Journal of Education and Practice
ISSN 2222-1735 (Paper) ISSN 2222-288X (Online)
Vol.14, No.9, 2023

Recurrent Retroperitoneal and Subhepatic EGIST – Case Report

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Abstract
Background
GISTs (gastrointestinal stromal tumors) are a common mesenchymal tumor of the gastrointestinal tract. It is a diverse pathology that can occur in any portion of the gastrointestinal tract, from distal esophagus to anus. However, the stomach is the most common origin. GISTs evolve from small neoformations, to a large, metastasizing sarcoma. Their pathogenesis is linked with the KIT or PDGFRA mutations. GIST is to be suspected in cases where a round mass is encountered in close relations to the stomach, intestine or lower esophagus. It has to be differentiated with other neuroendocrine tumors, lymphomas and other rare cancers by means of a biopsy.
Case presentation
The 62 y/o male patient was diagnosed 4 years prior to the current events with GIST for which a surgical procedure of distal pancreatectomy, splenectomy and partial resection of curvatura major of the stomach was performed at another hospital. This was followed by chemotherapy as advised by the treating doctors. The patient presents to our clinic with the complaints of left flank pain for over a week, dysphagia and weight loss. CT shows a giant formation of the left flank, at the splenic lodge, with dimensions of 30 × 32 cm, with close proximity to the left kidney, abdominal aorta, the lienal flexure of colon, the stomach, superior mesenteric artery. Besides this, another 8 × 9 cm subhepatic formation is noted, which has a close vicinity to the portal vein and hepatic hilum. He underwent the surgical procedure for the extirpation of the both masses, also partial resection of curvatura major, vagotomy, Heineke-Mikulicz pyloroplasty, segmental hepatic flexure colon resection and end-to-end anastomosis. The procedure was tolerated well and he was discharged in good health. The following pathology report confirms GIST.
Discussion
Treatment of GISTs and EGISTs consists on the R0 resection of the mass, without the need for radical removal of healthy tissue since these types of tumors do not normally infiltrate adjacent tissue. If other organs are invaded it is recommended to perform an en-bloc resection, taking care to avoid the rupture of the mass, as to minimize the chances of peritoneal dissemination. A prompt adjuvant therapy including the use of selective tyrosine kinase inhibitors (imatinib) is strongly advised. The patient should be followed with periodic controls for recurrence.
Conclusion
There is a clear role surgery can play in improving the outcomes in such patients. However, some authors still debate whether surgery improves results in such patients. For this purpose, it is necessary to involve a team of oncologists, imaging specialists and experienced surgeons in the treatment plan of GIST patients to provide the best treatment.
Keywords: General Surgery, Retroperitoneal EGIST, Hepatic EGIST, GIST.
DOI: 10.7176/JEP/14-9-06
Publication date: March 31st 2023

1. Introduction

GISTs (gastrointestinal stromal tumors) are a common mesenchymal tumor of the gastrointestinal tract. It is a diverse pathology that can occur in any portion of the gastrointestinal tract, from distal esophagus to anus. However, the stomach is the most common origin. GISTs evolve from small neoformations, to a large, metastasizing sarcoma. Their pathogenesis is linked with the KIT or PDGFRA mutations. These two are surface tyrosine kinase receptors that normally are activated upon ligand binding. Mutated KIT or PDGFRA may auto-phosphorylate and induce cell proliferation.

1.1 Epidemiology

It has a low rate of incidence of 14 – 20 per million in northern European countries. The average patient age is

60 – 65 years. Regarding the organ of origin, most of GISTs occur in the stomach (50%), followed by the small intestine (30%), duodenum (5%), rectum (5%) and esophagus (less than 1%). There is a significant number of GIST cases (10%) that are detected in an advanced, metastatic stage, where the organ of origin is not easy to be found. Some authors reject the possibility of GISTs occurring outside the gastrointestinal tract, such as pancreas, liver and gallbladder.

1.2 Clinical signs and diagnosis

The most frequent clinical presentation of GISTs are gastrointestinal bleeding and dyspepsia. GI bleeding may be chronic and develop anemia or with acute events of hematemesis and melena. Other manifestations include palpation of abdominal mass, intestinal obstruction or hemoperitoneum due to rupture.

