Gastro-Intestinal Stromal Tumor Complicated by an Acute Intestinal Occlusion: A Case Report

Abstract

GIST or Gastrointestinal stromal tumors are rare connective tumors, Derived from Cajal cells or one of their precursors. And they are often located in the stomach or small intestine.

Gastrointestinal stromal tumors (GISTs) are very rare neoplasms of the digestive tract, their clinical symptomatology is very variable according to the region affected or the size of the tumor, they could be responsible for abdominal mass, pain, or, digestive bleeding, as well as an alteration in the general state of health. And not very often, the diagnosis is made on the occasion of an intestinal occlusion.

the GIST complicated by occlusion due either to invagination of the tumor mass in the small bowel or to exophytic development of the tumor.

In CT scans, they are manifested by a localized parietal tumor thickening of variable size with most often an exophytic development, the diagnosis is based on molecular biology.

The main treatment is radical surgery, but The Imatinib has revolutionize the prognosis of patients with unresectable and/or metastatic GIST.

We report the case of a patient with a stromal tumor complicated by occlusion operated in our emergency department of visceral surgical in Ibn Rochd UHC.

1-Introduction:

Gastrointestinal stromal tumors (GIST) account for up to 2% of gastrointestinal neoplasms. They may appear anywhere in the gastrointestinal tract and are most frequently found in the stomach (60%). The diagnosis is made based on the expression of the tyrosine-protein kinase KIT CD117.[1]

The clinical symptomatology is different according to the segment of the digestive tract affected or the size of the lesion, the diagnosis of an intestinal obstruction is very rares [2]. The CT scan is the average paraclinical diagnosis [3].

Surgery is the treatment of reference in localized forms, and must be done urgently in GISTs complicated by occlusion. Imatinib is the standard first-line treatment in metastatic gastrointestinal stromal tumors, as well as adjuvant treatment after surgery [4].

We report the case of a patient with a complicated stromal tumor of occlusion operated in emergency at the visceral surgical department of Ibn Rochd UHC.

2-Case presentation:

A 36 years old male, without any particular pathological history, had presented for the last 48 hours an occlusive syndrome associated with vomiting and hypogastrium pain, without any external hemorrhage.

At the physical examination in admission, the patient was conscious, stable on the hemodynamic and respiratory level, with normocolored conjunctiva, the abdominal examination had revealed a distended abdomen, with the palpation of a mass in the hypogastric region.

the abdomen X rays showed hail water aerial levels, the abdominal CT scan showed intestine

distension of 44mm upstream of a parietal thickening and the presence of a tissue process of 87x60x71mm.

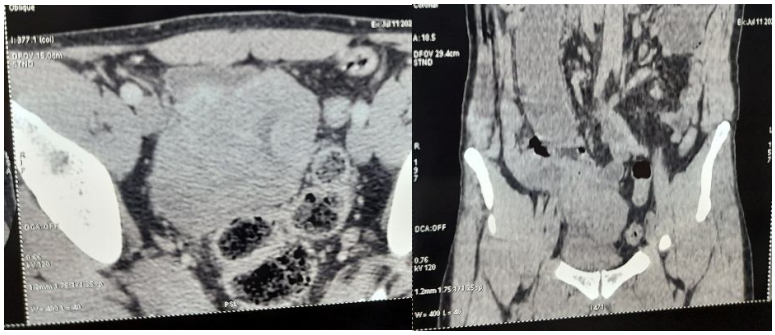

Figure 1

the surgical exploration had revealed the presence of an intestinal tumor with 8 cm in major axis, located 2m8d0 from the duodeno-jejunal angle responsible for an intestinal distension upstream at 4cm (Fig.2), the operation consisted of an intestinal resection including the tumor with a termino-terminal anastomosis , the postoperative follow up was simple, the patient leaved hospital 6 days after, the anatomo-pathological examination showed a morphological and phenotypical aspect (expressing C-Kit, DOG 1 and AML in an intense way) of a gastrointestinal stromal tumour measuring 11cm of major axis with a mitotic index of 5 mitoses for 50 fields, and healthy intestinal exeresis limits and healthy circumferential exeresis limits.

