

## Case Report

# Gastrointestinal stromal tumor at an unusual site with an unusual presentation

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## ABSTRACT

Gastrointestinal tumors (GIST) are malignant and rare forms of soft tissue sarcoma arising from interstitial cells of Cajal. With most common site of origin being stomach, Jejunal GIST accounts for 0.1–3% of all GIST. Due to non specific signs and symptoms diagnosis is often delayed and patient mostly presents in advanced stage. 50 year old female presented with decreased appetite from 5 months and lump in upper abdomen from 3 months associated with dull abdominal pain from 15 days. CECT abdomen showed a large soft tissue mass in left lumbar region. Exploratory laparotomy showed a highly vascular mass of size 12x10x8 cm abutting mesentery of transverse colon lodged between middle colic artery and superior mesenteric artery. Resection of mass with a segment of jejunum was done. Histopathological examination showed spindle cell tumor with high mitotic index with CD 117+. Adjuvant chemotherapy was given and patient is currently asymptomatic after 9 months of postoperative follow up. Spectrum of clinical presentation is broad so a preoperative diagnosis is difficult. GIST primarily presenting as an abdominal lump is rare and only 25 cases have been reported so far. Surgery is the primary mode of treatment but imatinib has transformed the treatment. Newer drugs are under phase II trials and hopefully will change the course of management in near future.

**Keywords:** Abdominal lump, GIST, Jejunal gastrointestinal tumours

## INTRODUCTION

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract.<sup>1</sup> These are rare forms of soft tissue sarcoma of the digestive tract. Interstitial cell of Cajal are presumed to be the origin.<sup>2</sup> 3000-4000 cases are reported annually in United States.<sup>3</sup> The most common site of origin of GISTs is stomach (60%-70%) followed by small intestine (25%-35%). Rectum and colon (5%-10%), mesentery (7%) oesophagus (5%) and duodenum (4%) are the other rare sites of origin.<sup>4</sup> Worldwide the incidence of GIST is reported to be 2 per 1 lakh population whereas GIST

located on jejunum are extremely rare accounting for 0.1-3% of all gastrointestinal tumors.<sup>5</sup>

Often GIST presents with very indolent symptoms and mostly presents in advanced stages and sometimes at unresectable stages due to which diagnosis is delayed. Commonly presenting with pain abdomen and GI bleeding but lump abdomen can be a presentation when initial presentation is very late. 30% of GIST is diagnosed incidentally during exploratory laparotomy or radiological studies. Worldwide GIST primarily presenting as a mass per abdomen is extremely rare with only 25 cases being reported till 2011. In this case report we present another case of GIST arising primarily from

jejunum presenting initially as a mass in abdomen managed at a teaching hospital in Uttarakhand, India

## CASE REPORT

A 50 year old female presented with complaints of decreased appetite from 5 months associated with incidentally noticed lump in left upper abdomen from 3 months and dull aching intermittent pain from 15 days. There was no history of dyspepsia, vomiting, fever, weight loss. General physical examination was normal. Per abdomen examination revealed a mobile, non tender firm mass of size 10×9cm with well defined margins in left hypochondrium and epigastrium region. Complete hemogram and routine biochemical parameters were normal. Computed tomography showed a large soft tissue mass with heterogeneous enhancement with small areas of necrosis seen in left lumbar region and periumbilical region (Figure 1 and 2).

Exploratory Laparotomy was done which showed a 10×10×9cm highly vascular mass adhered to antimesenteric border of proximal jejunum abutting mesentery of transverse colon lodged between middle colic artery and superior mesenteric artery but not involving them (Figure 3). Mass was also adhered to anterior wall of pancreas. No evidence of lymph nodes and peritoneal metastasis were noted. So resection of mass with segment of jejunum was done and primary closure of duodenal stump was done with end to side jejunoduodenal anastomosis.



