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A Case Report of Extra luminal Large Gastrointestinal Stromal Tumor in an Old Male Presenting as Perforation: A Rare Case

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Abstract: Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors that make < 1% of all gastrointestinal neoplasms and 20% of small bowel neoplasms. The most common acute presenting symptom of these tumors is gastrointestinal hemorrhage with obstruction being rare. This report describes a 65 year old male who presented with features of perforation in emergency. On his USG abdomen, he was found to have an ill-defined multiseptated lesion containing necrotic areas showing vascularity in the right iliac fossa with inflammation of surrounding mesentery and moderate collection in peritoneal cavity. He underwent exploratory laparotomy in emergency and was found to have 10-cm pedunculated lesion in the distal jejunum with rotation of the bowel on its mesenteric root in a meso-axial manner with attachment to base of mesoapendix. The literature on this rare modality and its management is briefly reviewed.
Keywords: Gastrointestinal stromal tumors (GISTs), multiseptated lesion.

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are rare but represent the most common type of mesenchymal neoplasms that arises from the gastrointestinal (GI) tract[1-2]. They account for 1–3% of all gastric neoplasms, 20% of all small bowel tumors, and 0.2– 1.0% of all colorectal tumors with an incidence of 10 to 20 per million population. Symptoms due to GIST are not typical and depend on the localization and the tumor size. About 10-30% of GIST is completely asymptomatic, and is discovered accidentally during the endoscopy or radiological evaluation as well as during surgical interventions performed for various other reasons. The diagnostic evaluation of GIST is based on imaging techniques however the most important diagnostic tools are the histological and immunohistochemical examinations.

Some gastric sub mucosal tumors that are considered to be gastrointestinal stromal tumor (GIST) The most common acute presenting symptom requiring surgical intervention in GIST tumors is GI bleeding with obstruction being much less common. Here we describe a male who presented in emergency with features of perforation peritonitis that was successfully treated operatively.

CASE PRESENTATION

A 65 year old male presented in the emergency with chief complaints of severe abdominal pain, abdominal distension and inability to pass flatus and motion since 4 days. On his physical examination he had tachycardia with a pulse rate of 134/min, BP 80/54 mm hg, and crepitation in his chest with a spo2 of 92 on Oxygen. On his per abdomen examination he had profound tenderness, generalized distension, guarding and rigidity all over his abdomen. After his emergency resuscitation he underwent supine x-ray abdomen showing some mildly dilated loops of small bowel. On USG abdomen, he was found to have an ill-defined multiseptated lesion containing necrotic areas showing vascularity in the right iliac fossa with inflammation of surrounding mesentery and moderate collection in peritoneal cavity. He was taken in emergency for exploratory laparotomy in which abdomen was opened from midline. He was noted to have about 800 ml of toxic fluid in peritoneal cavity which was drained and a 10-cm spheroidal mass lesion in the distal jejunum with rotation of the bowel on its mesenteric root in a meso-axial manner with attachment on base of mesoappendix.

The proximal bowel was dilated, and decompressed bowel was noted distal to the stalk of the lesion, which was constricting the bowel at the site of attachment. The bowel was untwisted, and the lesion was transected at its stalk across normal jejunum and mesoappendix with grossly negative margins and concomitant appendectomy was performed and sent for histopathological examination.

The patient did well postoperatively, and was discharged on postoperative day 10 with pathological reports evaluating it as spindle cell tumor compatible with a gastrointestinal tumor.

