

LAPAROSCOPIC RESECTION OF GASTROINTESTINAL STROMAL TUMORS (GIST) OF THE SMALL INTESTINE: A CASE REPORT

Daniel Navarini^{1,2,3}, Antônio Benincá Albuquerque¹,
André Luca Boeira Rovani¹, Diego Reffatti^{2,3},
Paulo Roberto Reichert^{1,2}, Carlos Augusto Scussel Madalosso^{2,3}

ABSTRACT

Although gastrointestinal stromal tumors (GISTs) are a rare type of cancer, they are the commonest mesenchymal tumors of the gastrointestinal tract (GIT). GISTs can affect any segment of the GIT, but the usual location is the stomach, followed by the small intestine. Surgical resection of the tumor is the gold standard treatment for localized GISTs, and in patients with inoperable and metastatic disease, imatinib mesylate is the standard treatment. Pathological diagnosis is based on morphology and immunohistochemical findings. We report the case of a 55-year-old man with jejunal GIST presenting with endophytic and exophytic growth, located in the proximal jejunum. He had history of melena, anemia and one episode of enterorrhagia, and was treated with surgical resection of the lesion.

Keywords: *Gastrointestinal stromal tumor; gastrointestinal neoplasms*

Clin Biomed Res. 2019;39(1):97-100

1 Faculdade de Medicina, Universidade de Passo Fundo (UPF). Passo Fundo, RS, Brasil.

2 Departamento de Cirurgia, Hospital São Vicente de Paulo (HSVP). Passo Fundo, RS, Brasil.

3 Departamento de Cirurgia, Clínica Gastrobese. Passo Fundo, RS, Brasil.

Corresponding author:

Daniel Navarini
danielnavarini@hotmail.com
Faculdade de Medicina, Universidade de Passo Fundo (UPF)
Rua Teixeira Soares, 817.
99010-080, Passo Fundo, RS, Brasil.

Although gastrointestinal stromal tumors (GISTs) are a rare type of cancer, they are the commonest non-epithelial tumors of the gastrointestinal tract (GIT)¹. The worldwide incidence and prevalence of GIST are estimated to be approximately 1-1.5 per 100,000 and 13 per 100,000 per year, respectively, and account for approximately 0.1-3% of all gastrointestinal tumors^{2,3,4}. GISTs can affect any segment of the GIT. While most GISTs occur in the stomach, only 25% appear in the small intestine⁵. Although they are sometimes asymptomatic, many GISTs may be identified clinically because of symptoms including gastrointestinal bleeding and subsequent anemia, early satiety, abdominal distension, and discomfort and/or pain due to tumor compression⁶.

The pathological diagnosis is based on morphology and immunohistochemical findings. The definite diagnosis depends on KIT positivity (major defining feature for the diagnosis of GIST) associated with a tumor that has morphological features compatible with GIST. When there is KIT negativity, as in approximately 5% of GISTs, DOG1 staining, followed by CD34 staining, is considered diagnostic^{5,6}. The standard treatment for localized GISTs is complete surgical excision of the tumor, with no dissection of clinically negative lymph nodes^{1,7}. In patients with inoperable and metastatic disease, imatinib mesylate, a tyrosine kinase inhibitor, is the standard treatment^{5,7}.

CASE PRESENTATION

A 55-year-old male patient presented with history of melena, anemia – requiring blood transfusion (three times within one year) – and one episode of enterorrhagia. There was no history of abdominal pain, vomiting, nausea, loss of appetite, weight loss or altered bowel habits. On physical examination, he was in good general condition, lucid, attentive and coherent, with normal colored and anicteric mucosa. Abdominal auscultation showed normal bowel sounds, periumbilical pain on palpation, and no signs of peritoneal irritation. Initial laboratory evaluation revealed hemoglobin level of 7.6 mg/dL, mean corpuscular volume of 83.8 fL, mean corpuscular hemoglobin of 25.5 pg, mean

corpuscular hemoglobin concentration of 30.5% and red cell distribution width of 20.0%.

