VOLUME - 11, ISSUE - 03, MARCH - 2022 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Surgery

A BIZARRE CASE OF GIANT JEJUNAL GIST PRESENTING AS A MALE ABDOMINOPELVIC MASS - CASE REPORT

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ABSTRACT Gastrointestinal stromal tumours(GISTs) are uncommon neoplasms, with 31.8% occurring in small bowel, and jejunal GIST being the rarest subtype among all types of GISTs, accounting for 0.1%–3% of all GIT tumours. GIST presenting as a palpable abdominopelvic mass, as is being described in this case report, is exceedingly rare and only 25 such cases have been previously reported in the world literature, with a higher female preponderance. Since this patient had a Giant GIST which was locally-advanced, it can be effectively treated with Imatinib for 3-6 months followed by surgery and adjuvant therapy, thereby improving disease-free survival.

KEYWORDS : Gastrointestinal Stromal Tumour (GIST), Jejunum, Imatinib, Neoadjuvant chemotherapy, Tyrosine kinase inhibitor

INTRODUCTION & BACKGROUND:

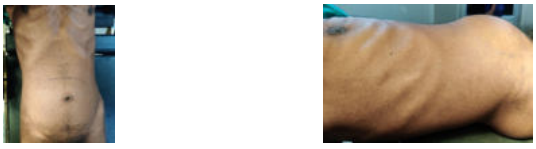
Gastrointestinal stromal tumours (GISTs) are uncommon tumours accounting for 1% of all gastrointestinal tract (GIT) neoplasms with an incidence of 2 in 1,00,000, and 14-20 cases per million population.[1–4] Most common location is gastric (55%) followed by the small bowel (31.8%), colon (6%), oesophagus (0.7%) and also occur in extraintestinal locations like mesentery and omentum. with jejunal GIST being extremely rare accounting for 0.1%–3% of all GIT tumours.(4–7) The tumour shows no strong sex predilection and usually occurs in adults, with a mean reported age of 50.6 years.[8]

GISTs are believed to arise from the interstitial cells of Cajal or related stem cells. Mutation of C-KIT oncogene with CD117 positivity is observed in 95% of cases. Platelet-derived growth factor receptor alpha (PDGFRA) mutations play a role in GIST pathogenesis.[2,9,10] Benign GISTs outnumber the malignant ones by a margin of 10:1. Clinical presentation depends on the site and size of the tumour and may include upper abdominal ulcer-like pain, dyspepsia, iron-deficiency anaemia, gastrointestinal bleeding, nausea, vomiting, palpable abdominal mass, and weight loss. However, small tumours may be asymptomatic.[11]

The signs and symptoms of GIST depend on the tumour's location and size. Gastrointestinal bleeding is one of the most common symptoms. GIST presenting as a palpable abdominal mass is extremely rare. At least 10 to 30% of GISTs are discovered incidentally during laparotomy, endoscopy, or other imaging studies, with 15% to 50% of GISTs presenting with metastatic disease.[12] GISTs initially presenting as an abdominal mass are exceedingly rare, and only 25 such cases have been reported in the world literature with a higher female preponderance.[13] 'Giant GIST' is tumour size >10 cm. This tumour has tendency to liver metastasis and peritoneal recurrence, however; the primarily lymph node involvement or metastasis is rare.[14] Imatinib mesylate is the first effective systemic treatment for advanced GISTs, and yields a benefit of 50%-80%.[15]

CASE REPORT:

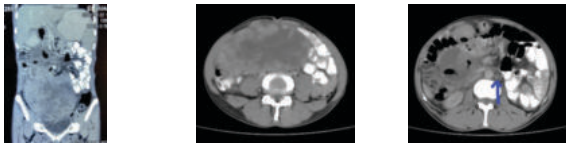
A 40-year-old male presented to our institution with complaints of abdominal pain for 15 days accompanied by fever for 2 days and non-bilious vomiting for 1 day. History of loss of appetite and loss of weight was present. Patient was moderately built and poorly nourished and Pallor was evident. On inspection, fullness was seen in lower abdomen, bilateral flanks were full. Hard, fixed, tender mass was palpable, extending 5 cm above umbilicus and lower border was not palpable as it extended into the pelvis. Dullness over the mass and peritumoral resonant tone was present. No shifting dullness was evident initially and developed following a week after hospital admission. Bowel sounds were present.