A significant portion of GISTs are diagnosed incidentally during routine imaging and endoscopic procedures, and in rare instances in rectal examination.

In advanced stages, the most common location of metastases are the peritoneal cavity and the liver.

GIST is to be suspected in cases where a round mass is encountered in close relations to the stomach, intestine or lower esophagus. It has to be differentiated with other neuroendocrine tumors, lymphomas and other rare cancers by means of a biopsy.

1.3 Surgical treatment of GISTs

For a gastric GIST it is recommended to perform a wedge resection in clear margins, whereas for intestinal GISTs it is appropriate to execute a segmental resection.

Total gastrectomy may be necessary for unusually large, multiple or recurrent GISTs.

Many studies confirm that a laparoscopic approach may be as effective as an open surgery in terms of outcome, however, it has the advantage of shorter hospital stay.

It is advised to avoid rupture of the tumor mass or peduncle as to minimize the risk of peritoneal dissemination.

2. Case presentation

2.1 History of present illness

The patient is a 67 years old male with the following medical history and presentation:

He was diagnosed 4 years prior to the current events with GIST (gastro-intestinal stromal tumor) for which a surgical procedure of distal pancreatectomy, splenectomy and partial resection of curvatura major of the stomach was performed at a hospital in Greece. This was followed by chemotherapy as advised by the treating doctors.

The patient presents to our clinic with the complaints of left flank pain for over a week, dysphagia and weight loss. Fibrogastroscopy and IV contrast CT exams were performed (Fig.1). CT shows a giant formation of the left flank, at the splenic lodge, with dimensions of 30 × 32 cm, with close proximity to the left kidney, abdominal aorta, the lienal flexure of colon, the stomach, superior mesenteric artery. Besides this, another 8 × 9 cm subhepatic formation is noted, which has a close vicinity to the portal vein and hepatic hilum.

Consent was obtained from the patient for the proposed laparotomy, and he is prepared for surgery.

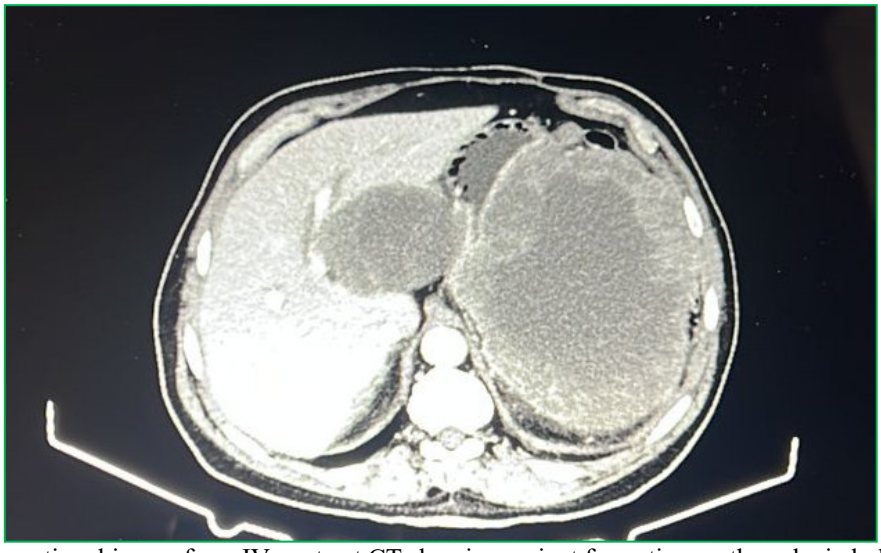

Figure 1. Cross sectional image from IV contrast CT showing a giant formation on the splenic lodge and another mass on the inferior aspect of the liver.

2.2 Details of the surgical procedure

The procedure begins with a general endotracheal anaesthesia. A superior and inferior median incision was made. The peritoneal cavity is accessed, where we evidence a giant abdominal mass on the left portion of the superior abdomen (Fig. 2). It has close contact with the left diaphragm, left kidney, splenic flexure of colon, pancreas body, curvatura major of the stomach, advancing until the cardias portion and esophagus.

A second formation of 8 × 9 cm dimensions is positioned at the subhepatic level, with close contact to the portal vein and vena cava, without infiltrating them (Fig. 3).

