ABSTRACT: GIST may present as an abdominal mass, obstruction, haemorrhage in the gastrointestinal tract or perforation. Literature on perforated jejunal GIST is scarce. Here by, reporting 2 cases of jejunal GIST. Perforated jejunal GIST in an elderly lady, who presented with mass abdomen. En-bloc resection of the tumor giving a clear margin was done. Immunohistochemistry showed a GIST of aggressive behavior and a case jejunal GIST in a young patient who presented with pain in abdomen. Resection of the jejunal segment containing mass and end to end anastomosis done. Immunohistochemistry showed intermediate type. Both the patients are undergoing adjuvant therapy with imatinib mesylate. A comprehensive review on GIST is being attempted.

KEYWORDS: gastrointestinal stromal tumor, perforated jejunal GIST, Small bowel GIST.

INTRODUCTION: BACKGROUND: The gastrointestinal stromal tumor (GIST) is the most common sarcoma of the gastrointestinal (GI) tract. It accounts for 0.2% of the gastrointestinal tumors. The stomach is the commonest site followed by small bowel, rectum and the esophagus. It may remain asymptomatic and may only be detected incidentally. Literature gives very few examples of a perforated small bowel GIST. Here, we discuss a case which presented as an abdominal mass and was found to be a jejunal GIST with perforation.

CASE PRESENTATION:

CASE 1: A seventy three year old lady presented with lower abdominal pain for the past one year. She gave history of weight loss and decreased appetite. She was on regular treatment for systemic hypertension. On physical examination she was poorly nourished and had a tender intra-abdominal mass of 10x7 cm occupying right iliac, lumbar and umbilical region with restricted mobility. Other system examinations were normal.

Contrast enhanced CT abdomen showed a well-defined large lobulated circumferential exophytic wall thickening of small bowel with multiple areas of necrosis and calcification without luminal narrowing. It also showed severe adhesions to surrounding structures.

Fig. 1: CECT shows well defined large lobulated circumferential exophytic wall thickening of small bowel with multiple areas of necrosis and calcification without luminal narrowing
CT screening of chest was normal. Colonoscopy revealed extraneous compression over the recto-sigmoid and upper GI endoscopy was normal.

Fig. 2: A 12x 10 cm mass in the ante mesenteric border of the jejunum

Explorative laparotomy was done through a lower midline incision. A 12x 10 cm mass in the ante mesenteric border of the jejunum with severe adhesions to jejunal mesentery, laterally to peritoneal folds and gonadal vessels, inferiorly to pelvic peritoneum and roof of bladder and anteriorly to rectus and recto-sigmoid.

Right ureter was pushed to the opposite side. Adhesions were released and the tumor was resected en-bloc along with a part of the jejunum. Tumor was adherent to the wall of the bladder and was opened during dissection. End to end anastomosis of jejunum done and bladder closed in layers over a supra pubic catheter. Postoperatively, she had hypoalbuminemia and electrolyte imbalances, which were tackled appropriately.

Fig. 3: The resected tumor along with a part of the jejunum

Cut section showed focal areas of cystic and hemorrhagic changes and an area of mucosa was found to be perforated and contained by the tumor surrounding it. Histopathological examination was suggestive of a high grade GIST (G2 tumor) of mixed cell type with high mitotic rate (12/5mm²)
and areas of necrosis and the margins were clear of tumor cells. Immunohistochemistry showed vimentin, CD34, c-KIT positivity which was consistent with GIST of aggressive behavior.

The patient was started on imatinib mesylate 400 mg daily and is on regular follow up.

**CASE 2:** A twenty-nine year old lady presented with lower abdominal pain for the past 4 months. She gave history decreased appetite. No comorbid illness. On physical examination she was thin built and had tenderness in lower abdomen. Other system examinations were normal.

USG abdomen showed a well-defined isoechoic lesion in left adenexal region. Both ovaries not separately visualized from lesion, possibility of subserosal fibroid or ovarian lesion. Contrast enhanced CT abdomen showed a hypodense lesion measuring 44 x 42 mm on the left side of uterus with in homogenous enhancement, possibility of ovarian mass.

Fig. 4: CECT showing hypo dense lesion measuring 44 x 42 mm on the left side of uterus with inhomogeneous enhancement, possibility of ovarian mass.

Diagnostic laparoscopy was done. A stalked polypoidal mass of size arising from the jejunum was found.

Fig. 5: Laparoscopic view of the solid tumor from antemesentric border of jejunum
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Explorative laparotomy was done through a midline incision. A 6x5x4cm polypoidal mass in the ante mesenteric border of the jejunum. Polyp attached to intestine by a short stalk measuring 0.5cm in length. Resection of jejunum containing GIST and end to end anastomosis of jejunum done.

