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Lesser Curvature and Omentum-Dependent Spindle Cell Tumor: Literature Review Regarding a Case

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ABSTRACT

GIST tumors are a group of rare neoplasms, having a reported incidence of 6/10 million people annually. 70-80% of the GIST are benign, however there are prognostic factors which can help us to predict its behavior. The stomach and the small intestine are the most frequent sites of location of these tumors. Diagnosis is not only based on clinical suspicion, we must also use imaging tools and pathology to confirm it. Surgery is the treatment that offers a permanent cure for localized primary GIST.

KEYWORDS: GIST tumor, stomach, omentum, spindle cell.

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INTRODUCTION

Gastrointestinal stromal tumors (GIST) were first classified as independent tumors in 1983 by pathologists Mazur and Clark who gave the term to non-epithelial tumors of the digestive tract, which lacked smooth muscle and had Schwann cell immunohistochemical characteristics. 1 Are rare and account for 0.1%-3.0% of all gastrointestinal neoplasms and 5-7% of sarcomas. [2,3]

Histopathologically, they were initially considered smooth muscle tumors, leiomyomas, leiomyoblastomas and leiomyosarcomas, however with the help of electromyoscopy and immunohistochemistry it was shown that they were made up of smooth muscle and Schwann cells. [5,6] Nowadays, GIST are defined as a epithelioid, spindle cell or occasionally pleomorphic mesenchymal tumors of the gastrointestinal tract which expresses de KIT protein (CD117) detected at immunohistochemistry, excluding the above-mentioned gastrointestinal smooth-muscle tumors; the exact cellular origin of GIST has been proposed to be the interstitial cell of Cajal, an intestinal pacemaker cell, identified as a complex cell network within the bowel wall dispersed in the muscularis propria. [7]

Most of them are benign (70-80%), however the transition from benign to malignant could be predicted according to the mitotic frequency and tumor size. [8] Benign behavior is characteristic in gastric tumors and outnumber malignant ones by a 3–5:1 ratio. They can occur anywhere in the gastrointestinal tract, from the esophagus to the anus; however, the stomach (39 to 60%) and the small intestine (30 to 42%) are the most frequent sites of location of these tumors, and they can even be present as primary tumors in the omentum and mesentery. [9]

These are often discovered incidentally during a surgery, an image study like tomography (TAC) or an endoscopy of the upper gastrointestinal tract. [9]

The computed tomography (CT) can well assess the involvement of the lumen, the wall, and the serosa. They are seen as well-defined exophytic masses with different densities based on their inner structure. [10,11]

GIST tumors are classified into four subtypes based on their relation to the muscularis propria: type I protrude into the digestive lumen with a narrow attachment, type II protrude with a broad connection, type III is centrally located on the gastric wall, and type IV extend outward, protruding into the serosa of the gastric wall. [29]

CASE PRESENTATION

This is a 63-year-old female patient, with an 8-year history of type 2 diabetes under control with metformin 850 mg every 12 hours, and 1-year history of arterial hypertension in management with losartan 50 mg every 24 hours, surgical history of bilateral tubal occlusion 33 years ago.

Her condition began 1 month prior to the evaluation with persistent nausea without apparent cause, sporadically accompanied by epigastric pain, with an intensity of 3/10, colic-like, without radiation, reason why she went to a private

physician who treated her with proton pump inhibitor, without improvement of the symptoms and then she decided to go to this medical unit.

During her consultation, an abdominal ultrasound was requested, which reported a rounded image with defined edges in the head/neck of the pancreas, with a heterogeneous pattern of predominantly solid, measuring 32x31x40 mm, with a volume of 22 ml (figure 1).

