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Gastrointestinal Stromal Tumor (GIST) with Surgical Treatment and Embolization: Clinical Case and Literature Review

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ABSTRACT

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal neoplasms, accounting for 1% to 2% of all malignancies of the digestive tract. Gastrointestinal stromal tumors are rare mesenchymal tumors (0.1% to 3% of all malignant gastrointestinal tumors) that originate within the interstitial cells of Cajal. We present a case of an 83-year-old patient with melena and hematochezia of one week's duration with hemodynamic instability. Complete resection with laparotomy and embolization of the 5 x 3.5 cm GIST was performed. The histopathology report showed spindle cells, a low mitotic index and the immunohistochemistry report: DOG-1+/CD117+/ H.CALDESMON + FOCAL. Treatment with imatinib is started. In this case report we describe the clinical presentation, diagnosis and treatment of gastric stromal tumor.

Keywords

Severe lower gastrointestinal bleeding, GIST tumor, Gastrointestinal stromal tumor, Embolization.

Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal neoplasms, accounting for 1% to 2% of all digestive tract neoplasms. Approximately 6,000 new cases of GIST are diagnosed each year in the United States [1]. In epidemiological studies of GIST, the estimated mean age at diagnosis is the sixth decade of life and the frequency of onset is 6.8 to 14.5 cases per million individuals per year [2]. Gastrointestinal stromal tumors (GISTs) are rare mesenchymal tumors (0.1% to 3% of all malignant gastrointestinal tumors) that originate within the interstitial cells of Cajal [3].

In the early literature, these tumors used to be classified as leiomyomas, cellular leiomyomas, leiomyoblastomas, and leiomyosarcomas, but in the 1960s, electron microscopic studies revealed the lack of typical smooth muscle differentiation in some of these gastric tumors. Subsequently, leiomyomas, which did not show ultrastructural features of smooth muscle cells and lacked immunohistochemical features of Schwann cells (i.e., did not express S-100 protein), were classified as "gastric stromal tumors" by Mazur and Clark in 1983 [4]. GISTs are most commonly located in the stomach (60–70%), followed by the small intestine (20–30%), colon and rectum (5%), and esophagus (<5%) [4]. When they reach a significant size, abdominal GISTs may be associated with complications, such as bleeding, intestinal obstruction, or spontaneous rupture. Although the curative option remains surgical resection [3]. Histopathological examination (HP) along with immunohistochemistry (IHC) and molecular testing are necessary for the differential diagnosis of mesenchymal tumors. More than 95% of GISTs harbor the c-Kit mutation, which can be assessed by positive IHC staining for cluster of differentiation (CD)117. In addition, GISTs are often CD34 positive and are found in GIST1 (DOG1) [5].

Differential diagnosis includes other gastric subepithelial lesions such as lipoma, neuroendocrine tumor, leiomyoma, neural stromal tumors (schwannoma, neuroma, neurofibroma), ectopic pancreas (pancreatic remnant), and extrinsic compression [1].

In the following review article, a case of GIST will be presented and its diagnosis and treatment will be discussed.

Case Report

An 83-year-old patient with a personal history of liver abscess 40 years ago, postoperative complicated appendectomy 15 years ago, has 3 covid vaccines. We are consulted due to presenting melena stools and hematochezia on multiple occasions during 1 week of evolution with poor general condition, with hemodynamic alterations, requiring fluid resuscitation and hospitalization. During hospitalization, endoscopy and colonoscopy are performed (Figure 1).

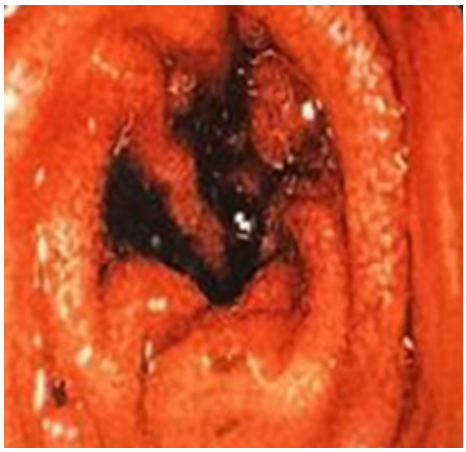

Figure 1: Endoscopic image of the ileal portion with abundant blood remains.

The results of endoscopy and colonoscopy showed Los Angeles grade A esophagitis, bleeding from the lower digestive tract proximal to the terminal ileum, with extrinsic compression of the cecum. A capsule endoscopic examination was requested, which reported abnormalities in the portion of the terminal ileum (Figure 2).