(a) (b)

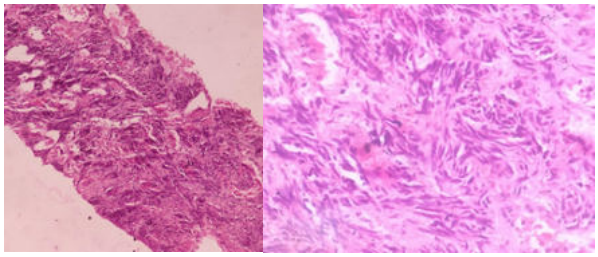
Fig.1: Clinical pictograph of the patient in Supine and Lateral views showing the abdominopelvic fullness.

Initial haematological investigations revealed severe anaemia. Basic metabolic panel and liver function tests and PT/INR were unremarkable. CECT of abdomen and pelvis showed heterogeneously enhancing necrotic soft-tissue density lesion(17.2×9.5×18.1cm) arising from jejunum, extending into the pelvis with multiple enlarged para-aortic nodes. USG-guided core biopsy showed Spindle cell neoplasm with Immunohistochemistry study testing positive for CD117 and negative for S100, CD34 and Desmin. Ascitic fluid aspirate analysis showed a SAAG of 3.6 (Transudate), without any malignant cells. Metastatic workup showed no disease elsewhere.



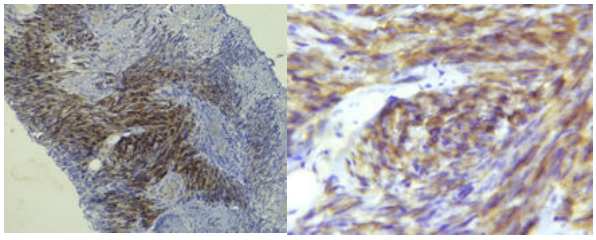
(a) (b) (c)

Fig.2: Contrast-enhanced Computed Tomography of Abdomen and Pelvis showing a heterogeneously enhancing necrotic soft-tissue density lesion(17.2×9.5×18.1cm) arising from jejunum, extending into the pelvis with multiple enlarged para-aortic nodes.



(a) (b)

Fig.3: Photomicrograph of Haematoxylin & Eosin staining showing GIST with fibrocollagenous tissue infiltrated by fascicles & bundles of spindle cells with congested blood vessels in intervening stroma and sparse areas of haemorrhage. Magnification: ×100(a) and ×400(b).



(a) (b)

Fig.4: Photomicrograph of Immunohistochemistry staining showing GIST positive for CD117. Magnification: ×100(a) and ×400(b).

After obtaining tumour board consensus, patient was started on Tab.Imatinib mesylate 400mg daily and is planned for response assessment after 3 cycles.

DISCUSSION:

Although GISTs are relatively uncommon (<1% of all gastrointestinal neoplasms) compared with adeno carcinoma, they are the most common mesenchymal tumours of the GIT. According to SEER data from 1992 to 2000, the estimated incidence in the USA is 6.8 per million, with a much higher incidence of occult micro-GISTs <1 cm.[16,17] GISTs presenting with palpable abdominal mass is rare and only 25 such cases have been published in world literature.[13]

Tumour size and the mitotic count are the most commonly used prognostic factors in addition to anatomic site, histologic variant, and type of mutation to estimate the outcome and risk of recurrence of the tumour, with those arising from the stomach having a more favourable prognosis compared to other sites.[18] Symptomatic patents are noted to have a higher incidence of high-risk tumours and metastases at presentation. When the size of the mass measures >10 cm, the GIST is referred to as giant GIST. The risk of malignancy of GISTs varies between very low and high based on the mitotic rate and size.[3,19]

Treatment strategy for GIST depends on the tumour site, size, and presence or absence of metastasis. Small GIST (less than 2 cm) can be conservatively observed or endoscopically resected. Surgical resection is the mainstay of therapy for a resectable, non-metastatic GIST, regardless of tumour location. The goal of surgery is to remove the tumour macroscopically with an intact pseudocapsule and microscopically, thereby giving an R0 resection. Lymph node dissection is routinely unnecessary because lymph node metastases are extremely rare. Organ– preserving and function– preserving surgery is oncologically allowed if negative resection margins can be achieved.[20,21] But if the GIST is large in size, unresectable or metastatic, shrinkage of the GIST can be done by giving Tyrosine kinase inhibitors(TKI) therapy prior to surgery with Imatinib at the standard dose of 400 mg daily, permitting conservative and less invasive R0 resection, especially for GIST located at the esophagogastric junction, duodenum and rectum. NCCN guidelines state that neoadjuvant chemotherapy is considered if surgical morbidity could be reduced by downstaging the tumour.[9,22]

