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Case Report

Gastro-intestinal stromal tumours of stomach and small intestine: Report of two cases

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Gastrointestinal stromal tumors (GIST) are rare gastrointestinal tract (GIT) tumors that originate from the interstitial cells of Cajal. Melena or hematemesis and anemia are the most common presentations and CT scan is the investigation of choice in demonstrating the endoluminal and exophytic extent of tumor. Surgery remains the mainstay of treatment in GIST and chemoradiation has not proved to be very effective. We have encountered two such cases of GISTs in our clinical practice. 62-years and 32-years old males presented with epigastric and paraumblical pain respectively. Clinical examination revealed a well defined epigastric mass of approximate size 12*10 cms in former and a vague paraumblical mass in the later. Contrast Enhanced CT Scan of abdomen and pelvis suggested the origin of masses in each case and exploratory laparotomies performed in both of them revealed a mass in relation to the stomach and ileum respectively. Histopathology and immunohistochemistry confirmed the diagnosis. Histopathology and immunohistochemistry are very useful aids in making a diagnosis of GIST and surgery remains the choice of treatment even in cases of larger GISTs.

Keywords: Gastro-intestinal stromal tumours, stomach, small intestine, interstitial cells of Cajal, c-kit proto-oncogene.

INTRODUCTION

Gastro-intestinal stromal tumours (GIST) comprise less than 1% of all gastro-intestinal tract (GIT) tumors but the most common mesenchymal tumours of the GIT. Usually it affects middle -aged people with equal incidence in both males and females Deshpande and Munshi, (2007). These are mostly solitary, well circumscribed tumors with a pseudocapsule and arise from embryological mesoderm of the GI tract. They were considered as smooth muscle tumors however their resistance to chemotherapy and morphological and immunohistochemical features dissimilar to smooth muscle puts them into a distinct category of mesenchymal tumors of GIT. The commonest presentation is Upper GI bleeding however, GIST as a cause of upper GI bleeding is rare. They can also present with abdominal pain, dyspepsia and vomiting or incidental findings during endoscopy, imaging or surgery

We present two such cases of GISTs in our clinical practice, one of them presenting as an upper abdominal mass which was related to stomach and another with paraumblical mass related to the small intestine.

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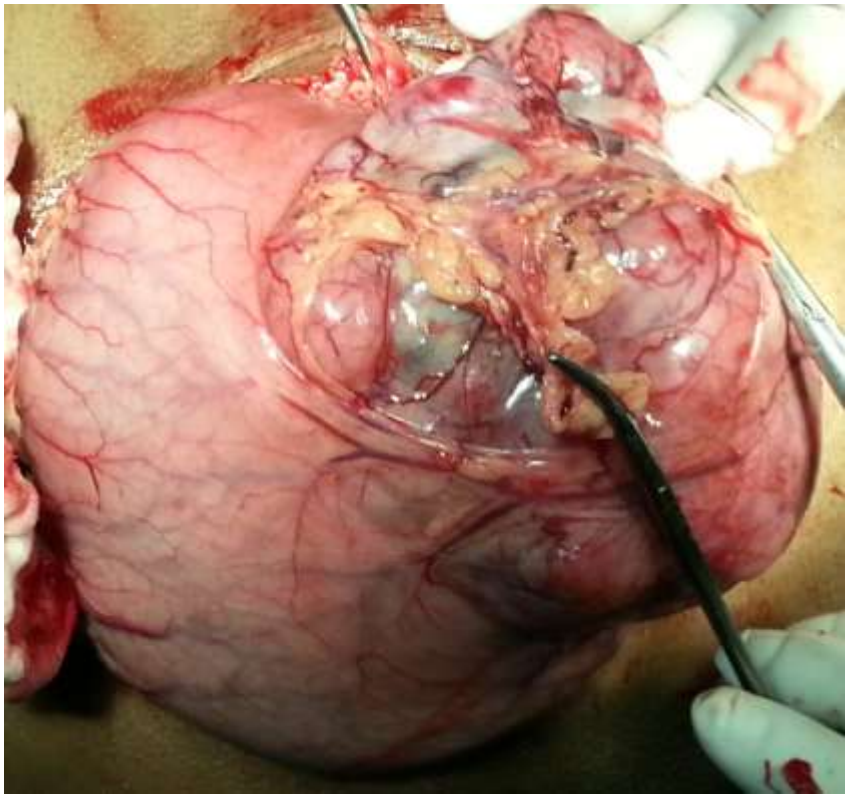
Case 1

