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Stromal Tumor of the Jejunum Complicated by Hemorrhagic Shock-A Case Report

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Authors' contributions

This work was carried out in collaboration between all authors. Author YF designed the case study, and wrote the first draft of the manuscript. Authors AEB and DK managed the discussion of the case study. Author DK managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Stromal tumors of the small bowel, commonly known as GIST (Gastrointestinal Stromal Tumors) are mesenchymal tumors of uncertain prognosis. They develop in the wall of the digestive tract. They are usually asymptomatic, incidentally discovered during endoscopy or during surgery. Identifying gastrointestinal stromal tumor is facilitated by a relatively specific marker, c-kit. Diagnosis is confirmed by histological examination of the surgical specimen. They pose two problems first is to confirm the diagnosis and second problem is to assess their evolutionary potential and customize the therapeutic management.

We report the case of a patient aged 34 years with no history of prior illness. He was admitted a year ago in the emergency room in a state of hemorrhagic shock due to lower gastro intestinal (GI) bleeding for three days. The patient underwent conservative therapy with I.V fluids and blood transfusions. An endoscopic assessment consisting of upper and lower GI(UGIE and colonoscopy) was done but that did not reveal any abnormalities.

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Surgical exploration revealed a tumor 1 meter distal to the ligament of Treitz. Rest of the exploration was unremarkable. The tumor along with small bowel was resected and end to end anastomosis was performed. The postoperative course was uneventful.
Histological study of the surgical specimen confirmed that this was a stromal tumor with low potential of malignancy. The staging did not reveal any secondary lesions. Stromal tumors are often asymptomatic, lower GI bleeding is an exceptional way of presentation. Complete resection of the tumor is the treatment of choice. The recent development of targeted therapies and molecular biology is a new hope in the treatment of these tumors. The aim of this case study is to review the diagnostic and therapeutic aspects of this disease, and explain that hemorrhagic shock can be due to jejunal stromal tumor.

Keywords: Small bowel; lower GI bleeding; c-kit; imatinib; GIST.

1. INTRODUCTION

Gastrointestinal stromal tumors (GIST) are mesenchymal tumors that develop from connective tissue of the wall of the digestive tract organs. They account for 85% of the mesenchymal tumors of the gastrointestinal tract. These rare tumors are often localized in the stomach (70%) and small intestine (20-30%).

They grow from Cajal cells which form a network interposed between the muscular and nervous plexus of the digestive tract involved in the regulation of cellular motility.

Stromal tumors may be asymptomatic, incidentally discovered during a morphological examination or surgical exploration, they can be revealed by hemorrhage. If there is a high index of suspicion then the diagnosis can be confirmed by the typical echoendoscopic appearance.

The diagnosis is confirmed by histological study. These tumors have a known potential for malignancy and their prognosis is poor when the tumor size is large and the mitotic index is high.

2. CASE

We report the case of a patient aged 34 years that had no past medical history or surgical operation. He was admitted in the emergency room in a state of hemorrhagic shock due to lower GI bleeding that had started three days before his admission.

Clinical examination revealed a patient in poor general condition, generalized cutaneous and mucosal pallor, thready pulse, respiratory rate was 40 cycles / min. The rest of the physical examination was unremarkable. The patient was stabilized with I.V fluids and blood transfusions. Endoscopic assessment (Upper GI echoendoscopic & colonoscopy) did not reveal any abnormalities.

Lower gastrointestinal bleeding persisted and an arteriography was indicated but could not be done due to its unavailability in the hospital.

Surgical exploration was decided. The exploration revealed a tumor 3 cm in its long axis, 1 meter distal to the ligament of Treitz. During surgery, possibility of a stromal tumor was considered, the rest of the exploration was unremarkable. Resection of small bowel along with the tumor was performed. Gut continuity was obtained by end to end anastamosis (Fig. 1).

Postoperative course was uneventful. There was cessation of lower gastrointestinal bleeding with clinical improvement of the patient.