We extend the incision to the left side, perpendicularly to the median one. We begin dissection of the formation from the diaphragmatic part, splenic flexure, left kidney (Fig. 4,5). We carefully divide it from the esophageal part, where the vagus is also cut. At the stomach level a partial resection of the curvatura major is performed.

At this point the mass is luxated and we confirm the cleavage plane with pancreas, the superior mesenteric artery and vein, and abdominal aorta (Fig. 6).

A partial colon resection at the splenic flexure and an end to end, two layered colo-colic anastomosis is performed (Fig. 7).

Next, we carefuly dissect at the hepatic hilum, where the second subhepatic formation is divided from the elements of hepato-duodenal ligament (Fig. 8).

Consequently, after cutting the vagus nerve, a Heineke-Mikulicz pyloroplasty is executed.

The procedure ends with lavage and three abdominal drains.

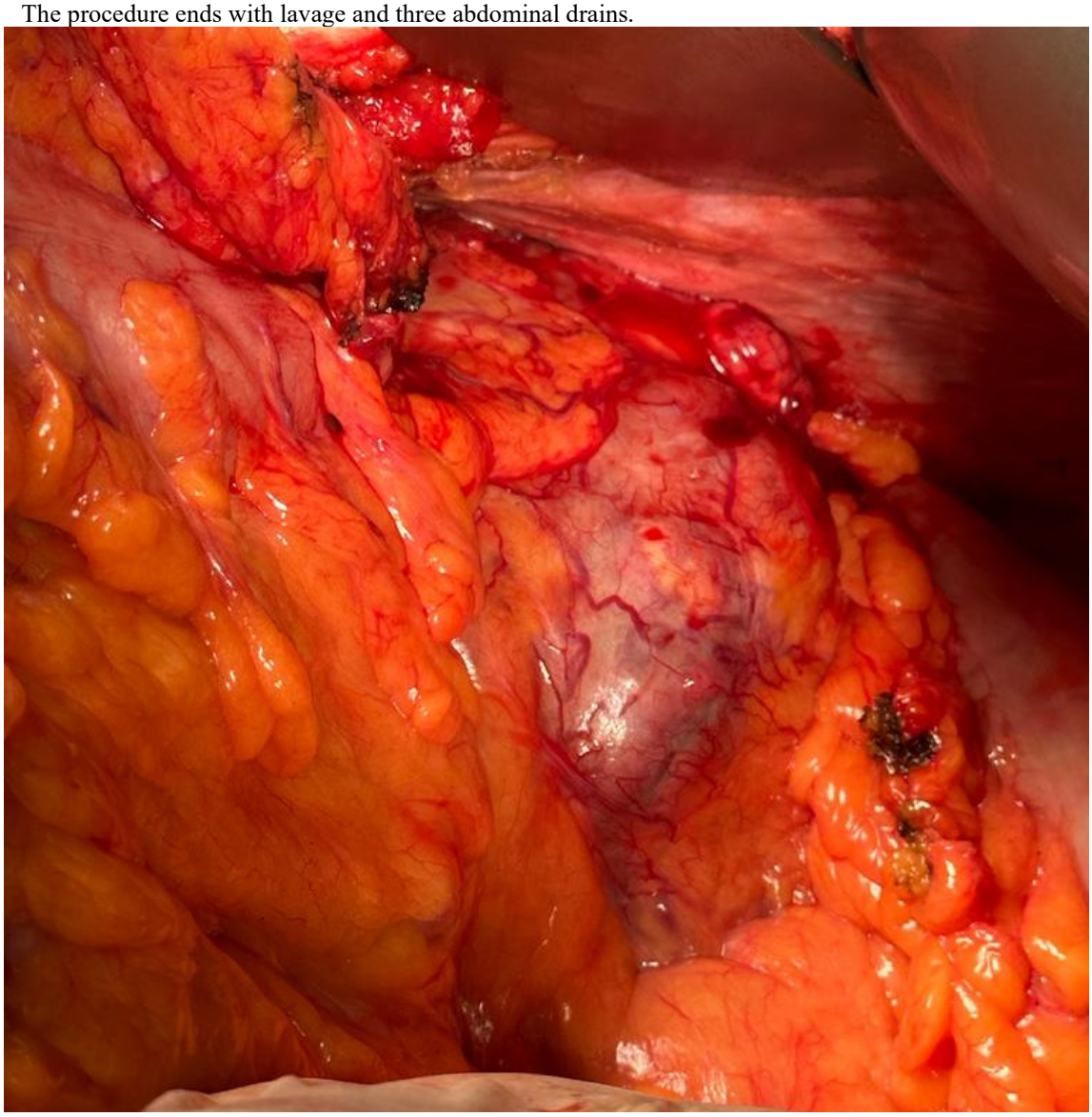

Figure 2. A large retroperitoneal mass on the superior left abdominal portion (splenic lodge) with close contact to the diaphragm, colon, stomach.

