Giant extraintestinal gastrointestinal stromal tumor: Case report and bibliography review

Abstract
Giant extraintestinal gastrointestinal stromal tumors (GIST) are rare tumors of the digestive tract. Its most frequent locations are the stomach, small intestine, colon, and rectum. Its appearance in other places outside the gastrointestinal tract such as the mesentery, omentum, or retroperitoneum is infrequent. Computerized axial tomography (CT) and magnetic resonance imaging (MRI) are the imaging studies of the first choice. Surgical recession is the gold standard for localized tumors and advanced or metastatic tumors are treated with imatinib. This study presents the case of a male patient of 53 years with no history of previous pathologies. The patient was admitted with a clinical condition of generalized abdominal pain, weight loss of approximately 20 kg, abdominal distention, melena, hematemesis, and asthenia. Physical examination revealed a distended abdomen and palpation revealed epigastric and mesogastric hardening and left colonic frame. The abdominal tomography revealed a tumor mass with an infiltrative appearance of apparent gastric origin, with extragastric growth and infiltration of the spleen, pancreas, mesenteric root, greater omentum, transverse colon, thin intestinal loops, and infiltration in the hepatic hilum, and liver metastases. Moreover, the condition was related to the severe acute respiratory syndrome coronavirus type 2 (SARS-CoV-2). An ultrasound-guided percutaneous biopsy was performed in the left upper quadrant and histology reported a GIST. In this article medical condition, diagnosis, and treatment of the Giant extraintestinal gastrointestinal stromal tumor, is reviewed.

Keywords
Gastrointestinal stromal tumor, Extragastrointestinal tumor, Giant tumor.

INTRODUCTION
Gastrointestinal stromal tumor known or GIST is the most common mesenchymal neoplasia of the digestive tract; however, their frequency represents only 0.1% to 3% of gastrointestinal neoplasms. Their incidence is 10 to 20 cases per million people, and approximately 10% to 30% are clinically malignant. Only 5,000 to 6,000 new cases per year are reported in the United States. They may appear at any age but predominate in people aged 40 to 70 years. About 65% of GISTs are found in the stomach, 25% to 40% in the small intestine, and 5% to 10% in the colon or rectum; its location in other digestive sites, such as mesentery, omentum or retroperitoneum, sites outside the gastrointestinal tract are infrequent and are reported to be less than 5%. Surgical recession remains the gold standard for localized...
tumors, and pharmacological treatments are indicated for advanced stages of the disease. Those currently available have radically changed prognosis(6).

CASE PRESENTATION

A 53-years-old male patient, with no personal, family or pathological surgical history, with a clinical picture of 2 years of evolution characterized by diffuse abdominal pain, abdominal distension, and weight loss of approximately 20 kg, which is why he went to private practice (Figure 1).

The patient was admitted to the emergency room due to a 7-day clinical picture characterized by melena-type upper gastrointestinal bleeding 3 times a day without hemodynamic instability. It was also accompanied by abdominal pain, unquantified fever, headache, asthenia, loss of smell and taste for 5 days. On physical examination, pale skin and mucosa were found. On auscultation there were crackles in both lung fields, a distended abdomen with a visible and palpable mass in the epigastrium, mesogastrium, and left colonic frame.

The results of the laboratory tests were as follows: Hemoglobin: 2.60 mg/dL; Hematocrit: 10.30%; leucocytes: 9,600/mm³; neutrophils: 75%; platelets: 557,000; C-reactive protein (CRP): 10.6 mg/dL; albumin: 2.7 mg/dL; total proteins: 5.50; amylase: 94 U/L; lipase 552 U/L; D-dimer: 2989.7 µg/L; procalcitonin: 1.87 ng/mL; ferritin: 138 µg/dL; electrolytes: Sodium: 131 mEq/L, Chlorine: 95 mEq/L; normal renal function; normal clotting times; normal bilirubin; serological test for non-reactive syphilis (VDRL); prostate-specific antigen (PSA): 0.88 ng/mL; carcinoembryonic antigen (CEA): 1.06 ng/mL; alpha-fetoprotein (AFP): 1.57 ng/mL; carbohydrated antigen 19-9 (CA 19-9): 14.60 U/mL; tuberculin skin test or mantoux negative test; serology for non-reactive hepatitis B virus (HBV), hepatitis C virus (HCV), and human immunodeficiency virus (HIV); rapid test for 2019 coronavirus disease (COVID-19), immunoglobulin G (IgG), and M (IgM). Imaging tests included an abdominal ultrasound that reported a solid hypoechoic mass with regular contours arising from the greater curvature measuring approximately 23 x 13 cm with Doppler ultrasound, and increased vascularity.

