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GASTRIC GIST WITH RETROPERITONEAL EXTENSION: CASE REPORT

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Abstract: Gastrointestinal stromal tumors (GISTs) are rare mesenchymal neoplasms of the gastrointestinal tract. GISTs that arise from the bowel wall usually present as subepithelial neoplasms in the stomach and small intestine; however, they can also arise in any portion of the gastrointestinal tract and, occasionally, in the omentum, mesentery, and peritoneum. They are rare neoplasms that represent approximately 1 to 2% of primary gastrointestinal (GI) cancers. Despite their rarity, GISTs are the most prevalent mesenchymal (ie, non-epithelial) neoplasms of the GI tract, with the stomach being the most common primary tumor site (40 to 60%). [1] This study aims to describe a case of low prevalence of gastric GIST with involvement of the retroperitoneum, focusing on its management and surgical approach. The case in question was conducted by the General Surgery Residency Service of the UFMS at the" Hospital Universitário Maria Aparecida Pedrossian". Taking into account the scarce records of the approach to this type of tumor in the literature, especially with retroperitoneal involvement, this report details the management of the case, from the initial diagnosis, anatomopathological and histopathological confirmation, with clinical evolution throughout the hospital stay.
Keywords:GIST. Neoplasm. Retroperitoneum.

INTRODUCTION

Within the GI tract, primary tumor sites are most common in the stomach (40 to 60%) and jejunum/ileum (25 to 30%). Less common sites include the esophagus (≤1%), duodenum (5% percent), colorectal (5 to 15%) and anus (<0.5%). [1]

The presentation of GISTs varies depending on the location of the primary tumor. For example, GISTs involving the upper GI tract (stomach, small intestine, or esophagus) may present with GI bleeding, dysphagia, or

obstructive jaundice, whereas those involving the colon or rectum may present with constipation, bowel obstruction, or urinary hesitancy in men (due to a tumor rectum adjacent to the prostate. Approximately 10 to 20 percent of patients have metastatic disease. [1]

CASE DESCRIPTION

Female patient, 71 years old, was admitted to the emergency room complaining of diffuse abdominal pain for about 1 year, with cramps, more intense in the left hypochondrium and mesogastrium, radiating to the lumbar region, with associated nausea and vomiting. On abdominal examination, a globular abdomen, normotensive, bowel sounds, massive percussion in the left upper quadrant with pain on percussion and palpation of the epigastrium, in addition to a palpable solid mass in the LUQ, with no signs suggestive of peritonitis. Other items of the physical examination remained unchanged. Upon admission, regarding the history of diseases, systemic arterial hypertension was reported using Amlodipine, Enalapril, Atenolol and Simvastatin; Previous surgeries: cholecystectomy, tonsillectomy, cesarean section and hysterectomy, with no reported complications. Sedentary, with no history of alcoholism or smoking.

EVOLUTION OF THE CASE

After hospitalization and clinical stabilization of the patient, CT scan of the abdomen with contrast was performed (Figures 1, 2 and 3) which showed a solid-cystic mass located in the left hypochondrium, with heterogeneous contrast enhancement, hypoattenuating in the non-contrast phase. Parietal thickening of the gastric antrum, without a clear cleavage plane with the aforementioned formation, as well as intimate contact with part of the body and tail of the pancreas. Largest measures: 19.5 x 13.4 x 11.3 (CC x LL x AP). It presents peripheral lobulations, predominantly without a clear plane of cleavage with the solid-cystic mass, whose imaging characteristics allow considering the possibility of lobulated contours of the mass and, as a differential, possible locoregional lymph node blocks (loss of lymph node capsular integrity). Lymph nodes with atypical dimensions or morphology characteristic of other lymph node chains are not observed. No characteristic signs of distant metastases were observed in the present study. It presents contact with the celiac trunk at less than 90 degrees of vessel circumference, associated with blurring of the surrounding fat, without thrombus. It maintains contact with the splenic vein between 90 and 270 degrees.

