International Surgery Journal
*Paruthy S et al. Int Surg J. 2021 Apr;8(4):1325-1328*
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20211007

**Case Report**

# An unusual presentation of a retroperitoneal gastrointestinal stromal tumors

**Samir Paruthy[1,2], Shivani B. Paruthy[3]***

[1]Consultant Surgeon, East Delhi Medical Centre, Shahdara, Delhi, India
[2]Holy Child Nursing Home, Krishna Nagar, Delhi, India
[3]Department of Surgery, Safdarjung Hospital and VMMC, New Delhi, India

**Received:** 25 Febraury 2021
**Revised:** 07 March 2021
**Accepted:** 08 March 2021

***Correspondence:**
Dr. Shivani B. Paruthy,
E-mail: shivani_paruthy@yahoo.co.in

**Copyright:** © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

## ABSTRACT

Gastrointestinal stromal tumors (GIST) are soft tissue mesenchymal tumor originating from intestinal cells of Cajal which are pace makers of the intestine. GIST commonly arises from stomach and intestine although they seem to appear occasionally in mesentery, omentum and esophagus, but origin from retro-peritoneum is very infrequent. We herein report case of a huge retroperitoneal GIST measuring 16x13x9 cm size, in a 52-year-old lady occupying whole abdomen arising as pelvic mass. GIST was initially mistaken with associated large intramural fibroid uterus and a large pedunculated sub-serosa broad ligament fibroid. Patient was taken for abdominal open hysterectomy with complete tumor excision after all adhesions from adjoining intestine was separated. Histopathological examination of the mass confirmed it was retroperitoneal GIST with positive immune-histochemistry. Patient responded well to Imatinib therapy, and for last 4 years there has been no residual disease or recurrence as evidenced with regular follow up.

**Keywords:** Retroperitoneum, GIST, Platelet-derived growth factor receptor α polypeptide

## INTRODUCTION

Gastro intestinal stromal tumor is a rare tumor of mesenchymal origin. It can present at any age, being common in elderly more than 50 years of age with stomach commonly involved in 60-70% of cases, followed by small intestine in 20-30%, but GIST of abdominal cavity, mesentery and omentum, and retro-peritoneum is a very rare presentation.[1]

GIST is a mesenchymal tumor of unknown malignant potential arising from interstitial cells of Cajal, which are pace makers of intestine responsible for peristalsis and segmentation of smooth muscles.[1-3]

GIST in mesentery arises from gastrointestinal tract.[1,4] At molecular level, GIST reportedly harbors mutation in the KIT proto-oncogenes or platelet-derived growth factor receptor α polypeptide (PDGFRA).[5,6]

## CASE REPORT

We report a case of 52 years old female patient who presented in our surgery clinic with pain, and a gradually increasing lump in the lower abdomen associated with menorrhagia. Physical examination revealed a huge pelvic mass measuring 16x20 cm occupying almost whole of the lower abdomen and extending to right hypochondrium. The lump was bimanually palpable along with fibroid uterus and broad ligament fibroid. Ultrasound radiography revealed a huge mass of mixed echogenicity in the pelvis and another large multi-lobulated mass arising from right iliac fossa to right lumbar region reaching up to right hypochondrium.

CECT scan revealed a well-encapsulated, enhancing mass lesion measuring 13x13x11 cm involving the uterine body. Another lobulated iso-dense mass lesion 16x13x9 cm was noted on the right iliac fossa and lumbar region suggestive of a large intramural fibroid with a large pedunculated sub serosal broad ligament fibroid. CT-guided fine needle aspiration cytology (FNAC) was done which revealed spindle cells with abundant cytoplasm in a hemorrhagic background with suspicion of Leiomyoma.

Against this background information of clinical presentation history, examination and imaging studies, the patient was planned for total abdominal hysterectomy with bilateral salpingo-opherectomy. A huge mass was seen lying adjacent to the caecum arising from retro-peritoneum and adhering to the small intestine. The retroperitoneal pedunculated mass was of the size of 16x13x9 cm, with a loop of intestine entangled with the mass; approximately 3 feet away from ileocecal junction. The mass was completely excised, taking care not to breach the capsule along with the loops of ileum encasing the mass. Both ureters were identified and bowel continuity established and checked for any serosal avulsions. Immediate postoperative period was uneventful. After hysterectomy retroperitoneal mass is depicted in Figure 1.

