International Surgery Journal
Ramesh BS et al. Int Surg J. 2022 Apr;9(4):920-922
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20220962

Case Report

Rare case of giant extra gastrointestinal stromal tumor of wild variant

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Received: 17 February 2022
Accepted: 10 March 2022

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are mesenchymal neoplasms of gastrointestinal tract. The interstitial cells of Cajal (pacemaker cells) act as normal counterpart of tumor. They are rare cancers with incidence of 1.5 in 100000. KIT and PDGFRA gene mutations are the most common pathogenetic causes, whereas a minority of GISTs do not harbour either of the mutations, referred to as wild type GISTs. A minority of cases are extra gastrointestinal (EGIST), arising from mesentry, omentum and retroperitoneum. We present a wild type EGIST measuring 32×24×15 cm and weighing approximately 6 kg.

Keywords: GIST, EGIST, Mesenchymal neoplasm, Wild type

INTRODUCTION

Gastrointestinal stromal tumors GISTs are rare cancers occurring at any age, with median occurrence at 60-65 years. A small minority affect children and adolescents, most of which are WT for KIT and PDGFRA. More than half cases arise from stomach, one fourth from small bowel, others from rectum and oesophagus.[3]

One fourth of GISTs are diagnosed as clinical emergency, often leading to surgical explorations. One fourth of GISTs are discovered incidentally during ultrasound, endoscopy or CT scan done for other reasons. Remaining are diagnosed because of symptoms of compression from an abdominal mass, chronic anemia or fatigue.

The hallmark of GIST is their positivity for KIT and DOG1.[4] The 10-15% are WT for KIT and PDGFRA which are classified as i. SDH deficient; ii. NF1 related; iii. Others.[5]

Risk classification systems are based on mitotic count, tumor size and site of origin.

CASE REPORT

We report 80-year-old female, known case of diabetes mellitus, on treatment, presented with complaints of distension of abdomen since 4 weeks, insidious in onset, gradually progressive in nature, associated with pain in right iliac fossa. Associated with constipation, breathlessness on lying down occasionally, relieved on lying down on sides, weight loss about 5 kg in 1.5 months and increased frequency of micturition.

On examination, a solid, mobile mass noted extending from right half of abdomen to involve almost whole of left half of abdomen. Mass mobile in horizontal direction. Per rectal examination was normal. Bilateral lower limb oedema was present.

USG abdo and pelvis suggestive of a fairly lobular heterogenous lesion noted in peritoneal cavity about 37×20 cm. Superiorly lesion extends to epigastrium, inferiorly to pelvic cavity and on either side to lumbar region with prominent internal vascularity. CECT abdo and pelvis showed findings similar to USG findings. Liver, spleen and pelvic structures normal. Thus, features

were suggestive of intraperitoneal neoplastic etiology (Figure 1).

Intra operatively the tumor was noted to be arising from mesentery and ileum and caecum was noted to be stretched over the tumor (Figure 2 and 3). It weighed about 6kgs and measured 32×24×15 cm. The patient underwent exploratory laparotomy with R0 resection (ileal resection with caecum resection) with end jejunostomy since the patient was hemodynamically unstable. Post operative period was uneventful.

Histopathological reports suggested a mixed type GIST with mitotic rate of >5/5 sq.mm with immunohistochemistry negative for KIT and DOG1.