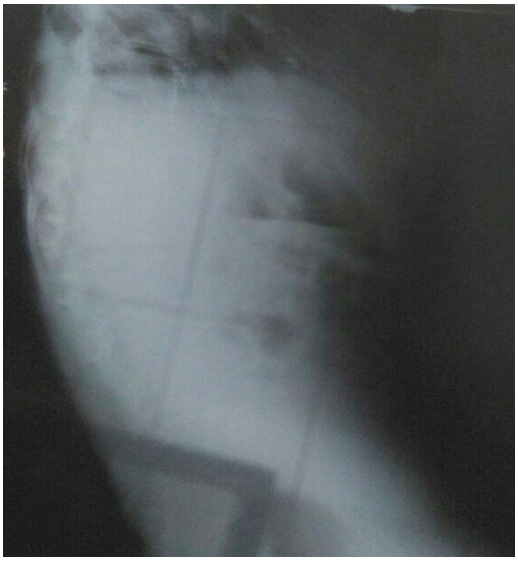

Fig-1: X-ray abdomen supine showing diffuse haziness with air shadow anteriorly

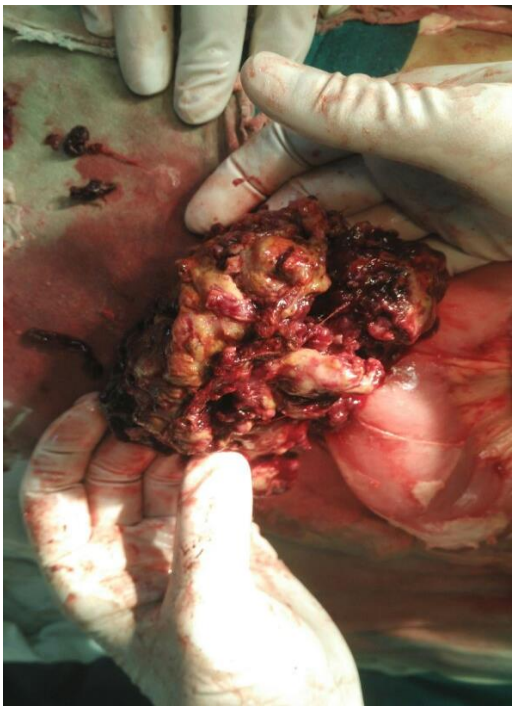

Fig-2:Ruptured mass lesion being shown arising from jejunum

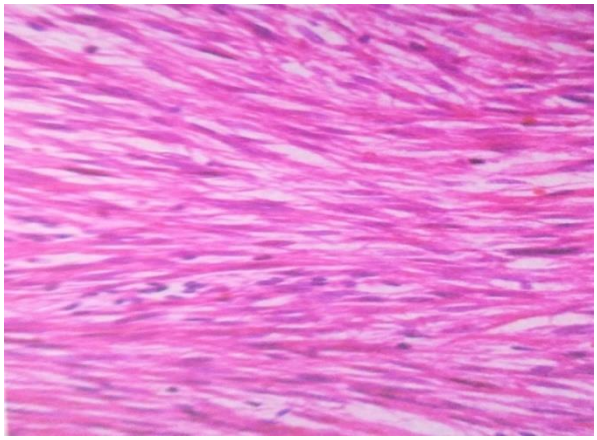

Fig-3: Histopathological examination showing Spindle cells showing elongated nuclei with eosinophilic cytoplasm arranged in fascicles suggestive of gastrointestinal stromal tumor

DISCUSSION

GISTs belong to a group of cancers called soft-tissue sarcomas that develop in the mesenchyme of our gut. Majority of them arise in the stomach or small intestine (95% of them) [3] but can occur anywhere in the GIT. Vague, nonspecific abdominal pain or discomfort comprises the most common symptom after bleeding (due to central necrosis of the tumor in large GIST), sensation of abdominal fullness (more common with jejunal GIST), Palpable abdominal mass (rare), Malaise, fatigue, or exertional dyspnea and sometimes focal or widespread signs of peritonitis. Obstruction may result from intraluminal or extra luminal growth of the tumor which may present with site specific features. Fever, anorexia, and weight loss are rarely observed, and GISTs originating in the jejunum seldom cause perforation and acute diffuse peritonitis. Bleeding into the peritoneal cavity due to a ruptured GIST can lead to diffuse peritonitis for which emergency surgery is the treatment of choice. In this case, GIST was found to be originating in the jejunum with rotation of the bowel on its mesenteric root in a meso-axial manner with attachment to base of mesoapendix.

Complete surgical resection is the mainstay and the only radical treatment for GIST [4]. Completeness of the primary resection (R0 v. R1 or R2) influences the prognosis. This is mainly important in low-and intermediate-risk GIST, whereas the prognosis for malignant GIST is primarily dictated by tumor aggressiveness [5]. In this case, complete resection of the tumor was performed with margins of around 4 cm and was sent for histopathological correlation and jejuno-jejunal anastomoses & appendectomy with derotation of gut was done after which peritoneal cavity was thoroughly washed with warm normal saline so as to minimize the risk of tumor contamination. After histopathological confirmation of the tumor mass, Patient was given adjuvant chemotherapy with imatinib mesylate [6]. But there was no documentation available

that a tyrosine kinase inhibitor improves patient survival. Our patient was a high risk case and prognosis was dismal as the tumor presented with signs and symptoms of perforation or rupture with a tumor size of more than 5cm and tumor necrosis with mitotic counts of 3/hpf. However the patient was alive even after 10 months without any evidence of active disease.

CONCLUSION

GIST in itself is a rare entity and Spontaneous rupture is a very rare presentation of GIST. The diagnosis should be ascertained if patient presents with abdominal mass, pain, fever and signs of peritonitis. In this case of ruptured GIST into the peritoneal cavity, a thorough resection of the mass lesion with grossly negative margins should be performed with thorough intraperitoneal lavage to reduce the risk of tumor spillage and Manipulation of the lesion should be kept to a minimum after which patient should be given imatinib for adjuvant chemotherapy but there has been no proof for it to improve patient survival.

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