Asian Oncology Research Journal

3(3): 1-5, 2020; Article no.AORJ.56587

Extramural Jejunal GIST: A Rare Case Report

Richa Chauhan[1*], Gyanendra Singh[2] and Upendra Prasad Singh[2]

[1]Mahavir Cancer Sansthan, Patna, India.
[2]Sanjeevani Hospital, Patna, India.

Authors' contributions

This work was carried out in collaboration among all authors. Author RC designed the study, wrote the protocol and wrote the first draft of the manuscript. Authors GS and UPS managed the analyses of the study. Author GS managed the literature searches. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Dinithi C. Peiris, University of Sri Jayewardenepura, Sri Lanka.
(2) Dr. Vaishali Kapoor, Washington University in St. Louis School of Medicine, USA.
Reviewers:
(1) Amal Halim, Egypt.
(2) Shailja Puri, SRL Diagnostics, India.
Complete Peer review History: http://www.sdiarticle4.com/review-history/56587

Case Study

Received 02 March 2020
Accepted 07 May 2020
Published 18 May 2020

ABSTRACT

Gastrointestinal stromal tumors (GISTs) of jejunum is a rare tumor arising from the interstitial cells of Cajal (ICC) present in the myenteric plexus of the muscularis propria in the intestinal wall. They usually present as small asymptomatic mass and a large exophytic mass causing pain is a relatively rare presentation of jejunal GIST. The tumor mass mainly consists of spindle cells arranged in layers or sheets and stain positive for CD117, CD34 and Vimentin on immunohistochemistry. CECT scan is the most common imaging modality used for staging of the tumor. Surgical resection with negative margins is the primary treatment for localized GIST, followed by adjuvant Imatinib in high risk cases. We present the case of a 65 year old male presenting with large extramural jejunal GIST.

Keywords: GIST; jejunum; abdominal lump; CD117; imatinib.

1. INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal tumors of the gastrointestinal tract. They comprise only 0.2% of all gastrointestinal tumors and are mainly found in the stomach. Small intestinal GISTs comprise only 20% of cases, with jejunal GISTs being the rarest

**Corresponding author: E-mail: chauhan_richa@outlook.com;*

subtype, accounting for 0.1-3% of all GI tumors [1]. They are known to arise from the gastrointestinal pacemaker cells, the interstitial cells of Cajal (ICC) present in the myenteric plexus of the muscularis propria [2]. GISTs usually occur in a sporadic form, but may present as a feature of multiple neoplastic syndromes [3]. They present as a spectrum of morphological and clinical features ranging from benign to malignant; with 10–30% of cases progressing to malignancy [4]. The diagnosis is established with the help of immuno histo chemistry (IHC) markers CD117, CD34, vimentin and mutational analysis for c-kit and platelet-derived growth factor receptor alpha (PDGFRα) genes [5]. The primary treatment for localized GISTs is complete surgical resection with microscopic negative margins [6]. Adjuvant treatment with Imatinib mesylate is recommended for patients with high risk features to decrease the rate of recurrence [7]. The risk stratification is done as per the National Institutes of Health (NIH) consensus criteria and UICC/AJCC TNM staging system [8-9]. Patients with unresectable, metastatic, or recurrent GISTs are also treated with imatinib mesylate [10]. We present a rare case of jejunal GIST with an unusual large size of presentation and with history of pain abdomen.

