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A Case Report: A Rare Case of Intestinal Gastro Intestinal Stromal Tumour with Peritoneal Metastasis.

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I. Introduction

Gastrointestinal stromal tumor (GIST) is an uncommon mesenchymal tumor which arise mostly from the gastrointestinal tract. It accounts for 0.1-0.3% of all gastrointestinal neoplasm. The term GIST was first used in 1983 by Mazur and Clark to encompass gastrointestinal nonepithelial neoplasm that lacked the immunohistochemical features of schwann cells and did not have the ultrastructural characteristic of smooth muscle cells. The tumors arise from intestinal cells of cajal(ICC) which serve as a gut pacemaker as they create the basal electrical rhythm leading to peristalsis and segmentation of the smooth muscle. The most common site of GISTs occur in the stomach(50-60%), followed by small intestine(30-40%). The other rare site are colorectal, esophagus,mesentery, retroperitoneum and omentum.

GISTs can arise at any age, but occur predominantly in the middle age or elderly individual with peak incidence in fifth and sixth decades of life. More then 80% GISTs are reported in individuals older than 50 years.

II. Etiology and Pathogenesis.

Most of GISTs are sporadic and no risk factors are established. Central to GIST's pathogenesis are two mutation: that of the KIT gene and platelet derived growth factor receptor alpha (PDGFRA) gene. Approximately 95% of GISTs are positive for KIT(CD117), the receptor for stem cell factor(SCF) and 5-7% of GISTs have activating mutations in PDGFRA, the gene encoding a receptor for PDGF-alpha, which results in constitutive activation of PDGFRA tyrosine kinase activity in the absence of its ligand.

Approximately 10-15% of GISTs do not show detectable KIT or PDGFRA mutation and are called wild-type tumors. Approximately 60-70% GISTs are also positive for CD34 and 30-40% for smooth muscle actin. Other markers are found only rarely, including S100 protein in 5%, desmin in 1-2% and keratin in 1-2%.

PATHOLOGY

Macroscopically, GIST appears as exophytic, intramural or intraluminal growth. They are smooth gray and white lesion which are well circumscribed, usually with pseudocapsule. Larger lesions may undergo significant necrosis and cyst formation.

Three morphological patterns are seen in GISTs: spindle cell(Fig.1b), epithelioid(Fig.1a) and mixed. GISTs in small intestine are more often spindle than epithelioid and may show paragangliomatous pattern. Other immunohistochemistry done for CD 117(KIT), CD 34, DOG-1 and PDGFRA etc.

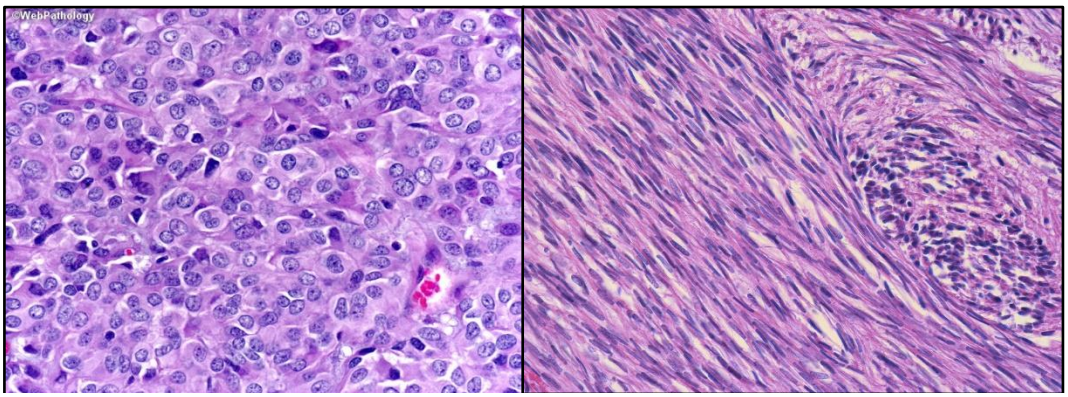

Fig. 1a: Epithelioid type. Fig. 1b: Spindle cell type.