Chest computed axial tomography (CAT) revealed alveolar consolidation with ground glass infiltration in both lung bases and septal thickening (Figure 2). The CAT scan of the abdomen and pelvis revealed an infiltrative tumor mass of apparent gastric origin measuring approximately 25 x 25 x 17 cm with extensive destruction of the wall and mucosa, with endoluminal infiltration and extragastric growth, liver metastasis (hepatomegaly with two hypoattenuating lesions of secondary appearance, measuring 72 x 41 mm and 86 x 75 mm respectively in hepatic segments IV and V, and infiltration of the hepatic hilum, resulting in dilation of the intrahepatic bile duct) infiltration of the spleen, pancreas, the mesenteric root, the greater omentum, the transverse colon, the small intestinal loops, the
lymphoplasmacytic inflammatory infiltrate, proliferative cryptitis, and vascular congestion. Videocolonoscopy was incomplete due to extrinsic compression in the descending colon that prevented the passage of the equipment; diverticular disease was observed in the sigmoid colon with no signs of inflammation or bleeding. An ultrasound-guided

portal and the periaortic and pericaval lymph nodes, the largest measures 13 mm (Figures 3 and 4).

Concerning endoscopic studies, an upper digestive video endoscopy was performed in which an infiltrative lesion was observed at the level of the gastric body with a gastric ulcer scar with a histopathological report of gastric atrophy,
percutaneous biopsy was performed on the left hypochondrium. The histology report informed a gastrointestinal stromal tumor (GIST), mitotic rate: mitosis/5 mm²; necrosis: unidentified, histological grade: low grade (mitotic rate ≤ 5/5mm²), evaluation risk: moderate (10%), immunohistochemistry: CD117 antigen (C-kit: receptor tyrosine kinase) with positive staining.

During his hospital stay, he received treatment for SARS-CoV-2 respiratory infection (COVID-19 pneumonia) with lopinavir/ritonavir 500 mg every 12 hours for 7 days, azithromycin 500 mg every day for 7 days, and ivermectin 6 mg 3 tablets (single dose). A satisfactory improvement was obtained.

A multidisciplinary team managed the treatment made up of internists, gastroenterologists, oncologists, pathologists, surgeons, imaging specialists, and nutritionists. It was determined that since it was an advanced disease with metastasis to different organs and without resection planes, he was a candidate for pharmacological treatment; therefore, it was started with imatinib and had a good response. He was discharged without complications, and he has monthly controls in external oncology consultations currently.

**DISCUSSION**

GISTs are rare non-epithelial tumors of the digestive system. Mazur and Clark introduced the term *stromal tumor* in 1837, and it is currently proposed that GISTs originate from the interstitial cells of Ramon and Cajal[7]. GIST can be morphologically classified as fusiform (70%), epithelioid (20%), and mixed (10%), and may show muscle or neural strain. Unlike other mesenchymal tumors, GISTs express CD117 antigen (part of the C-kit receptor), which immunohistochemistry can demonstrate.

GISTs are submucosal lesions that appear to arise from the muscle of the intestinal wall, those of intramural origin are often projected exophytic extraluminal or endophytic intraluminal and may have ulceration of the overlying mucosa. Its size can be extremely variable, from small incidental cases to large masses, which almost always exceeds its vascular supply and leads to extensive areas of necrosis and hemorrhage[8-9].

The clinical picture is variable, depending on the size of the tumor and the location. The most frequent symptoms are anemia, weight loss, gastrointestinal bleeding, abdominal pain, dysphagia, and symptoms related to the mass effect; they may present acute abdominal symptoms, obstruction, perforation, rupture, and peritonitis[10]. Concerning diagnosis, at least 10% to 30% is made incidentally, while 70% of patients have a wide range of clinical manifestations[10].

CAT and magnetic resonance imaging (MRI) are the first-choice imaging studies for GISTs. CAT with intravenous contrast allows the identification of possible hemorrhages, intratumoral calcification, preoperative staging, and evaluation of metastatic disease, as in hypervascular liver lesions. MRI is used to visualize tumors in the pelvic area and study the mesenteric and peritoneal extension. Although these imaging options serve to establish a pre-
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immunohistological analysis and reverse-transcriptase polymerase chain reaction (RT-PCR) analysis for KIT mutations, will generally confirm the diagnosis followed by histologic confirmation. In most cases, the diagnosis is fortuitous, and the histologic confirmation is not always required. The definitive diagnosis is obtained by histopathological examination, which includes immunohistochemistry and reverse-transcriptase polymerase chain reaction (RT-PCR) analysis for KIT mutations. In most cases, the diagnosis is fortuitous, and the histologic confirmation is not always required. The definitive diagnosis is obtained by histopathological examination, which includes immunohistochemistry and reverse-transcriptase polymerase chain reaction (RT-PCR) analysis for KIT mutations.

GISTs are rare tumors, and symptoms are usually nonspecific. In most cases, the diagnosis is fortuitous, and the histology combined with immunohistochemistry and reverse-transcriptase polymerase chain reaction (RT-PCR) analysis for KIT mutations confirms the diagnosis. We present a case of giant GIST with extraintestinal and unresectable localization that was diagnosed by percutaneous biopsy and was treated with imatinib with a currently good response.

CONCLUSIONS

GISTs are rare tumors, and symptoms are usually nonspecific. In most cases, the diagnosis is fortuitous, and the histology combined with immunohistochemistry and reverse-transcriptase polymerase chain reaction (RT-PCR) analysis for KIT mutations confirms the diagnosis. We present a case of giant GIST with extraintestinal and unresectable localization that was diagnosed by percutaneous biopsy and was treated with imatinib with a currently good response.

Conflicts of interest

There are no conflicts of interest.

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Authors’ own.

REFERENCES