Case Series

A case series on gastro intestinal stromal tumors at various sites

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Abstract

Gastrointestinal stromal tumors (GIST) represent only 0.1 to 3% of all gastrointestinal malignances. They are neoplasms with varying malignant potential ranging from virtually indolent tumors to rapidly progressing tumors. GISTs harbor an activating mutation in either kit or platelet derived growth Factor A (PDGTKA). Confirmation of diagnosis was mainly made by immunohistochemistry. We encountered 3 cases of GISTs with varying presentation. First one was a case of chronic abdominal pain for which the patient was evaluated and found to have mesenteric GIST and was operated electively after thorough work up. Second one presented as a case of acute abdomen. There was a small bowel gist causing intraluminal obstruction. The third one was a case of epigastric pain with recent episode of hematemesis and melena for which she was evaluated with upper Gastro intestinal scopy and found to have gastric gist with low malignant potential. Patient underwent elective wide local excision of tumor with follow-up.

Key words

GIST, Stromal tumors, Epigastric pain.

Introduction

Gastrointestinal stromal tumors (GIST) represent only 0.1 to 3% of all gastrointestinal malignances. But constitute 20%of soft tissue sarcomas [1]. They are neoplasms with varying malignant potential ranging from virtually indolent tumors to rapidly progressing tumors. It may occur in any site of intestinal tract [2]. GISTs harbor an activating mutation in either kit
or platelet derived growth Factor A (PDGTFKA). Confirmation of diagnosis mainly made by immunohistochemistry [3]. But molecular analysis will be helpful for appropriate therapy which include tyrosine kinase inhibitor (TKI’s) therapy which improve survival rate drastically in both localized and advanced disease adjuvant therapy with imatinib is mainly needed for gist with high risk of recurrence [4]. We encountered 3 cases of GISTs with varying presentation. First one was a case of chronic abdominal pain for which the patient was evaluated and found to have mesenteric GIST and was operated electively after thorough work up. Second one presented as a case of acute abdomen [5]. There was a small bowel gist causing intraluminal obstruction [6]. The third one was a case of epigastric pain with recent episode of hemetemesis and malena for which she was evaluated with upper Gastro intestinal scope and found to have gastric gist with low malignant potential. Patient underwent elective wide local excision of tumor with follow-up [7].

Figure – 1: GIST.

Case series
Case report - 1
46 years female with history of chronic abdominal pain with no other positive history and examination finding evaluated with basic and radiological investigations. Her ultrasonogram of abdomen showed exophytic growth in gastric region and ugiscopy was normal. Contrast enhanced CT scan of abdomen showed large heterogeneously enhancing exophytically located mass from gastric wall to consider GIST (Figure - 1). Elective exploratory laparotomy showed mass of 4x3 cm arising from lesser omentum and confined to it without any nodal involvement in perigastric region. Tumor removed in toto (Figure - 2) and biopsy obtained was omental GIST with low malignant potential and was positive for CD 117. Adjuvant TKI therapy was not considered as the prognostic factors revealed low risk.

Figure – 2: Tumor removed.

Case report - 2
70 years male presented to emergency room with features of acute abdomen having history of vomiting, abdominal distension and obstipation. On examination, there was diffuse distension of abdomen with guarding and rigidity. Bowel sounds were absent and rectum was roo my. CT abdomen showed dilated small bowel loops and collapsed large bowel loops along with a mass lesion from small bowel with extensive pneumoperitoneum (Figure - 3). Emergency laparotomy done and was found to have mass lesion of size 7x5cm arising from jejunum intraluminally along with a perforation of jejunum of size 1x1cm nearby to it. Resection of jejunum with tumor with 2 cm clearance both proximally and distally and anastamosis was done. Patient improved well post operatively and started on orals on 6th postoperative day. DT removed on 8th postoperative day. And histopathological report was small bowel gist of high risk group. Patient was started on imatinib therapy and is on regular follow-up. Serial ultrasonogram and contrast enhanced CT Abdomen was normal with no evidence of recurrence.

**Figure – 3:** CT abdomen.

**Figure – 4:** Tumor from greater curvature of stomach.

**Case report - 3**

53 years female came with complaints of epigastric pain along with hemetemesis and malena with no positive findings on examination. She was proceeded with upper gastro intestinal scopey which revealed large sub mucosal mass arising from greater curvature of stomach. And biopsy from it showed as gastrointestinal stromal tumor. Patient was electively explored and a tumor of 10x5 cm was seen arising from greater curvature of stomach (**Figure - 4**). There was neither infiltration of mass into surrounding structures nor any evidence of metastasis or lymphadenopathy. The tumor was excised with 2 cm sleeve of greater curvature of stomach and repaired classically and histopathologically it was low malignant positional GIST and so patient was not started with imatinib. She was on regular follow-up.

**Discussion**

GISTs occur throughout the gastro intestinal tract most commonly arise from stomach small intestine, colon and rectum, mesentry or omentum and esophagus in order. Rarely it may arise directly from abdominal cavity with no apparent connection to GI tract. Such GISTs are known as extra GI-GISTs [8]. It may occur in any age group mainly between 40-80 years. Its presentation may from acute abdomen to incidental while imaging or doing autopsy or surgery for other conditions. They are vascular tumors which may bleed in to abdominal cavity or bowel resulting in abdominal tachycardia, abdominal pain, hemetemesis or malena [9]. Other symptoms may depend on size and location which may cause mild abdominal pain to bowel obstruction in later stages [10]. Most common spread is to adjacent organs such as abdominal cavity and other intra-abdominal organs mainly liver. Lymphatic spread is rare. Around 3% GISTs arise as a component of syndromic GISTs which may include neurofibromatosis type1 and Carney stratakis syndrome and Carney’s triad which is due to germ line mutation of succinate dehydrogenate complex genes [11]. Diagnosis is mainly based on contrast enhanced computed tomography of abdomen and pelvis. Primary GISTs are typically well circumscribed mass within the wall of hollow viscera. Endoscopically primary gist may appear as a sub mucosal lesion with or without ulceration present in upper or lower GI tract. Preoperative biopsy role is controversial as it may cause rupture and dissemination of suspected GIST [12]. For neoadjuvant therapy or metastatic disease, biopsy is appropriate. Laparotomy and surgical resection, remains the mainstay of treatment in which complete microscopic and macroscopic clearance of tumor should be achieved and thorough search for local and distant metastasis to be done. Prognosis mainly depends on tumor size, mitotic index and site of origin especially small bowel GISTs have bad prognosis [13]. Since the lymphatic spread is rare no need for lymphadenectomy. And advanced and metastatic disease also needs cytoreductive surgery and therapy. In addition to the above, molecular study says insertion of kit exon appears to have favorable diagnosis whereas aneuploidy, telomerase expression, kit
exon mutations and kit exon11 deletions have adverse prognosis [14]. So following surgery, patient with high risk of tumor recurrence and metastatic disease may consider adjuvant treatment with imatinib, in resistant cases sunitinib, as a second line of treatment. Nowadays neoadjuvant with imatinib is considered for inoperable tumors to downsize and to allow organ preservation to prevent extensive surgery [15].

Conclusion
GISTs may occur at any site of gastrointestinal tract and the presentation may vary according to the site from where it arises. Upper gastro intestinal tract GIST may present with features of UGI bleed. Lower gastro intestinal GIST may present with acute abdomen features. Extra gastro intestinal GISTs may be silent or present with vague clinical features. And management of GIST vary depending on the site and presenting features apart from its malignant potential and other prognostic factors.

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References


