

# **Primary Extra-gastrointestinal Stromal Tumour (GIST) Arising from Mesentery of Small Bowel and Presenting as Abdominal Mass**

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## **Abstract**

**Majority of mesenchymal tumours of gastrointestinal tract are gastrointestinal stromal tumour (GIST). It is, however, a rare tumour, accounting for less than 1% of primary gastrointestinal (GI) neoplasms. Gastrointestinal stromal tumours originate from the interstitial cells of Cajal.**

**We report here a case of primary extra-GIST tumour arising from mesentery of small bowel in a 45 years old male patient.**

**The patient presented with abdominal distension and a palpable mass in upper abdomen for past 2 months.**

## **Introduction**

The most common malignant neoplasms of the small bowel in the approximate order of frequency are adenocarcinomas, carcinoid tumours, malignant gastrointestinal stromal tumours (GISTs) and lymphomas.

Malignant GISTs which arise from mesenchymal tissue, constitute about 20% of malignant neoplasms of the small bowel. It is estimated that the frequency of these tumours is 10 - 20/1,000,000 population<sup>1,2</sup> and the possibility of presence of malignancy is 20 - 30%. Malignant GISTs are greater than 5 cm at the time of diagnosis in 80% of patients. GISTs mostly arise from the muscularis propria and generally grow extramurally. Most common indications for surgery include bleeding and obstruction,

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although free perforation may occur as a result of haemorrhagic necrosis in large tumour masses. Typically, GISTs tend to invade locally and spread by direct extension into adjacent tissues and haematogenously to the liver, lungs, and bone; lymphatic metastases are unusual. The most useful indicators of survival and the risk for metastasis include the size of the tumour at presentation, the mitotic index, and evidence of tumour invasion into the lamina propria.

## **Case Report**

A 45 year old male patient presented with history of distension of abdomen without any other significant complaints for last 2 months. When evaluated clinically patient was found haemodynamically stable and his per abdominal examination revealed a mass of size 17 x 10 cm which was mobile, non-tender with bosselated surface and firm in consistency involving epigastric, left hypochondrium and umbilical region.

His laboratory workups were unremarkable and contrast enhanced computed tomography (CECT) of

abdomen depicted a heterogeneous mesenteric mass of size 20 x 17 cm (Figs. 1, 2). On laparotomy 20 x 17



Fig. 1: Shows the GIST



Fig. 2: Shows the extent of GIST

cm mass involving the small bowel and reaching the root of the mesentery near the jejunio-ileal junction was found without any peritoneal deposits, ascites and lymph node enlargement (Figs. 3, 4). Resection of



Fig. 3: Showing the intraoperative size of GIST



Fig. 4: After excision of GIST

the tumour along with the small bowel and ligation of the feeding vessels and double layer jejunio-ileal anastomosis was carried out and specimen was sent for histopathological examination (Fig. 5).

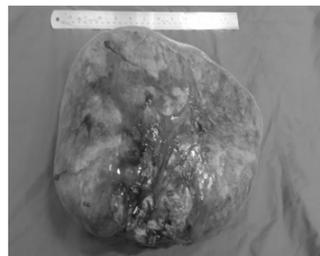


Fig. 5: Excised specimen

Histopathological examination grossly revealed grey white soft tissue mass of size 20 x 17 cm with encapsulated and nodular external surface. Microscopical examination showed features of malignant spindle cell tumours arranged in short fascicles and focally forming whorls with myxoid areas and multiple foci of necrosis. Individual tumour cells revealed moderate nuclear pleomorphism, brisk mitosis (>10/HPF). On the basis of immunohistochemistry, the tumour cells were found to be positive for Vimentin/smooth muscle actin (SMA) and CD-117.

The patient was discharged on 12th of post operative day on Imatinib mesylate adjuvant chemotherapy and is on regular follow up.

### Discussion

GISTs predominantly occur in patients around the sixth decade of life and can be found in any site of gastrointestinal tract with no significant difference in distribution between males and females.<sup>2</sup> The symptoms, which depend on tumour size and location are usually non specific.<sup>3</sup> Small GISTs are usually asymptomatic and are detected either during investigations or surgical procedures for unrelated disease. Incidental discovery accounts for approximately one third of the cases.

