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Research Article

GIANT MASS OF THE SMALL BOWEL: DIAGNOSIS AND TREATMENT (A CASE REPORT AND LITERATURE REVIEW)

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A R T I C L E I N F O

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A B S T R A C T

The giant masses of the small intestine have the problem of diagnosis of the etiologies and therefore the therapeutic management because of the endoscopic examination limitation and then diagnostic delay. They can be benigns or malignants, mucosal or mesenchymal tumors. The clinical symptoms of GISTs vary from mild to severe, and complications include vague abdominal pain, hematemesis, and intestinal obstruction. Computed tomography is the key exam for diagnosis, extension assessment and monitoring. The early diagnosis of small intestine gastrointestinal stromal tumors (GISTs) is difficult. Hereby we present a 36-year-old male patient who presented a giant mass of the small intestine of 25 cm of the size without metastases or any sign of tumor spread. The aim of this case study is to notify the difficulties encountered to set up the etiology of giant masses of small intestine and the contradiction of our case with literature which emphasizes on the presence of metastasis when GISTS are more than 10 cm of size.

INTRODUCTION

Considered as a "black box" in the GI tract, the small bowel has been longtime inaccessible to the endoscopist because of its anatomy, location and tortuosity. Small bowel tumors are relatively rare and account for 3%-6% of all digestive neoplasms, and represent only 1.1%-2.4% of gastrointestinal malignancies (1) [Arata er al]. This low incidence may be ascribed to its unique physiological features, which include an alkaline environment, fluidity, low bacterial count and a high level of IgA in the small intestine. The diagnosis and management of small bowel tumors are formidable tasks for physicians. Approximately two-thirds of GISTs in the small intestine are 5 cm or more in diameter at the time of diagnosis and rarely less than 2 cm (2) [chen-2013-double balloon].

Observation

It is a 36-year-old male patient, with chronic tobacco and alcoholic intoxication, who presented abdominal pain located in umbilical region, with upper GI tract hemorrhage of medium abundance, asthenia, weight loss. The physical examination found an abdominal mass in umbilical region of firm consistency, mobile in the abdominal cavity and measuring about 20 cm of diameter, with normal rectal examination. The abdominal CT with PCI injection and gastrographin ingestion found a circumferential and irregular mass measuring 25 cm and thickening of the small intestine wall with pseudo aneurysmal dilation of the loop (Figure 1). The extension assessment was negative and the tumor markers CA 19.9 and ACE were normal. The patient underwent a surgical treatment. Under supine position and general anesthesia a laparotomy was performed and the surgical exploration found a giant mass (figure 2) of the small intestine of 25 cm (jejunum) after omentum retraction, at the anti-mesenteric side (figure 3), the resection of the giant mass with a jejunum portion was performed (figure 4) with end to end anastomosis and abdominal wall closure with 3/0 silk suture without abdominal cavity drainage. The post-operative was uneventful. The patient retakes the intestine function at D3 and was discharged from hospital at 5th day. The patient was seen after 15th day, a month and three month completely recovered.

The histopathological result of the specimen found a spindle cell tumor. There were 10 mitotic rate indexes. The immunohistochemical study showed that the tumor cells were marked by the anti CD 117 and anti CD 34 Ab. there were no vascular emboli or perineural sheathing of the tumor.

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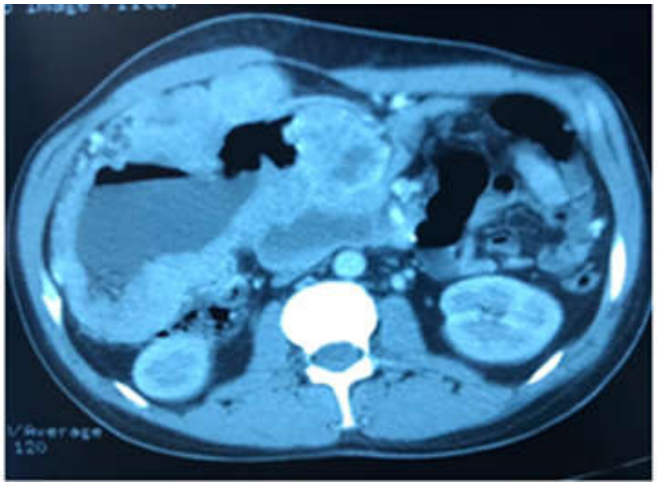