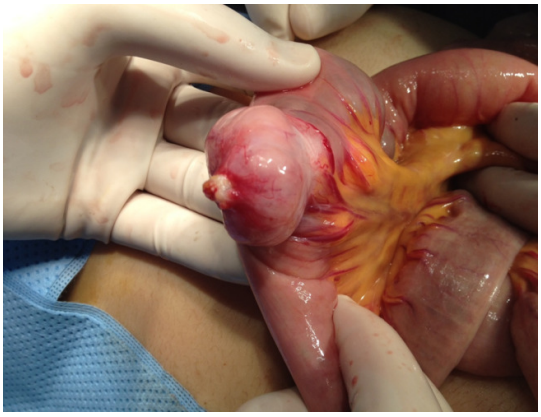

Fig. 1. Jejunal tumor located one meter from the first jejunal loop

Pathological study of the surgical specimen: Macroscopic examination showed a submucosal tumor 30mm in its long axis, nodular, with ulcerated surface. Histological examination revealed a proliferation of spindle cells with

eosinophilic cyotoplasm. Regular but poor mitosis. Mitotic index was less than five mitoses per 50 high power fields. Tumor cells expressed CD117 with diffuse cytoplasmic staining in the immunohistochemical study. The staging did not reveal any secondary lesions (Figs. 2 and 3).

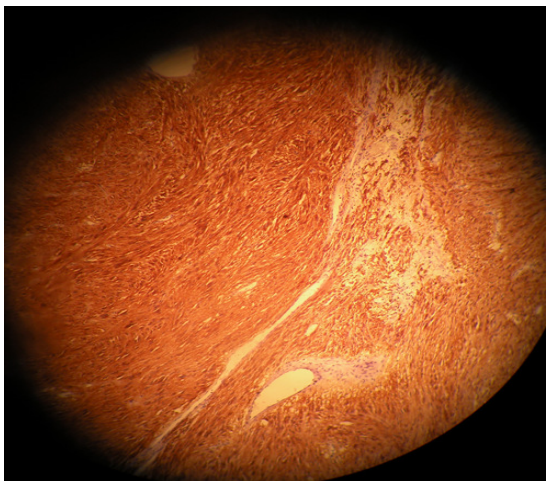

Fig. 2. Histologicalstudy: Proliferation of spindle cells with eosinophilic cyotoplasm
(magnification X 40)

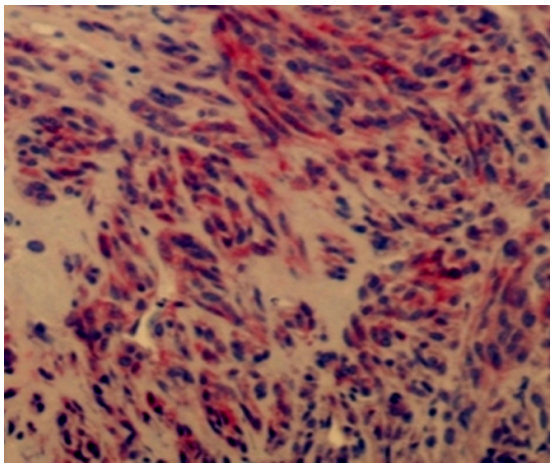

Fig. 3. Immunohistochemical staining of the jejunal stromal tumor by protein cd 117
(magnification X 80)

3. DISCUSSION

Gastrointestinal stromal tumors are circumscribed solid tumors, that develop in the wall of the digestive tract, the age of onset of these tumors is between 50 and 70 years, with a male preponderance.

Neurofibromatosis TYPE 1 and triad carney are predisposing risk factors. We must emphasize the existence of familial forms [8].

They can grow on the entire digestive tract with a predominance of gastric and small bowel locations. Other extradigestive locations such as greater omentum, mesentery, retroperitoneum are known but rare. Stromal digestive tumors (SDT) occur often in the jejunum, ileum and duodenum.

The presentation of gastrointestinal stromal tumors varies according to their origin and their aggressiveness. Therefore, the SDTs may be asymptomatic and discovered incidentally or manifest as gastrointestinal bleeding or abdominal pain, or more rarely by obstructive symptoms or perforation of the gastrointestinal tract [1-13]. The anemia associated with occult bleeding can be a call sign [10]. They can produce a biological inflammatory syndrome due to cytolysis and cholestasis can occur if there is secondary liver damage, mild leukocytosis is seen if there is an infectious complication of the tumor. Imaging has an important place in the management of SDT. Despite their various presentations, these tumors have a very clear diagnostic and therapeutic approach for appropriate management. Image guided transparietal biopsy should be avoided because it can become a source of peritoneal contamination. Imagery after treatment is useful to evaluate therapeutic efficacy and detect relapses [2]. Abdominal ultrasound can show a SDT if it is large.

