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# Incidental Finding of an Extraluminal Gastric GIST in Cholecystectomy

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## ABSTRACT

Gastrointestinal stromal tumors are the most frequent mesenchymal neoplasm found in the gastrointestinal tract. In the last decades, molecular and immunohistochemical characteristics of gastrointestinal stromal tumors have been elucidated, allowing their differentiation from other neoplasms, an established diagnosis, therapeutic management, and prognosis. We describe a case of a 65-year-old man with an extraluminal gastrointestinal stromal tumor, diagnosed incidentally during a laparoscopic cholecystectomy. The tumor had a location in the greater curvature of the stomach. In the histopathological report the tumor had a smooth muscle differentiation, with the expression of the following markers: DOG-1, CD117, CD34, actin and H-Caldesmon. Before the laparoscopic cholecystectomy, a computed tomography was performed and no signs of tumor in the greater curvature were appreciated. The treatment of the gastrointestinal stromal tumors was surgical, avoiding the rupture of the tumor wall, which is associated with tumor recurrence in the future.

**Keywords:** Gastrointestinal Stromal Tumors; GIST; Extraluminal growth; Laparoscopy; Cholecystectomy, Extraluminal

## INTRODUCTION

Gastrointestinal stromal tumors or GIST are neoplasms that originate from the interstitial cells of Cajal, which represent the gastrointestinal tract pacemaker. GIST is the most frequent mesenchymal neoplasm found in the GI tract with an incidence of approximately 1.5 per 100,000 people, occurring most frequently in adults from 50 to 70 years with equal gender distribution.[1,2] Most GIST are discovered incidentally, the most common location is the stomach 55.6%, followed by the small bowel 31.8%, and colorectal 6.0%.[3]

The most common clinical manifestations include bleeding, pain, symptoms of obstruction, acute abdomen, and in 5% of the cases asymptomatic. Historically, the first line treatment has been surgical, considering that the tumor is not sensitive to conventional chemotherapy and radiotherapy. Although with the introduction of tyrosine kinase inhibitors like Imatinib, new treatment options are available and median survival rate has drastically improved. GIST tumors can be classified based on immunohistochemical staining and recently Skandalakis created a classification based on macroscopic growth types into four different types: endoluminal, exoluminal, intramural and mixed.[4] Several gene mutations are associated with GIST and activation of these tumors growth, including Platelet-Derived Growth Factor Alpha (PDGFRA) and a tyrosine kinase receptor KIT gain of function mutation. Therefore, expression of the CD117 (C-kit) marker results essential in GIST diagnosis.[5] Here we describe a case of an extraluminal gastric GIST, which was found incidentally on a laparoscopic cholecystectomy.

## CASE PRESENTATION

A 65-year-old man presented to the hospital with a chief complaint of right upper quadrant pain with an intensity of 8/10 posterior to the ingestion of cholecystokinetics. In the physical examination, the patient had a positive Murphy sign, decreased peristalsis and painful decompression of the right upper quadrant. In the physical examination the patient had a small supraclavicular tumor (lipoma) and nonspecific gastric symptoms; therefore an abdominal computed tomography was requested. The abdominal computed tomography revealed chronic lithiasic cholecystitis with no dilatation of the biliary tract, hepatic cyst, hepatic and splenic granulomas and the presence of bilateral inguinal hernias (Figures 1 and 2). In the interpretation the radiologists did not identify at first hand the tumor, therefore when performing the gallbladder surgery, the surgeons had no knowledge of the GIST.

