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

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***Authors' contributions***

*This work was carried out in collaboration among all authors. All authors have contributed to this study and have read and approved the final manuscript.*

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***Case Study***

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## ABSTRACT

Gastrointestinal stromal tumours (GISTs) are rares connective tumors, usually located in the stomach or small intestine. Derived from Cajal cells or one of their precursors.
Gastrointestinal stromal tumors (GISTs) represent 1% to 2% of all neoplasms of the digestive tract, The average age is between 50 and 60 years old, most often occurring in the stomach, Their clinical symptomatology is variable according to the region affected or the size of the lesion, they are responsibles for digestive bleeding, pain, or abdominal mass, as well as an alteration in the general state of health. More rarely, the diagnosis is made on the occasion of an intestinal occlusion. the GIST complicated by occlusion due either to invagination of the tumour mass or to exophytic development of the tumour. In CT scans, they are manifested by a localised parietal tumour thickening of variable size with most often an exophytic development. Molecular biology plays an important role in this disease.
The main treatment is radical surgery .The Imatinib has radically changed the prognosis of patients with unresectable and/or metastatic GIST.
We report the case of a patient with a stromal tumour complicated by occlusion operated in emergency in the department of visceral surgical emergencies of Ibn Rochd UHC.

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## 1. INTRODUCTION

Gastrointestinal stromal tumours (GISTs) are rare, usually sporadic, connective tumors, usually located in the stomach or small intestinel. However, they are the most common sarcomas. [1], They are derived from Cajal cells or one of their precursors [2].

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 [3]. The CT scan is the average paraclinical diagnosis [4].

Surgery is the treatment of reference in localised 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 [5].

We report the case of a patient with a complicated stromal tumour of occlusion operated in emergency at the

## 2. CASE PRESENTATION

A 70-years-old man, a chronic smoker weaned 10 years ago, had presented 3 days before his hospitalisation an occlusive syndrome stopping of stool and gas, associated with vomiting and abdominal pain, without external digestive haemorrhages. At the clinical examination in admission, the patient was conscious, stable on the haemodynamic and respiratory level, with normocoloured conjunctiva, the abdominal examination had revealed a distended abdomen with periumbilical abdominal tenderness, the abdomen X rays showed hail water aerial levels, the abdominal CT scan showed intestine distension upstream of a parietal thickening with pneumatosis in a distended handle (Fig. 1), the surgical exploration had revealed the presence of a intestine tumor with 12 cm in major axis, located 50cm from the duodeno-jejunal angle responsible for a intestine distension upstream (Fig. 2), the operation consisted of a intestine resection including th tumor with a termino-terminal anastomosis , the postoperative follow-up was simple, the patient leaved hospital 5 days afte, the anatomo-pathological examination showed a morphological and phenotypical aspect (expressing CD117 and CD34 in an intense way) of a gastrointestinal stromal tumour of the mesentery invading the intestinal wall measuring 10cm of major axis with a mitotic index of 48 mitoses for 25 fields, and healthy intestinal exeresis limits and healthy circumferential exeresis limits the patient had received adjuvant treatment with imatinib and the scan control was normal.