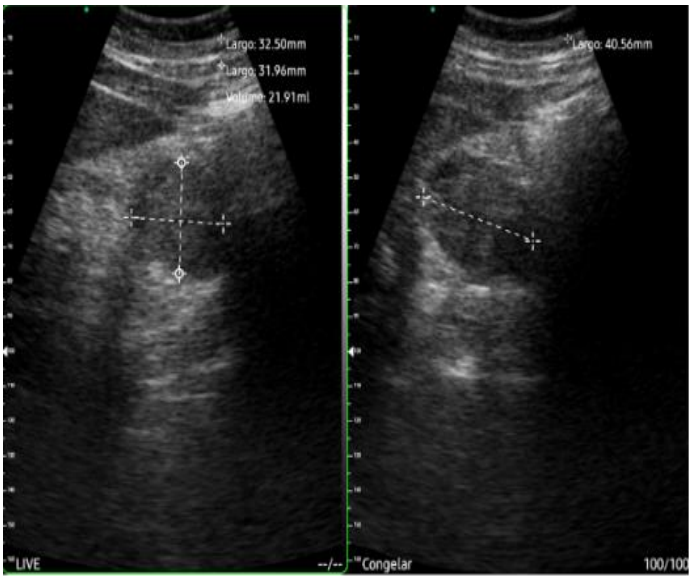

Figure 1. Abdominal ultrasound.

Based on this finding, an abdominal computed tomography scan was requested, which ruled out pancreatic lesions. However, a solid lesion was found in the lesser omentum, adjacent to the pancreas and lesser curvature, capsular type, heterogeneous, and persisting in the late portal and venous phase, measuring 41x32 mm, without associated regional adenopathy (figures 2 and 3).

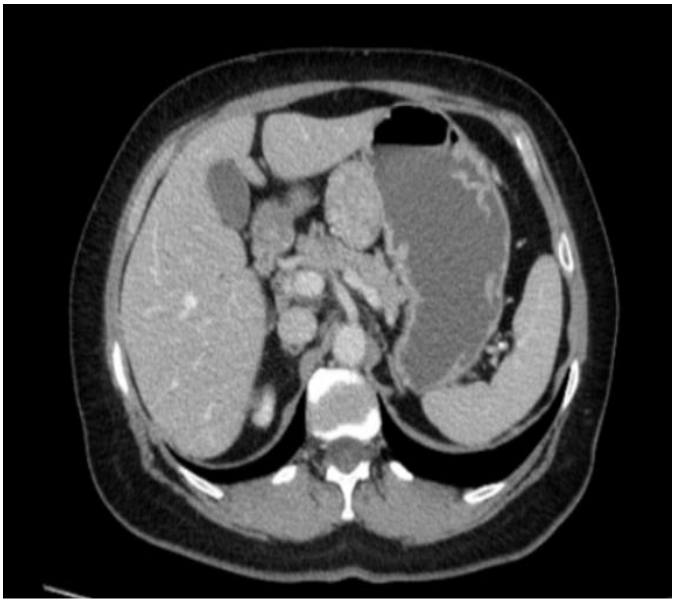

Figure 2. Axial view of computed tomography.

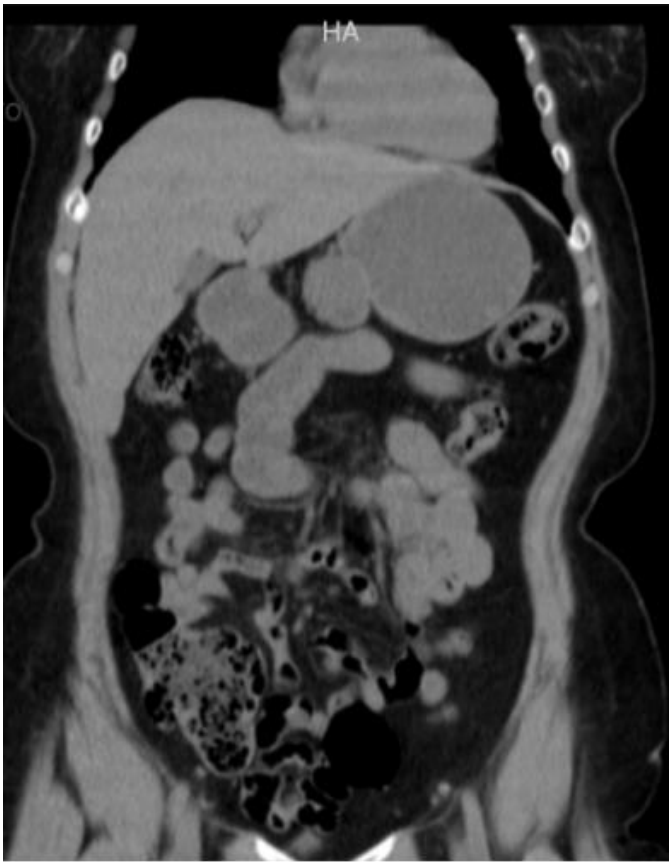

Figure 3. Coronal view of computed tomography.

