Journal of the Postgraduate Institute of Medicine 2025; 12(2): E269 1-4

https://doi.org/10.4038/jpgim.8555

Case Report

Jejunal gastrointestinal stromal tumor (GIST): Presenting as perforation and peritonitis – A Case Report

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Key words: **Jejunal gastrointestinal stromal tumor, GIST, perforation, peritonitis**

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Received: 05 Feb 2025, Accepted revised version: 14 Mar 2025, Published: 02 Sep 2025

Competing Interests: Authors have declared that no competing interests exist

Introduction

Gastrointestinal stromal tumors (GIST) are rare mesenchymal neoplasms of the GI tract, accounting for 0.5 -1% of all GI tumors [1]. Believed to originate from Cajal's cells, important in regulating gastrointestinal motility, these tumors mainly appear in the stomach (60–70%) and, to a lesser extent, in the small intestine (20–25%). Most GISTs are benign but their clinical presentation can vary. The most common symptom is intestinal bleeding, occurring in about 40% of cases. However, there are instances of critical emergencies, such as the rare jejunal GIST perforation leading to acute diffuse peritonitis [5]. GISTs are morphologically classified into spindle cell, epithelioid cell and mixed cell types. Diagnosis relies heavily on histopathological examination, particularly the detection of the CD117 protein. The pathogenesis of GISTs involves crucial molecular pathways, with genetic mutations in oncogenes like KIT and PDGFRA playing a significant role in tumor growth and progression. These insights are essential for targeted treatment approaches, which have notably improved outcomes in advanced cases.

Here we report a case of a 63-year-old male presenting with peritonitis secondary to a ruptured jejunal GIST.

Case presentation

A 63-year-old male, with previously known chronic obstructive pulmonary disease (COPD) presented to our emergency treatment unit with a 3-day history of fever associated with chills and rigors, lower abdominal pain and vomiting.

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On admission, the patient showed signs of septicaemia with low blood pressure and tachycardia. His WBC was14000/mm3 and CRP was 80mg/L with a normal haemoglobin level. Initially, he was resuscitated with intravenous fluids and started on intravenous broad-spectrum antibiotics. He underwent an urgent contrast enhanced computed tomography of abdomen and pelvis (CECT AP) and was found to have a 7.1x7.2x7.5cm sized, heterogeneously enhancing bowel mass which was likely to be arising from the distal jejunal or proximal ileal loops in the pelvic cavity. It was associated with regional infiltration into adjacent small bowel loops, fat stranding, a moderate amount of free fluid and pneumoperitoneum. CT features were highly suggestive of a small bowel mass with bowel perforation and acute peritonitis.

The patient underwent emergency exploratory laparotomy and, intra-operatively, a large jejunal tumor with perforation and peritonitis was found. The tumour was resected with adequate margins and end to end anastomosis was done. The patient was sent to the ICU for further post operative management. While in the ICU the patient developed an aspiration pneumonia but recovered fully on post op day 16. He was discharged from our ward post op Day 21.

Histology revealed a neoplastic lesion composed of spindle to elongated cells with minimally pleomorphic nuclei arranged in short intersecting fascicles. Many congested blood vessels were present, and tumor necrosis was evident. All the findings were in favor of GIST and immunohistochemistry for CD-117 confirmed the diagnosis. The patient was sent for further oncological management and was started on adjuvant chemotherapy with imatinib to be continued for 2 years. The patient was reviewed on post op Day 60 and was doing well without any post-surgical complications.

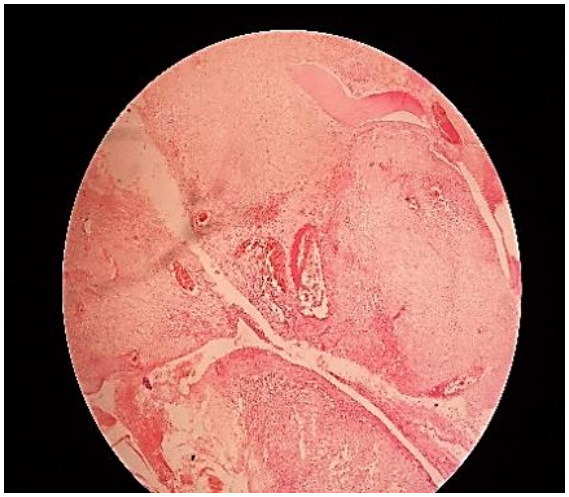

Figure 1: Microscopic specimen shows sub serosal lesion with spindle shaped short fascicles and nuclear pleomorphism