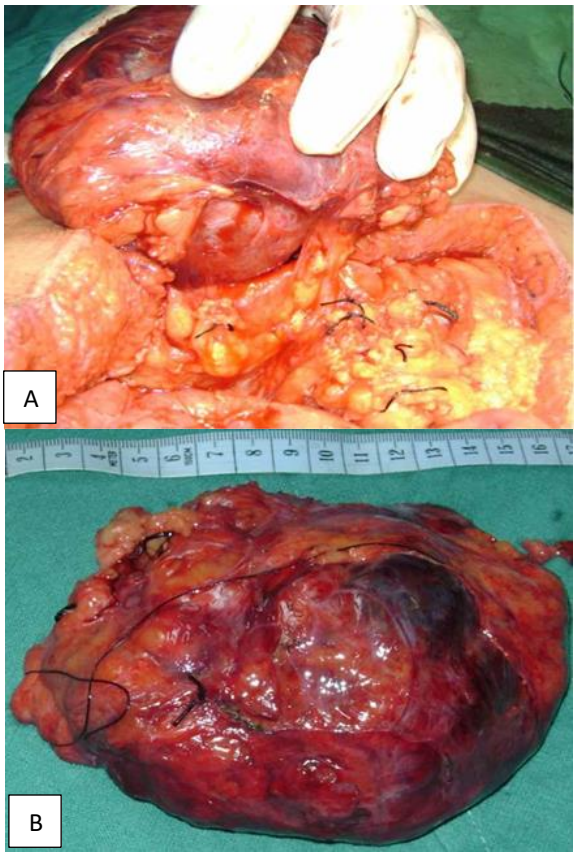


**Figure 1 (A and B): Left- intra-operative photograph of tumor; right- resected specimen.**

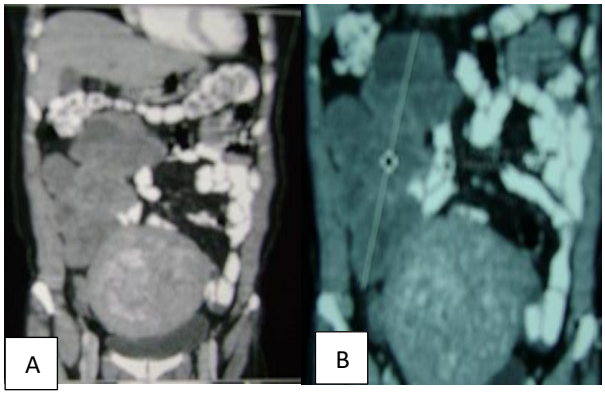


**Figure 2 (A and B): Preoperative CECT of huge tumor arising from pelvis.**

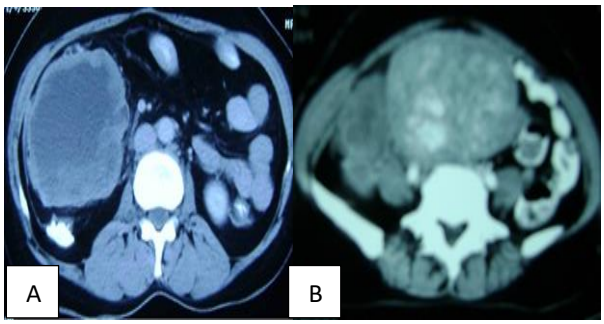


**Figure 3 (A and B): Tumor abutting lateral abdominal wall (left) and anterior abdominal wall (right).**

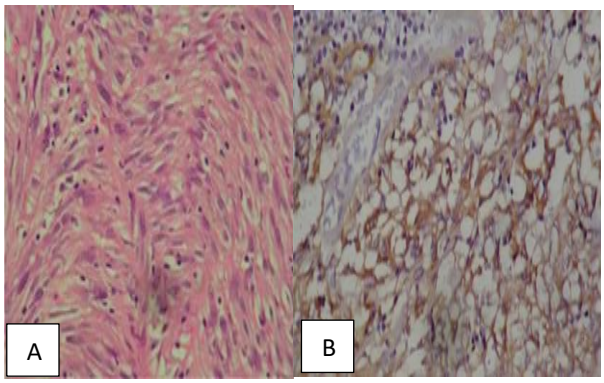


**Figure 4: Histopathological slide of abundant spindle cells (left) and immunohistochemistry of positivity to CD-117 (right).**

Surprisingly, histopathology of the resected retroperitoneal mass revealed as GIST confirmed by immunohisto-chemistry. The tumor was CD117 positive with immune-reactive score 4+ in the tumor cells. Post-operative PET scan did not show any metastasis. The patient responded to imatinib mesylate for four years, and since then she is on a regular follow up with no evidence of recurrence, metastasis or flare up of any residual disease.