2. CASE REPORT

A 65 year old male presented with pain and fullness of abdomen for the last 1 month along with decreased appetite and mild breathlessness on walking for the last 10 days. The clinical examination showed an approximately 16 x 10 cm well defined firm lump in lower abdomen and pelvis with mild tenderness. All the routine biochemical investigations were normal. An Ultrasonography of abdomen showed 15.7 x 7.9 cm size well-defined solid mass occupying major part of lower abdomen causing displacement of the bowel loops. A subsequent CECT scan was done which showed large 17.4 x 17.1 x 9.5 cm heterogeneously enhancing hypodense soft tissue lesion involving pelvis and rt. Lower abdomen with an ill-defined fat plane with a short segment of distal ileum/ proximal jejunum. [Fig. 1] An USG guide FNAC was done which was suggestive of benign mesenchymal tumor. So, a laparotomy was planned and done through rt. paramedian incision. There was a huge tumor arising from the mesenteric border of jejunum which was abutting sigmoid colon. Gross tumor along with the segment of jejunum from which it was arising was resected and end to end anastomosis was done. The abdomen was closed in layers and the mass sent for histopathological examination. [Fig. 2] The post-operative period was uneventful. Gross pathological examination showed a large 14 x 13 x 9 cm nodular mass with separate multiple large areas of hemorrhage and necrosis, partly encapsulated by fibrous tissue. The jejunal segment showed a patch of grey-white thickening in the mucosa of size 0.8 x 0.8 cm. Microscopic examination showed a highly cellular spindle cell tumor with cells arranged in fascicles forming organized pattern and with abundant eosinophilic cytoplasm. Focal areas also showed spindle cells in sheets with sprinkled mononuclear cells and mitotic rate >5/HPF with 20-30% necrosis. The jejunal section showed stromal tumor arising from muscularis propria and reaching serosa. This was suggestive of Gastrointestinal stromal tumor (GIST) and the resection margins were uninvolved. The immunohistochemistry test was positive for CD117, DOG1, CD34 and negative for S-100., which confirmed the diagnosis of GIST.

3. DISCUSSION

The incidence for jejunal GIST is extremely rare, accounting for only 0.1-3% of all GI tumors [1]. More than 80% of GISTs are primarily located in GI tract but may occur outside, known as extra-intestinal GIST in sites like omentum, mesentery, retroperitoneum, gallbladder and urinary bladder [8-11]. The most common site in the GI tract is the stomach with 70% of the cases of GIST, followed by small- intestine with 20-30% cases and remaining 10% occurs in esophagus, colon and rectum [12]. Over 90% of GISTs occur in adults over 40 years old with a slight male predominance but no association with geographic location or ethnicity [13]. GISTs usually arise from the intermuscular myenteric plexus region of the intestinal wall and may exhibit either an intramural expansile, extramural (peritoneal) or polypoid (exophytic) growth pattern or a combination, thus resulting in an extremely diverse gross and clinical presentation. They are usually small, asymptomatic and diagnosed incidentally on imaging or colonoscopy. Rarely, few cases grow large enough to present with abdominal pain, lump or intestinal obstruction. Other symptoms may include bleeding, anaemia, early satiety or bloating [14]. Large exophytic growths are usually diagnosed and staged by CECT scans. The differential diagnoses include retroperitoneal mesenchymal neoplasms. The diagnosis is established by a combination of histological

features showing spindle cells and immunohistochemical assay for CD117, an epitope of c-kit receptor tyrosine kinase. However, other GI tumors that may test positive for CD117 include metastatic melanoma, clear-cell sarcoma (30% to 50%), Ewing's sarcoma (50%), childhood neuroblastoma (30%), angiosarcoma (50%), and some carcinoma. Besides, these GI tumors other differential diagnosis for CD117 positive cancers include mastocytoma, seminoma, pulmonary small cell carcinoma and extramedullary myeloid tumors [15]. GISTs are known to arise from activating mutations of proto-oncogenes c-KIT or PDGFRα which increase tyrosine kinase receptor activity, resulting in uncontrolled proliferation of cells [5]. Surgical resection without lymph node dissection is the primary treatment of choice [16]. Tumor size and mitotic index are the two most important risk defining factors. With a tumor size > 10 cm and mitotic rate >5/HPF, our patient was in the high risk group as per NIH consensus and stage III B as per 7th UICC/AJCC staging guidelines [8-9]. So, he was started on adjuvant oral Imatinib mesylate (400mg/day) for 1 year. Imatinib, a small molecule inhibiting the tyrosinase kinase activity of c-kit, PDGFRα, and BCR-ABL, is the first and only approved treatment for GIST.