Laboratory studies were requested which reported glucose 164 mg/dl, BUN 39 mg/dl, creatinine 1.4 mg/dl, total bilirubin 0.6 mg/dl, direct bilirubin 0.3 mg/dl, indirect bilirubin 0.3 mg/dl, ALT 73 U/l, AST 46 U/l, LDH 355 U/l, amylase 72 U/l, lipase 57 U/l, alkaline phosphatase 104 U/l, calcium 7.2 mg/dl, magnesium 1.5 mg/dl, sodium 136 mg/dl, potassium 3.7 mg/dl, chloride 101 mg/dl, leukocytes 6.9 $x10^3$, neutrophils 11.8 $x10^3$, hemoglobin 16 g/dl, hematocrit 48.6%, platelets 367 $x10^3$.

Patient who underwent surgery for excisional biopsy, finding a tumor in the lesser omentum dependent on the lesser curvature of the stomach measuring 4x3x3 cm.

The specimen was sent to pathology, where it was described as a spindle cell neoplasm with nodular growth, with fusiform to epithelioid cytoarchitecture, slightly eosinophilic to pale cytoplasm, and vesicular chromatin residing within uniformly rounded nuclei with incipient atypia, associated with a focally hyalinized stromal component, with the presence of thin-walled blood vessels, some focally branched. Recent multifocal hemorrhage and moderate chronic inflammatory infiltrate were observed. The conclusion was a pseudoencapsulated spindle cell neoplasm with areas of hemorrhage and associated chronic inflammation, with a surgical margin focally in contact with the lesion (figure 4).

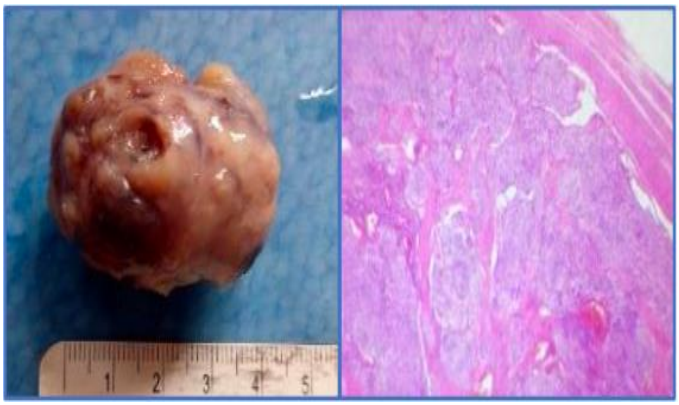

Figure 4. Lesser curvature and omentum-dependent tumor.

Immunohistochemistry was requested as a complement to the diagnosis for the adequate classification of the lesion, since in this scenario the diagnostic considerations are gastrointestinal stromal tumor (GIST), leiomyoma or solitary fibrous tumor, for which immunohistochemistry was performed with DOG-1, CD117, CD34 being all positive, and Ki-67 with 5%, thus concluding gastrointestinal stromal tumor.

DISCUSSION

The stromal gastrointestinal tumors (GIST) are a group of rare neoplasms, defined as mesenchymal cell tumors such as spindle cell, epithelioid and occasionally pleomorphic tumors of the gastrointestinal tract. According to American literature, there is an incidence of 6 cases per 10 million people annually [15] and they are predominantly male, however, the series of M. Miettinen et al. reports that the incidence is 10-20 cases per 10 million people, and the most frequent age of presentation is around 50-60 years [16] agreeing with the age of our patient.

Although a higher incidence has been reported in male patients, the incidence in some studies shows that there is no difference between both genders. [17] In Mexico, few have been reported related to the subject, however, Alcantara et al. remark that the average age of presentation is 54 years in the Mexican population [18] falling within the age range of our patient.

