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CASE REPORT OF GASTRO-INTESTINAL STROMAL TUMOR (GIST)

General Surgery

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ABSTRACT

GISTs are the most common sarcomatous tumors of GI tract, usually found in stomach and duodenum. Mostly GISTs arise as a de novo. Frequently present with hematemesis, melena and abdominal pain but sometimes diagnosed incidentally. CECT abdomen with pelvis is important to see the origin and extension of the tumor and to assess for metastasis. A 52 year old male admitted for left sided abdominal pain for 7 to 8 year and on examination lump is palpable in epigastrium, umbilical and left hypochondriac region. CT abdomen suggestive of mass arising from proximal jejunum. The patient underwent a surgical resection with free margins and had an uneventful recovery.

KEYWORDS

GIST, Sarcomatous tumor, Jejunum

INTRODUCTION

GISTs are the most common sarcomatous tumors of GI tract, derived from interstitial cells of Cajal, an intestinal pacemaker cells.[1,2]

They can appear anywhere in the GI tract but approximately 60% arise in the stomach and 30% arise in the small bowel; the other 10% arise in the esophagus, colon, rectum, gallbladder, appendix, omentum, mesentery, retroperitoneum, and pelvis.[2,5]

The cause is unknown and rarely associated with familial syndromes GIST-paraganglioma syndrome (CARNEY's triad), neurofibromatosis 1 and von-Hipple-Lindau syndrome but most developed DE NOVO.[1]

Most GISTs manifest in the form of melena, hematemesis, vague abdominal pain or discomfort. Tumor rupture with intra-abdominal hemorrhage is uncommon. While some patients remain asymptomatic and diagnosed incidentally.

Vast majority (more than 95%) of GISTs express CD117 (KIT) and are sensitive to tyrosine kinase inhibitor such as Imatinib and Sunitinib.[3]

Most important risk factors for recurrence are tumor size larger than 10cm and more than 5 mitoses per 50 high power fields.

CASE REPORT

A 52 years old male having left sided abdominal pain for 7 to 8 years. Patient's vital parameters were normal.

On examination approximately 10×8 cm, non-warm, non-tender, well defined, smooth, firm, non-pulsatile swelling was palpable in umbilical, epigastric & left hypochondriac region.

INVESTIGATIONS

Blood investigations were normal except Hb 8.8 gm/dl.
USG guided TRUCUT Biopsy revealed spindle cell tumor with possibility of gastrointestinal stromal tumor (GIST).

CECT(A+P+T) shows approx. 85×68×100 mm eccentric mass lesion with central necrotic area, air foci and air fluid level within arising from jejunum and abutting duodeno-jejunal flexure and proximal part of jejunum and displacing other small bowel loops.(Figure-1). Few enlarged adjacent homogenously enhancing lymph nodes were noted, largest measures approximately 14×22×15 mm.

Endoscopy suggested extra luminal compression at DJ flexure with normal overlying mucosa.

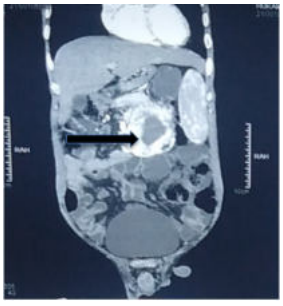

Figure 1: CECT (A+P) showing mass arising from jejunum (arrow)

OPERATIVE INTERVENTION

Planned laparotomy was done. Approximately 8×7×10 cm mass was present at mesenteric border of proximal jejunum, abutting the pancreas (Figure 2 and 3). Liver surface appeared normal.

Resection of jejunum with mass and end to end jejuno-jejunal anastomosis with feeding jejunostomy was done. Adjacent mesenteric lymph nodes were removed. Postoperative period was uneventful.

Adjuvant chemotherapy by Imatinib was started.
Patient was discharged on postoperative day 9 and followed up on POD 15, 30 days and 3 month with no late post-op complications.

Figure 2: Intra-operative image showing mass (arrow)

Figure 3: Resected mass with jejunum

BIOPSY REPORT

- HPE confirmed the diagnosis of gastrointestinal stromal tumor (GIST).
- The tumor showed cells with moderate degree of anaplasia with mitotic activity more than 5 per 50 HPF.
- Section from both surgical margins were free from tumor infiltration.
- Section from lymph node showed reactive hyperplasia.
- No evidence of metastasis was seen.
- IHC markers CD117, DOG1 and Vimentin were positive.

DISCUSSION

GISTs are potentially malignant tumors and the commonest mesenchymal tumors of the gastrointestinal tract.[5]

Familial GIST is an autosomal dominant condition caused by germline mutations of *cKIT* or PDGFRA and manifest at middle age. GIST may be a part of Carney's triad (gastric GIST, paraganglioma, and pulmonary chondroma) or Carney-Stathakis syndrome, which are caused by germline mutations in SDH. It is a well-known fact that von-Recklinghausen Neurofibromatosis type 1 (NF 1) is associated with an increased risk for GIST.[1,8]

- Most GISTs manifest symptomatically, typically with bleeding, vague abdominal pain or discomfort.
- Many patients remain asymptomatic and the tumors are discovered incidentally at the time of other surgery or increasingly during imaging performed for other indications.[4]
- GIST manifest in people with 50 to 70 years of age group and found equally in males and females, with slight male predominance.[6]
- CT of the abdomen and pelvis with an intravenous contrast agent is used to assess for metastatic disease.
- Confirmation by EUS guided FNAC is done if feasible.
- Mainstay of treatment is complete surgical resection with free margins.[1]

For completely resected primary GISTs, mitotic rate, tumor size, and tumor location are important risk factors for recurrence.[7]

Patient having tumor size larger than 10cm and more than 5 mitoses per 50 high power fields has a high risk for recurrence.

Gastric GIST has a favorable prognosis as compared to other location.[3]

Oral chemotherapy (Imatinib) is given for patients at high risk of recurrence and for patients with positive margins or lymph nodes, distant metastasis and recurrence.

Recurrence rates are approximately 40% and can occur 20 years later, so long term follow-up is warranted. Recurrences demonstrate metastasis to the liver, with one third having only isolated local recurrence.[1]

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