Asian Journal of Case Reports in Surgery

6(2): 11-14, 2021; Article no.AJCRS.64186

Ruptured Jejunal Gastrointestinal Stromal Tumor- An Unusual Presentation of a Rare Tumor

Nikhil Gupta[1], Raghav Yelamanchi[1*], Ekta Yadav[1] and C. K. Durga[1]

[1]Department of Surgery, Atal Bihari Vajpayee Institute of Medical Sciences and Dr. Ram Manohar Lohia Hospital, New Delhi, Pin-110001, India.

Authors' contributions

This work was carried out in collaboration among all authors. Authors NG & RY provided conceptualization, Investigation, data curation and wrote the original draft of the manuscript. Author EY performs investigation, data curation, review and edited the manuscript; Author CKD supervised the whole study and helped in write, review & edited the manuscript. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Ashish Anand, GV Montgomery Veteran Affairs Medical Center, USA.
Reviewers:
(1) Eduardo Blanco Faramiñán, Finis Terrae University, Chile.
(2) G. N. K. Suresh Babu, Prince Dr. K. Vasudevan College of Engineering and Technology, India.
Complete Peer review History: http://www.sdiarticle4.com/review-history/64186

Case report

Received 25 October 2020
Accepted 30 December 2020
Published 15 January 2021

ABSTRACT

Aim: We report an extremely rare case of jejunal GIST presenting with acute abdomen and shock due to tumor rupture with hemorrhage.

Case Presentation: A 52 year male presented to the surgical emergency with complaints of acute onset abdominal pain and abdominal distention for 12 hours. On abdominal examination, there was a tender ill defined lump in the umbilical region associated with free fluid. Hemoglobin was 5.4 gm/dl. Ultrasound guided diagnostic peritoneal aspiration revealed blood. Patient underwent exploratory laparotomy with resection and anastomosis of the jejunal segment containing GIST.

Discussion: GISTs (Gastrointestinal stromal tumors) are one of the rare tumors of the gastrointestinal tract. Jejunum is less common site for the origin of GIST accounting for <10% of all gastrointestinal tumors. Ruptured small intestinal GIST is an extremely rare cause of acute abdomen.

Conclusion: The above case report is a very atypical scenario of small bowel GIST presenting with an acute abdomen. Early operative intervention is required to control bleeding and to prevent sepsis in cases of ruptured GIST.

**Corresponding author: E-mail: raghavyelamanchi@gmail.com;*

Keywords: Gastrointestinal stromal tumor; Jejunal GIST; small bowel tumors; Imatinib mesylate.

ABBREVIATION

GIST: Gastrointestinal stromal tumors

1. INTRODUCTION

GISTs are one of the rare tumors with an incidence of 2 per 1,00,000 [1]. They arise from the interstitial cells of Cajal which are mesodermal in origin and regulate the peristalsis of the gastrointestinal tract. The stomach is the most common site for GIST accounting for two-thirds of the cases. The jejunum is less common site for the origin of GIST accounting for <10% of all gastrointestinal tumors [1]. About 10-30% of GISTs have malignant potential [2]. The neoplastic cells have increased tyrosine kinase activity resulting from mutation of either c-KIT gene or platelet derived growth factor α (PDGFRα) gene [3]. The incidence of GISTs increases with age with 90% of cases diagnosed in adults above the age of 40 years with a slight male predominance [3,4].

2. CASE REPORT

A 52 year male presented to the surgical emergency with complaints of acute onset abdominal pain and abdominal distention for 12 hours. There was no history of trauma, fever or any other symptoms. On examination, patient was pale and tachycardic with cold peripheries. Blood pressure was 80/50 mm of Hg. On abdominal examination, there was a tender ill defined lump in the umbilical region associated with free fluid. Hemoglobin was 5.4 gm/dl. Ultrasound guided diagnostic peritoneal aspiration revealed blood. Patient was resuscitated with intravenous fluids and blood and was taken up for emergency laparotomy.