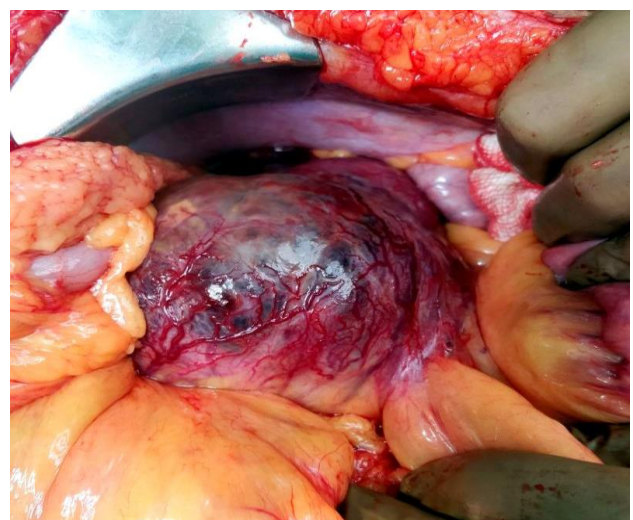
**Figure 1: Coronal image of CECT abdomen showing large soft tissue mass in left upper abdomen.**

Macroscopically well circumscribed encapsulated growth with congested blood vessels and areas of haemorrhages on its outer wall is identified at the proximal end of

jejunum measuring 140×100×80mm. Cut surface shows a variegated appearance and growth seems to be arising from submucosae (Figure 4). Microscopically sections shows well circumscribed spindle cell tumor with whorls and intersecting fascicles with mitotic index of 8-10/10 HPF with necrotic patches with minimal nuclear polymorphism and absence of mucosal infiltration (Figure 5 and 6). Two tiny reactive lymph nodes were also seen. Cells were found to be CD-117, SMA and Vimentin positive and Ki-67 was less than 5%. The above features both histopathological and immunochemistry proved it to be a high grade GIST (Size more than 10 cm and high mitotic index). The postoperative course of the patient is uneventful. Imatinib mesylate at a dose of 400 mg once a day was started as adjuvant chemotherapy as soon as the diagnosis was confirmed, and patient is currently asymptomatic without any evidence of recurrence after 9 months of postoperative follow-up.

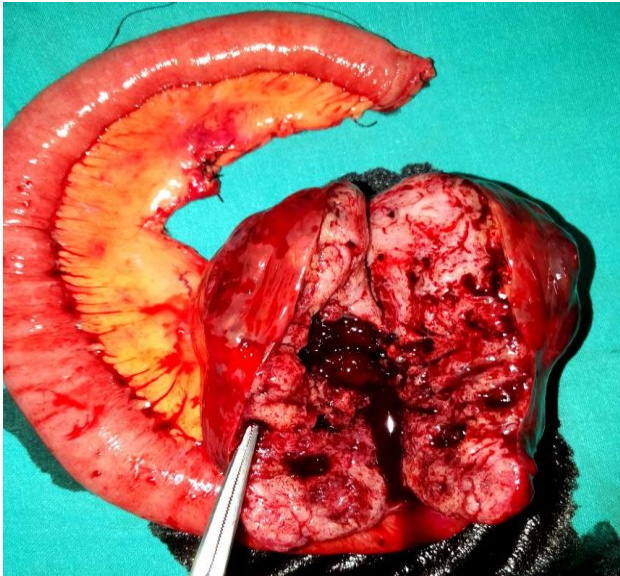


**Figure 2: Transverse section image of CECT abdomen showing large soft tissue mass in left upper abdomen with vascular enhancement.**

