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A Large Twisted Mesenteric Gist-Presenting as an Acute Abdomen- A Rare Case Report

T Lirangla Sangtam, Raghav Garg, Praveen M S Naik

Department of General Surgery, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi -110029, India

Abstract: ***Introduction: Gastrointestinal stromal tumors are the most common mesenchymal tumors of the digestive tract, and can arise from any part of the GI tract , frequently found in the stomach ~ 50%, followed by small intestine (25%) small intestine[1] , however, very rarely, may also occur in extraintestinal locations such as omentum, mesentery, pelvis or retroperitoneum[2]. GISTs present as a wide spectrum of disease with certain clinicopathological features such as tumor size, mitotic activity and site of origin; determining the aggressiveness of the disease. While surgery is the mainstay of the treatment in localized GISTs, achieving cure rate in 45-60 % cases [3]; Imatinib mesylate, a tyrosine kinase inhibitor (TKI) is an important drug for targeted therapy in locally advanced and metastatic GISTs and also in specific genotypes of GISTs. Case Presentation: We hereby report a rare case of an elderly 68 year old , Muslim woman with unusual presentation of a twisted mesenteric GIST in an emergency department, who presented with acute abdominal pain, vomiting, abdominal distension and obstipation. Emergency exploration was done in view of signs of peritonitis. A huge mass, origination from jejunal mesentery with areas of necrosis and ischaemia seen. The mass was resected along with adjacent bowel and anastomosis of the bowel with repair of mesenteric defect done and resected specimen sent for histopathological examination. A diagnosis of mesenteric GIST was confirmed on histopathological report. Discussion: While case of GISTs of stomach, small bowel and other GI tract are encountered infrequently in a patient on elective work up with vague abdominal symptoms; A GIST arising from mesentery, presenting as an acute abdomen isa rare clinical scenario. In this case report, we want to emphasize on the potentiality of a large mesenteric GIST which may undergo torsion and may present as an acute abdomen necessitating emergency exploration and hence differential diagnosis may be kept in mind. Conclusion: A large mesenteric GIST may undergo torsion with subsequent ischaemia and necrosis; and may present as an acute abdomen in the Emergency department. Complete excision of the cyst with wide margin must be performed with histopathological examination of the resected specimen and adjuvant therapy with Imatinib considered, if tumor aggressiveness is suggestive; as indicated by the clinicopathological and genetic profiling.***

Keywords: Mesenteric GIST,torsion, Acute abdomen , CD117, Adjuvant Imatinib.

1. Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the GI tract , with about 50% found arising from the stomach [1] , 25% in the small bowel and very rarely may occur in the extra intestinal locations; such as omentum, mesentery, retroperitoneum or pelvic cavity, as seen in our case. Average age of presentation is 60-70 years with no sex predilection [4], these tumours have variable clinical and biological behavior. Unlike other gastro-intestinal carcinomas, lymph node metastasis is rare and malignant forms spread usually via hematogenous route [5]. The mainstay of treatment for localized GISTs is complete surgical resection without regional lymphadenectomy; where asin locally advanced or metastatic GISTs, a tyrosine kinase receptor, Imatinib Mesylate (IM) is the first line of therapy[6].

2. Case Presentation

An elderly Muslim lady of 68 years old, presented in the emergency ward with complaints of acute onset severe abdominal pain and distension since, 48 hours,which was initially located in the lower abdomen and later spread to the whole abdomen. She had associated history of bilious vomiting, fever and obstipation. There was no history of malena, no urinary symptoms, bleeding per rectum or bleeding per vagina. She had attained menopause at the age of 48 and had with three children born by normal vaginal delivery. There was no history of previous abdominal surgery and had no other morbidity.

On examination, she was noted to be dehydrated, tachycardia with HR-116/minute and hypotensive with BP of 96/50 mm of Hg. She was febrile and chest auscultation revealed bilateral decreased air entry with coarse crepitation. Abdominal examination revealed gross distension, with generalized tenderness, guarding and rigidity. Deep palpation was avoided due to severe pain. Bowel sounds were absent. Per rectal and vaginal examination did not reveal any abnormality and all hernia sites were normal.

