

Gastrointestinal Bleeding due to the Small Bowel GIST. Multimodal Assessment of the Patient on Emergency. Case report and review of literature

Arvit Llazani¹, Ardian Nelaj¹, Leart Berdica², Merfa Aliaj¹,
Arvin Dibra³, Xheledin Draçini³, Edmond Çeliku³

¹Service of General Surgery, Faculty of Medicine, University of Medicine, Tirana, University Hospital “Mother Teresa”, Tirana.

²Department of Pathology, Faculty of Medicine, University of Medicine, Tirana.

³Department of Surgery, Faculty of Medicine, University of Medicine, Tirana.

INTRODUCTION

The term Gastrointestinal Stromal Tumour (GIST) was originally used to describe all mesenchymal gastrointestinal tumours, as a collective term used to include tumours of neural and muscle origin. GISTs have an incidence of 15 per million, more commonly occurring in middle-aged and older people with approximately equal sex distribution and having an overall 5-year survival of about 35%. The most commonly affected site is the stomach, followed by the small intestine, colon and rectum. A recent study on the epidemiology of GISTs including 1458 cases, demonstrated that these tumours commonly occur in those over the age of 40 year-old people. No cases were reported in individuals under the age of 20. GIST cells closely resemble the interstitial cells of Cajal (ICC), or gastrointestinal ‘pacemaker cells’. ICCs arise from precursor mesenchymal cells and express the trans-membrane tyrosine kinase receptor, KIT. Gene mutations in these cells result in KIT protein over expression, causing neo-plastic transformation and development of gastrointestinal stromal tumours.

This may be detected by immunohistochemistry, (IHC), using antibody CD117, which allows for accurate diagnosis of GISTs as well as of differentiations from non-GIST tumours of the gastrointestinal tract. Surgical resection of the tumour is the mainstay of treatment for primary disease. Lymph node dissection is not recommended since metastatic spread is rare. The tyrosine kinase inhibitor, Imatinib Mesylate (Glivec®) has also been used successfully in those cases not amenable to curative surgery or for recurrent and metastatic disease. While there is an abundance of literature on the causes and diagnostic management of per rectum bleeding in adults, very little has been documented with regards to young people. We report a case of GIST arising in the ileum, 70 cm from the ileocaecal junction, causing significant gastrointestinal bleeding to a 19-year-old.

Key words- stromal, tumours, rectum, bleeding, metastatic, disease, gastrointestinal, colonoscopy, metastatic, ultrasonography.

CASE REPORT

A 19 year-old woman was admitted by the general surgical team in the ER with a 3-day history of dark-red per rectum bleeding mixed with her stools, weakness, cold sweating, dazing. On physical examination conjunctiva was found to be pale. Blood pressure was measured as 90/50 mm Hg, heart rate 112/min and body measured temperature was within normal limits. On abdominal examination there was neither abdominal tenderness nor palpable mass.

The digital rectal examination showed liquid melena. Complete blood count showed haemoglobin level of 8.2g/dl, hematocrit of 27.5% and RBC of 3.1×10^6 while platelet count and biochemical test were normal. Patient refers that, for a similar clinical situation had suffered nine months before and was treated for erosive gastritis. The patient was hospitalized and reanimated while her vital signs were being monitored.

The following day an oesophagogastroduodenoscopy (OGD), up to the distal duodenum was performed but was thoroughly normal. A colonoscopy revealed blood throughout the colon and distal ileum. An abdominal ultrasonography examination demonstrated a hypoechoic mass (6X5cm) in the pelvic region. The patient underwent computed tomography with double contrast (oral and endovenous) which demonstrated a pelvic mass near the right ovary with regular margins, it also revealed the presence of cystic degeneration

inside, at 6.6X5.3cm in size. No other lesions were detected.

The patient was reanimated using liquid perfusions like 2 unit of blood and 4 unit of plasma before she underwent surgery.