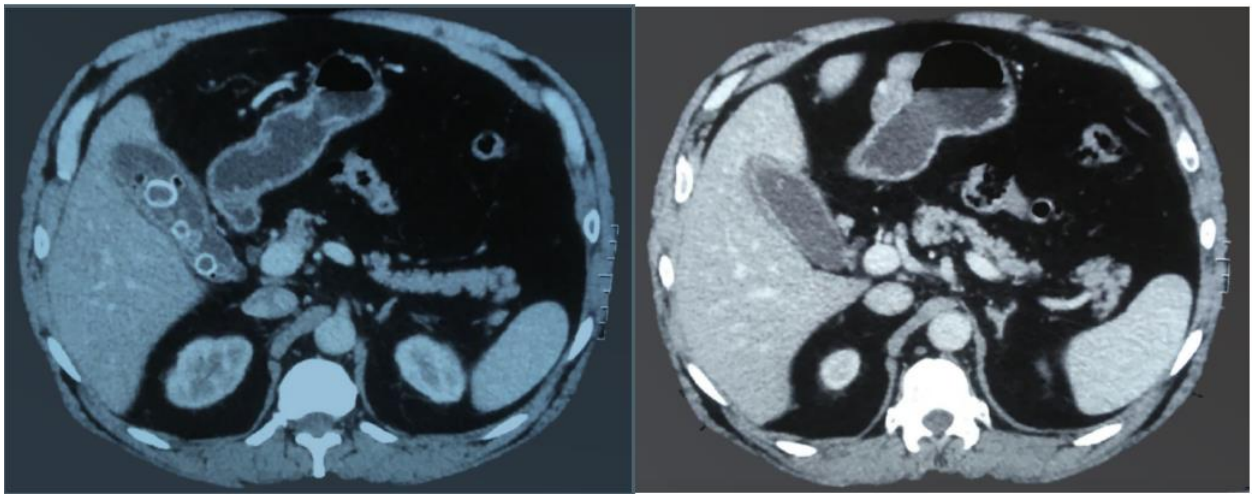

**Figure 1 & 2:** Axial abdominal computed tomography showing lithiasic cholecystitis.

A laparoscopic cholecystectomy was performed, finding incidentally an extraluminal tumor in the greater curvature of the stomach with no evidence of infiltration into surrounding no structures or lymphadenopathy (Figures 3, 4 and 5). The tumor was excised (Figure 6) and analyzed histopathologically, revealing a smooth muscle differentiation, with a positive expression of the following markers: DOG-1, CD117 (c-kit), CD34, actin and H-Caldesmon. A proliferation index of 2% (Ki67) was reported, CK AE1/AE3 marker was negative and the tumor margins were free. The management of the patient included resection of the tumor on the stomach greater curvature and a laparoscopic cholecystectomy.

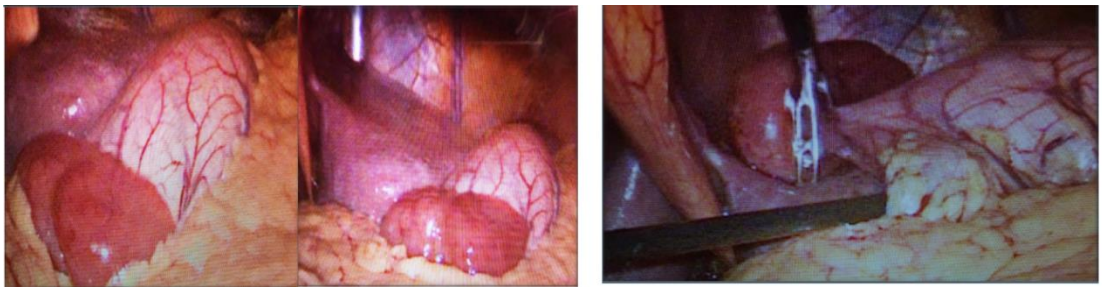

**Figure 3-5:** Extraluminal GIST visualization during laparoscopic cholecystectomy.

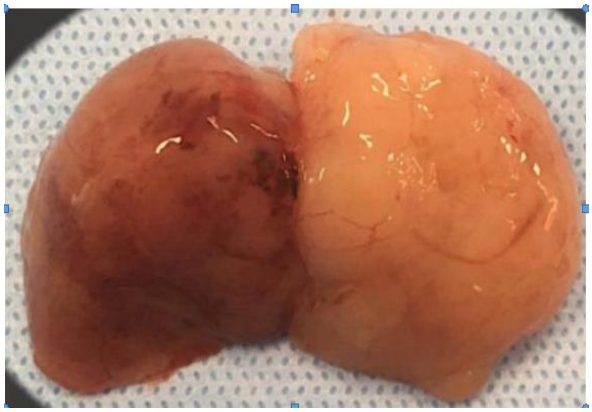

**Figure 6:** Excised tumor.

## DISCUSSION

GISTs are rare, accounting for 1-2% of gastrointestinal neoplasms, yet are the most frequent mesenchymal neoplasm in the GI tract.[4] The populations mostly affected by GISTs are adults between 50 to 70 years, with gender equal distribution. Although reported incidence of GIST is approximately 1.5 cases per 100,000 people, GIST real prevalence remains unknown.[4] GIST clinical presentation depends on size, location and macroscopical growth patterns (according to Skandalakis classification into four different types: endoluminal, exoluminal, intramural and mixed). Asymptomatic cases are mostly small sized neoplasms with an intestinal location.