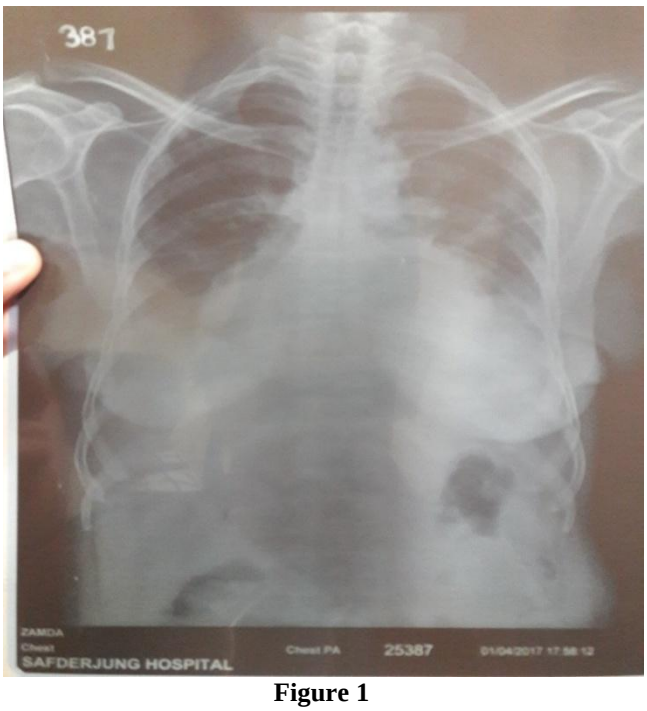


Figure 1

Immediate fluid resuscitation was done, nasogastric tube inserted, blood investigations sent and electrolyte correction started. Her Hb was 8.3 g/dl with leukocytosis of 16,800/cu mm. LFT and KFT were found to be normal. X-Ray chest showed bilateral mild pleural effusion (figure.1). plain X-Ray abdomen did not reveal any significant air fluid level or gas under diaphragm. Ultrasound whole abdomen suggested dilated bowel loops with features suggestive of intestinal obstruction. Provisional diagnosis of perforation peritonitis, sigmoid Volvulus and twisted Ovarian cyst was made and exploratory laparotomy was performed.

