Small bowel gastrointestinal tumour: An interesting case of presentation, diagnosis and treatment

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We report the diagnosis and surgical management of the interesting case of a patient with a gastrointestinal stromal tumour (GIST).

Case study

GISTs are rare, accounting for 0.1–3% of all malignancies of the gastrointestinal tract.[1] They can present with a number of local and systemic symptoms including acute or chronic blood loss.

A previously fit and healthy 35-year-old woman was airlifted to A&E after her husband found her collapsed and bleeding heavily on the bathroom floor. She had complained of flu like symptoms with muscle aches and night sweats for the previous three weeks and sudden onset severe abdominal pain 2 h prior to her haemorrhage. She had no relevant past medical history and denied any weight loss or alteration in bowel habit. In addition, there was no significant family history of bowel disease and she took no regular medications.

On inspection she was very pale. Examination of her abdomen was unremarkable. Digital rectal examination however revealed active bleeding but no palpable mass or further abnormality. Examination per vaginum was also unremarkable. Laboratory investigations revealed a haemoglobin level of 5 g/dL; all other haematological and biochemistry tests were within normal limits. She was immediately commenced on intravenous fluids and transfused with six units of blood. Subsequent upper GI endoscopy was normal and colonoscopy was inconclusive due to the large amount of blood and clots in the bowel.

Following these investigations, a computed tomography (CT) and CT angiogram were performed which showed a 'polypoidal filling defect in the jejunal lumen on the right side with intense enhancement which could signify a polypoidal small bowel adenoma'. At this point, the patient was taken to theatre for an exploratory laparotomy where a tumour was identified in the jejunum and duly excised. Histology subsequently identified the lesion as a GIST. The patient made an uneventful recovery and was discharged without further treatment, following discussion within the multidisciplinary meeting (surgeons, pathologists, oncologists and gastroenterologists).

Discussion

Bleeding from the small bowel is a rare cause of GI blood loss, accounting for only 3–5% of all GI bleeding.[2] Arterio-venous malformations (AVMs) account for 70–80% of small bowel blood loss while cancers, inflammatory bowel disease and infections account for a further 20–25%. GIST, a rare tumour of the gastrointestinal tract, is an unusual cause of GI bleeding accounting for 1–3% of all cases.[1]

GISTs are soft tissue sarcomas arising from the mesenchymal tissues of the GI tract. They are typically defined as tumours with cellular origins in the interstitial cells of Cajal and their pathogenesis stems from KIT proto-oncogene mutation.[3] Histologically, GISTs are usually either spindle cell type (70%) or epithelioid type (20%), the remainder being a mixture of both types.[4] There is a lack of data regarding the incidence of

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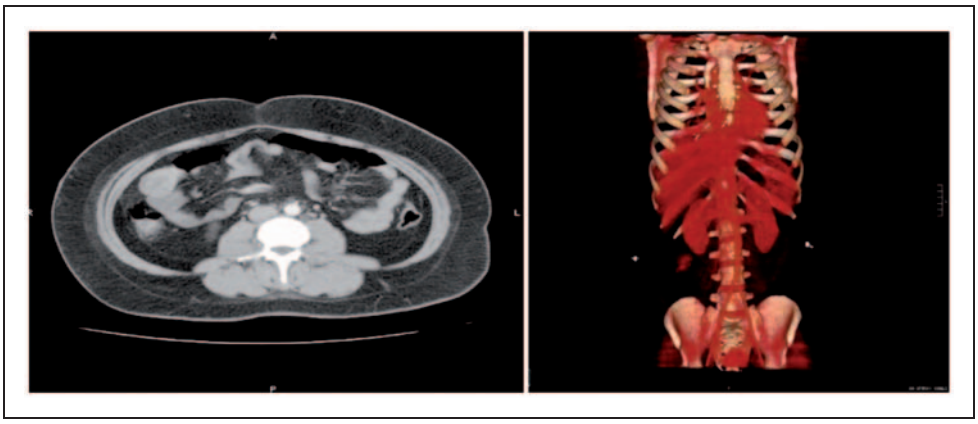

Figure 1. Computed Tomography cross sections showing a mass on the right side of the abdomen. CT Angiogram outlines a vascular mass in the jejunem, consistent with the appearance of a polypoidal tumour.

