



Gastrointestinal Stromal Tumor Inside a Jejunal True Diverticulum

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Abstract

Background: True diverticulum and malignant tumors are rare conditions of jejunum. This is the first published case of a Gastrointestinal Stromal Tumor (GIST) inside a true jejunal diverticulum.

Case Report: A 59-year-old male presented a sudden abdominal pain with emesis and abdominal distension. Computed tomography of the abdomen revealed a mechanical bowel obstruction caused by a tumor in the small bowel. At laparotomy, an isolated 8.0 cm × 4.5 cm jejunal diverticulum with a perforated tumor was found, about 80 cm from the Treitz ligament. The tumor was removed, as were the segment of the jejunum containing the isolated diverticulum and its correspondent mesentery, including the local lymph nodes. The histologic examination of the tumor revealed a GIST with no lymph node metastasis. After one year, the patient presented a new small bowel obstruction due to six nodules surrounding a jejunoileal segment, far from the first anastomosis. All nodules and a twenty-centimeter-jejunoileal segment were removed. GIST was diagnosed in five nodules. The patient was discharged from the hospital in good conditions on the third post-operative day. During the 38-month-follow-up from the second procedure, no adverse event has been identified. The patient is working, presents no complaints, and is under control with 800 mg imatinib.

Conclusion: A GIST may be found inside a jejunal true diverticulum. The treatment must be an en bloc resection of the diverticulum with a segment of jejunum and its mesentery, including all lymphatic tissue. Adjuvant imatinib chemotherapy is indicated to prevent recurrences.

Keywords: Jejunum; Small bowel diverticulum; Jejunum tumor; Gastrointestinal stromal tumor; Jejunum diverticulum GIST; Intestinal diverticulum GIST

Introduction

Approximately 2% of the population is affected by digestive diverticular disease characterized by the presence of all intestinal wall layers. Its least common site in the entire gastrointestinal tract is the small bowel [1]. According to previous studies, Jejunal Diverticulum (JD) is most likely the beginning of an intestinal duplication that did not occur [1,2]. This condition is different from the pseudo diverticulum, which is the pulsion herniation of mucosa and submucosa through a weakened muscular layer, recovered only by serosa and associated with intestinal dyskinesia, present mainly in the colon in more than 50% of the population, showing a prevalence in patients over 60 years of age [2]. The most frequent complications of JD are inflammation, perforation, bleeding, abscess, fistula, and obstruction, which commonly occur in 10% of cases. Most patients are asymptomatic and require no treatment, but in the presence of chronic abdominal discomfort, fullness, and anemia due to unapparent bleeding, surgical resection is recommended [1-3]. Small bowel tumors represent three percent of gastrointestinal neoplasms, and malignant tumors are much less frequent. Predisposing sporadic conditions are adenomas, Peutz-Jeghers syndrome, Meckel's diverticulum, Crohn's disease, and adult celiac disease [3-5]. The first report of a small bowel cancer appeared in 1746, when Hamburger described a duodenal carcinoma. About 50% occur in the distal small bowel, and less than one percent of the digestive cancers can be found in jejunum. More than half of these malignant tumors are adenocarcinomas, followed by carcinoids (23%), lymphomas (13%), leiomyosarcomas (7%), and melanomas (2%) [4,5]. The Gastrointestinal Stromal Tumor (GIST) is a tumor of mesenchymal origin and accounts for less than 1% of all small bowel malignancies. Its diagnosis is difficult because of its rarity and ambiguity of symptoms (abdominal pain, weight loss, and digestive bleeding), which are vague and nonspecific [4,6]. The treatment of choice is surgical resection, followed by adjuvant chemotherapy. The expression of the tyrosine kinase receptor KIT

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Received Date: 03 Feb 2020

Accepted Date: 27 Feb 2020

Published Date: 03 Mar 2020

Citation:

Petroianu A, Chaves Moreira W, Buzelin Nunes M. Gastrointestinal Stromal Tumor Inside a Jejunal True Diverticulum. *Clin Surg.* 2020; 5: 2749.