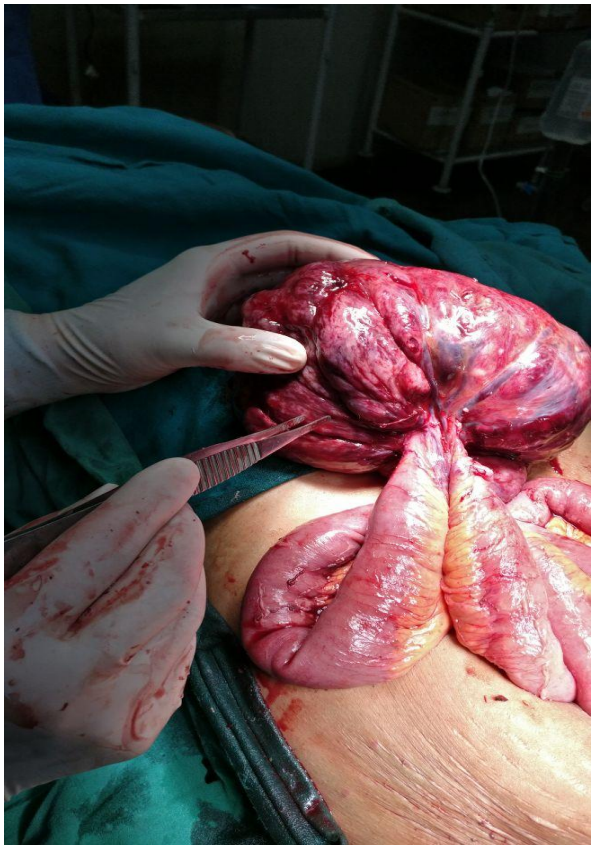


Figure 2

Intraoperatively, 500 ml of dark -brown, foul smelling, toxic fluid was drained from the peritoneal cavity, bowel loops were found to be dilated. A large, soft solid mass of 15 x 10x 8 cm was found arising from the small bowel mesenteryin close proximity to jejunal wall (figure.2). Dilated vascular channels extending from the mesentery to the mass was found to be twisted at the stalk . External surface appeared variegated and congested with areas of necrosis and haemorrhage (figure.3). Intraoperatively,a diagnosis of soft tissue tumor or possible mesenteric cyst was made. Complete resection of the mass along with adjacent bowel and jejunal anastomoses with repair of mesenteric defect done. Liver surface was found to be smooth, omentum appeared congested but no visible deposits detected. No growth was palpated in the colon or uterus. Bilateral ovaries were found to be normal in size. Peritoneal cavity was then washed with warm saline and abdominal drain placed in the pelvic cavity. Resected specimen was then sent for histopathological examination.

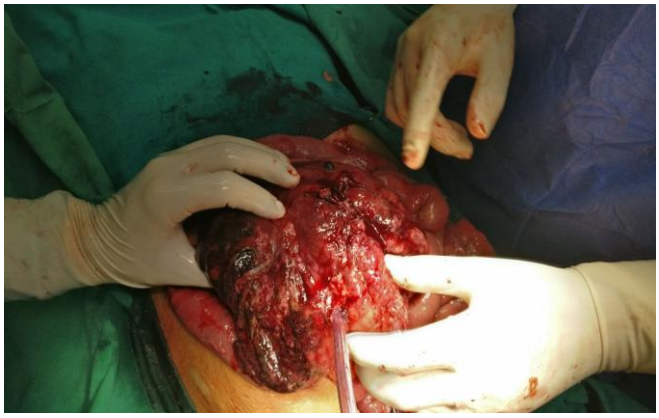


Figure 3

Postoperatively, patient was shifted to ICU and kept on ventilatory support for 24 hours due to poor respiratory function, later shifted to ward. Nasogastric Ryles tube was removed on 4th postoperative day after return of bowel sounds. Liquid feed was started by enteral route on 5thday followed by removal of abdominal drain. Patient was discharged on 9th postoperative day after respiratory condition improved with advice for follow up after a week.