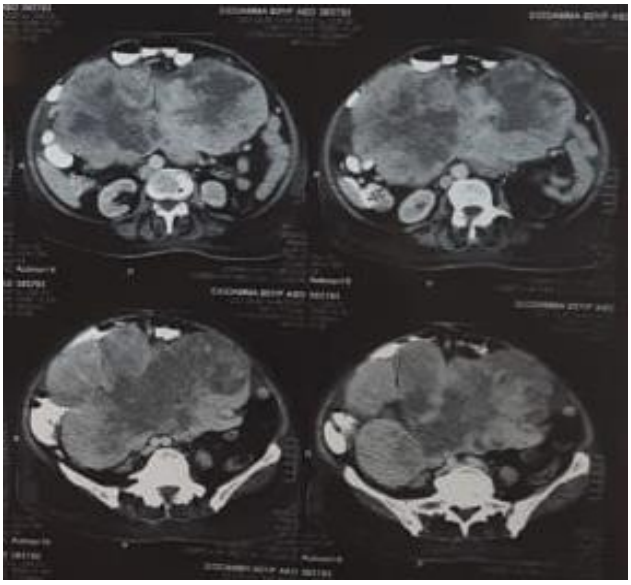

Figure 1: A lobular, heterogeneous lesion seen on CECT.

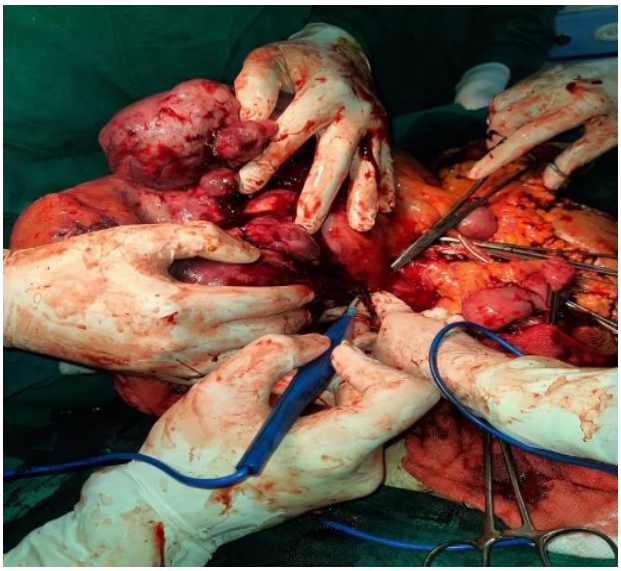

Figure 2: Intraoperative picture of the tumor being resected.

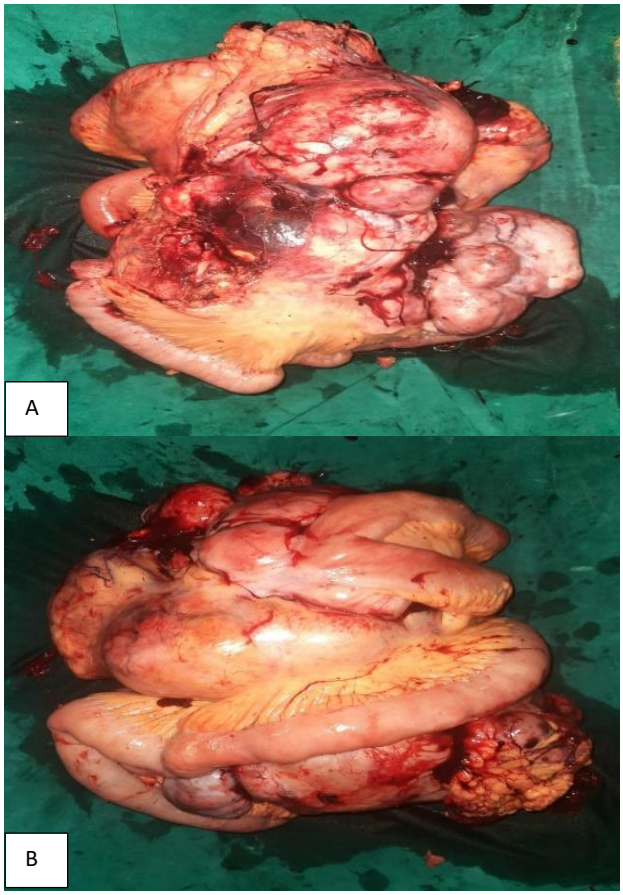

Figure 3 (A and B): Resected specimen of tumor with excised specimen of ileum and cecum noted to be stretched over the tumor.