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(CD 117 leukocyte antigen) and other markers (Ano-1, PDGFRA, CD 34, and protein kinase C theta), immunohistochemically demonstrated in the KIT-negative GIST tumor; confirm the diagnosis [5-8]. In an extensive review of the literature, GIST were found mainly inside the stomach and Meckel's diverticulum, but these were not related to any other diverticulum or even pseudo diverticulum [4-8]. Therefore, the present case report is the first of its kind concerning a GIST inside a jejunal true diverticulum.

Case Presentation

A 59-year-old male previously in good health with no significant comorbidities was referred to our surgical service with two days of sudden abdominal pain and nausea with emesis. During this time, progressive bowel obstruction manifestations were characterized by abdominal distension with decreased intestinal sounds and no stool passing. Physical examination revealed a tender and distended abdomen with hypoactive sounds. Laboratory analysis revealed hemoglobin at 12.6 g/dL, leukocytes 7.5×10^3 /dL, creatinine 1.2 mg/dl, glucose 113 g/dL, albumin 3.6 g/dL, alkaline phosphatase 91 U/l, SGOT 29 U/l, and SGPT 31 U/l. Computed tomography of the abdomen exhibited a fluid-filled, significantly dilated stomach, as well as dilated loops of small bowel, with air-fluid levels consistent with mechanical obstruction caused by a small bowel tumor. After initial fluid resuscitation, nasogastric intubation, and antibiotic administration, exploratory laparotomy presented an isolated 8.0 cm \times 4.5 cm jejunal diverticulum with a perforated tumor, located approximately 80 centimeters from the Treitz ligament. The tumor was not connected to the jejunal loop, but rather inside a lateral jejunal structure, which configured a diverticulum. (Figure 1A) No metastasis or other anomaly was found. The tumor was removed, along with the segment of the jejunum containing the isolated diverticulum and its correspondent mesentery, including the local lymph nodes. The histologic examination confirmed that the tumor was located outside the jejunum, inside a true diverticulum with all jejunal wall layers. The amount of diverticulum tissue with no tumor was very small, but enough to confirm its lateral location. The fusiform cells of the neoplasm were consistent with a GIST (Figure 1B). The diagnosis was confirmed by histochemistry stained for CD 117 leukocyte antigen expression. The hospital course was uneventful. On the first postoperative day, nasogastric intubation was discontinued, and on the third postoperative day, the diet was tolerated, and normal stool passing was confirmed. On the fourth postoperative day, the patient was discharged from the hospital in good shape. Antibiotics were discontinued on the 12th postoperative day. Chemotherapy using oral imatinib was started during the early postoperative period, initially with 400 mg and later with cycles of 800 mg. After one year, the patient presented abdominal distension due to small bowel mechanical obstruction. During the second laparotomy, peritoneal carcinomatosis was found, mainly in the lower abdomen and pelvis. One mesenteric and five peritoneal nodules surrounded and obstructed a jejunoileal segment far from the first anastomosis. A twenty-centimeter-obstructed jejunal segment en bloc with its mesentery and the six nodules were resected. A primary end-to-end anastomosis reestablished the digestive flow. GIST was histologically and immunohistochemically diagnosed in five of the nodules, and one nodule presented only inflammatory reaction. The patient had an uneventful follow-up and was discharged from the hospital on the third post-operative day, after accepting regular food. During the 38-month-follow-up from the second procedure, no adverse event has been identified. The patient is working and has a normal life with

