

## Case Report

# Giant jejunal leiomyosarcoma: a rare case

Jakkula Srikanth<sup>1\*</sup>, E. Rajesh Goud<sup>2</sup>, Sri Harsha Muvva<sup>1</sup>, Kanmathareddy Amulya<sup>1</sup>

<sup>1</sup>Department of General Surgery, <sup>2</sup>Department of Surgical Oncology, SVS Medical College, KNR University of Health Sciences, India

**Received:** 12 October 2018

**Accepted:** 01 November 2018

### \*Correspondence:

Dr. Jakkula Srikanth,

E-mail: [srikanthjakkula31499@gmail.com](mailto:srikanthjakkula31499@gmail.com)

**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

Unlike other gastrointestinal neoplasms, small bowel tumours are often rare. Of this, leiomyosarcoma of jejunum is extremely uncommon. Most of these patients are asymptomatic however in few cases may present with acute presentation such as intestinal obstruction, bleeding and perforation. A 35 years old female presented to the hospital with complaints of pain and vomiting. On palpation, an irregular 21x18cms lump with variable consistency and irregular borders which was intraabdominal and intraperitoneal. CECT revealed heterogenous, hypoechoic mass extending from epigastric to hypogastric region with probable diagnosis of mesenchymal tumour (probably GIST/LEIOMYOSARCOMA), however exact site of origin couldn't be traced. Patient underwent exploratory laprotomy, 25x20cms irregular mass arising from jejunum was resected and sent for histopathological examination. HPE and immunohistochemistry revealed grade 3 leiomyosarcoma.

**Keywords:** Asymptomatic, Bleeding, Chemotherapy, Immunohistochemistry, Nulliparous

## INTRODUCTION

Primary small bowel sarcomas are extremely rare, which accounts to 5% of GI malignancies.<sup>1</sup> Most common small intestinal tumours are adenocarcinoma, carcinoid, lymphoma and sarcoma in decreasing order of frequency. Histologically, most common variant being leiomyosarcoma most commonly occurs in 5<sup>th</sup> to 6<sup>th</sup> decade of life totalling to 2-9%.<sup>2</sup> Few cases were reported of primary small bowel leiomyosarcoma following vigorous immuno-histological techniques.<sup>3,4</sup> Small bowel sarcomas are usually asymptomatic at the initial stages, and difficult to visualise by oesophago-gastroscopy and colonoscopy.<sup>5</sup> In late stages, they may present as intestinal obstruction, bleeding and perforation with poor chances of survival.

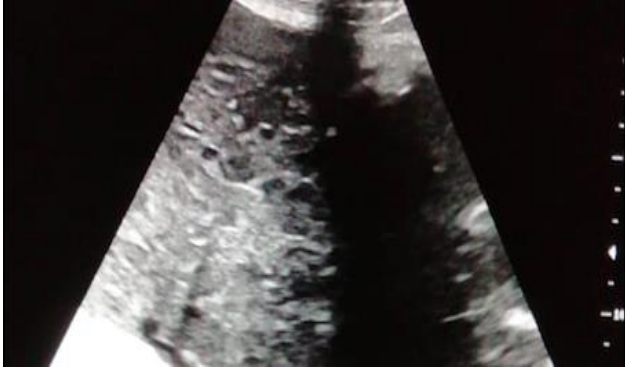
Authors give the account of extremely rare case of jejunal leiomyosarcoma in a 35 year old lady with no significant clinical history suggestive of mesenchymal tumor.