Figure 1 large GIST mass on CT of abdomen

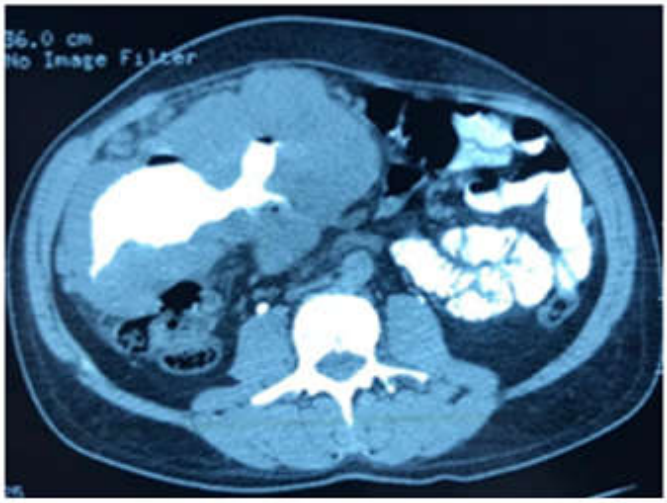

Figure 2 CT of the abdomen with contrast ingestion which shows a large mass depending to the small intestine with the contrast product inside

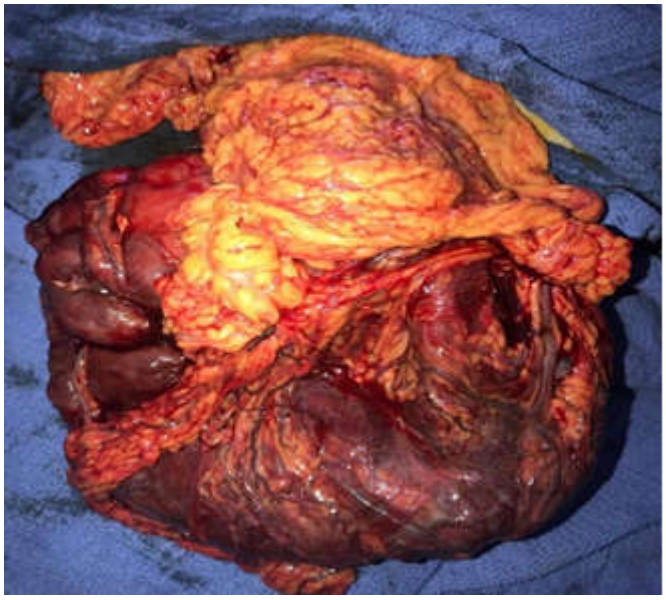

Figure 3 specimen of the mass and the small bowel after complete resection taking a small portion of great omentum

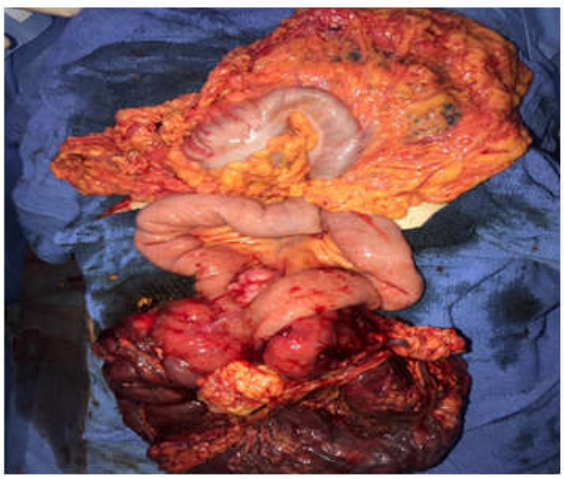

Figure 4 Peroperative image; note the great omentum with colon upper reclined.

DISCUSSION

The giant masses of the small intestine causes a problem of diagnosis and therefore of management because of the limit of paraclinical examinations. It can be a benign or malignant, mucous or mesenchymal tumor. Primary malignant tumors of the small intestine are rare, comprising less than 2% of all gastrointestinal tumors (3) [Takahashi *et al*. 2014]. Gastrointestinal stromal tumors (GISTs) are the most common submucosal mesenchymal tumor of the alimentary tract. Their preferred localization is commonly in the stomach (50–70%), followed by the jejunum and ileum (30–40%), duodenum (5%), colon (4%), and esophagus and appendix (1%).The GISTs occur at ages 50s to 60s and arise from Cajal interstitial cells, which are the pacemaker cells located at Auerbach's plexus of the alimentary tract.(4) [Inoue *et al*.]. GISTs rare tumors: 1 to 3% of gastrointestinal malignancies. They are usually sporadic; incidence is about 15 cases/million inhabitants/year, median age at diagnosis 60 years, and sex ratio 1. They are derived from Cajal cells or one of their precursor, and their phenotype is usually KIT + (95%) and DOG-1 + (95%). They typically harbor activating mutations of the genes encoding tyrosine kinase receptors KIT or PDGFRA. GISTs represent a heterogeneous set interms of molecular biology, clinical behavior and response to treatment. Oral tyrosine kinase inhibitors (TKI), originally imatinib, have revolutionized their management (5)[landi 2019]. The clinical symptoms of GISTs vary from mild to severe, and complications include vague abdominal pain, hematemesis, and intestinal obstruction. These non specific symptoms, coupled with the lack of physical findings, often cause a significant delay in reaching a diagnosis. The most frequent symptoms are bleeding into the bowel or abdominal cavity, anemia, and abdominal pain; some patients can present dyspepsia, nausea or vomiting, constipation or diarrhea, frequent urination and asthenia. Hemorrhage, tumor rupture, and bowel perforation or obstruction might need emergency surgery; however, symptoms are not predictive of patient survival (6)[Tziortzoti *et al*.-2016]. Because the small bowel has been a relatively inaccessible area to standard endoscopic techniques, contrast radiography has been regarded historically as the gold standard diagnostic modality (7) [abu-hamda-