Laparotomy with midline incision was performed. Abdominal exploration revealed a pedunculated mass of the ileum, 70 cm distant from the ileocecal junction. We didn't find other masses nor haemorrhage or other liquids. A primary bowel resection with 5 cm of safe surgical margins from the mass and an entero-entero end-to-end anastomosis was performed. No problems were confronted during postoperative period. The patient was discharged from the hospital with normal vital signs on the 5th postoperative day. Pathological examination confirmed a stromal tumour of the small bowel (H&E stained section of gastric spindle cell GIST; B by imunohistochemistry, the tumour cells are diffusely positive for CD 117 with cytoplasmatic and perinuclear staining. The patient was referred to oncologist to consider successive therapy evaluations.

DISCUSSION

GISTs are a rare cause of gastrointestinal tract bleeding. The most common presentation of GISTs is bleeding (50%), followed by abdominal pain (20–50%) and intestinal obstruction (20%). The most common causes of upper gastrointestinal bleeding in children older than 12 years old are oesophageal varices and gastritis. Lower gastrointestinal bleeding is most

commonly caused by polyps, inflammatory bowel disease, infectious diarrhoea and vascular anomalies. The majority of these disorders would usually be diagnosed by standard OGD and colonoscopy. This report highlights a rare case of GIST causing ongoing bleeding in a teenager, which resulted in diagnostic difficulties. Bleeding is considered to be obscure if a source of blood loss is not apparent from evaluation of the upper gastrointestinal tract and colon. Therefore, for a case of obscure gastrointestinal bleeding (OGIB), one must be able to image the small bowel effectively to establish a cause. Diagnostic limitations of imaging the small bowel create problems in accurate, early diagnosis. Effective use of video capsule endoscopy (VCE) and double balloon enteroscopy (DBE) could be useful diagnostic tools. A prospective study of patients with OGIB demonstrated a diagnostic detection rate of 80% and 60% for VCE and DBE, respectively, and these methods can be employed to detect GISTs presenting with OGIB. Radiological examination, especially careful evaluation of abdominal CT, is very important in making the diagnosis of the disease, although these introduce the associated risks of radiation exposure in the young patient. CT gives information on the localization and size of tumour, extension to liver and peritoneal involvement. It is the most effective imaging modality in the diagnosis of GIST. However, necrotic areas in the centre of tumour may not be differentiated from cystic degeneration and

abscess, which may lead to misdiagnosis. CT appearance of most GISTs is an exophytic lesion >5 cm in size which is well demarcated and has lobulated margins, necrosis, or haemorrhage at the centre but no calcification. CT examination of our case revealed a mass lesion which was 6.6X5.3 cm in size, originating near the right ovary. The main point of the treatment is surgical removal of the tumour, which is best to achieve cure. It is important to preserve pseudocapsule of tumour, while extensive intestinal resection is not necessary. Preservation of capsule is important in that it prevents intra abdominal seeding during surgery. Rupture of tumour negatively affects the prognosis. In our patient, the tumour was removed leaving safe surgical margins and the integrity of pseudocapsule was not disrupted.

Since GIST generally does not spread to lymph nodes, lymphadenectomy is usually not required in surgical treatment of GIST. In our case, no lymphadenopathy was detected preoperatively and lymph node dissection was not performed.

Literature data suggest that tumours smaller than 10 cm are less likely to metastasize, have lower mitotic index and no intraperitoneal spread, and may be treated by R0 surgery. Our case also had tumour less than 10 cm in size, which was excised according to the above criteria.

CONCLUSION

Although GISTs are uncommon, their incidence is probably increasing especially their emergency presentations. The emergency

surgeon must be acquainted with the disease, its emergency presentation and principles of surgery in the presence of GIST tumours. Early diagnosis and treatment would save life of many patients who presented with GIST related emergencies. Surgery is still the gold standard treatment in localized GIST, although the percentage of relapse is not low even after radical surgery. The prognosis is strictly related to size and completeness of surgical resection. We strongly advocate that all patients with a GIST should be carefully and regularly followed-up for an indefinite period. A multidisciplinary team that includes radiologists, medical oncologists, pathologists, and surgeons is paramount for the effective treatment of GISTs.

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