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Original Research Paper

General Surgery

A RARE CASE OF PERFORATED ILEAL GIST PRESENTING AS ACUTE ABDOMEN

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ABSTRACT A case of perforated gastrointestinal stromal tumor (GIST) of ileum causing acute abdomen is described, with a brief review of the literature. A male patient presented with symptoms of acute abdomen. After evaluation, a laparotomy was performed, where perforation of a tumor in the ileum was found. The tumor was attached to the ileal loop and also to the peritoneal attachment of the dome of urinary bladder. The tumor was resected with adjacent ileal loop and ileoileal anastomosis was done . On examining later revealed a perforation of small bowel at the site of tumor and histopathological examination showed that the tumor was GIST. Postoperative period was uneventful and the patient received treatment, using imatinib.

Gastrointestinal stromal tumors are relatively rare and often present with vague symptoms. Their first clinical manifestation as acute abdomen due to their perforation is extremely rare. In emergency laparotomy, a R0 resection is required and adjuvant therapy with imatinib must be considered.

KEYWORDS : Acute abdomen , perforation , gastrointestinal stromal tumors (GISTs) , imatinib

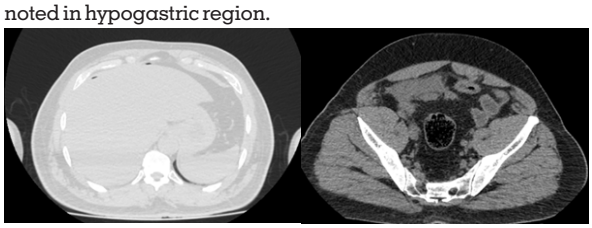
INTRODUCTION:

Gastrointestinal stromal tumors (GIST) is most common mesenchymal tumour of gastrointestinal tract (80%). Tumors originate from interstitial Cajal cells and account for 0.1–3% of all gastrointestinal tumors. The pathogenesis of GISTs is based on the oncogenic, mutational activation of KIT tyrosine kinase in >95% of cases, or less frequently, on that of platelet-derived growth factor receptor alpha . Tumors are usually located in the stomach and small intestine , but they can be located anywhere in the gastrointestinal tract, including the omentum and peritoneum. Generally, GISTs have a silent behaviour and are diagnosed incidentally. Presentations include abdominal mass (5–50%), haemorrhage (40%), obstruction (5%) and rarely perforation (0.8%).

Case study:

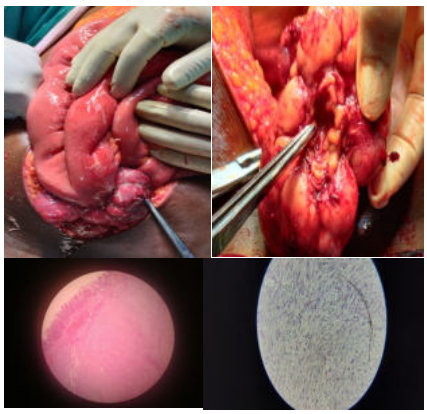
A 65 year old male was hospitalized following one day abdominal pain with vomiting and seven days melena. On admission, the patient had tachycardia and tachypneic. Abdomen examination revealed distension with diffuse tenderness and rigidity, inaudible bowel sounds. On routine blood investigation, leukocytosis (total count – 16,000cells/cumm), dyselectrolytemia (sodium-130meq/L, potassium- 2.9meq/L) was present, other values were within normal limits.

Ultrasound abdomen and pelvis shows hyperechoic mass measuring 6.7 * 6.4 cms with adjacent hyperechoic collection noted in hypogastric region.



CT abdomen and pelvis shows clumping of mid and distal ileal bowel loops giving mass like appearance, minimal free fluid in pelvis and minimal intra peritoneal free air. After resuscitation and stabilization, the patient was taken up for emergency laparotomy. Intra operative exploration identified a mass with maximum dimension of 8*7cm with lobulations and necrosis, which was attached to small bowel loops about 60cm from ileocecal junction and also attached to the peritoneal attachment of the dome of urinary bladder.