The patient underwent upper gastrointestinal endoscopy – which was normal – and colonoscopy, which demonstrated one angiodysplastic lesion in the terminal ileum. For diagnostic investigation, a capsule endoscopy was performed. The study showed signs of erosion in the proximal small bowel, with possibility of association with subepithelial lesion. Angiodysplastic lesions were also observed in visible segments of the colon. For further evaluation of the findings, abdominal magnetic resonance imaging (MRI) was performed (Figure 1). A nodular lesion was observed, with intermediate signs in T1 and T2, located in the proximal jejunum, measuring 3.6 x 3.2 cm, with regular outlines, well defined capsule, in endothelial situation, demonstrating intense impregnation by the paramagnetic contrast, suggestive of GIST. Localized adenopathy was identified along the fourth part of the duodenum in the mesentery, a lesion measuring 4 mm in its longest axis, which could be related to inflammatory changes. Two nodular lesions in the hepatic parenchyma, compatible with hepatic hemangiomas, were also identified. Based on endoscopic and MRI findings, the diagnostic suspicion was small intestine GIST.

The patient underwent videolaparoscopy, which detected a tumor at 8 cm from the ligament of Treitz. Mesenteric release was performed in the correspondent jejunum area to be resected. Lesion resection was performed with simultaneous anastomosis through double stapling with linear laparoscopic stapler (Figure 2). The resected surgical specimen was removed from the abdominal cavity through a small opening in the left hypochondrium, using a specimen retrieval bag (Figure 3). Histopathological examination revealed a fusiform cell neoplasm. Immunohistochemical study was performed using indirect immunoperoxidase method with diaminobenzidine. Microscopically, neoplastic cells exhibited positive antibodies for

DOG1, CD34, c-KIT (CD117) and Ki-67 (2%). There were no positive antibodies for actina, desmina and S100. Therefore, the diagnosis of GIST with fusocellular pattern was reached, with presence of 1 mitosis per 50 high-power field (HPF). No tumor necrosis was identified.

DISCUSSION

GISTs arise from the interstitial cells of Cajal, which act on the physiological control of peristaltic activity as an electric smooth muscle pacemaker. Men and women are equally affected by them. These tumors may be either benign or malignant, although benign tumors are much more common than the malignant ones^{1,8}. Their biological behavior is differentiated based on morphological parameters such as size of the lesion and number of mitoses per HPF⁹. Immunohistochemistry is important to differentiate a GIST from other mesenchymal tumors, as they show KIT (CD117) expression in 95% of the cases^{1,9}. On CD117-negative lesions, the DOG1 marker is useful for diagnosis as it is detected in approximately 97% of the cases⁹. Smooth muscle actin (SMA), CD34, S100 protein, PKC theta and desmin are other less commonly positive immunohistochemical markers^{6,8,9}. Complete surgical resection of the tumor is the gold standard treatment and should target macroscopic and microscopic negative margins.

In our patient, lesion resection was facilitated by using a double stapler with simultaneous anastomosis, and intestinal resection was the minimum required to guarantee a negative margin. Lymphadenectomy is not required for clinically negative lymph nodes, as their involvement is very rare^{1,6,7,10}. Laparoscopic approach is not recommended in patients with large tumors because of the risk of causing tumor capsule rupture⁷. In case of rupture, the disease progresses with peritoneal dissemination, then with relapse in 100% of the cases, at least at the peritoneal level¹¹.



Figure 1: Abdominal magnetic resonance imaging (MRI) showing a nodular lesion with intense impregnation by the paramagnetic contrast located in the proximal jejunum (arrows).