On laparotomy, there was about 800 ml of blood in the peritoneal cavity. There was a 10 x 8 cms exophytic growth arising from the jejunum about 3 feet distal to the duodenojejunal junction [Fig. 1]. The growth was perforated with active bleeding and involved the adjacent mesentery. The rest of the intra-abdominal organs and peritoneum were examined for any metastasis. As the growth was localized, it was resected with a 5 cm proximal and distal bowel margin with anastomosis of the resected ends. The patient had an uneventful recovery in the post-operative period. Histopathology report of the growth was suggestive of gastrointestinal stromal tumor (GIST) with a high mitotic index. Patient was started on imatinib mesylate 400 mg daily. After one year of follow up, patient is performing well with no radiological evidence of any recurrence.

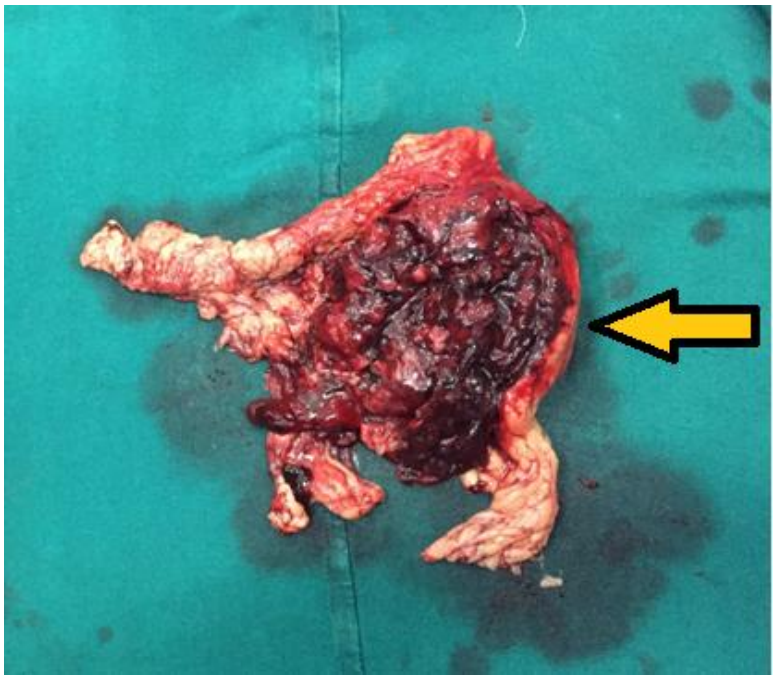

Fig. 1. Resected specimen of the tumor

3. DISCUSSION

Small GISTs less than 5 cms in size are indolent and asymptomatic. They are usually discovered incidentally during imaging done for unrelated symptoms. Symptoms arise when GISTs attain sufficient size even though many case reports of small GISTs causing symptoms are reported. Symptoms include gastrointestinal bleeding due to pressure necrosis of the overlying mucosa which may be acute or chronic leading to anemia. Other presenting symptoms include obstruction due to mass effect over the surrounding bowel loops, abdominal lump and abdominal pain. Ruptured small intestinal GIST is an extremely rare cause of acute abdomen with only a handful of cases reported so far in the literature [5,6]. Tumor rupture may lead to torrential hemorrhage as these are highly vascular tumors.

Imaging is the mainstay for the preoperative diagnosis of GIST. They appear as heterogeneously enhancing lesions on contrast enhanced computerized tomography (CECT) due to internal hemorrhage and necrosis. Small GISTs are frequently missed on imaging. Endoscopic techniques are futile for the diagnosis of GISTs as they are predominantly exophytic tumors. However, there are several case reports where small bowel GISTs are diagnosed on small bowel endoscopy done for gastrointestinal bleed [7]. Preoperative pathological diagnosis is not mandatory for suspected GIST. In most of the cases, the tumors are not amenable for image guided biopsy due to overlying bowel loops. Other soft tissue tumors of the small bowel are one of the close differential diagnosis for GIST.