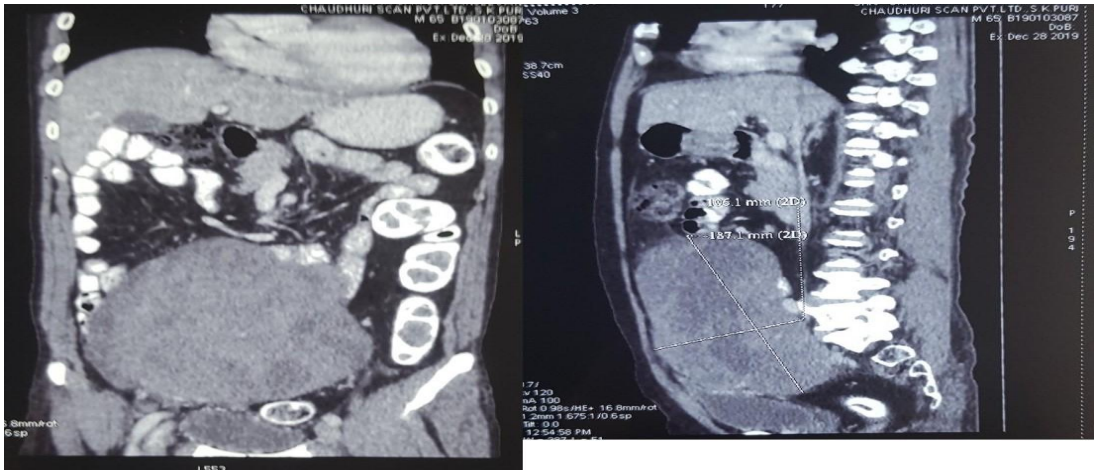

Fig. 1. Coronal and Sagittal CECT Scan images showing large 17.4 x 17.1 x 9.5cm heterogenously enhancing hypodense soft tissue lesion involving pelvis and rt. lower abdomen

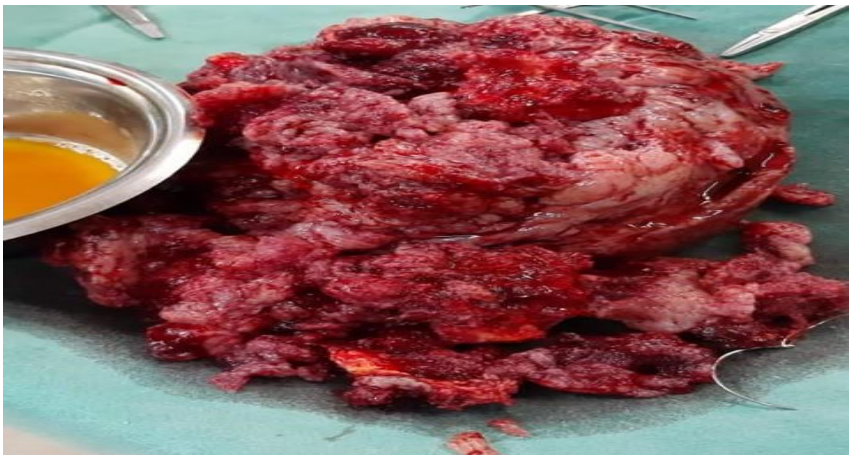

Fig. 2. Post-op specimen showing a large nodular mass with separate multiple large areas of hemorrhage and necrosis, partly encapsulated by fibrous tissue

It was first approved by the FDA in 2001 for unresectable, metastatic or recurrent GIST. The efficacy rate (complete response + partial response) of imatinib has been reported to be 53.8% and the disease-control rate (complete response + partial response + stable disease) to be 84% [10]. As per ACOSOG Z9001 trial the use of imatinib after radical surgery in high-risk cases gives a 14% absolute reduction in recurrence rate, achieving 97% recurrence-free survival[7]. The use of imatinib as an adjuvant therapy for patients after complete resection of localized, primary GIST was approved by FDA in 2008 [17].

