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Small Intestine Gastrointestinal Stromal Tumour—A Case Report

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Abstract

Gastrointestinal stromal tumours (GIST) are mesenchymal tumours that arise most commonly from the stomach. They are the 3rd most common tumour diagnosed behind adenocarcinomas and lymphomas. The majority of these tumours are asymptomatic and incidentally diagnosed in UGI scopy or contrast enhanced CT abdomen and pelvis studies. Obstruction, ulceration, gastrointestinal (GI) haemorrhage and perforation warrants an urgent surgical intervention. GIST is medically managed by tyrosine kinase inhibitors and surgically by resection and anastomosis. This case report highlights the diagnosis and management of a 42-year-old gentleman who presented with vague right iliac fossa mass. Informed consent was obtained from the patient for publication of this case. This case was chosen to be reported because of the rare incidence of small intestine GIST in South India and the effectiveness of minimally invasive laparoscopic surgery in management.

Keywords

Small, Intestine, Gastrointestinal, Stromal, Tumour, GIST, Ileum, Case, Report

1. Introduction

GISTs are classified under stromal tumours. In India, it affects middle aged 40-year-old men more commonly than women [1] [2]. Kindblom *et al.* in 1998 concluded that the pluripotent mesenchymal cells finally differentiate into the interstitial cells of Cajal, which is the origin of GIST. These cells are found in the muscularis propria that initiates and coordinates GI motility. They are the "pacemaker cells" of the gastrointestinal tract [3]. The differentiation from stem cell to interstitial cells of Cajal is regulated by KIT kinase. The association between

mutations in c-KIT proto-oncogene (85% - 95%) and GIST was described by Hirota and colleagues [4].

2. Case Presentation

A 42-year-old gentleman presented with complaints of vague right sided lower abdominal pain on and off for the past 6 months. He also complains of non-specific dyspepsia and abdominal distension. He has had multiple visits to the hospital and numerous consultations but to no avail. His general examination was unremarkable and per abdomen examination revealed tenderness in the right iliac fossa on deep palpation. Ultrasound of the abdomen and pelvis revealed heterogenous, peripheral enhancing lesion of 6.2 × 5.5 cm in size that seems to be arising from the small bowel. With the suspicion of GIST, a contrast enhanced CT scan of the abdomen and pelvis (**Figure 1**) revealed a soft tissue mass lesion arising from the mid ileal loops that showed peripheral enhancement and central non enhancement post contrast administration.

Patient was pre-operatively fit for surgery. Laparoscopic removal of the GIST with resection and anastomosis of the small intestine was planned. **Figure 2** shows a 6 × 6 cm cystic lesion was adherent to the mesentry and was exophytic in nature. **Figure 3** demonstrates the use of a Covidien 60 mm stapler to resect the specimen after adequate small bowel clearance (5 cm). Ileal anastomosis done using a Covidien 60 mm stapler (**Figure 4**). The specimen was retrieved though an upper midline laparotomy incision and sent for histopathological examination.

Gross histopathological examination revealed the tumour size of 6 × 5.5 × 5 cm with negative margins (**Figure 5**). Microscopic studies revealed spindle cell type of GIST with 0 - 1 mitoses per 50 high power field. The pathological classification (TNM) of the tumour was T_3N_x.

Post operatively, he was started on clear liquids on POD 3, normal diet on

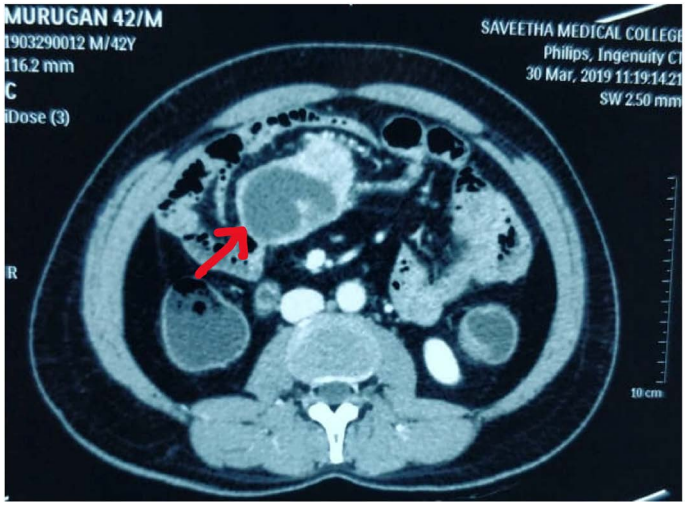

Figure 1. Shows the site of the small intestine GIST on contrast enhanced CT of the abdomen and pelvis (Red arrow).