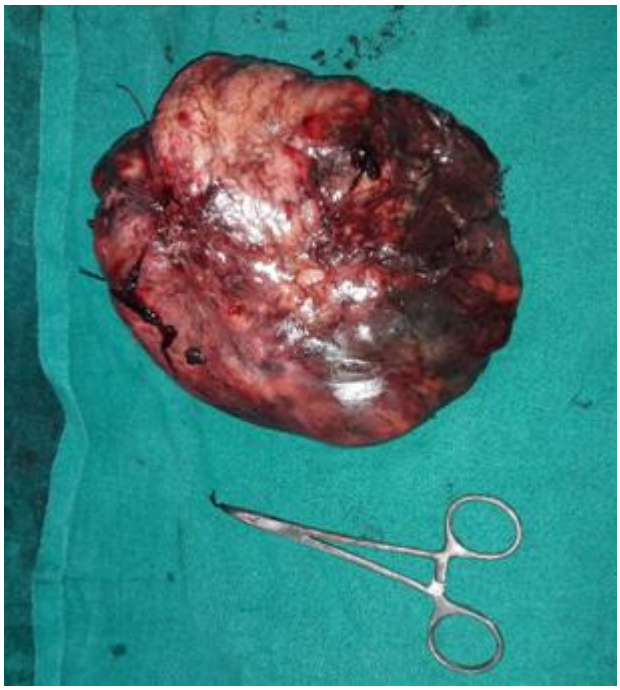
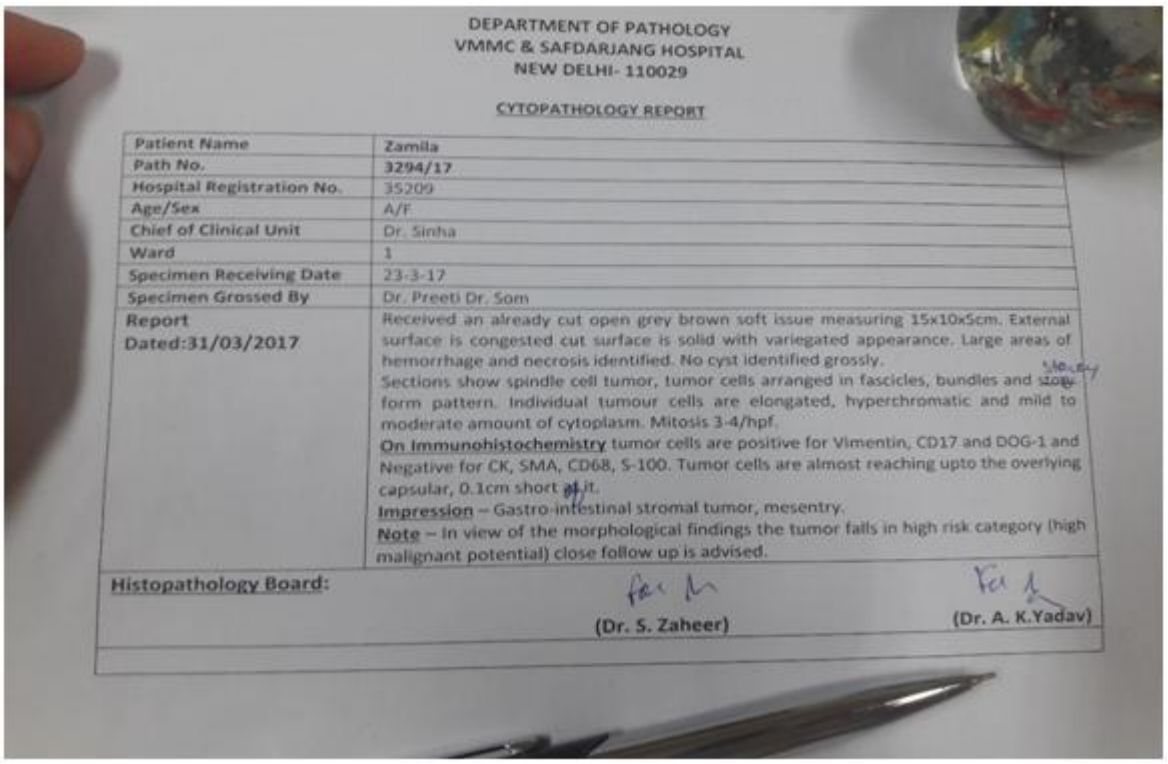


Figure 4

Histopathological report(figure.5)confirmed spindle cell tumor, hyperchromatic with mitosis of 3-4/50HPF. On immunohistochemistry, tumor cells were found positive for vimentin, CD117 and DOG1. Based on the clinicopathological evidence of large sized tumor of 15 x 10 x 8 cm and positive immunohistochemistry for GISTs, tumor fall under high risk category for aggressiveness; hence, patient was referred to medical oncology department for consideration of adjuvant Imatinib therapy.



DEPARTMENT OF PATHOLOGY
VMMC & SAFDARJANG HOSPITAL
NEW DELHI- 110029

CYTOPATHOLOGY REPORT

Patient Name	Zamila
Path No.	3294/17
Hospital Registration No.	35209
Age/Sex	A/F
Chief of Clinical Unit	Dr. Sinha
Ward	1
Specimen Receiving Date	23-3-17
Specimen Grossed By	Dr. Preeti Dr. Som
Report Dated:31/03/2017	Received an already cut open grey brown soft issue measuring 15x10x5cm. External surface is congested cut surface is solid with variegated appearance. Large areas of hemorrhage and necrosis identified. No cyst identified grossly. Sections show spindle cell tumor, tumor cells arranged in fascicles, bundles and storiform pattern. Individual tumour cells are elongated, hyperchromatic and mild to moderate amount of cytoplasm. Mitosis 3-4/hpf. On Immunohistochemistry tumor cells are positive for Vimentin, CD17 and DOG-1 and Negative for CK, SMA, CD68, S-100. Tumor cells are almost reaching upto the overlying capsular, 0.1cm short of it. Impression – Gastro-intestinal stromal tumor, mesentry. Note – In view of the morphological findings the tumor falls in high risk category (high malignant potential) close follow up is advised.

