

Case Report

Case report: successful treatment of a rare large jejunal gastrointestinal stromal tumour with surgery and using principle of short-term fasting

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ABSTRACT

Small intestinal gastrointestinal stromal tumours (GISTs) are rare mesenchymal tumours of gastrointestinal tract comprise 0.04% of small intestinal neoplasms. We presented a case of 61 years female presenting in emergency with pain in epigastrium, multiple episodes of vomiting, raised total leucocyte counts. CECT abdomen showed a large non-enhancing jejunal lesion. The patient had emergency small bowel resection and anastomosis. Postoperative patient started on imatinib and kept on short-term fasting. This case report highlights the rare presentation of jejunal GISTs, successful surgical outcome and good enhancing effect of short-term fasting on chemotherapy.

Keywords: Jejunal GISTs, Short-term fasting, Indian subcontinent

INTRODUCTION

GISTs (gastrointestinal stromal tumours) are rare mesenchymal tumours of the gastrointestinal system initially identified by Mazur and Clark in 1983.¹ Main causative factor includes gain in function mutations in proto-oncogenes i.e.; c-KIT or PDGFRA. These mutations enhance the activation of tyrosine kinase receptors, leading to unregulated proliferation of stem cells that develop into Cajal's intestinal cells.²

The great majority of GISTs are sporadic and solitary, however they can be seen in a variety of neoplastic diseases. While small, jejunal GISTs are usually asymptomatic and can be detected by CT, endoscopy, surgery, or symptomatic liver metastases. GI bleeding or non-specific GI symptoms like bloating or early satiety are common signs of enlargement.

Short-term fasting (STF) protects against toxicity while improving the efficacy of a number of chemotherapeutic drugs in the treatment of diverse tumour types, according to growing preclinical evidence.

CASE REPORT

61 years female presented in emergency with complain of pain in epigastrium for 20 days. Pain was dull aching, continuous, non-radiating, increased in intensity after food intake and relieved after oral analgesics. She also had multiple episodes of non-bilious 50-60 ml vomiting, around half an hour after food intake containing undigested food material for 4 days. There was no history of melena, haematemesis, loss of weight, loss of appetite.

Her abdominal examination revealed mild abdominal distension with hyperdynamic bowel sounds. There was a palpable vague lump around umbilicus. An abdominal X-ray showed multiple air fluid level which were suggestive of small bowel obstruction.

CECT abdomen (Figure 1) showed well defined exophytic soft tissue non-enhancing lesion measuring 82×60 mm just adjacent to jejunal loops. There was mild circumferential bowel wall thickening of jejunal loops and adjacent fat stranding suggestive of small bowel GIST. Her TLC counts were raised (14,000), albumin (4.2).

Midline laparotomy was done, around 200 ml pus was drained from left paracolic gutter. 8×8 cm hard mass (Figure 2) was present 60 cm proximal to ICJ, tumour was removed with 5 cm distal and proximal margin. End to end jejunojejunal anastomosis was done. Resected specimen shows feature of GIST, positive for CD117 and VIMENTIN. Focal expression of desmin also observed. The cells didn't show expression for SMA, S-100, HMB-45, CD60.

Post-operative period was uneventful. We kept her on intermittent short-term fasting in postoperative period with adequate protein and calorie supply as per principle of metabolic autophagy.^{3,4} She has been advised for follow-up colonoscopy after 5 years. We have started her on post-operative imatinib therapy for 2 years, as per the NICE guidelines.

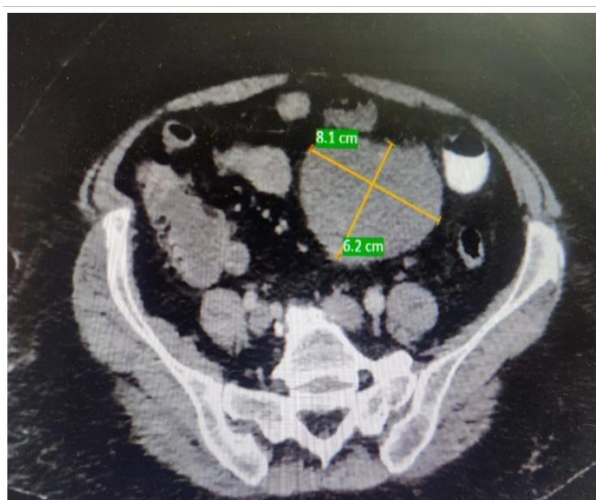


Figure 1: CECT abdomen showing large jejunal non-enhancing heterogenous lesion.



