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Extra Gastrointestinal Tumour (E-GIST): A Rare Case of Pelvic GIST

Rajendra Prasad Bugalia*[1], Raj Kamal Jenaw[2], Anuradha Salvi[3], Dhananjay Saxena[4], Jeevan Kankaria[5]
[1]Assistant Professor, [2]Senior Professor & Head, [4]Resident, [5]Associate Professor, Department of General Surgery, SMS Medical College, Jaipur, Rajasthan, India
[3]Senior Resident, Department of obstetrics & Gynaecology, SMS Medical College, Jaipur, Rajasthan, India

***Corresponding Author:**
Name: Dr Rajendra Prasad Bugalia
Email: drrpbugalia@gmail.com

Abstract: Gastrointestinal stromal tumors (GISTs) are rare tumors that can arise anywhere in the tubular gastrointestinal tract. A 60-year-old male presented with a pelvic mass measuring 12×10cm in diameter. He underwent exploratory laparotomy with excision of the pelvic mass. The mass was found in the pelvis adherent to bowel loop and sigmoid colon, was correctly diagnosed as a Extra Gastrointestinal stromal tumor (E- GIST) at surgery. It should be noted that preoperative diagnosis of E-GIST is uncommon, due to its rarity and the varying clinical presentation. Gynecologists need to be cognizant of extra-ovarian pathology in female patients presenting with an atypical pelvic mass.
Keywords: Extra gastrointestinal stromal tumors (E-GISTs), pelvic mass, sigmoid colon.

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are the most common sarcomas of the gastrointestinal (GI) tract [1]. They can be asymptomatic and are found incidentally during surgery or radiological studies. Symptoms include gastrointestinal bleeding, abdominal mass, and abdominal pain [2]. Nearly all GIST have mutations in the transmembrane tyrosine kinase (KIT), and most exhibit immunoreactivity to anti-c-kit immunohistochemical staining (CD117), a c-kit proto-oncogene protein [2, 3]. 10–30 % of GISTs are found incidentally during laparotomy, endoscopy, or other imaging studies [4]. Because of their histology GISTs presenting as pelvic masses can be mistaken for gynecological disease such as ovarian tumor or uterine leiomyoma. The majority of GIST have diameter of less than 10 cm. E-GIST are extra gastrointestinal stromal tumor with sites at omentum, mesentry, retroperitoneumor pelvic cavity and are rare tumour. We report here a case with a large pelvic mass with the characteristics of an E-GIST.

CASE REPORT

A 60 year old Hindu male presented in outpatient department of General Surgery with chief complains of constipation since 4 month and pain abdomen in left lower quadrant since 2 month, relieved by oral medications. There was history of use of purgatives in increasing dosage. Sudden weight loss of approximately 8-10 kg was seen since 1 month and loss of appetite was present. There wasno history of haemetemesis, malena, haematochezia or bleeding per rectum. Patient was a known Diabetic and Hypertensive since past 1 year, well controlled on oral medications. On per abdominal examination, a palpable, non tender, firm lump with irregular surface of about 12 × 10 cm size located in hypogastrium and extending into left iliac fossa. Patient was admitted in hospital and all routine blood investigation were within normal limit. Serum CEA was 1.53ng /ml. Ultrasound showed irregular hypoechoic mass in lower abdomen and pelvis of about 98 × 83 mm likely to be retroperitoneal mass. Core Needle Biopsy gave impression of Spindle cell neoplasm of uncertain diagnosis. Colonoscopy visualized uptoileoceacal junction and showed no obvious pathology. CECT Abdomen showed large lobulated centrally necrotic mass of size approximately 11.2×8.6×10.4 cm in pelvis involving multiple bowel loops with oral contrast in central necrotic area (Fig. 1 & 2).