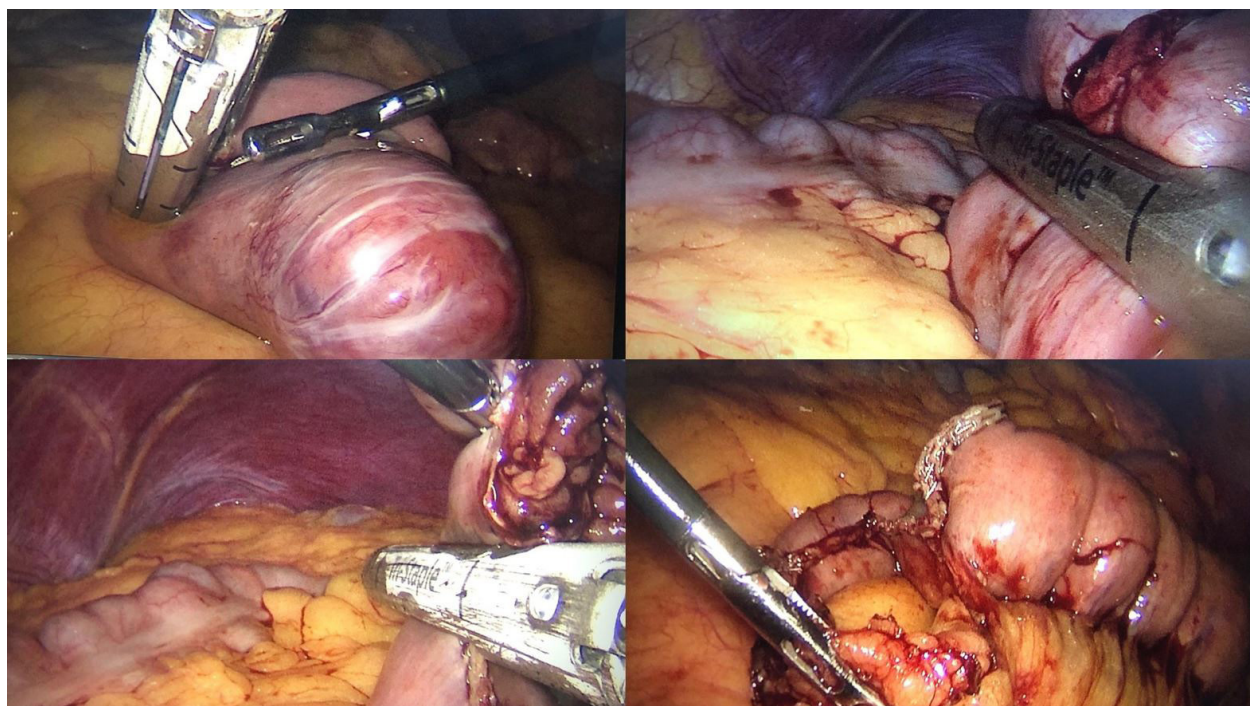


Figure 2: Tumor resection and anastomosis with double stapling.



Figure 3: Resected lesion showing exophytic and endophytic growth.

Imatinib mesylate therapy is indicated for inoperable, metastatic or recurrent GISTs. Its neoadjuvant administration can be used to reduce the size of the tumor and, consequently, to improve rates of R0 resections, disease-free survival and overall survival when compared to upfront surgery. Cases when R0 surgery is likely to result in major functional sequelae may be candidates for this therapy, in order to reduce surgical morbidity^{6,7,12}. In our patient, there was no indication of imatinib mesylate therapy because the lesions were smaller than 5 cm and had less than 5 mitoses per HPF. Size of the primary tumor and mitotic index are the most reliable prognostic factors for GISTs. Location of the primary GIST lesion is related to recurrence and survival rates. Gastric lesions have better prognosis than primary small bowel, colorectal and extragastrointestinal GISTs^{13,14}.

Conflicts of Interest

The authors declare no conflicts of interest.