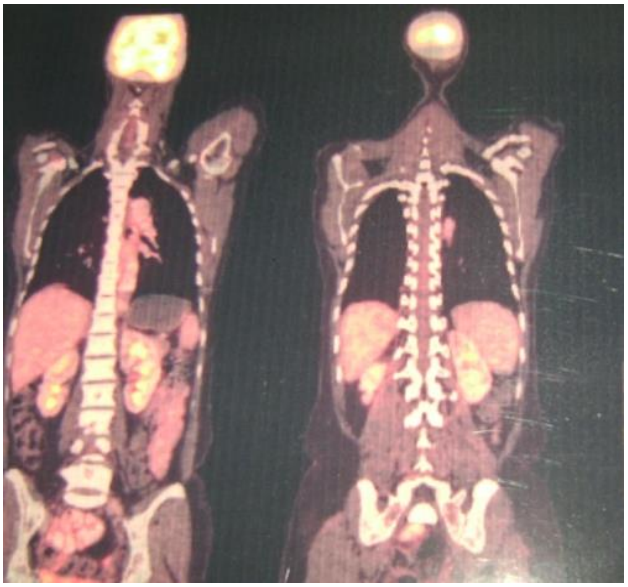

**Figure 5: PET scan of no abnormal FDG uptake.**

## DISCUSSION

GIST is an uncommon sarcoma of the gastrointestinal tract. These tumors are thought to arise from the interstitial cells of Cajal found in muscular layer of the gastrointestinal tract functioning as pacemaker. Based on appraisals from clinical observations, it has been noted that approximately 60% of GIST occur in stomach, 35% in small intestine and less than 5% in rectum, esophagus, omentum and mesentery.[7] Three key features associated with the disease from prognostic viewpoint include-mitotic rate, tumor size and site. Tumors that are small, less than or up to 2 cm in size and showing mitotic activity not exceeding 5 mitoses per 50 high-power fields (HPFs) reportedly have excellent prognosis, probably independent of site, although this has not been shown specifically for all sites.[8] Understanding GIST biology at molecular level led to evidence about gain of function mutation identified in KIT proto-oncogene and less frequently in PDGFRA (platelet-derived growth factor receptor α polypeptide) which are pivotal in initiating tumor genesis in the interstitial cells of Cajal ultimately culminating as GIST.[9-12.] Furthermore, FDA approved tyrosine kinase inhibitors such as- Imatinib mesylate (Gleevac™, Novartis pharmaceuticals, Basel Switzerland), is a specific inhibitor of the KIT, PDGFRA, ABL and BCR-ABL proteins. Therapeutically, Imatinib reportedly achieves restricted or stable response in 80% of GIST patients with metastatic disease having a median survival of 5 years.[13,14] With the knowledge of acquired resistance to chemotherapy, GIST patients also tend to develop chemo-resistance to Imatinib after median treatment duration of 2 years.[13,15]

Hence surgical resection is the first line of treatment for GIST followed by adjuvant therapy. Furthermore, most of these cases occur between the age 55-60 years presenting with non-specific symptoms of anemia fatigue or early satiety, and bloating.[1,14] If untreated, the tumor may rupture leading to massive bleeding or slow bleeding may ensue from intra-luminal tumor site, may present as acute abdomen.[1,15]

Beside surgical intervention and to overcome the challenge, additional FDA approved drug for use in GIST patient's developing resistance to Imatinib is Sunitinib Maleate (Sutent™, Pfizer, New York). Sunitinib inhibits KIT, PDGFRA, the vascular endothelial growth factor receptors, tyrosine kinase-3 receptor (FLT3), and the RET receptor and has shown a median progression free survival of 6 months.[16]

## CONCLUSION

Based upon clinical presentation and current understanding regarding diagnosis and management of GIST, we proceeded with exploratory laparotomy. GIST arising from retroperitoneum presenting with huge intramural fibroid, and sub-serosal fibroid uterus is a rare presentation. A high index of suspicion needs to be maintained and prompt surgical intervention is vital to prevent morbidity. Treatment of retroperitoneal GIST is not different from the GIST arising from other parts of the gastrointestinal tract. Surgery is the mainstay of treatment and in advance cases an integrated approach is mandatory.