Exploratory laprotomy was planned. Per operative findings showed large mass approximately 12×10 cm arising out of pelvis – greyish in colour, firm in consistency and dense adhesions were present between sigmoid colon and the mass. Loop of ileum was adherent to the mass superiorly along with adjoining necrotic debris, weak adhesions were present with urinary bladder which were easily separable. Thus Exploratory Laprotomy, with excision of pelvic mass, resection of sigmoid colon (Hartman‘s Procedure), resection of ileal segment and double Barrel Ileostomy was done. Patient was haemodynamically unstable during surgery hence primary anastomosis of large and small bowel was not done. Postoperative period was uneventful and patient discharged on day 7 with stable

vitals, with functional ileostomy and healthy suture line. Histopathological Report of multiple sections from pelvic mass revealed Spindle cell Mesenchymal neoplasm, Mild to Moderate Anaplasia with Mitotic activity and area of necrosis. Sections from adherent segment of ileum and colon showed congestion and chronic inflammation with fragments of mass adherent on serosal surface .On follow up, tumour marker study showed following findings (Table 1).

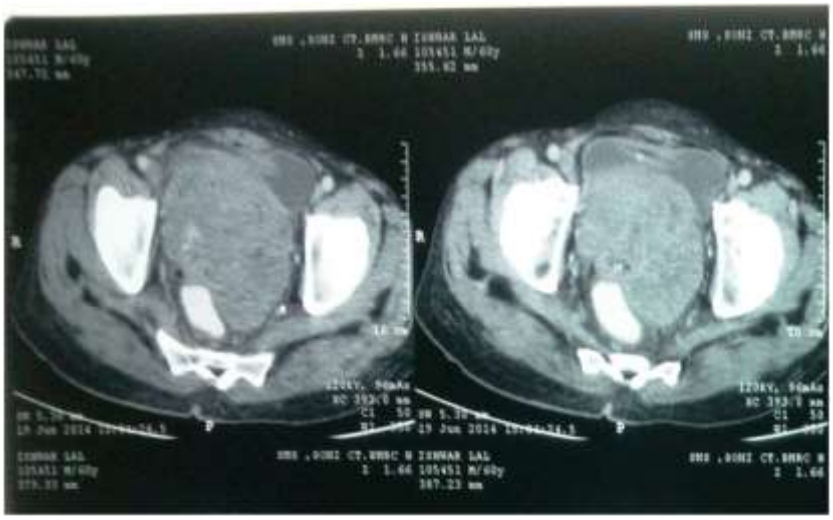

Fig. 1

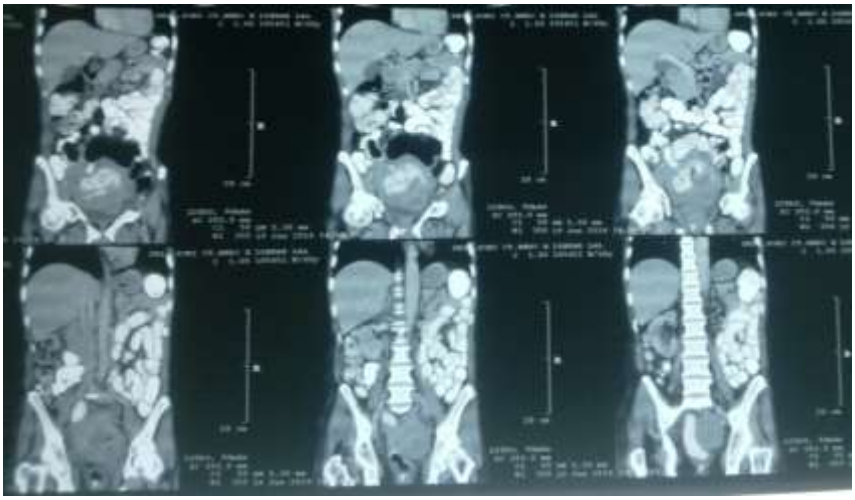

Fig.2

Fig. 1 & 2: CECT Scan of patient showing large lobulated necrotic mass of approximately 11.2×8.6×10.4 cm in pelvis involving multiple bowel loops with contrast in central necrotic area

Table 1: Immunohistochemistry (IHC) report

IHC Marker	Result
CD117/c-KIT	Positive
CD 34	Positive
Smooth Muscle Actin	Positive
KI- 67	Positive (3-4%)
S – 100 P	Positive

After 2 months, patient was taken for Laprotomy for maintaining large bowel continuity by using circular stapler. The patient is being treated with Imatinib Mesylate on an outpatient basis.