Histopathology Board:

(Dr. S. Zaheer) (Dr. A. K.Yadav)

Figure 5

3. Discussion

GISTS represents the most common primary mesenchymal tumor of the GI tract, but may also be seen in extravisceral locations[2], as seen in our case, which was found arising from the small bowel mesentery. They have a characteristic morphology which are generally positive for *CD117(c- KIT)* and are primarily caused by activation of mutation in *KIT* and *PDGFRA* genes[2].

These tumors have variable clinical and biological behavior, ranging from small benign tumors to aggressive forms that have dismal prognosis[2]. They have no sex predilection and average age of presentation is 60-70 years. In our case was an elderly lady of 68 years old. Despite the huge size of the tumor, she had no previous abdominal symptoms except vague discomfort until it underwent torsion. unlike other gastrointestinal carcinomas, lymph node metastases rarely occur in GISTs [5], hence complete surgical resection without regional lymphadenectomy is the mainstay of treatment in a localized GISTs [2]. In our case, the tumor, though large in size and lying in close proximity to the adjacent jejunum, was freely mobile, with no evidence of liver metastasis or mesenteric lymphadenopathy. Hence, resection of the tumor with adjacent jejunal segment was adequate.

Since, the patient was an elderly female, initial clinical suspicion was that of a twisted ovarian cyst or a possible sigmoid volvulus, as her abdomen was grossly distended with emergency ultrasound also suggesting bowel obstruction; but on exploration, both ovaries were found to be normal and no obvious colonic volvulus. Intraoperatively, atwisted mesenteric cyst could not be grossly differentiated from that of soft tissue tumor,as it had undergone ischaemic changes with areas of necrosis and haemorrhage.

Diagnosis was made on histological and immunohistochemical analysis, which was suggestive of mesenteric GIST. The diagnosis of GISTs relies on the histological findings of spindle-shaped cells proliferation in 70% cases and epitheloid cells in 20% [5] and on immunohistochemical studies, often showing positive for *KIT (CD117*) and *CD34*,variabily positive for *SMA*(smooth muscle actin), ocassionally positive for *desmin* and *vimentin* and rarely *S100* in small bowel GISTs [7] , As in our case which showed positive for *CD117, Vimetin, CD34* and *DOG1*.

GISTs have a complex biological behavior and their risk of malignant potential is difficult to predict. However, National Institute of Health (NIH) consensus criteria proposed in 2001 suggested two important histological features, namely: mitotic count and size of the tumor[8]. Joensuu [9] modified the criteria and included site of tumor and tumor rupture in the criteria. Tumor size in our patient was more than 10 cm which have high risk of malignant potential, hence surveillance with imaging may help in timely detection of relapse along with adjuvant imatinib. There is no well established guidelines for the frequency of surveillance imaging in GISTs. In a low risk tumors,resection is usually sufficient and do not require adjuvant Imatinib therapy. In a high risk GISTs, ESMO guidelines suggest imaging at an interval of 3-6months for the initial 3 years during adjuvant imatinib , followed by 3monthly in the next 2 years, then 6monthly after cessation of imatinib [10]

With the introduction of Imatinib Mesylate, the median survival of patients with advanced GISTs have increased from 18 to more than 60 months [11,14]. Imatinib,a *KIT, PDGFRA* and *ABL* kinase inhibitor given at a dose of 400 mg daily is the standard first line of treatment in a patient of unresectable or metastatic GISTs[1,3].