There is no consensus on the use of neoadjuvant therapy for the treatment of GISTs. Seven trials have been done in which neoadjuvant chemotherapy was studied and utilized including those by RTOG (Radiation Therapy Oncology Group) 0132 and Kurokawa et al. which had similar results which showed that neoadjuvant therapy along with surgery may be beneficial long term. Jakob et al showed that the rate of the negative surgical margin in patients after neoadjuvant therapy was significantly higher than that in patients without neoadjuvant therapy. The optimal duration of neoadjuvant imatinib therapy appears to be 3– 6 months, as reported in a phase II trial of imatinib for unresectable GIST.[23–26]

It remains controversial as to which is the best modality to evaluate the response of GIST to TKI. CECT is useful to assess not only tumour size, but also tumour viability, evaluated by blood supply. Choi criteria, which consider CT density, are useful to evaluate the response to imatinib for GIST. 18F-FDG PET is recently trending for early evaluation of response (after 2-4 weeks) that can result in a change of treatment strategy in GIST patients treated with neoadjuvant intent.[27,28] However, evidence of safety and efficacy for preoperative imatinib treatment remains to be established.

CONCLUSION:

To conclude, GISTs are highly aggressive tumours. Locally-advanced (as in our case) and metastatic tumours, can be effectively treated with Imatinib for 3-6 months followed by surgery and adjuvant therapy for patients with intermediate- and high-risk tumours with R0 resection, improving disease-free survival. Being vigilant about such rare presentations can help guide the treating surgeon avoid futile laparotomies and resultant morbidity to the patient.

REFERENCES:

1. Phongkitkarun S, Phaisanphrukkun C, Jatchavala J, Sirachainan E. Assessment of gastrointestinal stromal tumors with computed tomography following treatment with imatinib mesylate. World J Gastroenterol. 2008 Feb 14;14(6):892–8.
2. Skandalos IK, Hotzoglou NF, Matsi KCh, Pitta XA, Kamas AI. Giant extra gastrointestinal stromal tumor of lesser omentum obscuring the diagnosis of a choloperitoneum. Int J Surg Case Rep. 2013;4(10):818–21.
3. Wang Y, Peng J, Huang J. Giant and high-risk gastrointestinal stromal tumor in the abdomino-pelvic cavity: A case report. Oncol Lett. 2016 Mar;11(3):2035–8.
4. Kramer K, Siech M, Sträter J, Aschoff AJ, Henne-Bruns D. [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 Mar;43(3):281–8.
5. Søreide K, Sandvik OM, Søreide JA, Giljaca V, Jureckova A, Bulusu VR. Global epidemiology of gastrointestinal stromal tumours (GIST): A systematic review of population-based cohort studies. Cancer Epidemiol. 2016 Feb;40:39–46.
6. Mekni A, Chelly I, Azzouz H, Ben Ghorbel I, Bellil S, Haouet S, et al. Extragastrointestinal stromal tumor of the urinary wall bladder: case report and review of the literature. Pathologica. 2008 Jun;100(3):173–5.
7. Joensuu H. Gastrointestinal stromal tumor (GIST). Ann Oncol Off J Eur Soc Med Oncol. 2006 Sep;17 Suppl 10:x280-286.
8. Vij M, Agrawal V, Kumar A, Pandey R. Cytomorphology of gastrointestinal stromal tumors and extra-gastrointestinal stromal tumors: A comprehensive morphologic study. J Cytol. 2013 Jan;30(1):8–12.
9. Judson I, Bulusu R, Seddon B, Dangoor A, Wong N, Mudan S. UK clinical practice guidelines for the management of gastrointestinal stromal tumours (GIST). Clin Sarcoma Res. 2017 Dec;7(1):6.
10. Lasota J, Miettinen M. KIT and PDGFRA mutations in gastrointestinal stromal tumors (GISTs). Semin Diagn Pathol. 2006 May;23(2):91–102.
11. Omar I, Alsaati H, Waris E. A Huge Pelvic-Abdominal Malignant GIST Tumour in a Patient with Neurofibromatosis Type 1: Case Report and Literature Review. Case Rep Oncol Med. 2020;2020:6590307.
12. Roberts PJ, Eisenberg B. Clinical presentation of gastrointestinal stromal tumors and treatment of operable disease. Eur J Cancer Oxf Engl 1990. 2002 Sep;38 Suppl 5:S37-38.
13. Patil S, Jain S, Kaza RCM, Chamberlain RS. Giant gastrointestinal stromal tumor presenting as a palpable abdominal mass: an unusual presentation. ISRN Surg. 2011;2011:894829.
14. Shafizad A, Mohammadianpanah M, Nasrolahi H, Mokhtari M, Mousavi SA.