Mass along with part of ileum was resected and dissected from the peritoneal attachments of bladder. End to end ileal anastomosis was performed. Examination of the excised mass revealed a perforation in small bowel at the site of attachment of mass. On post operative pathological examination, the tumor shows well circumscribed malignant neoplasm just beneath the ileal mucosa consisting of spindle shaped cells with moderate cytoplasm. Mitotic figures >5/20 high power fields (HPF).



Margins uninvolved.
pTMNstaging- pT4Nx
Immunohistochemistry–CD117-intense cytoplasmic positivity in 70% of tumour cells.
S-100 – Negative.

DISCUSSION:

GISTs typically occur in patients around the sixth decade of life and can be found in any site of the gastrointestinal track. Small sized tumors (<2 cm) are usually asymptomatic, and are

discovered incidentally, while larger lesions present as large abdominal masses, with or without clinical manifestations.

The symptoms and signs are not disease-specific and as a consequence about 50% of GISTs have already metastases at the time of diagnosis, usually to the liver or the peritoneum.

Although the diagnostic procedure includes several examinations, such as barium examination of the gastro intestinal track , computer tomographyand angiography ,none of them can establish the correct diagnosis with 100% certainty . The preoperative percutaneous fine needle aspiration of the tumor for diagnostic purpose is not indicated because of the danger for potential intraperitoneal migration or tumor rupture. Recently, several studies pointed out the significance of endoscopic ultrasound-guided fine-needle aspiration for the diagnosis of GIST with a reported accuracy of 89% . On the other hand, positron-emission tomography (PET) with 18F-fluoro-2-deoxy-D-glucose is a very useful tool for the postoperative follow-up of patients receiving imatinib. GISTs can be categorized as low or high-risk tumors by taking into account the possibility of metastasis or recurrence. However, the main prognostic factor is the mitotic count.

The treatment of choice for GISTs is the surgical excision of the tumor. All tumors must be completely resected (R0 resection), where possible, including the tissues that are infiltrated, while systemic lymph node dissection is not recommended by many authors .Complete surgical resection is connected with 48-65% five-year survival. Partial resection must only be performed in case of large tumors, for palliative purposes or for the control of symptoms or complications such as compression of other organs, hemorrhage, or pain .

The prognosis is dismal when the tumor presents with symptoms or signs such as perforation, multifocal location or metastatic lesions. Patients with localized or locally advanced tumors have 46% five-year survival, in contrast to patients with metastatic tumors or multifocal tumors in whom the five-year survival is 0%. Perforation of the tumor lowers the five-year survival to 24%, probably due to peritoneal dissemination . These patients have a similar evolution as patients with incomplete tumor resection, with shorter disease-free survival and mean survival of 17 months .

GIST response to conventional chemotherapy is very poor (<10%), while radiotherapy is only used in cases of intraperitoneal hemorrhage, when the precise location of the tumor is known, or for analgesic purposes.

Imatinib was found to act as a powerful selective inhibitor of tyrosine- kinases (c-ABL, bcr-ABL), of PDGFR receptor (platelet- derived growth factor receptor) and of c-kit receptor. Imatinib is well tolerated by oral administration, and the suggested efficient doses must be >300 mg per day to achieve curative results . Imatinib is the first effective treatment for non-resectable or metastatic GISTs.

CONCLUSION:

GIST are relatively rare tumors . The clinical presentation of GIST is variable but perforation as in this case is rare. Patients with GIST have limited treatment options. A complete surgical resection without extensive lymph node sampling is still the primary treatment option, but even this has resulted in poor outcomes and recurrence. Although adjuvant and neoadjuvant therapies with imatinib is still investigational, it has considerable activity in patients with advanced disease such as perforated tumors and should be considered as an adjuvant to surgery.

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