Figure 2: Endoscopic image with capsule in the terminal ileum region where blood remains are observed, as well as a tumor 1.20 meters from the ileocecal valve.

After having all the laboratory tests, it was decided to perform exploratory laparoscopy. The tumor was removed through a medial incision, from the xiphoid process to the pubis. After identifying and ligating the multiple vessels that supplied the mass in the terminal ileum (Figure 3).

Figure 3: Macroscopic image of a tumor in the ileum in exophytic form.

Tumor embolization is performed (Figure 4), the lesion is removed with free surgical margins without lymphadenectomy, along with a portion of the small intestine (Figure 5). Material taken during the procedure is sent to pathology, which reports a low-risk gastrointestinal stromal tumor.

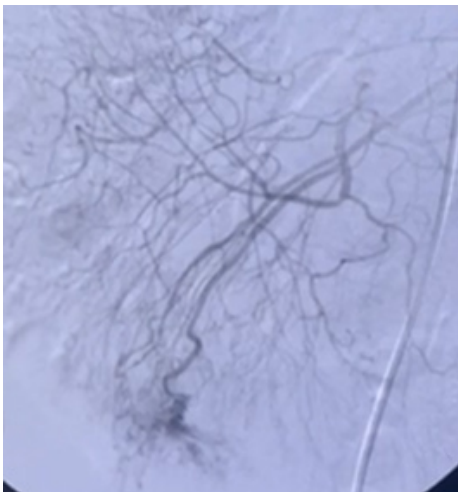

Figure 4: Arteriography where the superior mesenteric artery is canalized and the irrigation of the lesion, which depends on the ileal arteries, is observed.

Figure 5: Macroscopic image of gastrointestinal stromal tumor 5 x 3.5 cm, histological subtype: spindle cells, low mitotic rate.

There were no postoperative complications and the patient was discharged. The tumor was sent for immunohistochemistry, which yielded the following immunophenotype results: DOG-1+/ CD117+/ H.CALDESMON + FOCAL. (Figure 7) Treatment with imatinib was started.

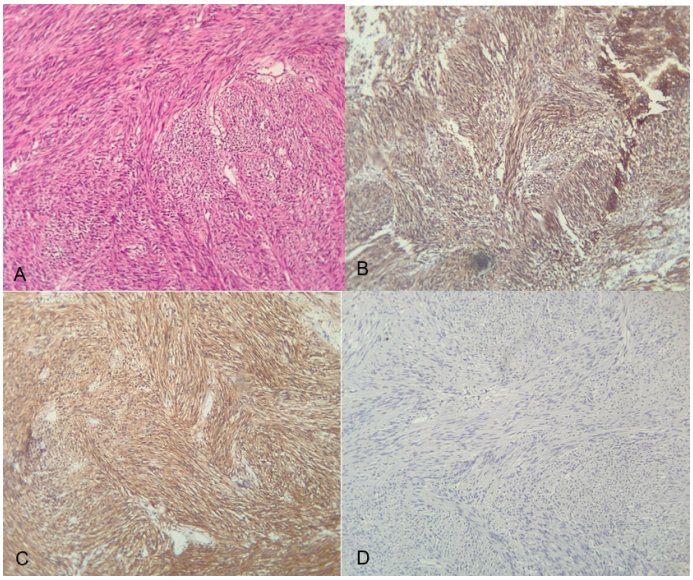

Figure 6: Microscopic images. Hematoxylin and eosin staining shows: A) the solid area of the tumor (60x magnification); Also shown are slides of immunohistochemical stains that demonstrate positivity towards: B) DOG1, C) KIT (CD117), D) KI 67 low mitotic activity

Discussion

Initial evaluation of the patient should include a computed tomography (CT) scan of the abdomen and pelvis. CT of primary GIST usually shows exophytic tumors, which are larger than 5 cm and enhance inhomogeneously (Figure 7) [6].

