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Original Research Paper

Surgery

JEJUNAL GASTROINTESTINAL STROMAL TUMOR - A RARE CAUSE OF HEMOPERITONEUM

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ABSTRACT Gastrointestinal Stromal Tumor (GIST) are the malignant mesenchymal tumor. Jejunal GIST are extremely rare. Most common clinical presentation of GIST is bleeding. Preoperative diagnosis is difficult. Diagnosis is confirmed on histopathology and immunohistochemistry. GIST originate from interstitial cells of Cajal, which are regulators of gut peristalsis and are preferentially located in the stomach and small intestine. We are reporting a 39 year old male presented with abdominal pain, malena and anemia. On evaluation patient was diagnosed as a case of Jejunal GIST. Laparotomy and complete surgical resection of tumor was performed. Postoperatively Tyrosine Kinase Inhibitor - Imatinib was given to the patient.

KEYWORDS : Jejunum, Exophytic, Gastrointestinal Stromal Tumor, Imatinib, Hemoperitoneum

INTRODUCTION :

Gastrointestinal Stromal Tumor is the most common mesenchymal tumor of digestive tract. It has malignant potential. It can occur from oesophagus to rectum. Most common location of GIST is stomach followed by small intestine. The incidence of GIST is very low i.e. 2 in 100000, while Jejunal GIST is extremely rare, accounting for 0.1% to 0.3% of all GIST tumors (1).

The most common clinical presentation of GIST is gastrointestinal bleed (2). which may be acute or chronic and results in anemia. Patient may also present with abdominal mass, obstruction and rarely perforation (3,4). GIST arises from interstitial cell of Cajal. These serve as pacemaker of stomach (5) . Complete surgical resection is the mainstay of treatment.

CASE REPORT :

A 39 year male presented to emergency department with history of abdominal pain from last 10 days, with off and on history of malena since 1 month. On clinical examination, patient was pale with pulse rate 94 per minute, blood pressure 90/60 mmhg, abdominal examination showed mild distension, diffuse abdominal tenderness with guarding. Laboratory investigation showed low haemoglobin (Hb 6.5 gm %), total leucocyte count (TLC) 11,000. Renal function was normal.

X-Ray Abdomen Erect did not show gas under right dome of diaphragm. Ultrasound abdomen showed 11 x 7 cm hypoechoic, heterogeneous and vascularised mass, (?) arising from bowel with free fluid in abdomen. CECT Whole Abdomen revealed exophytic mass arising from small bowel with hypervascularity. With proper consent and preanaesthetic workup, patient was planned for exploratory laparotomy. On exploration of the abdomen there was about 11 x 7 cm exophytic mass originating from proximal jejunum, about 35 cm distal to duodenojejunal flexure.

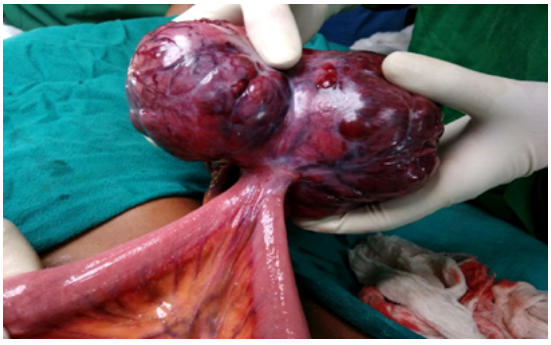

Fig.1 Intraoperative image showing exophytic mass from proximal jejunum.

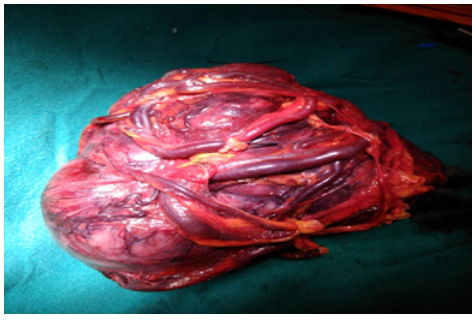

Fig.2 Excised specimen - hypervascular with dilated congested vessels and cystic degeneration.

The mass was firm, lobulated with areas of cystic degeneration and overlying distended veins with small hematoma. The tumor was hypervascular (Fig. 1 & 2). No evidence of visceral metastasis was found. Tumor was excised in toto and Jejuno-Jejunal anastomosis was performed. Histopathological analysis revealed densely crowded spindle and polygonal cell tumor showing moderate nuclear pleomorphism with 5 mitoses/HPF and the tumor spread into the mucosa and outwards into the serosa. There was no lymph nodal spread. Immunohistochemistry demonstrated positive for CD117 (eKit) and CD34 and negative for S-100n and chromogranin. Therefore, the final diagnosis was high-risk GIST of Jejunum. Postoperative recovery was uneventful and patient was discharged on the 7th postoperative day. Thereafter, adjuvant Imatinib Mesylate was administered.

DISCUSSION :

The term GIST was introduced by Mazur and Clark in 1983 (6). GIST can manifest at any age but mostly after 40 year. GIST manifest symptomatically typically with gastrointestinal bleeding, abdominal mass, intestinal obstruction and rarely perforation. Commonest site of GIST is stomach. As per literature, jejunal GIST are rarest type amongst all type of GIST (7).

GIST may have different clinical and biological behaviour ranging from small benign tumor to aggressive form that have a dismal prognosis. The main differential diagnosis for GIST are leiomyoma and schwannomas. Leiomyomas and schwannomas are biologically benign conditions with excellent prognosis. Hence, these must be differentiated from GIST. The non-specific and variable clinical symptoms of Jejunal GIST is a diagnostic challenge.

CT is the most commonly used diagnostic modality. Approximately 20-30% of all GIST are malignant at the time of initial diagnosis with the criteria of metastasis and adjacent organ invasion (8).

Endoscopic ultrasound guided FNA biopsy is a reliable and accurate method for evaluation of submucosal lesion in the gastrointestinal tract. When combined with cytological and immuno-cytochemical studies, EUS- FNAC is accurate and efficient in the diagnosis of GIST (9) . A complete surgical resection of the tumor is the only potentially curative treatment of the patient who have primarily resectable GIST (10) .

Histopathologically, GIST are composed of spindle (70%), epithelioid and round cells (11). On immunohistochemistry, 95% of GIST shows CD117 positivity and 70% shows CD34 (12). A Tyrosine Kinase Inhibitor - Imatinib, has revolutionised the treatment of unresectable, metastatic GIST. Imatinib is effective for the neoadjuvant and adjuvant chemotherapy.

There are no dichotomous criteria that are able to define benign versus malignant GIST, but most important risk factor for GIST labelled as malignant tumor are size more than 6 cm and more than 5 mitoses/50 HPF. In patients with metastatic disease or unresectable disease Imatinib 400 mg daily showed an overall 2 year survival of 70% compared with 25% for patient on traditional chemotherapy (13).

CONCLUSION :

Jejunal GIST are rarest type amongst all type of GIST. Symptoms are generally nonspecific. Surgery remains the mainstay of treatment in resectable tumor but absolute requirement is complete surgical resection. Advent of targeted molecular therapy - Imatinib has revolutionised the treatment of GIST. Imatinib is recommended in metastatic , residual or recurrent cases of GIST.

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