopy confirmed to have features of benign cellular spindle cell type GIST with minimal pleomorphism (mitotic figure < 5/50 HPF). At immunohistochemistry the tumour was positive for CD117 antigen.

Discussion

GISTs are rare lesions with an incidence of 2 in 100,000 but are the most common mesenchymal tumours of GI tract [2-4]. GISTs are uncommon in young people and the median age of diagnosis is 67 years [5]. The

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onset of symptoms is influenced by the size of the tumour. Most of them are localised in the stomach (50-70%) and have the best prognosis. GISTs in small intestine (20-30%), oesophagus, colon and retroperitoneal space are infrequent [5]. The most common clinical manifestation of symptomatic GISTs is occult GI bleeding from mucosal ulceration. About 64% of small bowel GISTs present with bleeding, whereas gastric, colonic and rectal GISTs have been associated with <50% incidence of bleeding. Abdominal pain, mass, obstruction and perforation are other manifestations of GISTs [6]. Size of the lesion increases the chances of malignancy. Jejunal GISTs are the rarest GI tumours and are usually asymptomatic. Jejunal GISTs may be diagnosed incidentally during cross sectional imaging, or surgery when they are small. Large lesions can present with GI bleeding (Occult or overt) and non-specific abdominal symptoms like bloating and early satiety. Abdominal pain, mass and obstruction secondary to intussusception are other with large tumours [7]. GISTs are believed to be originated from interstitial cells of Cajal with pacemaker activity. Diagnosis is confirmed after analysing histology and immunohistochemistry. Approximately 95% of tumours are positive for CD117 antigen. Morphologically there are three histological subtypes namely spindle cell (70%), epithelioid (20%) and mixed (10%).

Surgical resection with macroscopically clear margin is the mainstay of treatment for jejunal GISTs. Clear margins are vital in prevention of local recurrence. Risk of local recurrence depends on the tumour size, site and the mitotic figures. According to NIH-Fletcher criteria the index case was recognised as intermediate risk tumour with 24% risk of disease progression [8]. Currently there is no clear consensus on the need for routine lymph node dissection, in fact GISTs rarely involve lymph nodes. Inoperable tumours, malignant GISTs or metastatic disease are best treated with Imatinib mesylate (1st line therapy) and Sunitinib (2nd line therapy). Genetic phenotyping (KIT / PDGFRA) is useful in prognostication and guiding adjuvant treatments [9].



Figure 1: Peri-operative photograph of jejunal GIST with variegated appearance.

Conclusion

GISTs in small intestine can rarely manifest life-threatening GI bleeding. Resuscitative laparotomy indicated in such cases the haemodynamic instability does not warrant angiography and embolization. Resection of the tumour with macroscopically clear margin is the mainstay of treatment to prevent local recurrence. Confirmation of diagnosis, risk

stratification and genetic phenotyping is paramount in planning adjuvant therapy and follow up of these patients. The index case is perhaps the youngest patient with life-threatening jejunal GIST reported in English literature.

Conflicts of Interest

None.

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