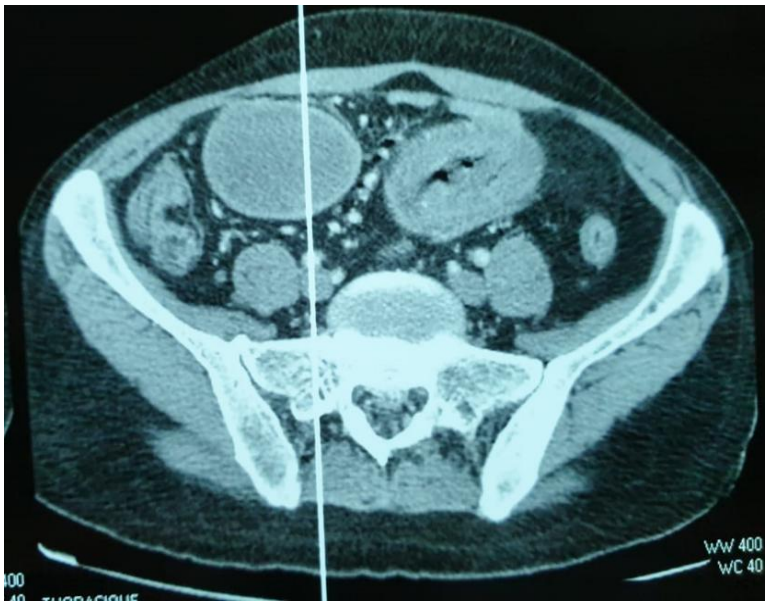


**Fig. 1. Abdominal CT scan showed intestine distension upstream of a parietal thickening**

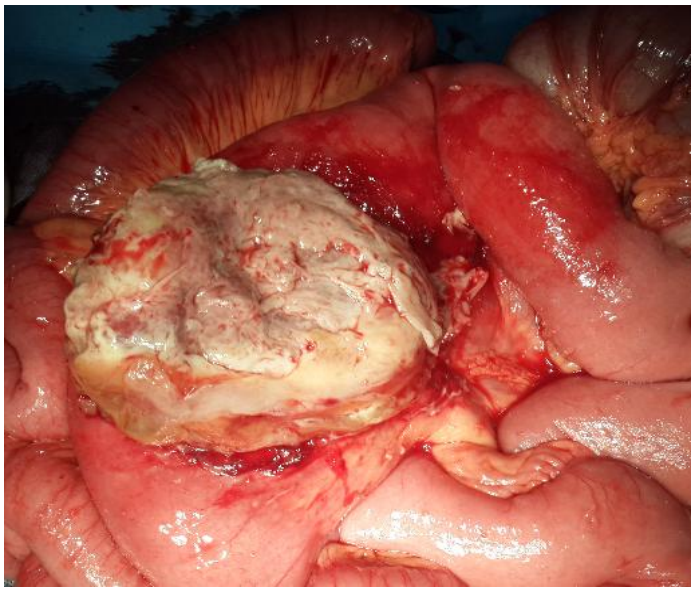

**Fig. 2. Peroperative image showing the stromal tumour with hail distension upstream**

## 3. DISCUSSION

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal neoplasms, accounting for 1% to 2% of all neoplasms of the gastrointestinal tract.[6].

The incidence of GISTs is estimated at 12 cases per million people per year [5]. 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 oesophagus (<5%) Concomitant metastases occur in 25-30% of cases, often in the liver and mesentery, and are often associated with a poor prognosis [6].

GISTs originate from the interstitial cells of Cajal that reside in the myenteric plexus in the muscular layer of the gastrointestinal tract [7]. Macroscopically, GISTs are generally a limited tumors, with extra-parietal development, of firm consistency, fish flesh colour with frequent haemorrhage [8].

GISTs are histologically characterised by a significant proliferation of cells, most often spindle-shaped or epithelioid. The diagnosis must be confirmed by immunohistochemical [8].

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 proto-oncogene similar to C-kit and mutations in this receptor similarly lead to the formation of GIST in 15% of cases [7].

Their clinical symptomatology varies according to the affected area or the size of the lesion. At the small inte stine level, they are discovered by digestive bleeding (haematemesis or melena ) in 40% of cases. They are also responsible for abdominal pain or mass, as well as an alteration in the general state of health. More rarely, the diagnosis is made on the occasion of an intestinal occlusion as in our case [3,9].

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 [10]. 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 [11]. In our case the occlusion was related to an exophytic development.

These tumors remain for a long time with few symptoms. As a result, 10 to 20% are discovered

accidentally and 15 to 50% are discovered at a metastatic stage [8].

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-abdomino-pelvic 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 [4].

The treatment in resectable primary localised GIST is radical surgery with negative margins [12]. Surgical resection can be performed using laparoscopic approaches with short hospital stays and low morbidity [13]. Surgical treatment may be required urgently in case of occlusion [11]. 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 [12].

The main criteria for aggressive behaviour 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 [12].

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 [14] Regorafenib is a third-line treatment in cases of failure of imatinib and sunitinib [9].

Locally advanced tumors often correspond to tumors more than 10 cm in diameter, which are extended to other neighbouring organs in more than half of cases [9], and which could potentially benefit from neoadjuvant imatinib treatment by reducing their size and vulnerability. If the tumor is located at a critical anatomical site, such as the gastroesophageal junction, juxtapancreatic duodenum or lower rectum, the surgical procedure can be reduced from a whole organ resection to a more limited surgical procedure [15,16].

As these tumors are rarely metastasic into the lymph nodes, lymph node removal is usually not necessary [9].

In 20% to 30% of patients, GISTs have already developed metastases in the viscera or peritoneum. The most common sites of synchronous metastases or subsequent recurrences are the liver, peritoneum or both. Pulmonary and bone metastases occur late in life. Due to the multifocality and diffuse nature of the recurrence, this stage of the disease is generally not amenable to surgical resection. Treatment with surgery alone for metastatic GIST is associated with short survival. Imatinib is the main treatment for metastatic or relapsing GIST. Some patients may benefit from surgical resection of a mass, omentectomy or metastasectomy to improve the prognosis, for some multiple liver metastases and/or intraparenchymal liver metastases radio frequency or embolisation is currently used [17].

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 [18,9].