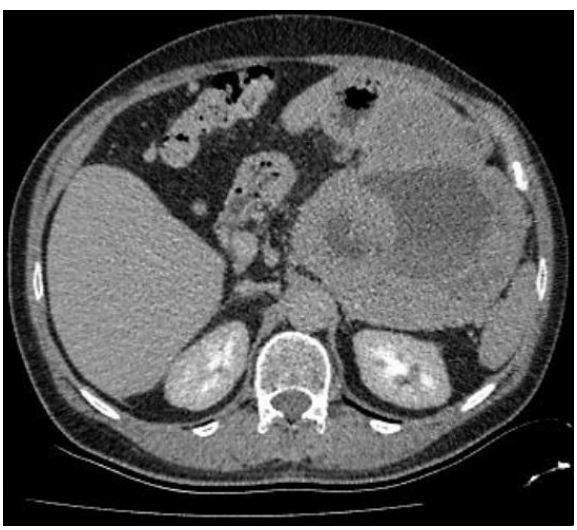

Figure 1. Extensive gastric mass extending to the spleen.

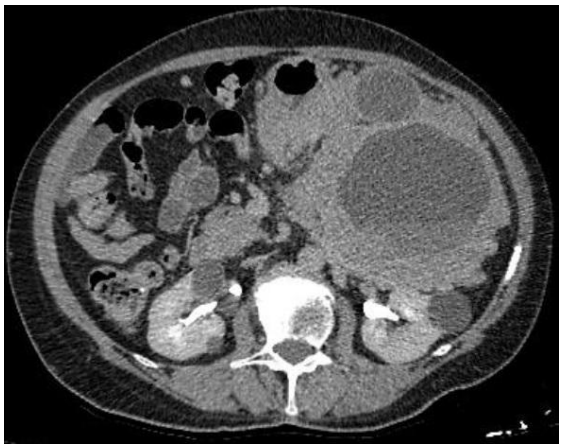

Figure 2. Gastric mass in contact with the retroperitoneum.

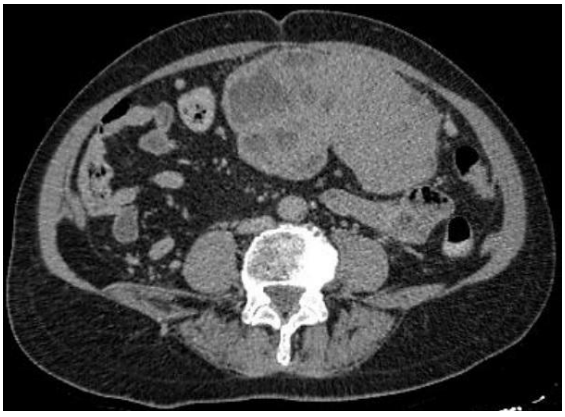

Figure 3. Caudal extension of neoplastic mass.

After the findings, it was considered a hypothesis of Gastrointestinal Stromal Tumor and opted for surgical approach by exploratory laparotomy.

During the surgical approach, the presence of a large, hardened mass was observed extending to the retroperitoneum, in contact with the pancreas, stomach, transverse mesocolon and spleen; proceeded with the release of mass from the mesocolon and omentum, from the greater curvature/ antrum by partial gastrectomy using a linear stapler with reinforcing suture, followed by pancreatectomy caudal body with a linear stapler with reinforcing suture in Greek bar and finally splenectomy.

After resection, the specimen was sent for anatomopathological analysis, which showed: mesenchymal neoplasm in connective and adipose tissue suggestive of gastrointestinal stromal tumor (GIST). Cell pattern: spindle cell. Visualized in gastric outer muscular layer. Size: 22.0x10.0x9.0cm. Histological pattern: solid. Presence of ectatic capillaries. Extensive areas of necrosis. Presence of foci with deposition of hyaline and eosinophilic material in the middle of the neoplasm. Mitosis /5 mm2. Mild nuclear atypia. Pancreas and spleen without injury.

Immunohistochemical examination showed GIST with co-expression of c-kit, CD34 and DOG1. Considering the morphological and topographic parameters found (high mitotic activity and probable gastric location), the neoplasm in question presents a high risk of aggressive behavior (tumor progression).

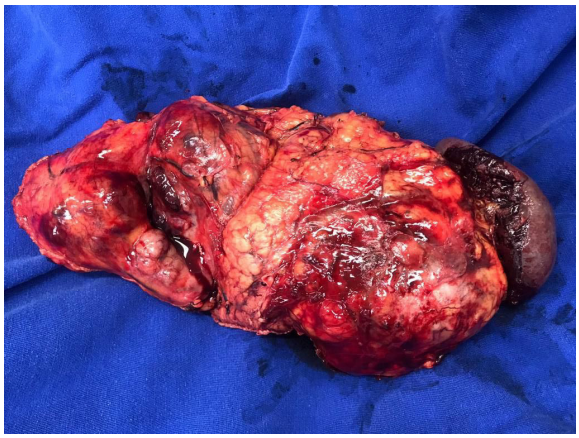

Figure 4: Surgical resection showing the gastric, pancreatic, spleen and omentum portions.