Most GIST tumors are located in the stomach (50%-60%) [19] which coincides with the location of the tumor in our case, as suggested by Skandalakis in a review of 725 cases, where the distribution of smooth muscle tumors along the gastrointestinal tract was primarily the stomach in 47.3% of cases. [20]

Histologically, GIST with the spindle cell type is the most frequent, which is the one presented by our patient, composed of cells with a spindle nucleus, pale and fibrillar cytoplasm, giving a lower risk of progression. [1,5]

The challenge in diagnosing this entity lies in the number of nonspecific symptoms that may appear 6 months prior to medical diagnosis and they are related to the size of the tumor. Small tumors are usually a surgical finding, while tumors > 5 cm are usually symptomatic. [19]

Symptoms include acute epigastric abdominal pain, weight loss, gastrointestinal hemorrhage, mass effect symptoms, nausea and vomiting. [1,13]

Costache et al. reports that at the time of diagnosis the symptoms are present in 75% of the cases. [19] Our patient presents abdominal pain as the main symptom, which is more frequently present in the Mexican population according to Medina et al. [20]

Due to the wide variety of symptoms, imaging modalities play an important role not only in diagnosis, but also in staging and treatment planning. These tools include computed tomography, ultrasound, magnetic resonance imaging and positron emission tomography (PET). [21]

Abdominal ultrasound can describe it as a large tumor with heterogeneous structure, areas of necrosis and floating inner echoes, however the origin is difficult to assess; As in our case, computed tomography is often used because it can show the structure involved, tumor size and extending outwards. Magnetic resonance imaging is particularly useful for large tumors (>6 cm) because it effectively characterizes the extent of the lesion and its internal structure, with the tissue component appearing hypointense on T1 and hyperintense on T2, apparent diffusion coefficient, perfusion parameters and degree of enhancement. [22, 23]

The role of PET consists in helping on differential diagnosis from non-GIST tumors, characterizing metastases and pathological lymph nodes. [22, 23]

The diagnosis of GIST is based on imaging and pathology. In our case, because the imaging reports a tumor <6 cm, we decided to perform a surgery with diagnostic and curative intentions. To confirm the diagnosis, we performed an immunohistochemical study using anti-CD117 antibodies (c-kit). [24]

Previously, there were no factors that determined the prognosis of GIST tumors; in fact, all tumors in this category have malignant potential. Currently, there are already prognostic factors described, such as mitotic activity, tumor size, location and necrosis within it, histological growth pattern (fusiform or epithelioid), and immunohistochemical pattern, which help us to predict its behavior. [25, 26]

In our patient, the tumor size was 4x3x3 cm, so according to the size it is categorized as a tumor with a very low risk of progression (1.9%), on the other side, tumors larger than 10 cm and a mitotic index of more than 5/5 mm^2 should be considered as tumors with a high risk of progression (10%) and should be treated in specialized medical centers. [27, 31]

Surgery is the treatment that offers a permanent cure for localized primary GIST. The aim for patients with localized lesions is to complete surgical resection with an intact capsule. Achieving wide surgical margins does not improve outcome, however a rupture of the tumor capsule is a significant adverse prognostic factor. [29]

Laparoscopic approach has emerged as the primary treatment for small and medium sized gastric submucosal tumors, but open surgery remains as the standard approach for large tumors. [29]

A good prognostic factor, according to Koelz et al., is a proliferation index <10% of KI 67, which is what our patient presents, since if it were greater than 10% it is associated with a worse prognosis. [28]

In our case, the tumor was surgically accessible and confined to the stomach, however when they are unresectable or with metastatic disease, tyrosine kinase inhibitor or Imatinib therapy should be considered. [29, 30]

As our patient had a low-risk tumor, we are following up with CT scan every 12 months for 5 years, but if the patient has had a high-risk tumor, the following up changes to CT scan every 3 months during 3 years with adjuvant therapy, then every 3 months for 2 years while discontinuing adjuvant therapy and finally every 6 months for 5 years without therapy. [30, 31]

CONFLICTS OF INTEREST

The authors declare no conflict of interest.

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INFORMED CONSENT STATEMENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A written copy is available upon request.

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