Figure 2: Resected jejunal segment with tumor.

DISCUSSION

GISTs are most prevalent in the stomach (60-70%), small intestine (25-35%), and rectum, oesophagus, omentum, and mesentery (less than 5%). The duodenum makes up 17.7% of the small intestine, whereas the jejunum makes up 47.6% and the ileum makes up 34.7 percent.⁵ Only 10-30% of cases evolve to malignancies. The clinical presentation is affected by the tumor's size. Small GISTs (less than 2 cm) are asymptomatic and can be discovered by accident during a laparotomy or endoscopy. Larger tumours might cause nonspecific abdominal pain, acute or chronic GI bleeding, intestinal blockage, or changes in bowel habits. Exophytic palpable masses with very big GISTs that are likely to be cancerous.⁵ In our case epigastric pain and vomiting was only symptoms. For diagnosis radiology investigations includes barium studies which identify around 80% of GISTs, capsule endoscopy 81.1%, CT scans 87% and MRI scans close to 100%.⁶ GISTs are diagnosed based on morphologic features supported by Immunohistochemical (IHC) studies. Histologically, GISTs can occur in three different types: spindle cell, which accounts for nearly 70% of tumors, epithelioid, and mixed.⁷ Rarely, GISTs may have myxoid stroma, neuroendocrine features or signet ring variant. Grossly GISTs are usually unencapsulated but well-circumscribed lumps on the surface. The sliced surface has a whorled fibroid-like look which is fleshier and varied.⁷ Cystic degeneration or central necrosis can be seen in large lesions. The underlying mucosa is often ulcerated.

Complete Surgical resection remains the mainstay of treatment for small intestinal GIST. An R0 resection should be the goal of surgery. Because lymph node metastases is uncommon, no lymph node dissection is necessary. Recurrence rates are roughly 40%, and the majority of patients had liver metastasis, with just one-third of patients having isolated local recurrence. Long-term disease-free survival is estimated to be approximately 50%. Tumors larger than 10 cm and tumours with more than 5 mitoses per 50 HPFs are risk factors for malignancy and recurrence. The mitotic index of most benign tumours is modest (5 mitoses/50 HPFs). For metastatic or inoperable tumours, imatinib mesylate is the mainstay treatment.⁸ GISTs in the small intestine are more dangerous than those in the stomach, with 40 percent to 50 percent of small bowel GISTs demonstrating malignant activity compared to 20 percent to 25 percent of gastric GISTs. The National Cancer Institute advises adjuvant imatinib for GISTs larger than 10 mm. Imatinib reduces recurrence rates by 14% in absolute terms, resulting in a 97 percent recurrence-free survival rate.⁹

GISTs with a low risk of recurrence may not require routine follow-up. Follow-up for low-risk tumours can be done every 6-12 months for the next 5 years using abdominal CT scanning or MRI. High-risk individuals should have an abdominal CT scan or MRI every 3-6 months for 3 years during adjuvant therapy, then every 3 months for 2 years after discontinuing adjuvant therapy,

then every 6 months until 5 years after ceasing adjuvant therapy, then annually for a further 5 years.¹⁰

Short-term fasting (STF) protects against toxicity while improving the efficacy of a number of chemotherapeutic drugs in the treatment of diverse tumour types, according to growing preclinical evidence.⁴ Nutrient shortage causes healthy cells to shut down growth-promoting pathways in order to re-invest energy in maintenance and repair pathways. Greater cellular defence develops, contributing to increased resistance to various stresses such as chemotherapy and radiation. Tumor cells, on the other hand, are unable to activate this protective response due to: (1) uncontrolled activation of growth pathways and self-sufficiency in growth signals caused by oncogenic mutations or autocrine growth factor production, and (2) loss of anti-proliferative signals caused by tumour suppressor gene mutations. As a result of gaining the capacity to grow faster, tumour cells lose their ability to adapt to tough circumstances, such as nutrition restriction. Furthermore, tumour cells' constant increasing development rate necessitates an abundance of resources. As a result, STF raises tumour cell DSS in response to a variety of chemotherapeutic drugs, radiation, and Tyrosine kinase inhibitors (TKIs).⁴

CONCLUSION

This case was noteworthy for a number of reasons. First, large jejunal GISTs are extremely rare. Second, uncommon presentation of patient. Finally, use of short-term fasting during chemotherapy and the patient had made a remarkable recovery without reoccurrence till date.

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