62- years old male presented with history of epigastric mass which was progressively increasing in size for the last two and half months. It was not associated with fever, vomiting, haematemesis or malena. On examination patient was anaemic. Abdominal examination showed well-defined mass of approximately 12*10 cms was present in the epigastrium, extending to right hypochondrium. His routine blood investigations were within normal limits except for low hemoglobin level.

Ultrasound examination of abdomen revealed assymetrical solid mass lesion measuring 13 cms present in the posterior wall of the stomach. Contrast Enhanced CT Scan of abdomen and pelvis was done which revealed a large well defined lobulated exophytic mass of size 13.3 cms x13.8 cms x7.8 cms with areas of cavitation and air present in relation to lesser curvature of stomach bulging into gastro-hepatic recess.

Figure1. Intraoperative picture showing Large Gastro-intestinal stromal tumour arising from the lesser curvature of stomach.



Exploratory laparotomy was done through upper midline incision. A large mass measuring approximately 15 cms x 15cms was arising from the lesser curvature of stomach which was excised with a tumour free margin of 5-6 cms on both the sides. The mass was present approximately 8-10 cms from the gastroesophageal junction and the stomach was closed at the lesser curvature. A temporary feeding jejunostomy was done. [Figure1, 2].

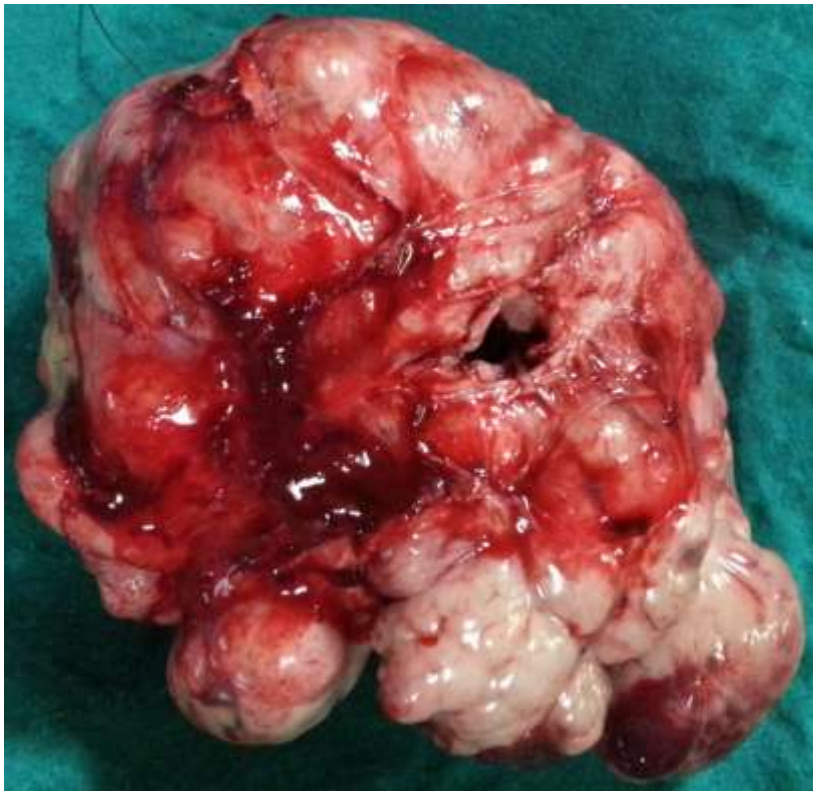
Histopathological examination revealed areas of haemorrhage and necrosis with a large cavity of 6.5 cms on cut section. Microscopically it was a highly cellular tumour composed of proliferating spindle cells in an interlacing bundle pattern. Individual tumour cells vary from spindle to polygonal shape and myxoid degeneration and peripheral pallisading was prominent. The mitotic figures were at the rate of >5/50 HPFs with large areas of necrosis, inflammatory infiltrate, giant cell and haemorrhage. Cells showed positivity of CD117 and CD34 on immunohistochemistry. After surgery the patient was put on Imatinib therapy and the patient was doing well in the follow-up period of 7 months.