## CASE REPORT

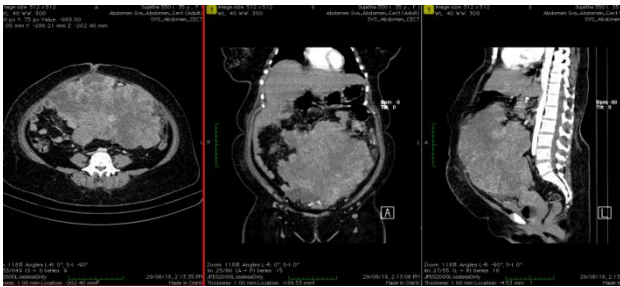
A 35 years old nulliparous female, homemaker by occupation presented to the hospital with chief complaints of pain abdomen since 1 week and vomiting since 2 days. Pain was dull aching, non-progressive, insidious in onset without any radiation, postural and diurnal variations with no specific aggravating and relieving factors. Complaints of non-projectile, vomiting since 2 days, 4-5 episodes per day, straw color. Contains partly digested food particles-20 ml in each bout. doesn't contain blood. Known case of hypertension and type 2 diabetes taking regular medication since 7 years.

On palpation- tenderness present all over abdomen with no guarding and rigidity. A single lump of 21x18cms extending vertically from epigastric to hypogastric region and horizontally extending to both lumbar regions. Lower border not felt separately. Surface was irregular with irregular borders. Firm too hard in consistency. Lump not

moving with respiration with restricted mobility present in both directions. Lump is intra-abdominal and intra peritoneal with no hepatosplenomegaly and no ascites. On prevaginal examination- Fullness noted in lateral fornices. Uterus felt separately from the mass. There is no movement of the cervix on moving the abdominal lump on bimanual examination. No bleeding P/V. Per rectal examination-A bulge is felt through anterior wall of rectum. No intraluminal mass felt. No bleeding. P/R. Gloved finger stained with faeces. On systemic examination, other systems were found to be normal.



**Figure 1: USG- heterogeneous mass.**



**Figure 2: CECT-axial, coronal, sagittal sections.**

Haematological investigations were within normal limits. USG revealed heterogenous hypoechoic mass covering all quadrants of abdomen taking flow on Color Doppler (Figure 1). CECT abdomen revealed, intaperitoneal mass measuring approximately 19x21x16cms showing heterogenous mild enhancement with few necrotic areas, extending from epigastric to pelvic region and also sideways to both iliac fossa, causing mass effect on adjacent bowel loops. Probable diagnosis of mesenchymal tumour arising from jejunum was made (Figure 2).

Patient underwent exploratory laparotomy by midline incision. Linea alba and peritoneum were incised. A 25x20cms irregular mass with lobulations and high vascularity was found with surrounding omental adhesions. Mass was found to be arising 10cms distal to ligament of treitz from the jejunum (Figure 3). Mass was excised with 5cms margin of jejunum (Figure 4 and 5). A single layer end to end jejunojejunostomy was done. Specimen was sent for HPE. Haemostasis secured.

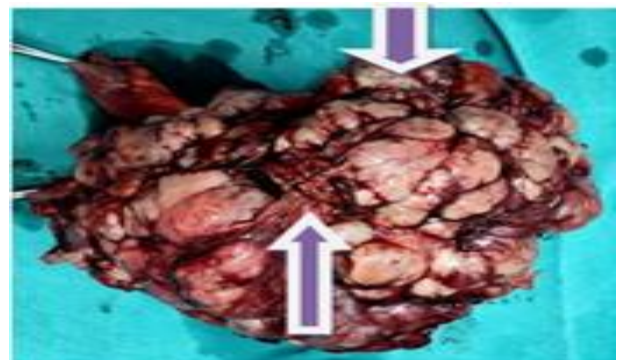
Abdominal drains placed in sub-hepatic and pelvic spaces. Uterus and both ovaries appear to be normal. Liver and spleen normal abdominal closure done in layers. Post op recovery uneventful.