The most common presentation is

bleeding related to mucosal erosion (approximately 50%),<sup>3</sup> which may be either chronic, with anaemia, or acute, necessitating emergency treatment (approximately 40% of the cases presenting with haemorrhage). Some 20% of patients present abdominal discomfort, or even pain, which is generally associated with larger size GIST. In the small intestine obstruction is also frequent. Bowel perforation is infrequent. The symptoms and signs are not disease specific and as a consequence about 50% of GISTs have already metastases at the time of diagnosis, usually to the liver or the peritoneum.<sup>1,4,5</sup> In order to carry out detection, staging, surgical planning and follow-up of patients with GIST, CT, MRI and fluorine-18-fluorodeoxyglucose (FDG) positron emission tomography (PET) are considered to be the imaging modality of choice. The majority of GISTs appear to be well-defined, extraluminal or intramural masses with varying attenuation on CT. Small lesions, which are usually benign, tend to be well-defined and relatively homogeneous, While larger lesions normally show well-defined or ill-defined margins, inhomogeneous density both on unenhanced and on contrast enhanced scans and a tendency to spread to surrounding structures. Large tumours (> 6 cm) frequently show central areas of necrosis or haemorrhage.<sup>6</sup>

The first step in the treatment of GISTs is the surgical R0 resection of the primary tumour. Several reviews have reported that small GISTs (< 2 cm) can be treated

adequately by wedge (gastric) or segmental (bowel) resection. Larger GISTs may require more extensive resection including adjacent structures or organs if involved. Surgical resection is associated with a 5-year survival rate of 48 - 70%. There is no indication for chemotherapy and radiation therapy after surgical resection of GISTs as these tumours are notoriously unresponsive to such treatment. Radiotherapy is only used in cases of intraperitoneal haemorrhage, when the precise location of the tumour is known or for analgesic purposes.<sup>1,7</sup> In advanced tumours, a neoadjuvant strategy to reduce the tumour load before surgical resection is under study. The molecular status of GISTs turns out to be relevant for the response to targeted treatment with imatinib a powerful and relatively selective inhibitor of all ABL tyrosine kinases of platelet-derived growth factor receptor (PDGFR) and of c-kit receptor. Imatinib has also become the first line of treatment for recurrent and/or metastatic GISTs and it's very well tolerated (Oral administration of 400 mg daily for some months).

Longer duration of imatinib treatment in patients with advanced GIST increase the risk of resistance to therapy. Several groups describe an increasing frequency of secondary mutations in KIT or PDGFRa. Several other mechanisms as for example, gene amplification or activation of other tyrosine kinase pathways are still under research.<sup>8</sup>

### **Conclusion**

Surgical resection without extensive

lymph node sampling is the treatment of choice in the treatment of GISTs. But even this has resulted in poor outcome and recurrence in patients with malignant potential GISTs or with complications such as perforated tumours, metastatic and unresectable tumours and locoregional recurrence. In advanced cases the gold standard is a targeted therapy with tyrosine kinase inhibitors leading to response rates of up to 80%.

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#### How the salt reduction strategy was derailed

The food we eat is not the biggest cause of death and ill health in the UK, owing to the large amounts of salt, saturated fat, and sugars added by the food industry. Graham MacGregor, Feng He, and Sonia Pombo-Rodrigues discuss the Food Standards Agency's successful salt reduction strategy and how Andrew Lansley and the coalition government's responsibility deal has stalled its progress. They call for urgent action to protect and improve our nation's health.

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#### Minimally invasive surgery for lumbar spinal stenosis

*As good as open laminectomy, but no better*

Their study compares the "old" standard surgical treatment for spinal stenosis (open laminectomy) with a newer and less invasive alternative (microdecompression). Microdecompression is a procedure to decompress the lumbar nerves by removing a minimal amount of bone and the ligamentum flavum but leaving the spinous process and the supraspinous and interspinous ligaments intact.

In this case, however, we still have no evidence that minimally invasive surgery works any better in the long term for patients than more traditional open decompression techniques.

**Wouter A Moojen, Wilco C Peul, BMJ, 2015, Vol 350, 9**