Case 2

A 32- years old male presented with history of paraumbli-cal mass and intermittent pain in the abdomen without any history of haematemesis or malena. On examination abdomen was soft, non tender and a vague mass could be appreciated in the paraumblical region. His routine blood investigations were within normal limits. Ultrasound examination of abdomen revealed a hetrerogenously hypoechoic predominantly solid mass with central areas of necrosis in relation to small bowel mesentry and communicating with small bowel loops. Contrast Enhanced CT Scan of abdomen was done which revealed an exophytic lobulated mass of size 9 cms x 6 cms x 4 cms with eccentric soft tissue component and fluid-debris level in relation to jejunum. [Figure-3]

Exploratory laparotomy was done through midline incision and a mass of size 10 cms x 7 cms was seen in relation to the jejunum. Resection of the involved portion of jejunum was done followed by end to end anastomosis. Histopathological examination of the specimen showed a cellular tumour located at the serosa and the tumour was c+posed of interlacing bundles of smooth muscle cells with interspersed many blood vessels. Individual cells were oval to spindle, pleomorphic with high mitotic activity (>5/50 HPFs), and at places showing pallisading arrangements. Myxoid and cystic degeneration was also evident and the cells were positive for CD117 on immunohistochemistry. The postoperative

Figure 2. Postoperative picture of excised specimen of Gastro-intestinal stromal tumour from stomach.



period was uneventful and the patient is on Imatinib therapy 400 mg/day.

DISCUSSION

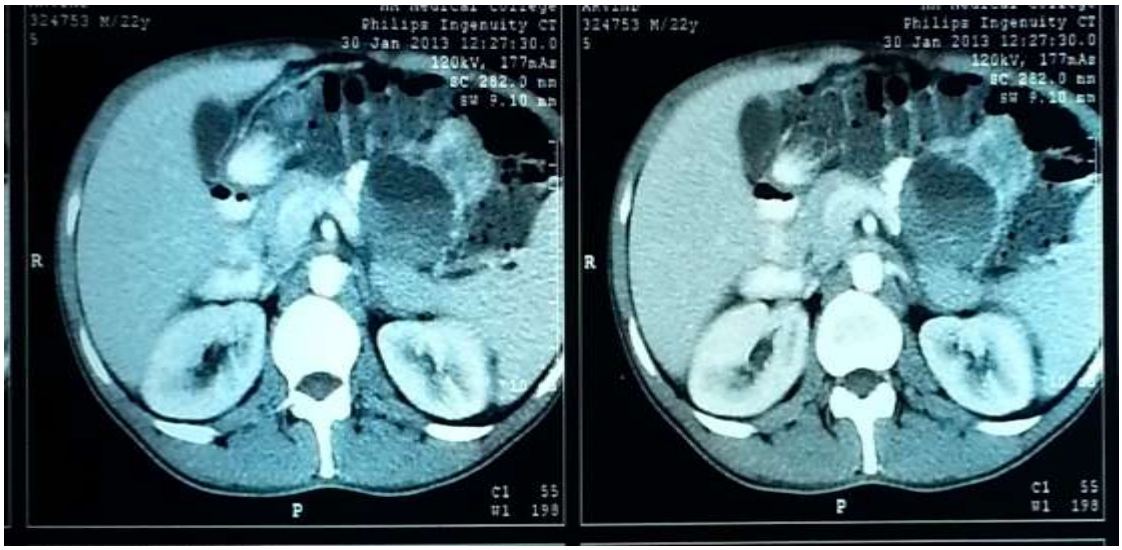
Gastrointestinal stromal tumors (GIST) are a distinct group of rare gastrointestinal tract (GIT) tumors that originate from the interstitial cells of Cajal. These cells regulate gut peristalsis and express CD117. It is a product of the c-kit proto-oncogene that encodes a tyrosine kinas receptor and regulates cellular proliferation in GIST Kindblom, et.al(1998).