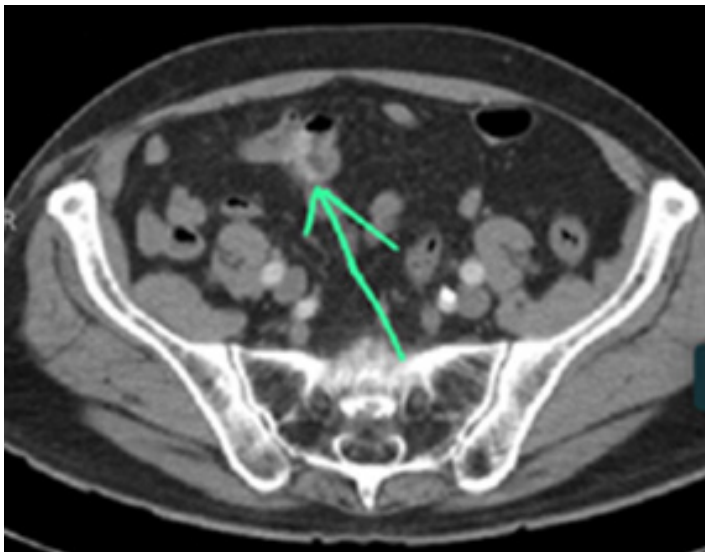

Figure 7: Contrast-enhanced abdominopelvic CT scan. In arterial phase, polypoid formation in the terminal ileal region with heterogeneous enhancement and exophytic growth of submucosal origin. (green arrow) Gastric GIST.

Endoscopic ultrasound may be of additional use, but biopsy is not mandatory prior to surgery, except to rule out differential diagnoses such as lymphoma or other malignant or benign neoplasms. Biopsies may be taken endoscopically (deep biopsy reaching the muscularis layer) or as core needle biopsies (guided by ultrasound or CT) [4]. GISTs do not present with specific endoscopic or endoscopic ultrasound findings, and the diagnosis is difficult to achieve by histopathologic examination using hematoxylin and eosin staining alone. Immunohistochemical analysis, such as that involving measurement of KIT, CD34, or DOG1, is essential for a definitive diagnosis. First, all subepithelial lesions (SELs) are examined by endoscopic ultrasound, and SELs mentioned in the ultrasound section that are conclusively diagnosed by endoscopic ultrasound findings alone are excluded. Second, endoscopic ultrasound with fine-needle aspiration using immunohistochemical analysis is performed for the remaining hypoechoic solid masses to differentiate GISTs from other tumors [2]. In the Japanese clinical practice guidelines for GISTs, biopsy is recommended to exclude cancer-like subepithelial lesions when a subepithelial lesion is diagnosed endoscopically [7]. The definitive diagnosis of GIST is made by histopathological and immunohistochemical study. In 3 of our patients, positivity for CD117 was demonstrated, a marker present in 95% of GISTs [8]. GISTs typically appear as solid, hypoechoic lesions arising from the muscularis layer. Most gastric GISTs arise from the muscularis propria; only rarely do they arise from the muscularis mucosae. GISTs 2 to ≤5 cm in size and >5 mitoses per 50 high-power fields (HPFs) located in the stomach show progression in 10–15%, similar to tumors >10 cm but with mitotic rates ≤5/50 HPFs. Tumors >2 to ≤5 cm in size are at high risk for clinical progression even with ≤5 mitoses per 50 HPFs [9].

Simultaneous endoscopy and laparoscopy have proven highly effective in localizing these submucosal tumors for surgical resection. Although a prospective randomized trial remains to be conducted, this study provides additional evidence suggesting that early removal of GISTs, 5 cm or less in size, provides better disease-free survival than later removal of the larger tumor [10-12].

Vincenzo et al. demonstrated in their study a comparison between performing laparoscopy and laparotomy [13]. It was shown that the LAP group had fewer complications, faster gastrointestinal recovery, reduced use of analgesics and a shorter postoperative hospital stay ($p<0.05$ in each case). Radiological embolization is increasingly used as a treatment for bleeding in different locations because it allows hemodynamic stabilization of patients. Thus, arterial embolization as a treatment for severe gastrointestinal bleeding achieves hemodynamic stabilization of patients and reduces the need for transfusion [13]. Adjuvant cancer therapy is considered the next step after surgical resection for both high- and intermediate-risk patients. Available data have defined patient risk based on high mitotic rate, tumor size and location, Ki67 expression index, as well as the presence of metastasis within the diagnosis [14].

Conclusions

Diagnosis of gastric GIST is difficult due to its characteristic

submucosal growth, which occurs incidentally in a large percentage of cases and does not present typical symptoms. Tumor resection should be tailored to the location, symptoms, and condition of the patient to consider laparotomy or laparoscopy in order to preserve adjacent organs and provide better management. Laparoscopy is considered the first option in uncomplicated gastric stromal tumors.

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