DISCUSSION

The GISTs are predominantly diagnosed in elderly patients and rarely occur in patients under the age of 40 years. They affect both sexes equally, with the exception of pediatric patients, in which they are more common in females in a 2:1[1] ratio.

The interstitial cells of Cajal are thought to be the probable origin of GISTs. These regulate peristalsis through autonomic innervation of

the visceral wall and smooth muscle. They are located below the epithelium, in the intestinal wall, which is the primary location of GISTs.[2]

The immunohistochemical profile distinguishes GISTs from leiomyomas and leiomyosarcomas. The presence of the CD117 antigen (transmembrane KIT receptor tyrosine kinase) has almost universal expression in GISTs, being typically negative in these other tumors. Most GISTs present mutation in the KIT gene, causing abnormal activation of this and allowing the cell's oncogenic signaling.[3]

A small portion of patients diagnosed with GISTs are associated with genetic syndromes. They may be related to Neurofibromatosis type I, Carney-Stratakis Syndrome, Carney's Triad and primary familial GIST Syndrome.[5]

In primary familial GIST Syndrome, we see inherited mutations in the KIT or platelet-derived growth factor receptor alpha (PDGFRA) genes. Skin hyperpigmentation, dysphagia or gastrointestinal autonomic nerve tumors may be observed in these cases. It presents early and has multiple primary sites.[5]

In those with Neurofibromatosis Type I, GISTs are predominantly multifocal and occur most frequently in the small intestine. Carney-Stratakis Syndrome are characterized as the dyad of paragangliomas associated with GISTs. Diagnosis occurs predominantly in children and young adults. The Carney Triad is added to the Carney-Stratakis Triad due to the presence of pulmonary chondromas, affecting young patients.[5]

The primary tumor of GISTs affects the GI tract, and may also be found extra-intestinally (may not be associated with other locations or even as sites of distant metastasis). Metastatic disease occurs in approximately 10-20% of cases. The most common sites of metastasis are the liver, omentum and peritoneum.[4,5]

Clinically, adults manifest symptoms associated with GI bleeding (28% of cases), may be asymptomatic in 13-25% or may develop abdominal pain (8-17%), acute abdomen (2-14%) and asymptomatic abdominal masses (5 %). Paraneoplastic syndromes rarely occur in adults with GISTs, but may present as hypoglycemia and consumption hypothyroidism.[4,5]

The diagnosis of GIST is established through histopathology and immunohistochemistry. Histologically, GISTs present epithelioid to fusiform variation. The markers present by immunohistochemistry characterize KIT (CD117) as the almost universal marker of GIST (approximately 95% are positive). Other markers present are PDGFRA, DOG-1, PKC-theta, CD34, smooth muscle actin, S-100 protein, and desmin or keratin mutations. Approximately 15% of GISTs do not have a mutation in the KIT gene, these are called wild-type KIT.[1,3]

Endoscopy and imaging are also required for patient evaluation. Computed tomography is the method of choice to assess the lesion, using intravenous and oral contrast. Being able to observe solid mass highlighted by the contrast with soft contours.[5]

Endoscopic ultrasound can be used to better characterize high tumors. Magnetic resonance imaging can be used for preoperative evaluation and for those small-sized GISTs. PET-CT is highly sensitive for diagnosing GISTs but is not sufficient for preoperative diagnosis.[5]

Among the factors that influence the outcome of the disease, tumor size and mitotic rate are relevant, as well as the site of the primary tumor. In general, those found in the stomach present better evolution when compared to those originating in the rectum, mesentery, small intestine and colon. Another risk factor that must be taken into account also includes tumor rupture, whether during surgery or spontaneously, a fact that affects disease-free survival.[5]

Adjuvance and neoadjuvant can be performed as well as treating metastatic and advanced diseases through small molecule tyrosine kinase inhibitors. Initial therapy with Imatinib as a neoadjuvant can be performed in cases of tumors bordering on resection, advanced and unresectable. Adjuvance can be performed taking into account the risk of recurrence of the neoplasm, which takes into account tumor size, location, mitotic index and the occurrence of tumor rupture. [1]

CONCLUSION

GISTs are rare neoplasms and, in particular, in this case, with a high volume and involvement of multiple organs, this case becomes an unusual finding among the reports already listed in the literature.

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