4. CONCLUSION

Large extramural GISTs of jejunum are rare tumors. Surgery is the primary treatment for resectable cases. The diagnosis should be established with the help of both morphological and an immunohistochemical characteristic as its adjuvant treatment is different from other intestinal carcinomas and sarcomas. The treatment of GIST has been revolutionized with the use of Imatinib. Localized and resectable GIST has a good prognosis.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Dhull AK, Kaushal V, Dhankhar R, Atri R, Singh H, Marwah N. The inside mystery of jejunal gastrointestinal stromal tumor: a rare report and review of the literature. Case Reports in Oncological Medicine. 2011;4.
2. Kindblom LG, Remotti HE, Aldenborg F, Meis-Kindblom JM. Gastrointestinal Pacemaker Cell Tumor (GIPACT): gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. Am J Pathol. 1998;152(5): 1259–1269.
3. Li FP, Fletcher JA, Heinrich MC, et al. Familial gastrointestinal stromal tumor syndrome: Phenotypic and molecular features in a kindred. J Clin Oncol. 2005; 23:2735-43.
4. Choi H. Response evaluation of gastrointestinal stromal tumors. Oncologist. 2008;1(13): 4-7
5. Miettinen M, Majidi M, Lasota J. Pathology and diagnostic criteria of gastrointestinal stromal tumors (GISTs): A review. Eur J Cancer. 2002;38(Suppl 5):S39–S51.
6. Miettinen M, Makhlouf H, Sobin LH, et al. Gastrointestinal stromal tumors of the jejunum and ileum: A clinicopathologic, immunohistochemical and molecular genetic study of 906 cases before imatinib with long-term follow-up. Am J Surg Pathol. 2006;30:477-89
7. Dematteo RP, Ballman KV, Antonescu CR, et al. Adjuvant imatinib mesylate after resection of localised, primary gastrointestinal stromal tumour: a randomised, double-blind, placebo-controlled trial. Lancet. 2009;373:1097-104.
8. Fletcher CD, Berman JJ, Corless C, et al. Diagnosis of gastrointestinal stromal tumors: A consensus approach. Hum Pathol. 2002;33:459-65.
9. Agaimy A. Gastrointestinal stromal tumors (GIST) from risk stratification systems to the new TNM proposal: More questions than answers? A review emphasizing the need for a standardized GIST reporting. Int J Clin Exp Pathol. 2010;3:461-71.
10. Kubota T. Gastrointestinal stromal tumor (GIST) and imatinib. Int J Clin Oncol. 2006; 11:184–189.
11. Reith JD, Goldblum JR, Lyles RH, et al. Extragastrointestinal (Soft tissue) stromal tumors: An analysis of 48 cases with emphasis on histologic predictors of outcome. Mod Pathol. 2000;13:577-85.
12. Shanmugam S, Vijaysekaran D, Marimuthu MG. Gastro intestinal stromal tumor: A case report. Indian J Radiol Imaging. 2006;16:373-6.
13. Sornmayura P. Gastrointestinal stromal tumors: A pathology view point. J Med Assoc Thai. 2009;92:124–35.
14. Jaison J, Joshi SR, Pathak S, Tekwani D, Nagare M. Gastrointestinal stromal tumour at an unusual site jejunum: A case report. Int J Sci Stud. 2014;2(4):80-83.
15. Miettinen M, Lasota L. Gastrointestinal stromal tumors: Review on morphology, molecular pathology, prognosis and differential diagnosis. Archives of

Pathology and Laboratory Medicine. 2006; 10(130):1466–1478.

16. Demetri GD, Benjamin RS, Blanke CD, et al. NCCN Task Force report: Management of patients with gastrointestinal stromal tumor (GIST)--update of the NCCN clinical practice guidelines. J Natl Compr Canc Netw. 2007;5:S1-29.

17. John T Vetto. Role of imatinib in the management of early, operable, and advanced GI stromal tumors (GISTs). Onco Targets Ther. 2009;2:151–159.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/56587