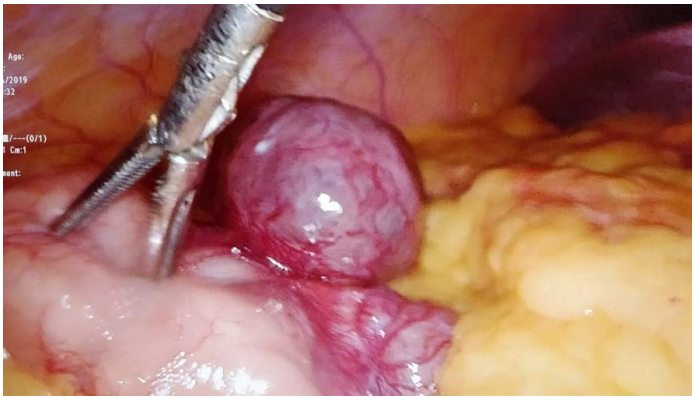

Figure 2. Shows the exophytic small intestine GIST (Intra-operative photo).

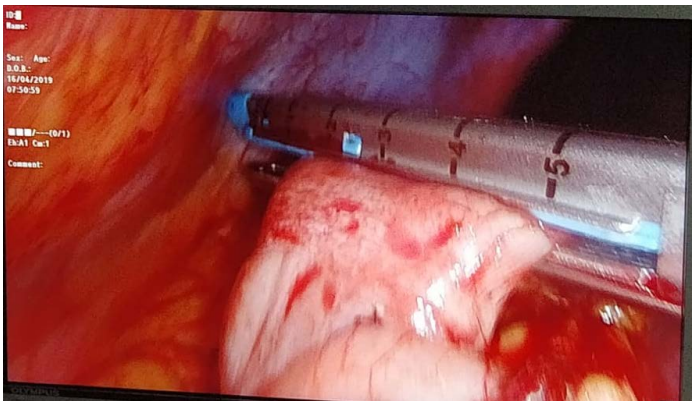

Figure 3. Shows the resection of the small intestine with a Covidien 60 mm stapler.

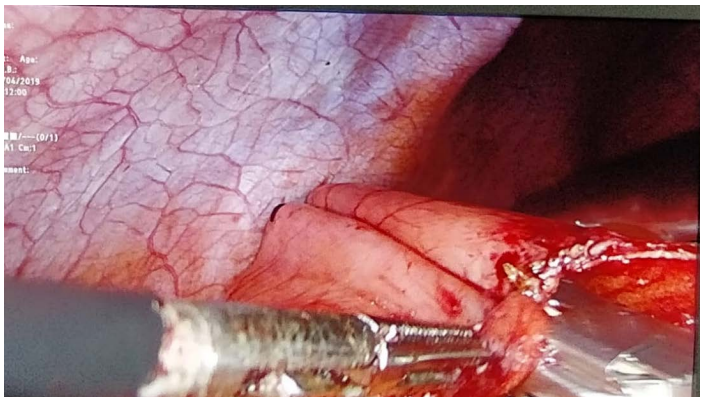

Figure 4. Shows the side to side anastomosis of small intestine with a Covidien 60 mm stapler.

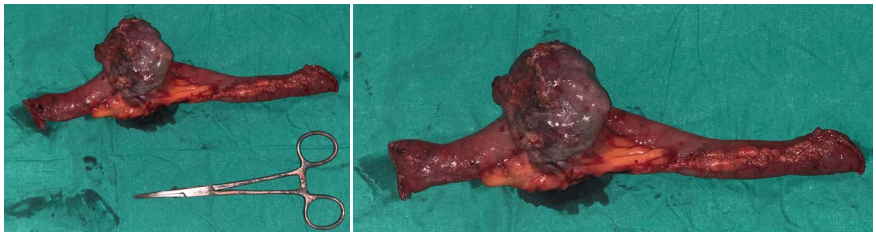

Figure 5. Shows the gross resected specimen.

POD 5 and was referred to Medical Oncology for further management. The medical oncology team advised Tab. Imatinib 600 mg for 6 months. Patient had serial contrast enhanced CT scans to monitor his response to the tyrosine kinase inhibitor. He is on regular follow-up and doing well. His last visit to the surgical out patients department was uneventful, his midline and port site scars healed and is leading his regular lifestyle.

3. Discussion

GISTs are either single or multiple with their size ranging from >1 cm to a maximum of 40cm in diameter [5]. They are commonly found in the body of the stomach (70%) [6]. The jejunoileal site is the second most common site followed by rare presentations in the esophagus, colon, rectum and extra-GI sites [7] [8] [9]. Extraluminal GISTs are asymptomatic and can grow for an extended period and present as a large abdominal mass. Intraluminal GISTs cause complications like obstruction, mucosal ulceration, GI hemorrhage, perforation and peritonitis. If left untreated, they metastasise to the liver. Lymph node metastasis is rare.