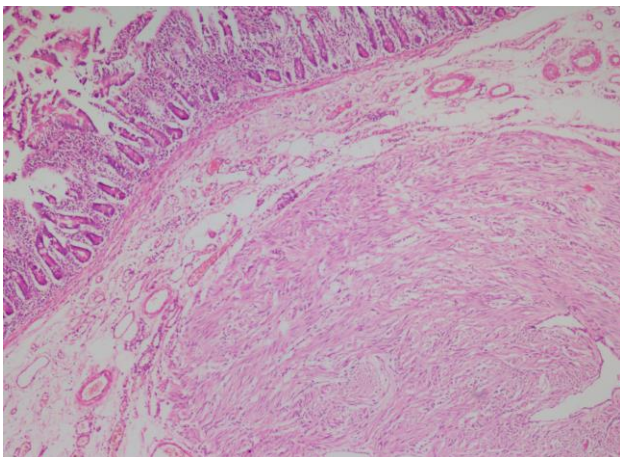


**Figure 3: Encapsulated growth with congested vessels and areas of hemorrhage on its outer wall.**

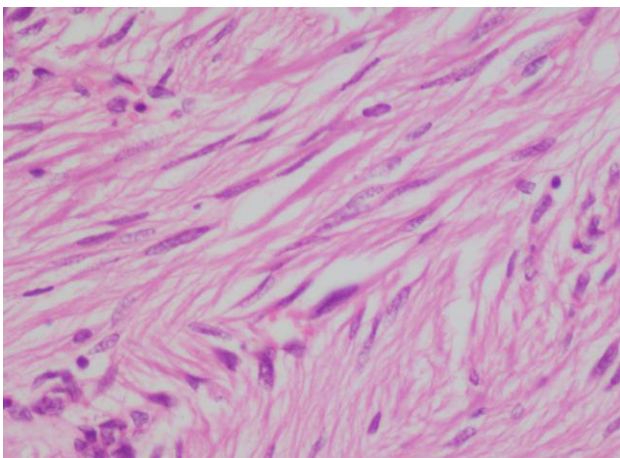




**Figure 4: Resected specimen showing resected part of bowel with soft tissue mass. Cut surface showing variegated appearance with hemorrhagic material.**



**Figure 5: Circumscribed spindle cell tumor with whorls and intersecting fascicles (4x).**



**Figure 6: Spindle cells with high mitotic index in sheets (40x).**

## DISCUSSION

GIST can occur at any age although they are more common in adults with a peak incidence in the fifth and sixth decade of life with slight male predominance.<sup>1</sup> Interstitial cells of Cajal are thought to be the cell of origin. Loss of heterozygosity of the NF1 gene and mutation in the c-kit leading to increased expression of KIT and PDGFR- $\alpha$  were thought to be pivotal. GISTs on the basis of histopathology characteristics are classified into three types spindle cell (70%), epithelioid (20%) and mixed (10%). 95% are CD117 positive on immunochemical examination. Abdominal pain and GI bleeding are most common presenting features. Given the non specific signs and the symptoms preoperative diagnosis is difficult. 20-30% of GIST is diagnosed incidentally or at autopsy. Contrast-enhanced CT scan is the imaging modality of choice.<sup>6</sup> Gastrointestinal schwannomas is the most common entity to be differentiated from a small size GIST and it is essential to differentiate as GI schwannomas are benign with a good prognosis.<sup>7</sup> Preoperative biopsy carries a risk of haemorrhage hence it is avoided but if biopsy is necessary fine needle aspiration cytology will generally confirm the diagnosis.<sup>8</sup> National Institute of Health developed the first system for risk stratification, the so-called Fletcher's criteria based on Tumor size, location and mitotic index.<sup>9</sup> Later "Armed Forces Institute of Pathology risk stratification System" (AFIP) was given which has advantage of calculating risk of relapse and progression. International union against cancer (UICC) for the first time in its seventh edition published on 2010 included a classification and staging system using the TNM system for uniform and standardized analysis but its efficacy needs to be further validated.

Surgery is the primary treatment of choice and till 2001 it was the only effective treatment modality available with overall 5-year survival rates of 45-55%. In 2001 Imatinib has revolutionized the treatment of high grade GIST and was recognized to be highly effective in metastasized GIST and currently it is only drug used for the treatment of GIST.<sup>10</sup> Imatinib is usually used in a starting dose of 400 mg per day but recent trials done on high dose imatinib (800 mg) showed slightly improved survival and less progression but leads to higher rate of side effects. Sunitinib, an inhibitor of KIT, PDGFRs is now approved as a second-line treatment for advanced GISTs.<sup>4</sup> Nilotinib, sorafenib, dovitinib, crenolanib, pazopanib, and dasatinib are newer monoclonal antibodies currently in phase II trials and in future these drugs might be first line drugs for treatment of GIST.

## CONCLUSION

GIST is a disease of growing concern. Having very less treatment options the primary mode of treatment is still surgery despite of high incidence of reoccurrence. Due to improvement of surgical techniques and cancer surgery guidelines it has been found that there has been decrease

in incidence of reoccurrence. Recent trials and studies have shown an increase in reoccurrence free survival after using low dose imatinib for 1 year. But there has been surge in cases with resistance with currently available drugs. Newer drugs currently under clinical trials will change the current management in the near future and will hopefully improve survival of the patients with GIST.

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