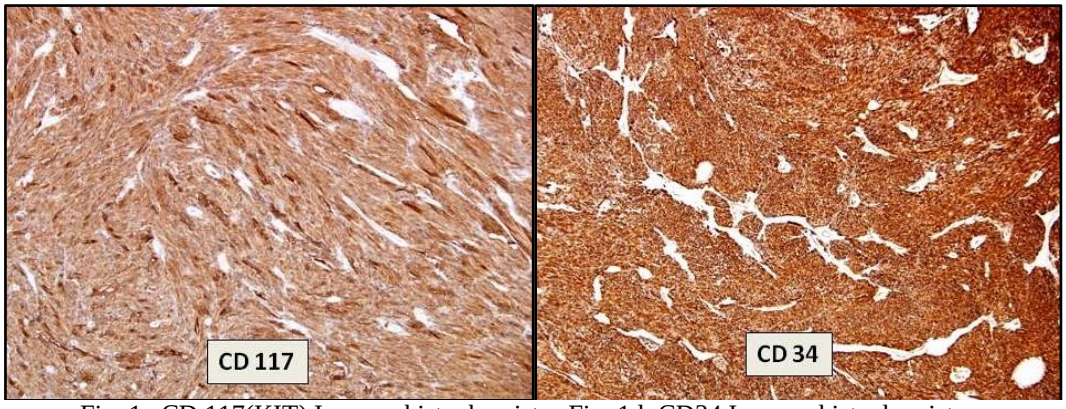

Fig. 1c:CD 117(KIT) Immunohistochemistry.Fig. 1d: CD34 Immunohistochemistry

III. Case Report

A 34 year old Hindu male patient presented to the OPD with complain of vague abdominal pain and nausea since 3 month. Also have complain of postprandial fullness and bloating. Patient have no complain of vomiting, diarrhoea, constipation, bleeding P/R, malena, weight loss.

No past history of similar complain, DM, hypertension, Tuberculosis and any other surgery. Sleep pattern and bowel-bladder habits are regular and undisturbed. No significant family history.

EXAMINATION

On General Examination, pulse: 80/min., Blood pressure: 118/70 mm of hg., Respiratory Rate: 18/min. and temperature was normal.

On per abdominal examination: soft, non-tender and no guarding rigidity. Approximately 8*6 cm non-tender lump palpable in infraumbilical region with well defined margin and firm in consistency. Not mobile with respiration.

INVESTIGATION

All routine lab investigation including CBC, Coagulation Profile, Liver Function test, Renal function test and cross matching done. Chest and abdominal x-ray with RT-PCR for COVID-19 done. Ultrasound of abdomen and pelvis done.

CECT(A+P) s/o large lobulated partly solid(enhancing) partly cystic(Non-enhancing) abnormal lesion(85*57*61 mm) in pelvis showing involvement of adjacent ileum loops, likely to represent **neoplastic mass most likely GIST**. There is multiple hyperdense hyperenhancing soft tissue deposits seen in left iliac fossa region(25*22 mm), in right hemipelvis(44*35 mm), left hemipelvis(35*34mm). s/o **metastatic soft tissue deposits.**

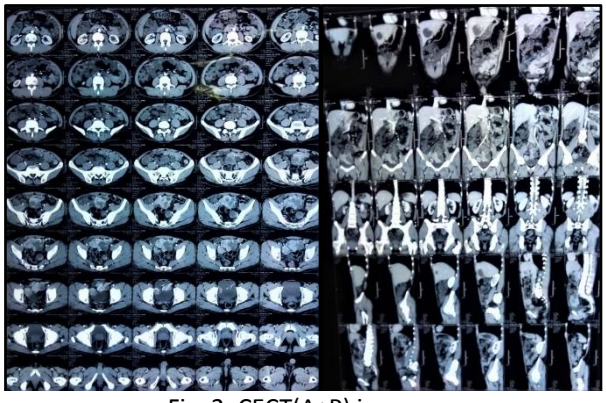

Fig. 2: CECT(A+P) image.

MANAGEMENT

The main aim of management in this patient was the excision of the tumor with negative surgical margins. The procedure opted was laparoscopic excision of tumor. Bowel preparation was done prior to the operation and patient was on liquid diet for two days before the date of surgery pre- operative anaesthetic assessment was done and patient was declared fit for the procedure.

OPERATIVE NOTE

Intraoperatively, approx. 8*7 cm sized tumor was found arising from antimesenteric border of ileum. With adherent omentum with hypervascularity. Another three small(approx. 3*3 cm) mass lesion adherent to abdominal wall in LIF, right and left hemipelvis which s/o metastatic deposits. Laparoscopic resection of abdominal deposits. Laparoscopic resection of ileal loop with 10 cm margin and ileo-ileal side to side anastomosis done with help of stapler. Approx. 7 cm infraumbilical midline incision kept and all specimens delivered and sent for HPE. Adequate normal saline wash given and haemostasis achieved. Romo ADK 20fr drain