**Figure 3: Intra operative.**



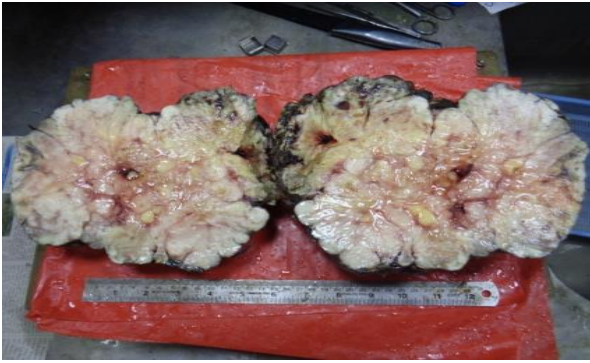
**Figure 4: Resected specimen and jejunum.**



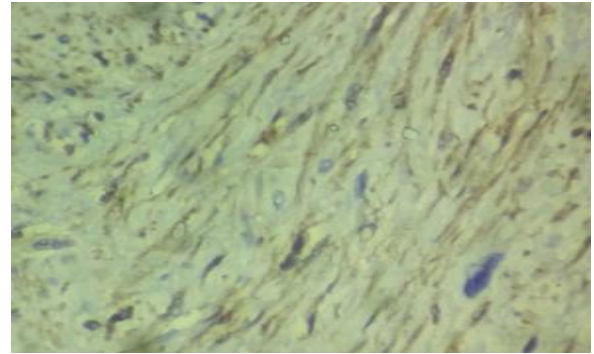
**Figure 5: Gross specimen.**

HPE revealed that tumour cells were spindle shaped to epithelioid in appearance (Figure 6 and 7). Polar vacuoles mimicking GIST with mitotic score 2 (10-19/10HPF). Tumour differentiation: score 3 (undifferentiated sarcoma/sarcoma of uncertain type). Immuno histochemistry markers were done which revealed KIT 117 negative (Figure 8), SMA, CALDESMIN, CALPONIN, DESMIN positive (Figure 9 to 12) indicates smooth muscle cell tumour-malignant leiomyosarcoma of jejunum.