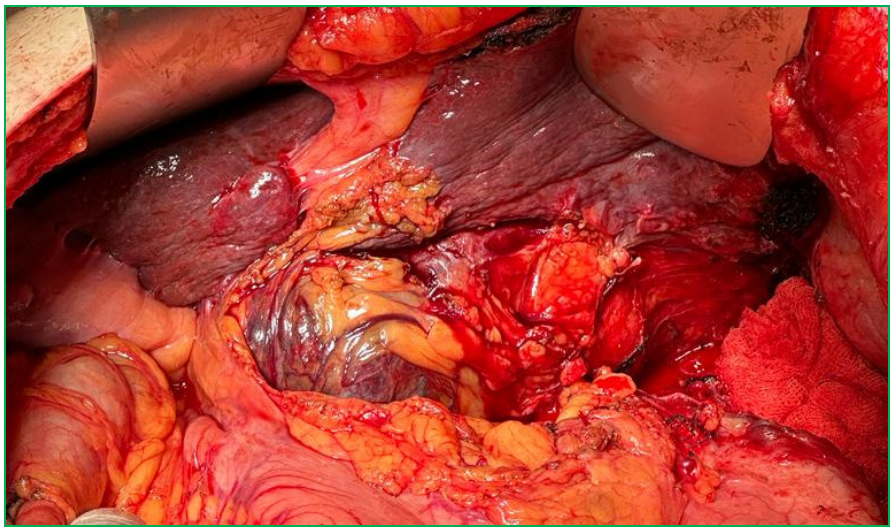

Figure 3. A subhepatic formation of 8 × 9 cm.

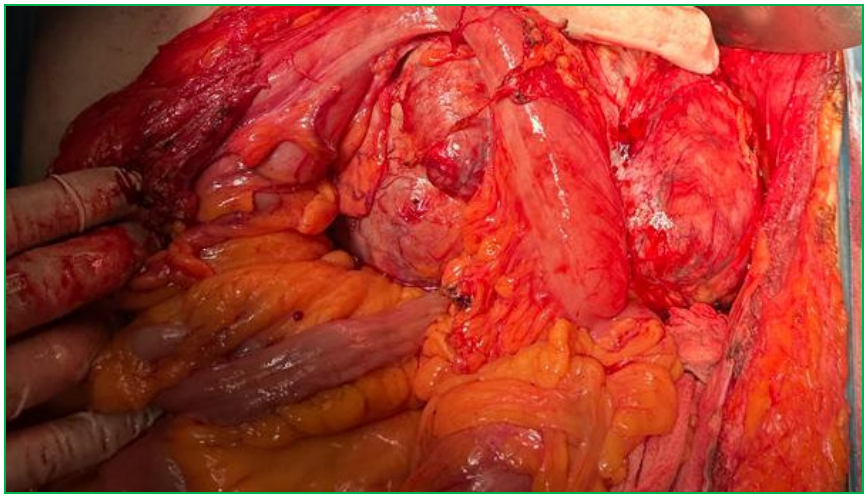

Figure 4. Starting the dissection of the larger mass.