When symptomatic, patients may present abdominal pain, nausea, vomiting, gastrointestinal bleeding, symptoms of obstruction, acute abdomen, among others.[4] The identification of these tumors is mostly done with computed

tomography scan and a biopsy should be performed. CT-guided biopsy is a strategy frequently used to make a diagnosis. Diagnosis in most cases is incidental when performing a gastrointestinal endoscopy.

Most tumors stain positive for CD-117 (C-Kit), CD-34 and DOG-1. GIST in most cases tends to have benignant behavior, yet around 10-30% have shown to progress to a malignant behavior. GIST can have different types of growth patterns, exophytic representing 79% of the tumors, intraluminal, mixed less frequently and extraluminal being the rarest of them all. GISTS with extraluminal growth, no more than 10 cases have been reported.[5] The gastrointestinal pacemakers better known as the cells of Cajal act as interface between the smooth muscle cells and the autonomic innervation. Most of these cells are found in the stomach (60-70%) and in the small intestine (20-30%).

When making a diagnosis of a GIST, differentiating between a benign and malignant tumor is essential, due to potential risk of metastasis. Currently, it is accepted that tumors measuring more than 10 cm and having a mitotic rate higher than 10/50 HPF are considered to have a more aggressive behavior. In case of diagnosing a GIST, some authors recommend the use of the risk group classification which was proposed by Miettinen, et al. 2006.[6] Where patients are given a risk (low, intermediate or high) depending on the size of the tumor and the mitotic index in a 50 HPF. Some adaptations have been made to Miettinen classification, in which a third component was added, the location of the tumor.[6]

Important differential diagnoses that should be considered include leiomyoma, leiomyosarcoma, schwannoma, malignant peripheral nerve sheath tumor, inflammatory myofibroblastic tumor, solitary fibrous tumor, neurofibroma and neuroendocrine tumors.[6] Currently the treatment of choice is surgical resection with disease free margins. When performing the surgery, the procedure should be done carefully, avoiding the rupture of the tumor wall, taking in consideration that rupture is associated with tumor recurrence.[7]

Discovery of molecular pathways and gene mutations associated with GIST, has allowed a new therapeutic approach to these tumors. The introduction of tyrosine kinase inhibitors such as imatinib, which target KIT, PDGFRA and Bcr-Abl (seen in some types of leukemia, such as CML), has drastically improved the median survival rate for patients, particularly in those with advanced localized or metastatic tumors.[4,6] Following the introduction of imatinib, the survival rate in advanced stage GIST has increased from 18 to more than 60 months. Although, it's important to take in consideration that around 12-15% of adults and 90% of pediatric GIST lack the PDGFR and KIT mutations, this is known as GIST wild type.[6] In case of diagnoses of GIST, molecular studies should be performed, being that the effectiveness of tyrosine kinase inhibitors depends on the presence of certain mutations. For example, in case of having a D842V substitution in exon 18 of the PDGFRA gene, resistance to imatinib has been shown, both with 400 mg and 800 mg doses. Luckily if resistance to imatinib is detected, other tyrosine kinase inhibitors can be used, such as sunitinib.[8]

Different series have reported the incidental finding of GIST tumors during surgery for another cause.[9,10,11] Most of the incidentally diagnosed GIST tumors were found during bariatric procedures where laparoendoscopic approaches are combined, and because 60% of the GISTs are located in the stomach.[12] In fact, there are authors who affirm that these tumors will be found incidentally in 1% of bariatric surgeries,[13] which suggests that the incidence of these tumors is much higher than that reported in the literature. There is no consensus on what the surgeon's behavior should be when encountering these tumors serendipitously, but different authors recommend that if the tumor can be resected without significantly altering the original surgical plan,[14] it should be done, as has been done in most cases reported in the literature.

## CONCLUSION

One of the main advantages of laparoscopic surgery is allowing the exploration of the abdominal cavity, therefore the surgeon should be prepared in case of finding an incidental diagnosis, such as gastrointestinal stromal tumors. GIST is the most common frequent mesenchymal neoplasm found in the GI tract. The most frequent location is the stomach, and the diagnosis is made through histopathological findings. The use of certain markers is helpful to confirm diagnosis, determine treatment eligibility with tyrosine kinase inhibitors, and prognosis. Nowadays, the first line treatment continues to be surgical. This is an extremely unique case; this is the second reported case of an incidental finding of a GIST in a cholecystectomy. Extraluminal GIST are extremely rare, this tumor has the uniqueness of being completely extraluminal.

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