Lymph Node Metastasis in Gastrointestinal Stromal Tumor (GIST): to Report a Case. Iran J Cancer Prev. 2014;7(3):171–4.
15. Demetri GD, von Mehren M, Blanke CD, Van den Abbeele AD, Eisenberg B, Roberts PJ, et al. Efficacy and safety of imatinib mesylate in advanced gastrointestinal stromal tumors. N Engl J Med. 2002 Aug 15;347(7):472–80.
16. T. Nishida, S. Hirota. Biological and clinical review of stromal tumors in the gastrointestinal tract. Histol Histopathol. 2000 Oct 1;(33):1293–301.
17. T. Tran, J. A. Davila, H. B. El-Serag. The epidemiology of malignant gastrointestinal stromal tumors: an analysis of 1,458 cases from 1992 to 2000. Am J Gastroenterol. 2005;100(1):162–8.
18. Fletcher CDM, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ, et al. Diagnosis of gastrointestinal stromal tumors: A consensus approach. Hum Pathol. 2002 May;33(5):459–65.
19. Sashidharan P, Matele A, Matele U, Al Felahi N, Kassem KF. Gastrointestinal stromal tumors: a case report. Oman Med J. 2014 Mar;29(2):138–41.
20. Feng F, Liu Z, Zhang X, Guo M, Xu G, Ren G, et al. Comparison of Endoscopic and Open Resection for Small Gastric Gastrointestinal Stromal Tumor. Transl Oncol. 2015 Dec;8(6):504–8.
21. Demetri GD, von Mehren M, Antonescu CR, DeMatteo RP, Ganjoo KN, Maki RG, et al. NCCN Task Force Report: Update on the Management of Patients with Gastrointestinal Stromal Tumors. J Natl Compr Canc Netw. 2010 Apr;8(Suppl 2):S-1-S-41.
22. von Mehren M, Randall RL, Benjamin RS, Boles S, Bui MM, Ganjoo KN, et al. Soft Tissue Sarcoma, Version 2.2018, NCCN Clinical Practice Guidelines in Oncology. J Natl Compr Canc Netw. 2018 May;16(5):536–63.
23. Eisenberg BL, Harris J, Blanke CD, Demetri GD, Heinrich MC, Watson JC, et al. Phase II trial of neoadjuvant/adjuvant imatinib mesylate (IM) for advanced primary and metastatic/recurrent operable gastrointestinal stromal tumor (GIST): early results of RTOG 0132/ACRIN 6665. J Surg Oncol. 2009 Jan 1;99(1):42–7.
24. Kurokawa Y, Yang H-K, Cho H, Ryu M-H, Masuzawa T, Park SR, et al. Phase II study of neoadjuvant imatinib in large gastrointestinal stromal tumours of the stomach. Br J Cancer. 2017 Jun 27;117(1):25–32.
25. Iwatsuki M, Harada K, Iwagami S, Eto K, Ishimoto T, Baba Y, et al. Neoadjuvant and adjuvant therapy for gastrointestinal stromal tumors. Ann Gastroenterol Surg. 2019 Jan;3(1):43–9.
26. Jakob J, Mussi C, Ronellenfitsch U, Wardelmann E, Negri T, Gronchi A, et al. Gastrointestinal stromal tumor of the rectum: results of surgical and multimodality therapy in the era of imatinib. Ann Surg Oncol. 2013 Feb;20(2):586–92.
27. Farag S, Geus-Oei L-F de, van der Graaf WT, van Coevorden F, Grunhagen D, Reyners AKL, et al. Early Evaluation of Response Using 18F-FDG PET Influences Management in Gastrointestinal Stromal Tumor Patients Treated with Neoadjuvant Imatinib. J Nucl Med Off Publ Soc Nucl Med. 2018 Feb;59(2):194–6.
28. Choi H, Charnsangavej C, Faria SC, Macapinlac HA, Burgess MA, Patel SR, et al. Correlation of computed tomography and positron emission tomography in patients with metastatic gastrointestinal stromal tumor treated at a single institution with imatinib mesylate: proposal of new computed tomography response criteria. J Clin Oncol Off J Am Soc Clin Oncol. 2007 May 1;25(13):1753–9.