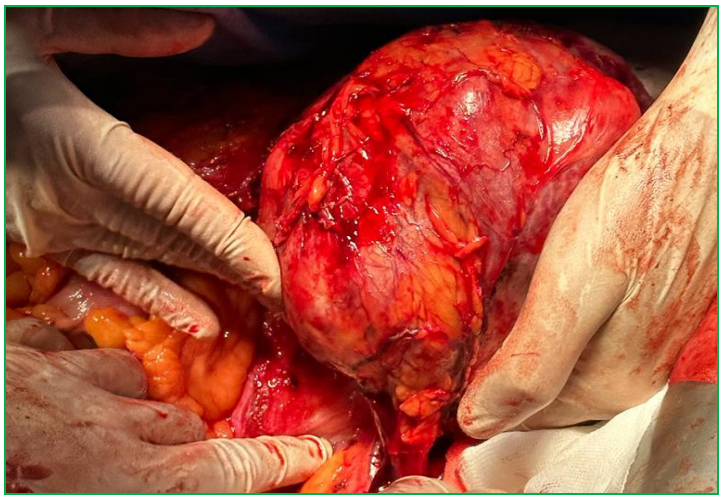

Figure 5. Luxating the larger abdominal mass.

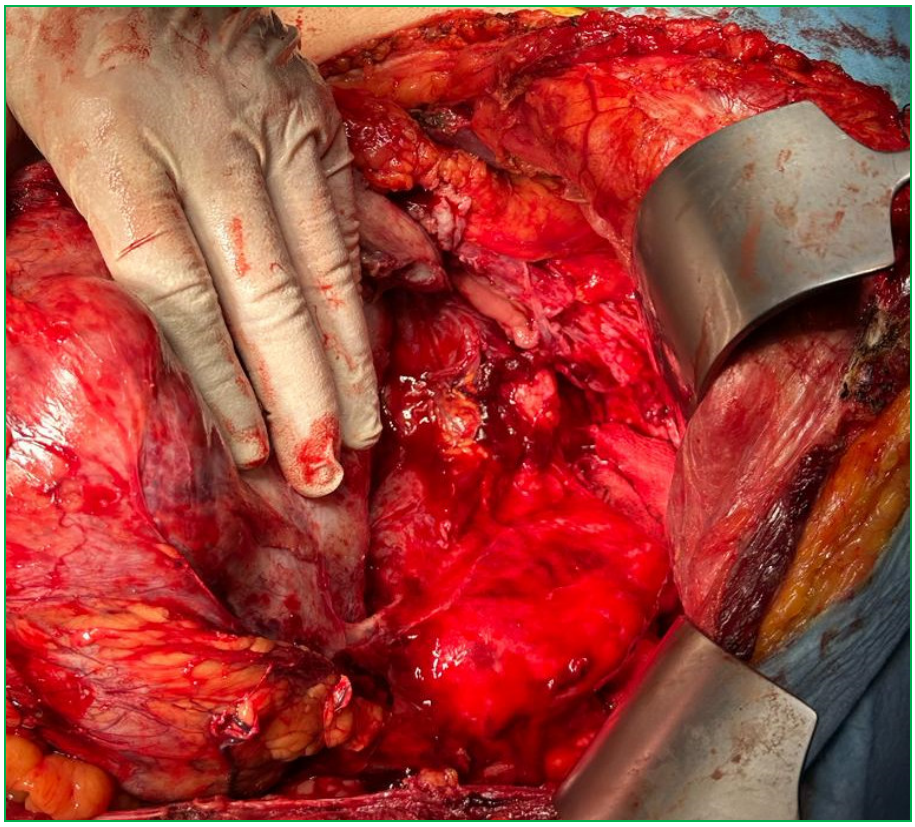

Figure 6. We confirm the cleavage plane with pancreas, the superior mesenteric artery and vein, and abdominal aorta.