DISCUSSION

GIST is a descriptive term for intraabdominalmesenchymal tumours that arise mostly in the GI tract were definitely not carcinomas and failed

to exhibit features of smooth muscle or nerve cells (MAZUR and CLARK ,1983) [4]. GIST cells bore histopathological similarities with interstitial cells of Cajal (pacemaker cells). Both cells expressed KIT receptor tyrosine kinase (KIT RTK /CD117) [3]. Incidence of approximately 15% /million. A peak incidence in the fifth and sixth decades of life is noted and slightly more common among males [3, 4]. GISTs are most commonly located in the stomach and proximal small intestine, but can occur in any portion of the alimentary tract including the omentum, mesentery, and peritoneum, although, as reported here GIST may also have unusual origins from sites such as the pelvis (E-GIST) or Extra gastrointestinal stromal tumour [4]. Exact incidence of E-GIST not known because of rarity of reporting and cells of origin are controversial. Majority of E-GIST are >10 cm and histologically resemble interstitial GIST. As in present case tumour arising from pelvis (E-GIST) in a 60 year old male. Most patients with GIST present with symptoms of varying severity ranging from dysphagia, abdominal discomfort, altered bowel movements and bowel obstruction, to acute or chronic gastrointestinal hemorrhage and peritonitis due to perforation [3, 4]. In the present case the patient had a visible palpable tumor with significant weight loss and reported chronic constipation. Preoperative diagnosis of a GIST is uncommon due to their rarity varying presentation and the lack of distinguishing characteristics on imaging studies. The diagnosis of GIST is based on cellular morphology and immunophenotype. Histologically there are three types of GIST: spindle cell (70 %), epithelioid (20 %), and mixed. Approximately 95 % of GISTs carry an activating somatic mutation of CD117 (c-kit) [3]. Positive immunohistochemical staining for CD117 is a defining characteristic of GIST [3]. Tumor size and mitotic index are the two most important prognostic factors used for risk stratification of GISTs [5]. Outcomes in patients with GIST depend very much on clinical presentation and the histopathological features of the tumor. Thus the overall 5-year survival rate for all patients with GIST ranges from 28–60 % but this range can be stratified based on disease progression. For patients presenting with localized primary disease the average survival rate is 5 years and those with metastatic or recurrent disease the average survival rate is approximately 10–20 months.

Contrast-enhanced CT scan is the first choice imaging modality for patients with suspected abdominal mass as it is useful for both preoperative staging and evaluation for metastatic disease [6]. There are no specific CT findings for GIST tumors although they typically appear as inhomogeneous masses with areas of necrosis and hemorrhage with viable tumor areas showing contrast enhancement [6]. FDG-PET use of ^{18}F-fluorodexoyglucose [^{18}FDG] Positron emission tomography (PET) adds to functional imaging data [6]. Complete surgical resection is the treatment of choice and biological therapy with imatinibmesylate is recommended for cases with incomplete resection or unresectable tumors and when tumors have progressed to metastatic disease [1]. A more recent agent, sunitinib which inhibits vascular endothelial growth factor receptor (VEGFR) in addition to KIT and platelet-derived growth factor receptor alpha (PDGFRα) has proven to be effective in patients intolerant or refractory to imatinibmesylate [1].

CONCLUSION

Our case suggests possibility of GIST must be kept in mind while evaluating any case of abdominal lump. Occurrence of E-GIST is rare and treatment options are similar. The advent of molecularly targeted therapies revolutionized the management. Surgery remains the first choice of early stage and resectable disease. In female patients gynecologists should consider GIST in the differential diagnosis of patients presenting with an atypical ovarian mass.

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