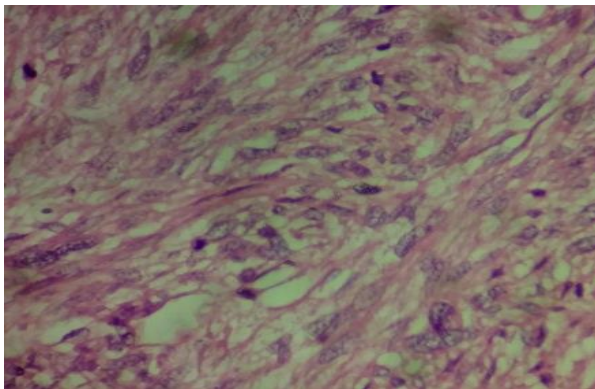




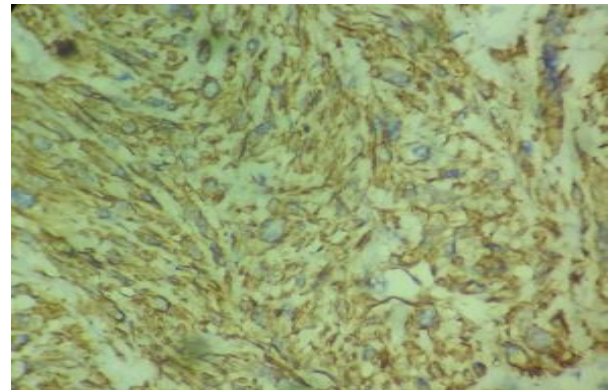
**Figure 6: Cut section.**



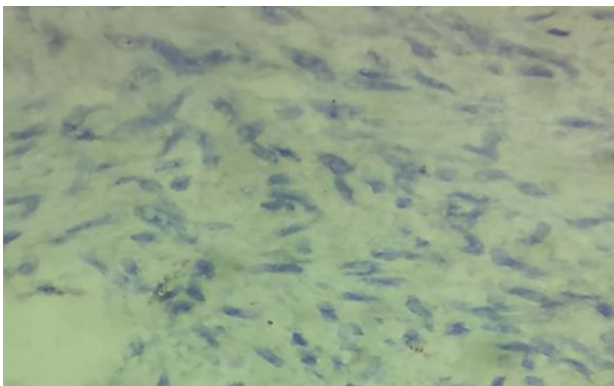
**Figure 10: Calponin.**



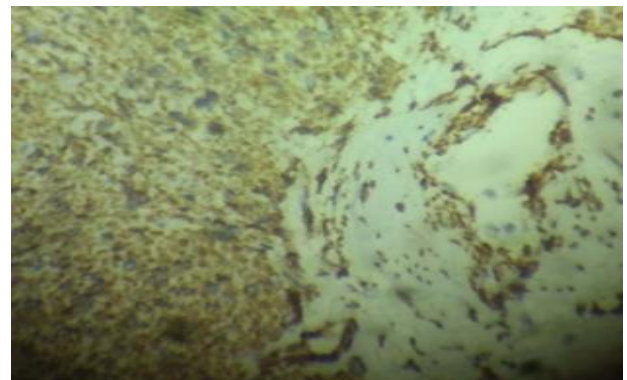
**Figure 7: Spindle cell morphology.**



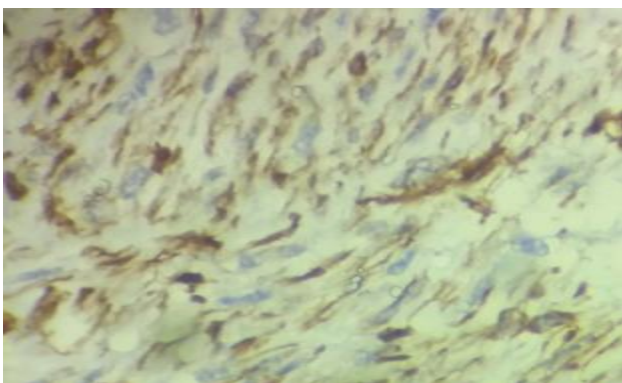
**Figure 11: Caldesmin.**



**Figure 8: KIT 117 negative.**



**Figure 12: Desmin.**



**Figure 9: SMA.**

## DISCUSSION

Small intestine neoplasms are rare, with an incidence of 22.7 per million per year and less than 30 case reports worldwide have been found in literature.<sup>6,4</sup> It accounts to less than 5% of all malignancies in the gastrointestinal tract.<sup>1</sup> Sarcomas rank fifth (1.2%) among all malignant small bowel tumors, after carcinoids (44.3%), adenocarcinomas (32.6%), lymphomas (14.7%), and gastrointestinal stromal tumors (GISTs) (7.2%). Leiomyosarcoma originates from smooth muscle cells between the muscularis mucosa and muscularis propria, with male-to-female ratio ranges from 1:1 to 2:1. The highest incidence of LMS is found in the 6<sup>th</sup> decade.<sup>7</sup>

Symptoms are usually absent. If present, they are nonspecific. Vague complaints, such as malaise, fatigue, and non-focal abdominal pain, are often described. Large leiomyosarcomas that undergo necrosis may bleed into bowel complaints of hematemesis and melena. The causes of leiomyosarcoma are unknown. Hill MA et al, reviewed more than 11,000 cases, 6% of the patients with leiomyosarcomas also had a history of Crohn's disease.<sup>8</sup>

Leiomyosarcoma of small intestine most commonly grow towards the subserosal side of the bowel without causing any intraluminal obstruction.<sup>9</sup> Therefore, patients remain asymptomatic in the early stages of disease. Later, they can present with iron deficiency anaemia, melena, abdominal pain, abdominal lump or intussusception (in case of ileal leiomyosarcoma).<sup>10</sup> Atypical clinical presentation of leiomyosarcoma makes difficult to visualize the small bowel by oesophago-gastroscopy and colonoscopy. Majority of tumors are discovered at advanced stage. Preoperative differentiation between benign and malignant tumors of small intestine remains very difficult.<sup>11</sup> 40% of these tumors are discovered incidentally. Contrast-enhanced computed tomography (CECT), MRI-enterography and enteroclysis are useful for diagnosis of intestinal leiomyosarcoma.<sup>12,13</sup> In acute setting, CT has advantage over MRI because of fast imaging, low cost and high sensitive for detecting the source of GI bleeding.<sup>14</sup> MRI acquires better soft tissue delineation more sensitive in detecting mucosal lesions and it has the advantage of differentiating between different tumors based on the T1 and T2 characteristics without the need for ionisation.<sup>4</sup> Endoscopy and wireless capsule endoscopy also useful in diagnosis of leiomyosarcoma. Endoscopy in acute setting has disadvantage of inadequate bowel preparation, low sensitivity and risks of sedation, perforation and bleeding.<sup>14</sup> Positron emission tomography can contribute in evaluating metastases however it is not in widespread usage.<sup>15</sup>