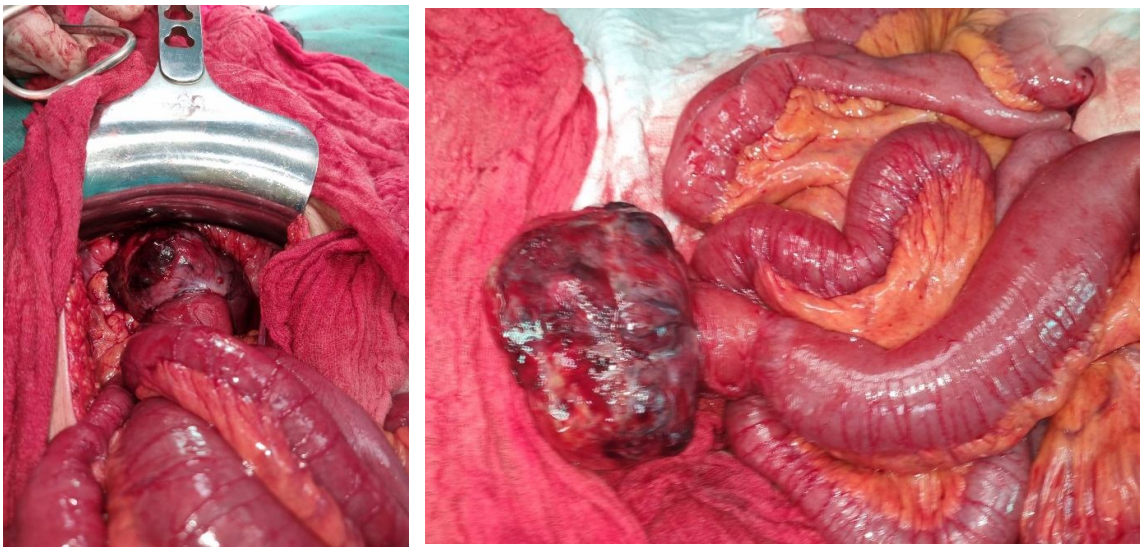

Figure 2

3-Disscussion:

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal neoplasms, accounting for 1% to 2% of all neoplasms of the gastrointestinal tract.[5]. The incidence of GISTs is estimated at

12 cases per million people per year [4]. The middle age of occurrence is between 50 and 60 years with an equal distribution between men and women.

GISTs occur most often in the stomach (50%-70%), followed by the small intestine (25%-35%), the colon and rectum (5%-10%) and the esophagus (<5%) Concomitant metastases occur in 25-30% of cases, often in the liver and mesentery, and are often associated with a poor prognosis [5].

GISTs originate from the interstitial cells of Cajal that reside in the myenteric plexus in the muscular ayer of the gastrointestinal tract [6]. Macroscopically, GISTs are generally a limited tumor, with extra-parietal development, of firm consistency, fish flesh colour with frequent hemorrhage [7].

GISTs are histologically characterized by a significant proliferation of cells, most often spindle-shaped or epithelioid. The diagnosis must be confirmed by immunohistochemical [7].

These cells regulate intestinal motility and act as stimulators of the gastrointestinal tract. Cajal interstitial cells are positive, or CD117 positive so CD117 staining is a diagnosis of GIST in 75% to 85% of patients. KIT is a gene that codes for C-kit, C-kit is a proto-oncogene which, when activated, leads to unregulated proliferation of precursor cells. The Platelet-Derived Growth Factor Alpha Receptor (PDGFRA) is a protooncogene similar to C-kit and mutations in this receptor similarly lead to the formation of GIST in 15% of cases [6].

In the literature, when intestinal GISTs are revealed by signs of obstruction, this is usually due either to invagination of the tumour mass or to exophytic development of the tumour [9]. In a series of 92 GISTs, Sorour et al. described 26 cases of small intestinal GISTs that presented as emergencies due to obstruction explained in 12 cases by invagination, in 10 cases by exophytic development of the tumour and in 4 cases by the development of a mesenteric mass [10]. In our case the occlusion was related to an exophytic development.

In imaging, small intestine GISTs appear in CT scan by a localised parietal tumour thickening of variable size with most often an exophytic development, sometimes hourglass-shaped but more rarely circumferential and which may be responsible for an upstream occlusion as in our observation. The tumor sometimes appears heterogeneous in relation to necrotic remodelling and endo lesional calcifications which are rare but evocative. An CT enteroclysis is beneficial, especially for smaller lesions. Satellite adenomegaly is rare. The thoraco-abdominopelvic scanner also carries out a general extension assessment, with first of all the search for secondary hepatic localisations, 25 to 50% of GISTs are being metastatic from the outset. The differential diagnoses are mainly small intestine adenocarcinoma and neuroendocrine tumors [3].

The treatment in resectable primary localised GIST is radical surgery with negative margins [11]. Surgical resection can be performed using laparoscopic approaches with short hospital stays and low morbidity [12]. Surgical treatment may be required urgently in case of occlusion [10]. like in our case. But usually GISTs are associated with a risk of recurrence, and about 40% of patients with potentially curable resections will eventually develop recurrent or metastatic disease. The identification of risk factors for recurrence after primary surgery is crucial for a reliable prognosis,a follow-up schedule, and the selection of patients likely to benefit from adjuvant treatment, aimed at reducing the recurrence of the disease [11].

The main criteria for aggressive behavior in GISTs are based on the presence of invasion of adjacent structures and/or the presence of metastases (obviously malignant cases), as well as on the primary tumor site, size, and mitotic index [11]

The management of gastrointestinal stromal tumors (GISTs) has been completely changed by the introduction of Imatinib, which is a tyrosine kinase receptor inhibitor (TKI) approved for the treatment of adult patients with unresectable positive KITs and/or metastatic GISTs, and which has radically

changed the prognosis of patients by prolonging survival. Sunitinib is the second line treatment after failure of imatinib [13]

Surveillance is an important element for operated, locally advanced or metastatic GISTs. Patients should have a clinical examination with imaging once every three months for two years, then once every six months for two years, and finally once a year. Patients on imatinib should be followed up every fortnight for the first month to assess treatment-related toxicity and then every three months thereafter [14,8].

4- Conclusion:

The stromal tumor are rare digestive tumors, and the bowel obstruction by a stromal tumor is even more rare, and it requires urgent treatment.

The radical surgery is the treatment but the introduction of the Imatinib has improved the prognosis, especially in the locally advanced and metastatic Gist.

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