Fig. 6: Cut section showed focal areas of solid and myxoid changes.

Cut section showed focal areas of solid and myxoid changes. Histopathological examination was suggestive of GIST of spindle cell type with mitosis (<5/50HPF) and the margins were clear of tumor cells. Immunohistochemistry showed CD34, CD117 positivity. The patient was started on imatinib mesylate and is on regular follow up.

DISCUSSION: The expression of c-KIT (CD117) distinguishes GIST from other gastrointestinal smooth muscle tumors. The initial ambiguity in its distinction from a leiomyoma or a leiomyosarcoma due to the phenotypic overlap has now tailed off. It was first described by Mazur and Clark in 1983.

The Surveillance, Epidemiology, and End Results (SEER) registry reports an incidence of 0.32 in 100,000 population per year, and a prevalence of 1.62 per 100,00 per year during a 15-year period [1]. The tumor shows 20-30% malignant potential[2] It occurs in the stomach (60%), jejunum and ileum (30%), duodenum (5%), colo-rectum (4%), and esophagus or appendix (1%).[3-9] It rarely presents as extra-gastrointestinal GIST in mesentery, omentum and retroperitoneum.[10]

These mesenchyme tumors are thought to originate from the interstitial cells of Cajal or related stem cell like precursors,[11] Though the exact pathogenesis remains unclear, Hirota et al.(1998) pointed to the gain of function mutation in c-KIT[12] and Heinrich et al towards mutations in in platelet-derived growth factor receptor α (PDGFRA).[13] Recent studies have shown a germ line mutations in succinate dehydrogenase (SDH), leading to complete loss of SDH protein.[14,15]

It is more common in adults above the age of 40 years with a slight male preponderance.[16] Seventy percent are symptomatic at presentation, 20% are asymptomatic and 10% are detected at autopsy.[17,18] Clinical presentations include abdominal mass (5-50%), obstruction (5%), haemorrhage and rarely perforation (0.8%).[19,20] Abdominal pain, loss of weight and appetite and anemia are also common features.

Owing to the lack of specific signs or symptoms, a preoperative diagnosis of GIST is grueling. Contrast enhanced CT scan is the preferred imaging modality in patients presenting with an
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abdominal mass. However, endoscopic ultrasound and fine needle aspiration is diagnostic in 89% cases.[21-23] Most common metastatic sites are liver and peritoneum and less commonly, the lungs. Lymphatic spread is rare.[24]

The treatment modality is primarily surgical, ensuring the complete removal of the tumor with involved neighboring structures giving a clear surgical margin. Roy et al.[24] suggests the use of distilled water for peritoneal lavage after surgery as it has a cytolytic activity on the tumor cells and hence prevent peritoneal spillage. The discovery of tyrosine kinase receptor mutations in the pathogenesis has led to the usage of tyrosine kinase inhibitor, imatinib mesylate as an adjuvant to surgery and in inoperable and recurrent cases.[25]

Sunitinib has been in use since 2006 for imatinib resistant GIST. Response of the tumor to conventional chemotherapy is poor.[26] The most common histologic subtype is the spindle cell variety. Others include epithelioid type and mixed spindle and epithelioid type.[27,28] Though there appears to be no correlation between the tumor size and the mitotic index and neither of them to the histological subtype, the former two factors are used in risk stratification and evaluating prognosis.[27] Complete surgical resection gives a chance of 48-65% five-year survival.[19] However, perforation of the tumor drops the five-year survival to 24%, probably due to peritoneal dissemination.[17]

Large size, increased mitotic activity and location of the tumor in the small bowel are associated with poor prognosis. Positron Emission Tomography (PET) is a very good tool for post-operative follow up of patients on imatinib.[26] The median survival period is 9 to 16 months in case of recurrence.

The c-KIT expression, which is pathognomonic of GIST, can be negative in 5% of the cases.[29] A recently detected marker, DOG1 (Discovered on GIST 1), a calcium-activated chloride channel is found to be highly expressed in GIST, with an overall sensitivity of 75-100%.

CONCLUSION: There are only 15 cases of perforated jejunal GIST reported in literature so far.[26] Our case was peculiar in that, the perforation was contained by the tumor itself. With the large size, location in jejunum, high mitotic index, advanced age of the patient and dense adhesions to surrounding structure, the prognosis of our case needs to be watched. Nevertheless, this case stands out for its rarity of presentation and anatomical location.

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