REFERENCES

1. Hoeppner J, Kulemann B, Marjanovic G, Bronsert P, Hopt UT. Limited resection for duodenal gastrointestinal stromal tumors: surgical management and clinical outcome. *World J Gastrointest Surg*. 2013;5(2):16-21. <http://dx.doi.org/10.4240/wjgs.v5.i2.16>. PMID:23515427.
2. Arellano-López PR, Solalinde-Vargas R, Guzmán-Mejía JI, González-Pérez LG, Zamora-García AC. Gastrointestinal stromal tumor of the stomach: case report. *Rev Med Hosp Gen (Mex)*. 2017; (In press)
3. Nilsson B, Bümming P, Meis-Kindblom JM, Odén A, Dortok A, Gustavsson B, et al. Gastrointestinal stromal tumors: the incidence, prevalence, clinical course, and prognostication in the preimatinib mesylate era—a population- based study in western

- Sweden. *Cancer*. 2005;103(4):821-9. <http://dx.doi.org/10.1002/cncr.20862>. PMID:15648083.
4. Gatta G, van der Zwan JM, Casali PG, Siesling S, Tos APD, Kunkler I, et al. Rare cancers are not so rare: the rare cancer burden in Europe. *Eur J Cancer*. 2011;47(17):2493-511. <http://dx.doi.org/10.1016/j.ejca.2011.08.008>. PMID:22033323.
 5. Navarini D, Albuquerque AB, Rovani ALB, Rohden TB, Refatti D, Oliveira AEL, et al. Justapapillar duodenal gastrointestinal stromal tumor (gist) local resection: a case report. *American Journal of Cancer Case Reports*. 2018;6:11-7.
 6. Nishida T, Blay JY, Hirota S, Kitagawa Y, Kang YK. The standard diagnosis, treatment, and follow-up of gastrointestinal stromal tumors based on guidelines. *Gastric Cancer*. 2016;19(1):3-14. <http://dx.doi.org/10.1007/s10120-015-0526-8>. PMID:26276366.
 7. 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;7:6. <http://dx.doi.org/10.1186/s13569-017-0072-8>. PMID:28465823.
 8. Miettinen M, Lasota J. Gastrointestinal stromal tumors: definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. *Virchows Arch*. 2001;438(1):1-12. <http://dx.doi.org/10.1007/s004280000338>. PMID:11213830.
 9. Rohde L, Osvaldt AB. *Rotinas em cirurgia digestiva*. 3. ed. Porto Alegre: Artmed; 2018.
 10. Naguib SF, Zaghloul AS, Marakby HE. Gastrointestinal Stromal Tumors (GIST) of the stomach: retrospective experience with surgical resection at the national cancer institute. *J Egypt Natl Canc Inst*. 2008;20(1):80-9. PMID:19847285.
 11. Poveda A, del Muro XG, López-Guerrero JA, Martínez V, Romero I, Valverde C, et al. GEIS 2013 guidelines for Gastrointestinal Sarcomas (GIST). *Cancer Chemother Pharmacol*. 2014;74(5):883-98. <http://dx.doi.org/10.1007/s00280-014-2547-0>. PMID:25193432.
 12. Ozkan EE. Radiotherapy for gastrointestinal stromal tumors. *Chin Med J (Engl)*. 2018;131(2):235-40. <http://dx.doi.org/10.4103/0366-6999.222344>. PMID:29336374.
 13. Lamba G, Gupta R, Lee B, Ambrale S, Liu D. Current management and prognostic features for gastrointestinal stromal tumor (GIST). *Exp Hematol Oncol*. 2012;1(1):14. <http://dx.doi.org/10.1186/2162-3619-1-14>. PMID:23210689.
 14. Zhang Q, Shou CH, Yu JR, Yang WL, Liu XS, Yu H, et al. Prognostic characteristics of duodenal gastrointestinal stromal tumours. *Br J Surg*. 2015;102(8):959-64. <http://dx.doi.org/10.1002/bjs.9831>. PMID:25980461.

Received: Jan 10, 2019

Accepted: Mar 12, 2019