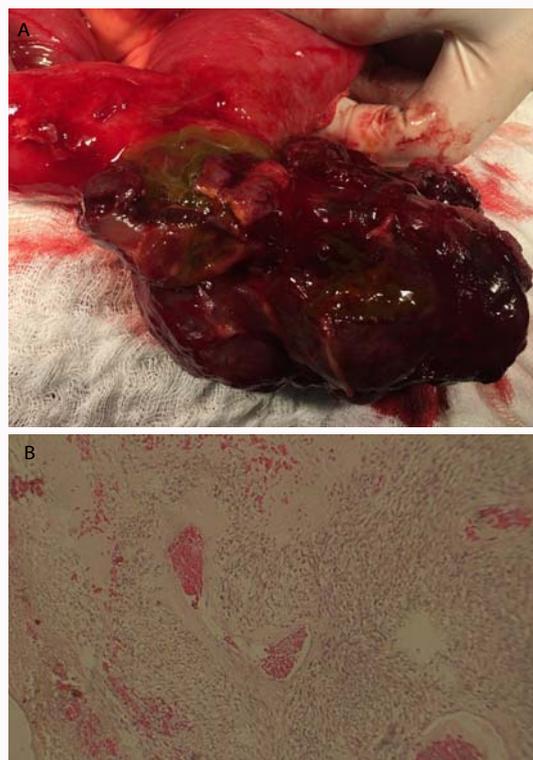


Figure 1: Gastrointestinal Stromal Tumor (GIST) inside a jejunal true diverticulum. A) Surgical view of the perforated tumor inside the jejunal diverticulum. B) Tumor microscopy showing a fusiform cellular neoplasm consistent with GIST, expressed by CD 117 leukocyte antigen immunohistochemistry. (HE, x100).

no medical complaints. The GIST is still under control with 800 mg imatinib.

Discussion

Small bowel diverticulum was first described by Soemmering and Baille in 1794, which today is not recognized as a true but a pseudo diverticulum. Since then, large series of this anomaly have been published [1,2]. However, the prevalence of true diverticulum is difficult to ascertain with the diversity of data without histological support. It is necessary to distinguish a true diverticulum from Meckel's diverticulum and small-bowel duplication [2-4]. True intestinal diverticula have been identified in only 0.06% to 4.6% of all autopsies and 0.02% to 5% of roentgenographic studies [1,2]. Considering the non-Meckeli diverticula, approximately 80% occur in the jejunum, 15% in the ileum, and 5% in both. Diverticula of the jejunum tend to be multiple and larger than those of the ileum, which are solitary [4-8]. Congenital diverticula are rare, and true diverticula contain muscularis propria, whereas acquired pulsion diverticula are false or pseudo diverticula. The pathogenesis of true diverticula is not well-established, although embryologic examination in the second month of gestation demonstrated small bowel diverticula occurring mainly on the anti-mesenteric surface [1-3]. Jejunal diverticulosis is often asymptomatic and is only occasionally found. When present, the symptoms and signs are non-specific, including abdominal pain, dyspepsia, anemia, and transient small bowel obstruction. Severe complications are rare and consist of diverticulitis, perforation, acute intestinal obstruction, and digestive bleeding [1,2,4,6,8]. No evidence of familial small bowel malignant disease was found in the literature. Laboratory abnormalities are not specific. Routine screening of

patients for occult blood in the stool may provide an earlier detection of these tumors. Upper gastrointestinal CT presents over 90% sensibility to detect a jejunal disease comprised of diverticula and tumors. Curative treatment of jejunal GIST entails a wide resection of the bowel and adjacent mesentery. Distant metastases, mainly in the liver and pelvis, must be investigated. The overall survival for GIST, as low as 30% after 5 years, is related in large part to a delay in diagnosis and to advanced stage tumors at the time of the surgical approach. However, if the operation is undertaken before lymph node metastasis, a five-year-survival is higher than 80% [4,6-8]. The patient described in this report presented a stage III GIST but without detected mesenteric lymph node metastasis during the first surgical approach. Long-term survival after resection and adjuvant chemotherapy indicates an adequate oncological control. Surgery, followed by imatinib chemotherapy, is the mainstay treatment for GIST. Imatinib, a tyrosine kinase receptor inhibitor of KIT activity, causes apoptotic GIST cell death.

Conclusion

A GIST may be found inside a jejunal true diverticulum. Due to the absence of symptoms or signs in the early stages of the tumor, the diagnosis tends to be delayed. The treatment must be an en bloc resection of the diverticulum, including a segment of jejunum and its mesentery, with all lymphatic tissue. Adjuvant imatinib chemotherapy is indicated to control this disease and prevent recurrences.

Acknowledgment

The authors gratefully thank the Research Support Foundation of the State of Minas Gerais (FAPEMIG) and the National Council for Scientific and Technological Development (CNPq).

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