A different day in the management of diffuse peritonitis: an elusive case of a perforated jejunal stromal tumor.

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Abstract
Background
Jejunal location of a gastrointestinal stromal tumor is a rare occurrence and its presentation in Emergency department in the form of peritonitis as a consequence of its perforation is even rare. We herein describe a rare case of exophytic growth at proximal jejunum with a perforation causing generalised peritonitis.

Case presentation
A 52/female presents with acute abdomen for a duration of 2 days. Examination was consistent with hollow viscous perforation. Lab values showing leukocytosis without renal function deterioration. Plain X-ray showed gas under right dome of diaphragm. On exploration, a perforated jejunal mass letting intestinal content into the peritoneal cavity was found. Segmental resection with primary anastomosis was performed. Histopathological examination revealed it to be Gastrointestinal stromal tumor.

Conclusion
Stromal tumors rarely present as acute abdomen. If guided by an abdominal mass with diffuse peritonitis, the possibility of jejunal GIST perforation should be considered, even though it is extremely rare. A high degree of suspicion is necessary in view of the high morbidity rates resulting from a delayed diagnosis of the disease.

Key words: GIST, Jejunum, perforation, peritonitis

Introduction
Gastrointestinal stromal tumors (GIST), originating from the interstitial cells of Cajal, are rare but the commonest mesenchymal tumors of the gastrointestinal tract accounting for 0.1–3% of all gastrointestinal tumors (1) (2) (3). These tumors express c-kit (CD 117) oncoprotein and have mutations in the c-kit or platelet derived growth factor receptor alpha (PDGFR-A).

Occurring usually in fifth decade of life, the stomach has been the commonest localisation (60-70%) followed in order by small intestine (25-30%, of which jejunum constitutes 10%), colon, esophagus and the rectum. Extraintestinal occurrence is very rare but reported pointing towards a multipotent mesenchymal stem cells. (3)
While the asymptomatic population can be large in whom the diagnosis is incidental, patients usually have symptoms of vague abdominal pain, a palpable mass, GI bleeds in the form of malena and/or hematemesis, intestinal obstruction, anorexia, weight loss and fever; but all being nonspecific a preoperative diagnosis can be elusive. (4) In this article we describe an acute presentation of GIST arising from the jejunum, that perforated and resulted in diffuse peritonitis requiring an urgent laparotomy.

Consent
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Case report
A 52 year female was admitted to the emergency department of BP Koirala Institute of Health Sciences with a 2-day- history of acute abdominal pain, nausea and an episode of bilious vomiting. She had no bowel movements in two days. Exempting an episodic history of epigastric pain, usually relieved by non-prescription antacids, she had no other illness in the past. She had never undergone surgical or endoscopic procedures, and denied any use of drugs for prolonged period. On admission, the patient was conscious, had blood pressure of 90/60 mm of Hg, heart rate 110/min, respiratory rate 22/min, while her physical examination revealed palor and tachycardia. Remarkable per abdomen were: distension, generalized tenderness and guarding and an obliterated liver dullness. No palpable mass was revealed on physical examination due to abdominal guarding, and bowel sounds were hypokinetic. A diagnosis of peptic ulcer perforation peritonitis was made and planned for urgent laparotomy, while she was being resuscitated with fluid.

Blood cell count was 13,800 cells/µl (Neutrophils 75, lymphocytes 25), haemoglobin 8 gram percent and a normal urea and electrolytes values. Chest X-ray in erect position showed free gas crescents beneath both domes of diaphragm.

At laparotomy, the peritoneal cavity had bilious contaminant fluid ~2L in volume. Duodenal region relatively uninflamed and clean. Exploration of the peritoneal cavity revealed a large
10X8 cm, irregular, bosselated exophytic and seemingly vascular tumor arising from the proximal jejunum, 8 cm from the DJ flexure with its lateral and inferior surfaces adherent to the parietal wall and to the descending colon, with a 1X1 cm perforation at its medial surface emanating bilious fluid.

The perforation was closed with 2/0 vicryl sutures preventing further contamination and the tumor was dissected free of its lateral and inferior adhesions. Macroscopically tumor free margins were achieved proximally by dissection at DJ flexure. Segmental resection and jejunal end to end anastomosis was performed in conventional two layers.

Patient had an uneventful recovery and was discharged on 7th post-operative day.

**Histopathological examination**

Gross examination revealed edematous jejunal mucosa, attached mass measuring 9X8X4 cm arising from anti-mesenteric border with irregular, bosselated grey white to grey brown surface. Cut surface shows capsulated lesion with areas of necrosis.

Microscopic examination reveals spindle shaped cells in fascicles and vague storiform pattern. Individual cells exhibiting moderate pleomorphism, plump nucleus, vesicular chromatin and inconspicuous eosinophilic cytoplasm. Mitotic activity 5/50 hpf. Resected
margins free of tumor. CD117 staining shows strong immunoreactivity of nuclear membrane and cytoplasm CD34 staining shows less than 50% tumor cells exhibiting immunoreactivity.

Following histopathological diagnosis the patient has been receiving Gleevec 200 mg OD, and has been tolerating the drug without adverse reactions in the first month follow-up.

Discussion
Jejunal GIST are uncommon tumors (10%) in the digestive tract. Patients usually suffer from abdominal pain or palpable mass, and also complain of early satiety or abdominal fullness. Jejunal GISTs may cause symptoms secondary to obstruction or hemorrhage. Intussusception due to Jejunal GIST, and GIST in a true jejunal diverticulum have also been reported. Pressure necrosis and ulceration of the overlying mucosa may cause gastrointestinal bleeding, and patients who experience significant blood loss may suffer from malaise and fatigue consequent to anemia. Obstruction may result from the intraluminal growth of the tumor or luminal compression from an exophytic lesion. Fever, anorexia and weight loss are
rarely observed (5) and GISTs originating from the jejunum seldom cause perforation and acute diffuse peritonitis (5) (6).

This patient had perforation of the tumor and resulting in diffuse peritonitis. Patients often undergo resuscitation and open surgical exploration when diagnosed as having hollow viscus perforation. Diagnostic laparascopy and laparoscopic repair attempts at peptic ulcer perforation in the Emergency operating room have been punctuated by several factors at our institute.

To date, surgery is the only potentially curative therapy for patients with primary, resectable GIST. Prognosis depends on several factors including size of the primary lesion, mitotic activity, metastatic disease at diagnosis etc. perforation of the tumor, necrosis within the tumor are markers of worsened prognosis (4) (7). Nonmetastatic GISTs greater than 2 cm should be resected. Tyrosine kinase inhibitor therapy has significantly improved overall survival in patients with advanced disease and should be continued indefinitely. Prior to the development of imatinib, recurrences were common even in patients undergoing surgery. Adjuvant imatinib for 3 years should be considered in patients undergoing resection for primary disease (7).

Conclusion

In conclusion, GISTs rarely present as acute abdomen. If guided by an abdominal mass with diffuse peritonitis, the possibility of jejunal GIST perforation should be considered, even though it is extremely rare. A high degree of suspicion is necessary in view of the high morbidity rates resulting from a delayed diagnosis of the disease.

Competing Interests:
The authors declare none of financial or nonfinancial interests.

Authors contribution
AP collected the information, kept records and prepared the initial manuscript. SR did literature review and helped in finalizing the manuscript. SA supervised and finalized the manuscript. All authors read and approved the final manuscript.

Bibliography