## 4. CONCLUSION

Stromal tumors are a rare digestive tumors. Their prognosis is bad when tumor size is larger and when the mitotic index is higher. They can be complicated by occlusion, which requires emergency surgical treatment. Radical surgery is

desirable. the introduction of Imatinib has radically changed the prognosis of patients with locally advanced and/or metastatic GIST.

## CONSENT AND ETHICAL APPROVAL

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

## REFERENCES


1. Bruno Landi, Tumeurs stromales gastro-intestinales (GIST): Diagnostic et traitements (hors traitements endoscopiques) ; POST'U ; 2019. disponible sur le site. Available :https://www.fmcgastro.org/wp-content/uploads/2019/03/275_019_Landi.pdf
2. Casali PG, Abecassis N, Bauer S, et al. Gastrointestinal stromal tumors: Esmoeurocan clinical practice guidelines for diagnosis, treatment and follow-up. Ann Oncol. 2018;0:1-11.
3. Demuynck F, Morvan J, Brochart C, Blanpain S, Brasseur A, Fuks D, Robert B, Chatelain D, Yzet T. Suivi radiologique des tumeurs gastro-intestinales stromales (GIST) sous traitement : A propos d'un cas, J Radiol. 2009;90:735-8.
4. Demuynck F, Morvan J, Brochart C, Blanpain S, Brasseur A, Fuks D, Robert B, Chatelain D, (5), Yzet T. Suivi radiologique des tumeurs gastro-intestinales stromales (GIST) sous traitement : A propos d'un cas, J Radiol. 2009;90:735-8
5. Sakina Sekkate, Mouna Kairouani, Halima Abahssain, Badr Serji, Saber Boutayeb, Hind Mrabti, Hassan Errihani. Tumeurs stromales gastro-intestinales, Presse Med. 2012;41:917–926.
6. Ravy K Rajravelu, Gregory G Ginsberg, Management of gastric GI stromal tumors:Getting the GIST of it, Gastrointe-stinal Endoscopy. 2020;91:823 -825.
7. Lauren Theiss, Carlo M. Contreras, gastrointestinal stromal tumors of the stomach and esophagus. Surg Clin N Am. 2019;99:543–553.
8. Jean-François Emile, Jean-Baptiste Bachet, Séverine Tabone-Eglinger, Sabrina Brahimi. Histology and molecular pathology of gastrointestinal stromal tumors (GIST), Anatomie et cytologie pathologiques. Revue Francophone Des Laboratoires. 2008;53-58.
9. Tumeurs stromales gastro-intestinales (GIST), info cancer; 2018. disponible sur Available:https://www.arcagy.org/infocancer/localisations/appareil-digestif/autres-cancers-du-tube-digestif/tumeurs-stromales-gist.htm
10. Fourati H, Hmaydia G, Harbi H, Mzali R, Mnif Z. Tumeur stromale gastro-intestinale (GIST) pédiculée révélée par une occlusion intestinaleaiguë. Presse Med; 2016. DOI:http://dx.doi.org/10.1016/j.lpm.2016.06.032
11. Sorour MA, Kassem MI, Ghazal AE-HA, El-Riwini MT, Nasr AA. Gastrointestinal stromal tumors (GIST) related emergencies. Int J Surg. 2014;12(4): 269–80.
12. Piotr Rutkowski, Daphne Hompes, Combined Therapy of Gastrointestinal Stromal Tumors. Surg Oncol Clin N Am. 2016; 25:735–759.
13. Nakamori M, Iwahashi M, Nakamura M, et al. Laparoscopic resection for gastrointestinal stromal tumors of the stomach. Am J Surg. 2008;196(3):425–9.
14. Patrick Scho, Olivier Mir, Bernd Kaspar, Zsuzsanna Papai, et al. Activity and safety of the multi-target tyrosine kinase inhibitor cabozantinib in patients with metastatic gastrointestinal stromal tumour after treatment with imatinib and sunitinib: European Organisation for Research and Treatment of Cancer phase II trial 1317 'CaboGIST'. European Journal of Cancer. 2020;134: 62-74.
15. The ESMO/European Sarcoma Network Working Group. Gastrointestinal stromal tumors: ESMO clinical practice guidelines for diagnosis, treatment and follow-up. Ann Oncol. 2012;23(7):49–55.
16. Margaret von Mehren, Robert S. Benjamin, Marilyn M. Bui, et al, Soft Tissue Sarcoma, Version 2. JNCCN. 2012;10:951–960
17. Valerie P Grignol, Paula M Termuhlen. Gastrointestinal stromal tumor surgery

and adjuvant therapy, Surg Clin N Am. 2011;91:1079–1087.
18. Network NCC. Soft tissue sarcoma. 2018;2. Available:https://www.nccn.org/professionals/physician_gls/pdf/sarcoma.pdf. Accessed November 21,2018.



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