Endoscopic ultrasonography can be used to determine the local extension in case of esophageal, gastric and rectal tumors. In the CT scan SDTs appear as exoluminal masses of variable sizes and sharp edges. Their density and tissue homogeneity are variable. Aggressiveness of SDT (stromal digestive tumor) is suggested by a large size, irregular contours, invasion of adjacent organs and densification of the adjacent fat [3]. MRI allows the study of mesenteric extension and search of liver metastases. Positron emission tomography [10] is a very sensitive test for the assessment of tumor metabolism. It can be used for initial staging and later to assess the therapeutic response to imatinib, [3,9]. Endoscopy is useful for diagnosing colic or gastric stromal tumors, and often is diagnostic if the growth has an intraluminal component.

The capsule video, entero MRI, CT enteroclysis, double-balloon enteroscopy are some more sensitive tests to visualize stromal tumors of small intestine [4].

The diagnosis is based on anatomopathological study. Macroscopically stromal tumors develop from the muscular layer of the gastrointestinal tract.

They are usually very circumscribed, formed by a fasciculated tissue, sometimes surrounded by a pseudo capsule.

They are often associated with ulceration of the mucosa that may explain the gastrointestinal bleeding.

They may have an endophytic component which grows into the intestinal lumen, exophytic component, or both components (mixed). Their size may vary from a few millimeters to more than 30 cm [11,3].

Microscopically the tumor consists of proliferation of spindle cells (rarely épitheloid cell); strongly positive immunohistochemistry for c -kit (cd117).

c-kit is responsible for receptor tyrosine kinases (kit or cd117) that is widely involved in genesis of GIST.

Searching mutations in the target genes is only necessary if the tumor is c-kit. Negative [1,7].

The malignant potential is difficult to assess for SDTs, even for small tumors less than 2 cm in size, several prognostic factors have been reported [10]:

Location:- Gastric stromal tumors have a better prognosis.
Tumor size:- Tumors larger than 5 cm are usually malignant.
Intratumoral necrosis:- Suggests adverse prognosis.

The size of the tumor cells, the number of mitoses, aneuploidy and cell proliferation index with a threshold of 2 to 3mitosis/ 10 fields with high power fields(x 400), all suggest adverse prognosis. In practice if there are less than one mitosis per 10 fields, tumors are considered benign, if there are more than 4 mitoses per 10 fields the tumors are considered to be malignant.

Mitotic activity should be evaluated in 20 high-power fields (5 mm sq). If there are 5 or less mitoses per hpf and the tumor is smaller than 2 cm: no risk; if the tumor is between 2 and 5 cm: low risk; if the tumor is between 5 and 10 cm: moderate risk; if the tumor is greater than 10 cm: high risk. If there are more than 5 mitoses per 20 hpf, the tumor is high risk, regardless of its size.

The tumor that we are presenting was 3 cm and was interpreted as low-risk; presumably it has less than 5 mitoses per 20 hpf.

Existence of metastases at diagnosis:-

Nevertheless, there are many tumors that are considered "border line". Each stromal tumor should be considered to have malignant potential.

The treatment of stromal tumors is complete surgical resection (the only potentially curative treatment). There is consensus on the optimal margins of resection. A margin of 1 to 2 cm is desirable. Extensive lymphadenectomy is not required because lymph node metastases are rare (less than 10%) and the risk of lymph node recurrence is limited (less than 5%) [5].

The surgical procedure depends on tumor site. In case of small bowel tumor more or less extensive resection is made with immediate end to end anastomosis. In our case we performed a limited resection with anastomosis.

IMATINIB is a selective inhibitor of tyrosine protein kinases, especially c- kit. It is indicated in stromal tumors locally advanced or metastatic.

Its benefit as an adjuvant or neo adjuvant to surgery is being evaluated [6,12].

The progress of aggressive stromal tumor is neither modified by radiation nor by the standard chemotherapy.

The recent development of specific and potent inhibitors of tyrosine kinases (st1570, su11248) is a good alternative.

4. CONCLUSION

Gastrointestinal stromal tumors are the most common mesenchymal tumors of the digestive tract. They may be asymptomatic or manifested by a complication. Jejunal localization is rare. The lower GI bleeding is a way of exceptional presentation of these tumors, because of their exoluminal development. With the current

immunohistochemical means, their diagnosis has become relatively easy. Their prognostic evaluation is not easy. Complete resection of the tumor is the treatment of choice. The recent development of targeted therapies by molecular biology is a new hope in the treatment of these tumors.

CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

All authors declare that this manuscript have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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