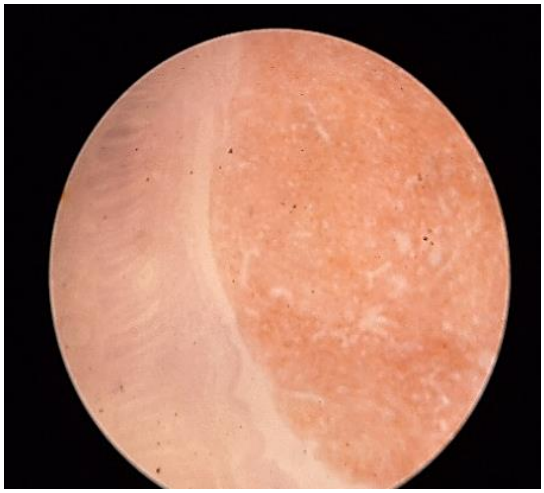

Figure 2: Shows Positive CD117 in IHC

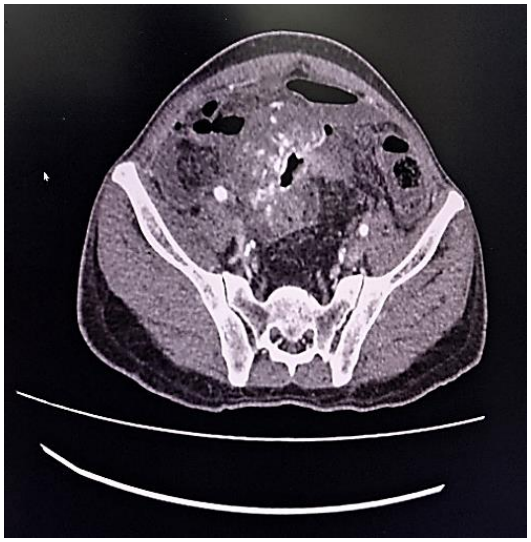

Figure 3: CECT image showing the ruptured tumor

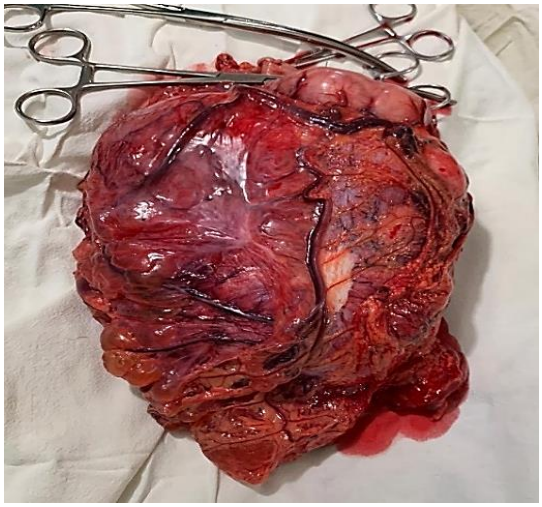

Figure 4: Macroscopic specimen of the GIST

Discussion

GISTs represent the commonest mesenchymal tumors of the GIT, accounting for 1-2% of all GIT Tumors. Jejunal GISTs are rare and only account for 5-10 % of them (1). GISTs are asymptomatic in 18% of cases, especially in smaller tumors [2]. Large tumors may obstruct the lumen by endophytic growth or by exophytic growth, obstructing the lumen from outside causing dysphagia, obstructive jaundice or constipation depending on the site. They also can present as a perforated mass causing peritonitis (as in this case) or with major GI hemorrhage [3].

Understanding the molecular basis of GISTs is crucial, as these neoplasms are driven by specific genetic mutations and alterations in signaling pathways. The interplay of these molecular factors not only influences the tumor's location and development but also its clinical behavior, including rare presentations like perforation. Jejunal GISTs are unique in their clinical presentation and prognosis due to their rarity and tendency for severe complications. Perforation of GISTs, particularly in the jejunum, is most often spontaneous and associated with a poor prognosis [4].

There are three types of GIST rupture described in the literature: closed perforation (abscess type), haemoperitoneum resulting from rupture of the hematoma capsule in the tumor (hemoperitoneum type), and free perforation (bowel perforation type). The latter, which includes cases like the jejunal GIST perforation in our patient, is the rarest and may result from obstruction with increasing intraluminal pressure or tumor erosion, leading to mural necrosis [4]. Adjuvant therapy for GIST has a place in the management and imatinib is found to be highly effective [6].

Conclusion

This case report of a patient with jejunal GIST perforation highlights the complexities and different presentations of gastrointestinal stromal tumors. The case highlights the importance of considering GISTs in the differential diagnosis, even in atypical presentations like acute diffuse peritonitis. Our case emphasizes the role of advanced imaging and histopathological analysis, including immunohistochemistry, in accurately diagnosing GISTs. The successful outcome in this case due to early diagnosis and surgical intervention, careful follow-up and adjuvant therapy with imatinib, demonstrates the potential for favorable prognosis even in rare and severe cases of GISTs when managed promptly and appropriately.

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