*Funding: No funding sources*
*Conflict of interest: None declared*
*Ethical approval: Not required*

## REFERENCES

1. Matthews BD, Joels CS, Kercher KW, Heniford BT. Gastrointestinal stromal tumors of the stomach. Minerva chirurgica. 2004;59:219-31.
2. Szanto P, Barbus A, Al Hajjar N, Zaharia T, Manciula D. Gastric stromal tumor: a rare cause of upper gastrointestinal bleeding. J gastrointestinal liver dis. 2007;16:441-3.
3. Sanders KM, Koh SD, Ward SM. Interstitial cells of cajal as pacemakers in the gastrointestinal tract. Ann review physiol. 2006;68:307-43.
4. Miettinen M, Monihan JM, Sarlomo-Rikala M, Kovatich AJ, Carr NJ, Emory TS, Sobin LH. Gastrointestinal stromal tumors/smooth muscle tumors (GISTs) primary in the omentum and mesentery: clinicopathologic and immunohistochemical study of 26 cases. Am J surgical pathol. 1999;23:1109-18.
5. Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ et al. Diagnosis of gastrointestinal stromal tumors: A consensus approach. Human pathol. 2002;33:459-65.
6. Miettinen M, Lasota J. Gastrointestinal stromal tumors: review on morphology, molecular pathology, prognosis, and differential diagnosis. Arch pathol lab med. 2006;130:1466-78.

7. Miettinen M, Lasota J. Gastrointestinal stromal tumors: pathology and prognosis at different sites. Seminars diagnostic pathol. 2006;23:70-83.
8. Miettinen M, El-Rifai W, Lasota J. Evaluation of malignancy and prognosis of gastrointestinal stromal tumors: a review. Human pathol. 2002;33:478-83.
9. Hirota S, Isozaki K, Moriyama Y, Hashimoto K, Nishida T, Ishiguro S et al. Gain-of-function mutations of c-kit in human gastrointestinal stromal tumors. Science. 1998;279:577-80.
10. Heinrich MC, Corless CL, Duensing A, McGreevey L, Chen CJ, Joseph N et al. PDGFRA activating mutations in gastrointestinal stromal tumors. Science. 2003;299:708-10.
11. Heinrich MC, Owzar K, Corless CL, Hollis D, Borden EC, Fletcher CD et al. Correlation of kinase genotype and clinical outcome in the North American Intergroup Phase III Trial of imatinib mesylate for treatment of advanced gastrointestinal stromal tumor: CALGB 150105 Study by Cancer and Leukemia Group B and Southwest Oncology Group. J clin oncol. 2008;26:5360-7.
12. Corless CL, Fletcher JA, Heinrich MC. Biology of gastrointestinal stromal tumors. J clin oncol. 2004;22:3813-25.
13. Blanke CD, Demetri GD, Von Mehren M, Heinrich MC, Eisenberg B, Fletcher JA et al. Long-term results from a randomized phase II trial of standard- versus higher-dose imatinib mesylate for patients with unresectable or metastatic gastrointestinal stromal tumors expressing KIT. J clin oncol. 2008;26:620-5.
14. Cirocchi R, Farinella E, La Mura F, Cavaliere D, Avenia N, Verdecchia GM et al. Efficacy of surgery and imatinib mesylate in the treatment of advanced gastrointestinal stromal tumor: a systematic review. Tumori. 2010;96:392-9.
15. Verweij J, Casali PG, Zalcberg J, LeCesne A, Reichardt P, Blay JY et al. Progression-free survival in gastrointestinal stromal tumours with high-dose imatinib: randomised trial. Lancet. 2004;364:1127-34.
16. Demetri GD, Van Oosterom AT, Garrett CR, Blackstein ME, Shah MH, Verweij J et al. Efficacy and safety of sunitinib in patients with advanced gastrointestinal stromal tumour after failure of imatinib: a randomised controlled trial. Lancet. 2006;368:1329-38.

**Cite this article as:** Paruthy S, Paruthy SB. An unusual presentation of a retroperitoneal gastrointestinal stromal tumors. Int Surg J 2021;8:1325-8.