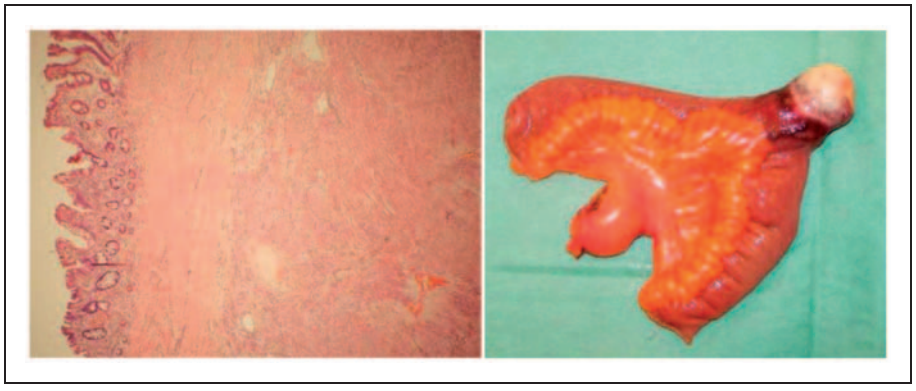

Figure 2. Histological slide confirming Gastrointestinal Stromal Tumour and same tumour resected from the jejunum.

GISTs in the UK; however, data from a recent population-based study in Sweden have proposed an incidence of 13 cases per million persons per year.[5]

GIST is the most common form of sarcoma, constituting more than 70 types of cancer, but in all forms constitutes less than 1% of all cancers.[1] Epidemiological studies performed by Tran *et al*. involving 1458 patients found the mean age at diagnosis was 63 years and a higher incidence rate in males and particularly in black males. Fifty-one per cent of cases were in the stomach, 36% in the small bowel, 7% colonic, 5% rectal and 1% oesophageal. GIST patients usually present with dysphagia, and there is often a history of vague abdominal pain or discomfort. In most cases, the tumour has been found to have grown significantly before the time diagnosis is made.[1]

Mesenteric angiography is a commonly employed investigation for gastrointestinal bleeding if the bleeding sites cannot be identified by endoscopic means. The minimal bleeding rate required for angiographic detection is 0.5 mL/min.[6] When the bleeding rate reaches 1 mL/min, angiography becomes optimally sensitive but vasospasm may occasionally account for a negative study shortly post bleeding.

The role of CT angiography in the detection of GI bleeding has been documented by various authors and has been proven to be effective. Studies to investigate the emerging role of multidetector CT angiography in acute GI bleeding

and to compare to other imaging techniques have found it to be a promising first-line modality for sensitive and accurate diagnosis. Furthermore, it provides a time-efficient method for directing and planning further management and surgery.[7]

The use of CT angiography for the detection of GISTs may therefore prove to be another valuable tool in their detection, especially if endoscopic means are inconclusive or unavailable.

The treatment for primary localized tumours is surgical and surgery remains the only known chance for cure. Resection of a GIST can usually be achieved by wedge resection of the stomach or segmental resection of the small intestine. While complete gross resection is possible in approximately 85% of patients with primary localized tumours, en bloc resection is necessary where the tumour has become densely adhered to adjacent organs.[3]

The discovery of the role of KIT proto-oncogene mutations in GIST pathogenesis and the subsequent potential therefore to specifically inhibit KIT tyrosine kinase is now being exploited by Imatinib Mesylate, a specific KIT tyrosine kinase inhibitor. Bearing in consideration that at least 50% of patients who undergo complete resection of localized GIST develop tumour recurrence, the benefits of adjuvant treatment with Imatinib are currently the subject of further investigation.

The treatment of unresectable, recurrent or metastatic GIST is primarily with Imatinib which is also being used currently as a debulking agent prior to surgery.[3]

Finally, the presence of a well-coordinated multidisciplinary team, consisting of the surgeons who undertake the excision of the tumour as well as other members such as oncologists who monitor and administer the pharmacologic therapy, is vital to control and prevent the recurrence of the disease.

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