DISCUSSION

Our patient presented with complaints of distension of abdomen since 4 weeks, insidious in onset, gradually progressive in nature with radioimaging suggesting intraperitoneal neoplastic etiology. The patient underwent laparotomy with R0 resection of tumor due to the severity of symptoms. Histopathology suggested high grade mixed type GIST.

When the disease is localised, surgery is the mainstay of treatment. All GISTs ≥2 cm in size should be resected with the goal of surgery R0 excision which is associated with better prognosis then a microscopically incomplete resection.[6]

Adjuvant therapy with tyrosine kinase inhibitors (TKI) is recommended for 3 years in patients with significant risk of relapse. Preoperative imatinib is considered if extensive surgery is required for 6-12 months.[7] When the disease is metastatic or locally advanced, imatinib is to be continued indefinitely.[8] Secondary resistance is the limiting factor for imatinib, with sunitinib and regorafenib being second- and third-line drugs respectively.

Follow ups after surgery are aimed at picking up relapses early. CT scan is most sensitive for peritoneal and liver metastases with maximum risk interval being 2-3 years after surgery.[9]

CONCLUSION

GISTs are rare tumors of gastrointestinal tract and can present as large abdominal masses. Hence, GIST should be considered as one of differential diagnoses among neoplasms of gastrointestinal tract. They are generally well defined and may present as an incidental finding on scanning or may even present as emergency. Resection should be performed to relieve symptoms and for determining presence of mutations. Appropriate follow up should be offered for chemotherapy and picking up relapses early.

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

REFERENCES

1. Kindblom LG, Remotti HE, Aldenborg F. Gastrointestinal pacemaker cell tumor (GIPACT): gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. Am J Pathol. 1998;152:1259-69.
2. Søreide K, Sandvik OM, Søreide JA. Global epidemiology of gastrointestinal stromal tumours (GIST): A systematic review of population-based cohort studies. Cancer Epidemiol. 2016;40:39-46.
3. Von Mehren M, Joensuu H. Gastrointestinal Stromal Tumors. J Clin Oncol. 2018;36:136-143.
4. Novelli M, Rossi S, Rodriguez-Justo M, Taniere P. DOG1 and CD117 are the antibodies of choice in the diagnosis of gastrointestinal stromal tumours. Histopathology. 2010;57:259-70.
5. Pantaleo MA, Nannini M, Corless CL, Heinrich MC. Quadruple wild-type (WT) GIST: defining the subset of GIST that lacks abnormalities of KIT, PDGFRA, SDH, or RAS signaling pathways. Cancer Med. 2015;4:101-3.
6. DeMatteo RP, Lewis JJ, Leung D, Mudan SS. Two hundred gastrointestinal stromal tumors: recurrence patterns and prognostic factors for survival. Ann Surg. 2000;231:51-8.
7. Tirumani SH, Shinagare AB, Jagannathan JP. Radiologic assessment of earliest, best, and plateau response of gastrointestinal stromal tumors to neoadjuvant imatinib prior to successful surgical resection. Eur J Surg Oncol. 2014;40:420-8.
8. Casali PG, Zalcberg J, Le Cesne A. European Organisation for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group, Italian Sarcoma Group, and Australasian Gastrointestinal Trials Group. Ten-Year Progression-Free and Overall Survival in Patients With Unresectable or Metastatic GI Stromal Tumors: Long-Term Analysis of the European Organisation for Research and Treatment of Cancer, Italian Sarcoma Group, and Australasian Gastrointestinal Trials Group Intergroup Phase III Randomized Trial on Imatinib at Two Dose Levels. J Clin Oncol. 2017;35:1713-20.
9. Plumb AA, Kochhar R, Leahy M, Taylor MB. Patterns of recurrence of gastrointestinal stromal tumour (GIST) following complete resection: implications for follow-up. Clin Radiol. 2013;68:770-5.

Cite this article as: Ramesh BS, Kumar PS, Khan HM, Shetty VV. Rare case of giant extra gastrointestinal stromal tumor of wild variant. Int Surg J 2022;9:920-2.