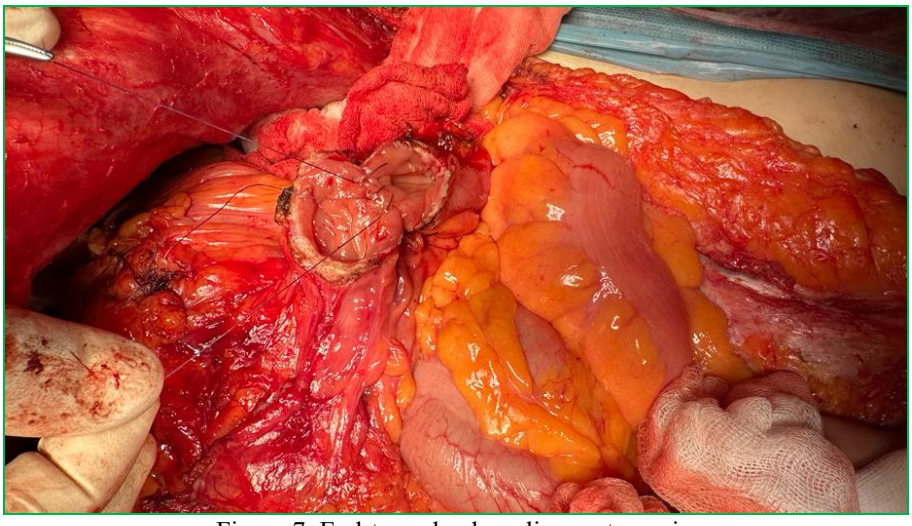

Figure 7. End-to-end colo-colic anastomosis.

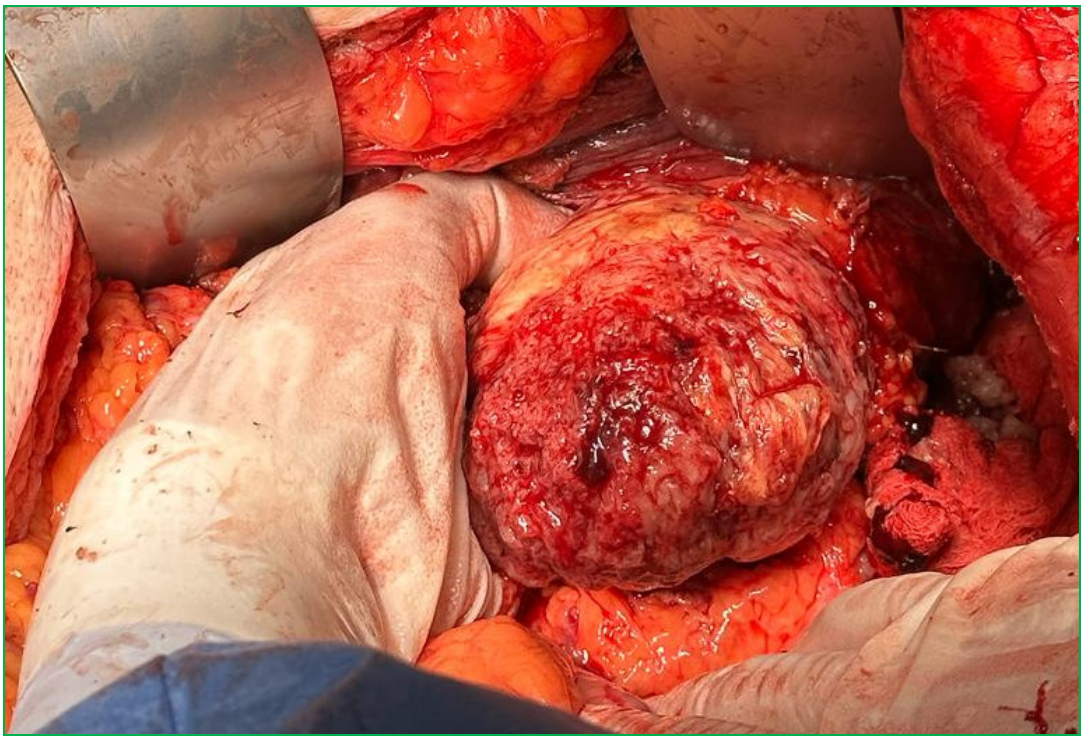

Figure 8. The second subhepatic formation is divided from the elements of hepato-duodenal ligament.