These tumors usually arise from mucosa or muscular is propria layers and exhibit an endophytic growth pattern leading to obstruction of bowel lumen. However in up to one third of patients, it may invade adjacent organs. They are most commonly found in stomach (70%), followed by small intestine (20-30%) and the rest in oesophagus, colon and rectum (10%). The clinical presentation of GIST varies depending upon the anatomic location, size and aggressiveness of the tumor. Melena or hematemesis and anemia are the most common presentations due to GI bleeding. It may also present with an acute abdomen due to tumor rupture, intestinal obstruction and pain mimicking acute appendicitis, fatigue, dysphagia and satiety Deshpande and Munshi, (2007); Kwon et.al,(2002)

Imaging studies are aimed at location of GIST lesions; evaluate local invasion and distant metastases. However any of the imaging studies like barium studies, ultrasound examination, Computerised Tomography (CT), Magnetic resonance (MR) cannot reliably distinguish between benign and malignant GIST unless there is associated metastases or local invasion Li SQ,et.al (2001). However CT scanning has 87% sensitivity for the detection of GIST and is ideal in demonstrating the endoluminal and exophytic extent of tumor. Smaller GISTs appear as smooth, sharply defined intramural masses with homogenous attenuation on contrast enhancement whereas larger GISTs with necrosis appear as heterogeneous masses with enhancing borders of variable thickness and irregular central areas of fluid, air, or oral contrast attenuation that reflect necrosis. It is also sensitive for the detection of metastatic liver, peritoneal, lung, and bone lesions.

The most important prognostic factors predicting malignant behaviour of GIST are tumor size and mitotic activity Malla, et.al (2011). It is favourable in patients with tumor size less than 5cm and without infiltration into

Figure 3. CT scan showing exophytic lobulated mass with eccentric soft tissue component and fluid-debris level in relation to jejunum.



adjacent organs. Furthermore, histologically, grade of GIST is dependent upon the number of mitotic figures. GISTs with less than 1 mitotic figure per 50 high-powered fields (HPFs) are correlated with benign behaviour, 1-5 mitoses per 10 HPFs suggests potential malignancy, more than 5 per 10 HPFs indicates malignancy and more than 10 per 10 HPFs denotes high-grade malignancy Miettinen, et.al (2003). Cytologically, the tumour cells show highly varied appearance of spindle shaped, round (epitheloid), plasmacytoid, mixoid, signet ring, granular or multinucleated with other benign or malignant cytologic features.

Surgery is the mainstay of treatment in GIST and chemoradiation has not proved to be very effective. However the finding that GIST are characterized by the expression of the transmembrane receptor tyrosine kinase KIT, has recently suggested new alternative treatment approach to advanced disease. Imatinib is a tyrosine kinase inhibitor that selectively inhibits various tyrosine kinases. The role of this drug in the preclinical experiences showed rapid inhibition of KIT phosphorylation, with decreased cellular proliferation and induction of apoptosis after exposure of GIST cells. Clinical application of this drug in metastasized disease has been encouraging Pidhorecky, et.al (2000).

CONCLUSION

The preoperative diagnosis and behaviour of GIST remains a dilemma even in presence of modern imaging aids and surgical resection still remains the primary form of treatment. However tyrosine kinase inhibitors should be given as an adjuvant therapy in tumors with high mitotic activity.

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