2003]. Computed tomography is the key exam for diagnosis, extension assessment and monitoring. The GIST typically presents itself as a mass with clear limits, with exolumen development and heterogeneous enhancement after intravenous injection of PCI, with sometimes necrotico-hemorrhagic aspect. Their spread is mainly hepatic and mesenteric (6,8). Tumor size is crucial in the prediction of progression of the disease and GIST risk stratification systems are mainly based on tumor size that leads to assessment of the malignancy and has a significant impact on overall survival. The Chinese epidemiological study recorded mean diameter of 5.78 cm (0.3e25 cm). An Egyptian study on gastric stromal tumors which included 16 GIST patients reported tumor sizes between 8.4 and 20 cm. In study of (9) [Al-Thani *et al*.], the median tumor size was 8 cm ranging from 0.4 to 18 cm and 62% of the cases were with tumor size greater than 5 cm. The diagnosis of certainty is histological, the cells are: spindle-shaped in 70% of the cases, epithelioids in 20% and mixed in rare cases. Immunohistochemical confirmation, which makes it possible to highlight an expression of c-KIT thus allowing differential diagnosis with other benign or malignant tumors, namely leiomyomas, leiomyosarcomas, schwannomas, fibrous tumors, carcinomas and tumors endocrines(10) (11).

The treatment of primary malignant tumors of the small bowel is essentially surgical. However, adjuvant chemotherapy may be warranted, depending on the type of tumor especially for GIST tumors. The treatment strategy of GISTs varies depending on size and tumor location. Complete surgical extirpation remains the cornerstone of GIST management and the only curative treatment. The tumor resection with no touch technique by oncologic surgical approach is the only potentially curative treatment(12,13). Segmental resection is the technique for the GIST of small bowel with laparotomy approach. The European Society for Medical Oncology 2004 consensus suggests that GISTs should be resected through a minimally invasive approach only if they are 2 cm or smaller, for there is a risk of capsular rupture using laparoscopic forceps (14)[baiu 2018]. However, tumors as large as 10 cm can be resected laparoscopically but for the large tumors, the laparoscopic is discouraged and laparotomy is the best approach of choice for surgical treatment, as GISTs of the small bowel can be large and vulnerable (15)[Boonstra *et al*. 2019]. Lymph-node dissection is usually not indicated (lymph node metastases risk: 1%) except in pediatric GISTs (5)[Landi *et al*.-2019]. Even though tumor size is related to the prediction of progression of the disease and a high risk of metastasis and overall survival, there is some particular cases where the giant GISTs can remain closed without rupture or metastasis even if there have more than 10 cm as in our case. For these cases, there is then a problem in decision for the therapeutic management whether to begin by surgical treatment or chemotherapy. These cases might be discussed in oncology staff for decision. Unresectable metastatic or recurrent GIST can be treated with a tyrosine kinase inhibitor, imatinib, with a remarkable response (50%e70%) and prolonged survival (median progression-free survival: 18-20 months; median overall survival: 51st 57th months). The standard approach in the case of tumor progression on 400 mg once per day is to increase the Imatinib dose to 400 mg twice per day as permitted by toxicity. Use of a second-line targeted agent, sunitinib, in patients with advanced GIST who fail (or are intolerant of) imatinib therapy is advised. The 5-year overall survival rate after complete resection of GISTs is 50%e-65% according to (16)[Lai *et al*.]. When Sorour *et al*. reported 3 and 5 years disease free survival for all GIST patients as 73.2% and 64.5% respectively (10)[H. Al-Thani *et al*.]. The prognosis is defined by the size, the mitotic index and the location of the tumor. The estimation of the risk of recurrence is essential for the adjuvant chemotherapy treatment, and to adapt the long term surveillance. Imatinib is indeed the standard chemotherapy treatment after a complete resection of a GIST with high or intermediate risk of recurrence (12,17).

CONCLUSION

The giant masses of the small intestine have the problem of diagnosis. Gastro intestinal stromal tumors are rare tumors of the GI tract (less than 3%) with non specific symptoms and coupled with the lack of physical findings which lead to a significant delay of diagnosis. The curative treatment is surgery. Lymph node resection is not usually recommended and the margins of 2 cm are supposed to be sufficient for oncologic resection. Some particular cases of GISTs can reach a great size without rupture or metastasis and their treatment management may be discussed.

Consent

As per international standard, patient's consent has been collected and preserved by the authors.

Ethical Approval

As per international standard, written ethical approval has been collected and preserved by the author(s).

Declaration of Interests

The authors declare that they have no conflicts of interest in connection with this article.

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