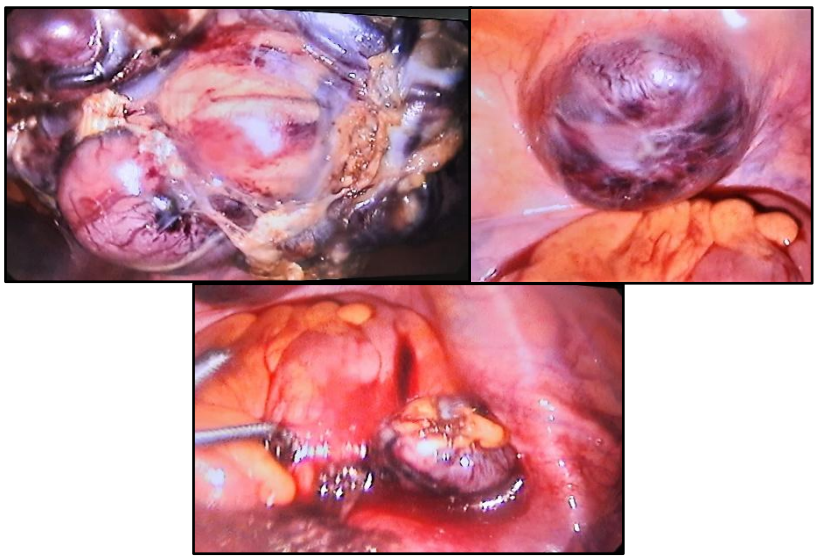

Fig.3: (1) laparoscopic view of ileal GIST on antimesenteric border with hypervascularity,(2)(3) laparoscopic view of peritoneal metastasis.

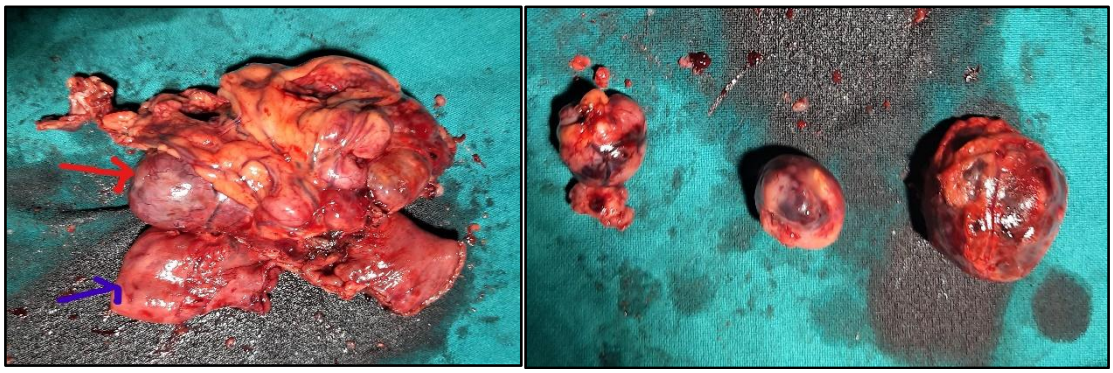

Fig.4: on gross examination (1) ileal loop(blue arrow) with both ends closed with stapler and mass(red arrow) with omentum present on antimesenteric border.(2) small peritoneal deposits.

kept in pelvis. Midline sheath closed with help of epimide loop no-1 and all skin incision closed with epimide 3-0 in vertical matterus manner.

BIOPSY

Biopsy report suggestive of spindle cell typegastrointestinal stromal tumors(GISTs) with malignant behaviour. There is moderate degree of pleomorphism with more than 5 mitoses per 50 high power field. There is evidence of fibrous capsule and large area of tumor necrosis. Section from both surgical margins show normal histology. No evidence of tumor infiltration. On immunohistochemistry tumor is positive for c-KIT(CD 117) and DOG1 and Focal positive for CD34.

IV. Discussion

GIST is aggressive lesion with recurrence rate of 40%. The most common site of metastasis in GIST is liver(65%) followed by peritoneum(21%). The treatment of choice for primary GISTs remain complete surgical resection with negative margins(R0 resection). The macroscopic margin of 1-2 cm is sufficient to achieve microscopically negative margins. Precaution is taken not to rupture tumorpseudocapsule as large lesion are

typically soft, fragile with thin capsule. If capsule breach and margin is not negative for cell then patient is considered for imatinib treatment. A laparoscopic approach may be considered for tumors in favourable anatomic location by expert surgeons, only in situation where a complete resection without capsule rupture is feasible and should be removed in a plastic bag. Both NCCN and European society for medical oncology recommend 3 years of adjuvant treatment with imatinib in high risk patients. The high risk tumor define as a gastric GIST with mitotic count more than 10/50 HPF or non-gastric GIST with mitotic count more than 5/50HPF or tumor presenting with rupture or metastatic lesion.