GISTs tumor harboring a *KIT exon 11* mutation seen in 70% cases, exhibit excellent response to Imatinib therapy, a tyrosine kinase inhibitor (TKI) and may allow subsequent Ro resection in 83% of cases with a median overall survival of 104 months, when given in a neoadjuvant setting in selective locally advanced GISTs [12]. A second line therapy with sunitinib (SU) can be given in cases unresponsive or resistant to Imatinib [13]

4. Conclusion

In an elderly patient with acute abdomen, a torsion of mesenteric GIST may also be a possibility. If encountered, must be completely resected with adequate wide margin and confirmed by histopathological examination and surveillance must be done for appropriate management; either by interval imaging or targeted therapy with Imatinib depending on genetic profiling and risk of malignant potential.

5. Consent

Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

6. Conflict of Interest

The authors have no conflicts of interest to declare.

7. Source of Funding

None

8. Author Contribution

All the authors have contributed significantly for the article.

9. Guarantor

R.K Soni

10. Ethical Approval

not required

11. Acknowledgement

none

References

[1] Nuria Mulet- Margalef and Xavier Gracia- Del- Muro. Sunitinib in the treatment of Gastrointestinal tumor: patient selection and perspectives .2016 Dec17. PMCID: PMC5171199
[2] Andrés Povedaa, Xavier García del Murob, Jose Antonio López-Guerreroc, Ricardo Cubedod, Virginia Martínez. GEIS guidelines for gastrointestinal sarcomas (GIST). http://doi.org/10.1016/j.ctrv.2016.11.011
[3] Supraclavicular lymphnode metastases from malignant gastrointestinal tumor of jejunum: a case report with literature. Chi Ma, Shao-Long Hao, Xin- Cheng Liu, Jin -Yao Nin, Guo-Chang Wu World J Gastroenterol.2017. march 14; 23(10)
[4] Bosman F.T, Carneiro F, Hruban R.H, Theise N.D. WHO classification of tumors of the Digestive system. 4th Tumours IWCo ; 2010.
[5] Coindre JM, Emile JF, Monges G, Ranchere- Vince D. Gastrointestinal stromal tumours : definition, histological, immunohistochemical, and molecular features and diagnostic strategy. Ann
[6] Gastrointestinal tumors who should get imatinib and for how long?Vinod Cancer Center. Clin Transl Oncol.2012 jul; 14(7):536-40. doi:10.1007
[7] Markku Meittinen , Leslie H S Sobin, Maarit Sarlomo-Rikal Immunochemical spectrum of GISTs at different sites and their differential diagnosis with their reference to CD117(KIT) Mod Pathol 2000 ; 30(10): 1134-1142
[8] Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ, et al. Diagnosis of gastrointestinal stromal tumors: a consensus approach. Hum Pathol. 2002;33:459–465. [PubMed] [Ref list]
[9] Joensuu H. Risk stratification of patients diagnosed with gastrointestinal stromal tumor. Hum Pathol. 2008;39:1411–1419.[PubMed] [Ref list]
[10] ESMO/European Sarcoma Network Working Group. Gastrointestinal stromal tumours: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. Ann Oncol. 2012;23(Suppl 7):vii49–vii55.[PubMed]
[11] Poveda A, Rivera F, Martin J ; SEOM guidelines for gastrointestinal stromal sarcomas
[12] Sabrina Rossi, Teresa Congedo, Riccardo Ricci, Maurizio Martini. Is surgery mandatory in locally advanced GISTs after imatinib ? A case report and literature review. Journal of gastrointestinal oncology. 2016.12.2
[13] Cytoreductive surgery in patients with metastatic gastrointestinal stromal tumor treated with sunitinib malate. Raut CP, Wang Q, Manola J, Morgan JA, Wagner AJ. Ann Surg Oncol.2010 feb;17(2):407-15
[14] DeMatteo RP. Lews JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF. Two hundred GISTs : recurrence patterns and prognostic factors for survival. Ann Surg. 2000; 231;51-58 [PubMed]