PERFORATED JEJUNAL GASTROINTESTINAL STROMAL TUMOR: A RARE PRESENTATION WITH PERITONEAL CARCINOMATOSIS

Patricia Palanca, DO; Adelso Tejada Jackson, MD; Judy Wong, MD; Scott Walter, MD; Brent Hochstein, DO; Christopher Abdo, MD; Nathan Kang, DO; Muhammad T Hassan, MS; Maheen Ali Khan, MS; Umair Haider, MD; Craig Biegel, DO; Dmitriy Kim, DO, FACS; Saint John's Episcopal Hospital; Ross University School of Medicine

Introduction

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract and account for 0.2% of all gastrointestinal neoplasms. Though most GISTs occur in the stomach, jejunal GIST is extremely rare and accounts for less than 0.04% of all GI tumors. These tumors often present with nonspecific symptoms, causing the diagnosis to be made when complications such as perforation or peritonitis develop. We present a case of perforated jejunal GIST.

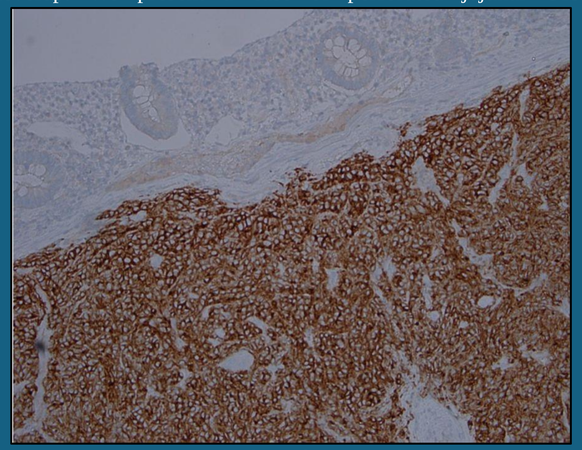


Figure 1. Histology of GI stromal tumor.

Case Report

A 69-year-old male with significant co-morbidities including coronary artery disease, diabetes mellitus, and gastric carcinoma was admitted from the nursing home for acute abdominal pain, hypotension, and melena. On arrival, he was hypotensive and febrile with laboratory results showing leukocytosis, anemia, and elevated C-reactive protein levels. Contrast-enhanced CT scan evidenced pneumoperitoneum, extensive peritoneal carcinomatosis with abnormal soft tissue in mesentery and omentum, and three large peritoneal masses. Patient subsequently underwent emergency exploratory laparotomy that demonstrated a perforated jejunal mass approximately 125 cm distal to the Ligament of Treitz. A resection of the perforated segment was performed, followed by a side-to-side anastomosis. Histopathological findings showed a high-grade GIST, with spindle and epithelioid cell morphology. Immunohistochemistry results were positive for CD117, DOG1, and CD34; the mitotic rate was above 5 mitoses per 5 mm^2, and all margins were tumor-positive. Final stage determined to be pT3N1M1. Postoperatively, the patient remained in critical condition. The prognosis remained poor due to the advanced stage of his disease, and the patient expired a few days after the surgery.

Discussion

Perforation, though rare, is a serious complication arising in GISTs and, as a rule, is followed by peritonitis and the diffuse spread of the tumor within the abdomen. Jejunal GISTs, with KIT exon 11 mutations, are well described to pursue an aggressive course, with a high rate of recurrence following surgical resection of the tumor. The presence of extensive peritoneal carcinomatosis, coupled with positive margins after the surgical removal of the perforated segment, considerably worsened the prognosis. The tumor was then staged as pT3N1M1, reflecting the advanced nature of the disease. High-risk GISTs receive Imatinib, recommended postoperatively in cases presenting residual or metastatic disease.

Conclusion

This case illustrates the aggressiveness of jejunal GISTs, particularly in the presence of perforation and peritoneal carcinomatosis. Surgical resection remains the cornerstone of treatment; however, poor prognosis is seen in advanced-stage disease with positive margins. Adjuvant therapy with Imatinib is of utmost importance in managing residual disease.

References

- Shoji M, Yoshimitsu Y, Maeda T, Sakuma H, Nakai M, Ueda H. Perforated gastrointestinal stromal tumor (GIST) in a true jejunal diverticulum in adulthood: report of a case. Surg Today. 2014 Nov;44(11):2180-6. doi: 10.1007/s00595-013-0732-0. Epub 2013 Sep 20. PMID: 24052432.
- Arata R, Nakahara H, Urushihara T, Itamoto T, Nishisaka T. A case of a diverticulum-like giant jejunal gastrointestinal stromal tumour presenting with intraperitoneal peritonitis due to rupture. Int J Surg Case Rep. 2020;69:68-71. doi: 10.1016/j.ijscr.2020.03.017. Epub 2020 Apr 1. PMID: 32283516; PMCID: PMC7154945.
- Dev S, Luitel P, Paudel S, Shrestha A, Dev B, Sah J. Acute abdomen due to perforated jejunal gastrointestinal stromal tumor: A case report. Int J Surg Case Rep. 2024 Aug;121:109922. doi: 10.1016/j.ijscr.2024.109922. Epub 2024 Jun 21. PMID: 38908159; PMCID: PMC11637173.