Histologically, leiomyosarcoma and gastrointestinal stromal tumor (GIST) shows a similar morphologic appearance so immune-histochemical methods should be applied to differentiate these tumors. The main antibodies to detect GISTs are CD117 (c-KIT), DOG1 and CD34. To detect leiomyosarcomas antibodies against smooth muscle are actin (SMA), desmin, caldesmin and calponin are required.<sup>16</sup> The tumour can be graded using the Trojani or French systems (FNCLCC) for soft tissue sarcomas.<sup>17,18</sup> High-grade tumors have 10 or more mitoses per 50 high-power fields (HPF).

Leiomyosarcoma metastasize hematogenously, especially to the liver (65%), other GI locations (15%), and the lungs (4%). In contrast to other sarcomas, it does spread lymphogenously (13%) and peritoneal route (18%).<sup>19</sup> Main modality of treatment for all small intestinal leiomyosarcoma are radical surgical resection with adequate margins.<sup>5</sup> Metastasectomy, if possible, should be considered.<sup>20</sup> In the case of hepatic metastases,

primary and repeated metastasectomy combined with the resection of extrahepatic metastases showed better survival than chemotherapy.<sup>21</sup> In case of unresectable tumors chemotherapy Whilst adjuvant chemotherapy agents like docetaxel, gemcitabine and trabectedin were used for unresectable leiomyosarcomas, their uses have not been proven in the case of small bowel leiomyosarcomas.<sup>22</sup> The prognosis of small intestine leiomyosarcoma is very poor. In low grade disease, the five-year survival rate was 55%, in contrast to 5-20% in high-grade disease.<sup>23</sup>

## CONCLUSION

Leiomyosarcoma of jejunum is a rare entity. In spite of advanced diagnostic modalities early diagnosis of tumour is difficult because of its asymptomatic presentation in the initial stages. Radical surgical resection is the treatment of choice. Role of chemo radiation has not yet been proved. Advancement of robust immuno-histological diagnostic methods is allowing differentiation of leiomyosarcoma from other mesenchymal tumours of small intestine.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

## REFERENCES

1. Jemal A, Siegel R, Ward E, Hao Y, Xu J, Murray T, et al. Cancer statistics, 2008. *CA Cancer J Clin.* 2008;58(2):71-96.
2. McGrath PC, Neifeld JP, Lawrence W Jr, Kay S, Horsley JS 3rd, Parker GA. Gastrointestinal sarcomas. Analysis of prognostic factors. *Ann Surg.* 1987;206(6):706-10.
3. Aggarwal G, Sharma S, Zheng M, Reid MD, Crosby JH, Chamberlain SM, et al. Primary leiomyosarcomas of the gastrointestinal tract in the post-gastrointestinal stromal tumor era. *Ann Diagn Pathol.* 2012;16(6):532-40.
4. Arts R, Bosscha K, Ranschaert E, Vogelaar J. Small bowel leiomyosarcoma: a case report and literature review. *Turk J Gastroenterol* 2012;23(4):381-4.
5. Coco C, Rizzo G, Manno A, Mattana C, Verbo A. Surgical treatment of small bowel neoplasms. *Eur Rev Med Pharmacol Sci.* 2010;14(4):327-33.
6. Bilimoria KY, Bentrem DJ, Wayne JD, Ko CY, Bennett CL, Talamonti MS. Small bowel cancer in the United States. *Ann Surg.* 2009;249:63-71.
7. Evans HL. Smooth muscle tumors of the gastrointestinal tract: a study of 56 cases followed for a minimum of 10 years. *Cancer.* 1985;56:2242-50.
8. Hill MA, Mera R, Levine EA. Leiomyosarcoma: a 45-year review at Charity Hospital, New Orleans. *Am Surg.* 1998;64(1):53-61.
9. Miettinen M, Kopczynski J, Makhlof HR, Sarlomo-Rikala M, Gyorffy H, Burke A, et al.