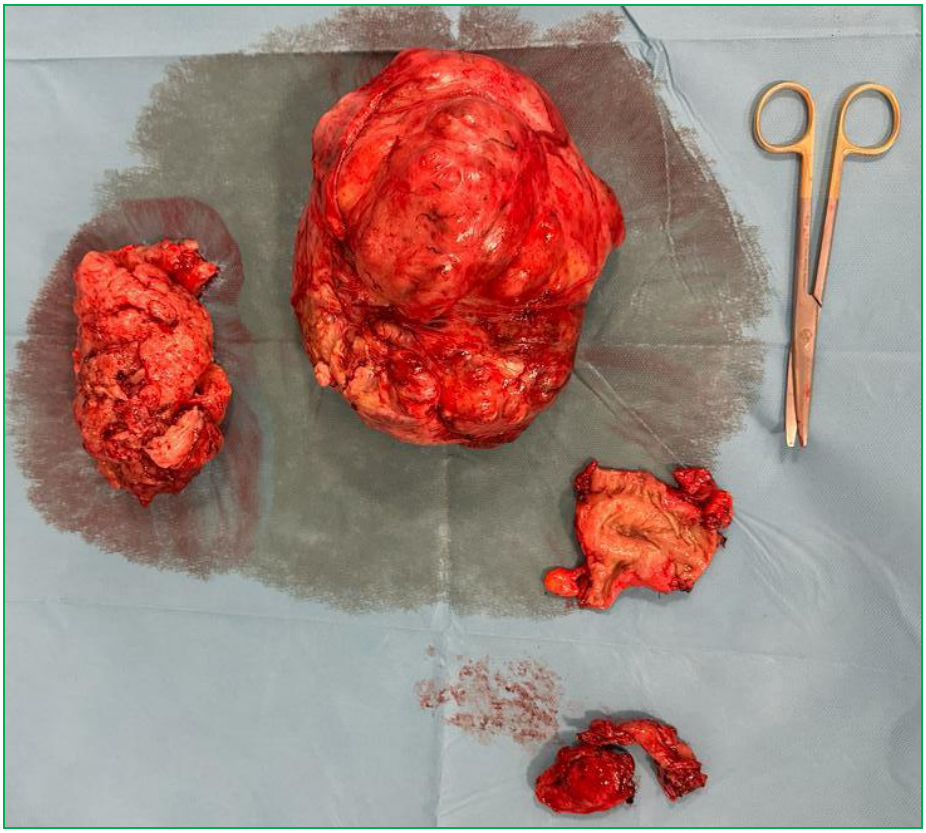

Figure 9. End result, the resected specimen.

2.3 Post-operative period

The patient tolerated the procedure well, with uneventful post-operative course. He received a total of 3 whole blood transfusion units during the treatment. He was discharged in good health.

The biopsy report confirms gastro-intestinal stromal tumor (GIST).

3. Discussion

The most frequent localization site for GISTs are the stomach and small bowel. Studies have shown that more distally located GISTs have a worse prognosis. On the other hand reported sites for EGISTs are the omentum, mesentery, retroperitoneal space and in rare cases in the pancreas, liver, cholecyst, abdominal wall, etc. In terms of size EGISTs tend to be significantly larger than GISTs, up to 35 cm in diameter.

The most common clinical features of EGIST patients are abdominal pain, palpation of an abdominal mass, GI bleeding, loss of appetite, dysphagia, bowel obstruction and fever. The most beneficial imaging modalities for EGISTs diagnosis are CT and MRI.

Treatment of GISTs and EGISTs consists on the R0 resection of the mass, without the need for radical removal of healthy tissue since these types of tumors do not normally infiltrate adjacent tissue. If other organs are invaded it is recommended to perform an en-bloc resection, taking care to avoid the rupture of the mass, as to minimize the chances of peritoneal dissemination.

A prompt adjuvant therapy including the use of selective tyrosine kinase inhibitors (imatinib) is strongly advised. The patient should be followed with periodic controls for recurrence.

We believe that in our case the current disease is a recurrence of his previous condition. He was adequately diagnosed and a strict surgical procedure was performed and after recovery he was consulted by the oncology team.

4. Conclusion

Surgery and adjuvant therapy are the two important modalities that are crucial for improving the prognosis of patients. Neoadjuvant therapy may be considered in cases of unresectable disease, to diminish the size of tumor.
In conclusion, there is no universally accepted consensus on the duration of adjuvant therapy after the surgical procedure and the role of surgery is debated in patients with metastatic or recurrent disease. There is a clear role surgery can play in improving the outcomes in such patients. However, some authors still debate whether surgery improves results in such patients.

For this purpose, it is necessary to involve a team of oncologists, imaging specialists and experienced surgeons in the treatment plan of GIST patients to provide the best treatment.

Conflict of interest

The author(s) declare(s) that there is no conflict of interest. The authors alone are responsible for the content and writing of the paper.

Financial disclosure

There is no financial support to this study.

Ethical aspect

Informed consent was obtained from all participants in the study and all procedures were conducted in accordance with the Declaration of Helsinki.

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