At the genetic level, mutations involving c-KIT and PDGFRA are known to be involved in the pathogenesis of GIST. Patients with an exon 11 mutation results in a gain of function KIT mutation [10]. KIT is involved in the phosphorylation of several downstream proteins that results in mitogenic activity and protein transcription. Mutations in KIT sends proliferative signals to the nucleus that evades apoptosis and hence leading to tumorigenesis [11]. PDGRFA mutations are less common and are associated with gastric and epithelioid GISTS [12]. Exon 18 is most commonly involved [13].

GISTs are asymptomatic and are usually incidental findings during endoscopic, surgical procedures or radiological investigations [14]. A previously healthy individual with an abdominal mass gradually increasing in size can present with abdominal discomfort, anorexia, nausea, vomiting and weight loss [15]. An intraluminal growth can cause site specific obstruction (for example, gastric GIST presenting as gastric outlet obstruction) whereas an extraluminal growth can cause luminal compression due to the external pressure effect exerted by the enlarging mass. A palpable mass can be felt in the abdominal cavity and is detected in exophytic GISTs.

The most common complication is upper GI haemorrhage that presents as hematemesis or melena. The mass causes pressure necrosis and ulceration of the mucosal surface which results in haemorrhage due to the disrupted blood vessels. Perforation is associated with signs of peritonitis and shock [16] [17]. Kim MS *et al.* reported a rare case of intussusception due to a GIST [18].

While routine blood investigations and organ specific tumour markers are non specific in diagnosing GISTs, imaging and endoscopic studies are more specific. Plain chest and abdomen radiographs help in identifying those patients with an obstruction or perforation but are nonspecific to GISTs. Dualim DM *et al.* highlighted the use of a capsule endoscopy and double balloon enteroscopy in

the diagnosis of a bleeding jejunal GIST [19]. Barium studies and enteroclysis provides information regarding the presence or absence of a mass but is not specific for GISTs. A filling defect that is sharply demarcated and elevated with the surrounding mucosa can be appreciated in barium studies [20].

Contrast enhanced computed tomographic (CT) abdomen and pelvis provides information regarding the size and location of the tumour and the surrounding structures [21]. Distant metastasis and infiltration of adjacent structures can also be identified on CT. Tumours between 5 - 10 cm present as irregular, heterogenous extraluminal or intraluminal masses that shows signs of biological aggression [22].

These tumours appear as spindle shaped cells with an increased cellularity. Factors like mitotic index, cellularity, nuclear-cytoplasmic ratio, amount of stroma and vascularity are taken into account for evaluating the prognosis of the disease.

GIST expresses CD117, which helps in differentiating GIST from other GI mesenchymal tumours [23] [24]. CD34 is expressed in 70% of GISTs and indicate the probability of a lesion being malignant or not. Presence of CD44 indicates a better prognosis.

A multidisciplinary team, in a tertiary care centre, including a surgeon, radiologist, pathologist and medical oncologist is necessary for complete treatment of the disease. Complete resection of the tumour with negative margins is the surgical treatment of choice [21]. Small intestine GISTs are treated by a segmental resection and anastomosis. Lymphadenectomy is not routinely performed because of the lack of involvement. Unresectable tumours are treated with imatinib. Recently, laparoscopic approaches to deal with GISTs less than 5cm in size have been successful. Piessen *et al.* reported that laparoscopic treatment of gastric GISTs was associated with significantly lower surgical and medical morbidity, and significantly better 5-year recurrence-free survival [25]. Faster recovery, shorter hospital stay and decreased analgesia are the advantages of laparoscopic surgery over open surgery. Tao *et al.* compared laparoscopic versus open surgery in the treatment of GIST and observed that operation time, time to first flatus and pre-operative hospital stay was shorter in the laparoscopic group. He concluded that laparoscopic intervention in experienced medical centers is best preferred for GISTs in unfavourable locations [26].

Newer minimally invasive surgical techniques that involve transrectal extraction of the resected GIST specimen have been highlighted by Wang X *et al.* After anal dilation, an incision was made in the upper rectum. A protective bag was inserted intra-abdominally into which the resected specimen was placed. The bag was pulled out making sure there is no capsular breach. The rectal stump was closed with a stapler. This method prevents an unwanted upper abdominal incision for retrieving the specimen [27].

Imatinib, a tyrosine kinase inhibitor, plays a major role in curing the disease post operatively. It can be administered pre-operatively to reduce the size of a

large tumour and to achieve negative margins. A dose of 400 mg daily, with the maximum dose of 800 mg/day, is given for up to 2 years post-operatively [28] [29]. The response is monitored with the help of serial CT scans which reveals a decreased tumour density and tumour shrinkage [21].

4. Conclusion

Clinically impalpable lesions presenting with vague abdominal complaints must alert a surgeon to consider the possibility of GIST. Early identification, surgical intervention and post operative tyrosine kinase inhibitor therapy are the treatment of choice. Minimally invasive surgical modalities should be implemented in elective and emergency cases of GIST in tertiary care medical centres.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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