- Gastrointestinal stromal tumors, intramural leiomyomas, and leiomyosarcomas in the duodenum: a clinicopathologic, immunohistochemical, and molecular genetic study of 167 cases. *Am J Surg Pathol*. 2003;27:625-41.
10. Luis J, Ejtehadi F, Howlett DC, Donnellan IM. Leiomyosarcoma of the small bowel: report of a case and review of the literature. *Int J Surg Case Rep*. 2015;6:51-4
  11. Blanchard DK, Budde JM, Hatch GF 3rd, Wertheimer-Hatch L, Hatch KF, Davis GB, et al. Tumors of the small intestine. *World J Surg*. 2000;24:421-9.
  12. Maglinte DDT, Kelvin FM, O'Connor K, Lappas JC, Chernish SM. Current status of small bowel radiography. *Abdom Imag*. 1996;21:24757.
  13. Megibow AJ, Balthazar EJ, Hulnick DH, Naidich DP, Bosniak MA. CT evaluation of gastrointestinal leiomyomas and leiomyosarcomas. *Am J Roentgenol*. 1985;144:727-31.
  14. Kim BS, Li BT, Engel A, Samra JS, Clarke S, Norton ID, et al. Diagnosis of gastrointestinal bleeding: a practical guide for clinicians. *World J Gastrointest Pathophysiol*. 2014;5:467-78.
  15. Punt SE, Eary JF, O'Sullivan J, Conrad EU. Fluorodeoxyglucose positron emission tomography in leiomyosarcoma: imaging characteristics. *Nucl Med Commun*. 2009;30(7):546-9.
  16. Joensuu H, Hohenberger P, Corless CL. Gastrointestinal stromal tumour. *Lancet*. 2013;382(9896):973-83.
  17. Coindre JM, Nguyen BB, Bonichon F, de Mascarel I, Trojani M. Histopathologic grading in spindle cell soft tissue sarcomas. *Cancer*. 1988;61(11):2305-9.
  18. Guillou L, Coindre JM, Bonichon F, Nguyen BB, Terrier P, Collin F, et al. Com-parative study of the National Cancer Institute and French Federation of Cancer Centers Sarcoma Group grading systems in a population of 410 adult patients with soft tissue sarcoma. *J Clin Oncol*. 1997;15(1):350-62.
  19. Skandalakis JE, Gray SW, Skandalakis LJ. Metastasis of malignant smooth muscle tumors of the gastrointestinal tract: pattern and process. *J Med Assoc Ga*. 1991;80:701-9.
  20. Gill SS, Heuman DM, Mihas AA. Small intestinal neoplasms. *J Clin Gastroenterol*. 2001;33:267-82
  21. Lang H, Nussbaum KT, Kaudel P, Frühauf N, Flemming P, Raab R. Hepatic metastases from leiomyosarcoma: a single-center experience with 34 liver resections during a 15-year period. *Ann Surg*. 2000;231:500-5.
  22. Duffaud F, Bui BN, Penel N, et al. A FNCLCC French Sarcoma Group-GETO multicenter randomized phase II study of gemcitabine versus gemcitabine and docetaxel in patients with metastatic or relapsed leiomyosarcoma. *J Clin Oncol*. 2008;26:10511.
  23. Edge SB, Byrd DR, Compton CC. *AJCC (American Joint Committee on Cancer) Cancer Staging Manual*. 7th ed. New York: Springer; 2010:285.

**Cite this article as:** Srikanth J, Goud ER, Muvva SH, Amulya K. Giant jejunal leiomyosarcoma: a rare case. *Int Surg J* 2018;5:4085-9.