Surgery is the mainstay of treatment for non-metastatic GIST. Prophylactic lymph node dissection is not recommended as GISTs spread predominantly through the hematogenous route. The most common sites for metastasis include the liver and the peritoneum. Neoadjuvant chemotherapy with imatinib mesylate may be considered for non-resectable tumors to downsize the tumor. Immunohistochemical markers such as CD117 and CD34 will help in the histopathological diagnosis of the tumor [3]. Tumor size and mitotic rate play a crucial role in deciding the need for adjuvant therapy. Imatinib mesylate is the most commonly used tyrosine kinase inhibitor in the adjuvant setting for GIST. The dose and duration of the drug will depend on the histopathological and genetic characteristics of the tumor [8].

4. CONCLUSION

To conclude, the above case report is a very atypical scenario of small bowel GIST presenting with an acute abdomen. Early operative intervention is required to control bleeding and to prevent sepsis in cases of ruptured GIST. Ruptured GIST requires adjuvant chemotherapy with imatinib mesylate as there are high chances of tumor recurrence in the follow-up period.

CONSENT

Informed **c**onsent was taken from patient for publication.

ETHICAL APPROVAL

Authors declare that they have followed the principles of the Declaration of Helsinki 1975 modified in 2008.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Kramer K, Siech M, Sträter J, Aschoff AJ, Henne-Bruns D. Hämorrhagischer schock bei doppelmalignom: Gastrointestinaler Stromatumor (GIST) und neuroendokrines Karzinom bei morbus recklinghausen [GI hemorrhage with fulminant shock induced by jejunal gastrointestinal stromal tumor (GIST) coincident with duodenal neuroendocrine carcinoma (NET) + neurofibromatosis (NF) -- Case report and review of the literature]. Z Gastroenterol. 2005;43(3):281-88.
DOI: 10.1055/s-2004-813810
2. Choi H. Response evaluation of gastrointestinal stromal tumors. Oncologist. 2008;13(2):4-7.
DOI: 10.1634/theoncologist.13-S2-4
3. Sornmayura P. Gastrointestinal stromal tumors (GISTs): A pathology view point. J Med Assoc Thai. 2009;92(1):124-35.
PMID: 19260254.
4. Sankey RE, Maatouk M, Mahmood A, Raja M. Case report: Jejunal gastrointestinal stromal tumour, a rare tumour, with a challenging diagnosis and a successful

treatment. J Surg Case Rep. 2015;2015(5):rjv050.
DOI: 10.1093/jscr/rjv050

5. Hirasaki S, Fujita K, Matsubara M, Kanzaki H, Yamane H, Okuda M, et al. A ruptured large extraluminal ileal gastrointestinal stromal tumor causing hemoperitoneum. World J Gastroenterol. 2008;14(18):2928-31.
DOI: 10.3748/wjg.14.2928

6. Attaallah W, Coşkun Ş, Özden G, Mollamemişoğlu H, Yeğen C. Spontaneous rupture of extraluminal jejunal gastrointestinal stromal tumor causing acute abdomen and hemoperitoneum. Ulus Cerrahi Derg. 2015;31(2):99-101.
DOI: 10.5152/UCD.2015.2877

7. Dualim DM, Loo GH, Rajan R, Nik Mahmood NRK. Jejunal GIST: Hunting down an unusual cause of gastrointestinal bleed using double balloon enteroscopy. A case report. Int J Surg Case Rep. 2019;60:303-06.
DOI: 10.1016/j.ijscr.2019.06.053

8. Balachandran VP, DeMatteo RP. Gastrointestinal stromal tumors: Who should get imatinib and for how long? Adv Surg. 2014;48(1):165-83.
DOI: 10.1016/j.yasu.2014.05.014

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/64186