

## Encountering a Gastrointestinal Stromal Tumor at Our Institution which is a Rare Entity: A Case Report and Literature Review

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**Received:** June 20, 2019; **Published:** August 16, 2019

### Abstract

A 50-year-old female, found to have a Gastrointestinal stromal tumor in her stomach, treated surgically, no recurrence was noticed upon follow up for 2 years with adjuvant therapy. Finding of Intraoperative gastric mass involving the anterior wall of the stomach is infrequently coping with at our institution. The Gastrointestinal mass was resected completely with histopathological examination supporting the diagnosis of GIST. The present case highlights the importance of considering GIST in an investigating a patient with history of GI bleeding mainly or vague on/off abdominal pain with a palpable epigastric mass.

**Keywords:** Gastrointestinal Stromal Tumor; Case Report; Vague Abdominal Pain; Gastric Tumors

### Introduction

Gastrointestinal stromal tumors (GISTs) are uncommon nonepithelial mesenchyme soft tissue sarcoma of the GI tract. These tumors start in very early forms of special cells in the wall of the GI tract called the interstitial cells of Cajal (ICCs). ICCs are cells of the autonomic nervous system, the part of the nervous system that regulates body processes such as digesting food. ICCs are sometimes called the “pacemakers” of the GI tract because they signal the muscles in the GI tract to contract to move food and liquid along. The incidence of GIST ranges from 11 to 15 per million per year, although are considerably underestimated [1].

More than half of GISTs start in the stomach. Most of the others start in the small intestine, but GISTs can start anywhere along the GI tract. A small number of GISTs start outside the GI tract in nearby areas such as the omentum or the peritoneum [2].

Some GISTs seem to be much more likely to grow into other areas or spread to other parts of the body than others. Factors determined whether a GIST is likely to grow and spread quickly, such as how large the tumor is, where it's located in the GI tract, and how fast the tumor cells are dividing (its mitotic rate).

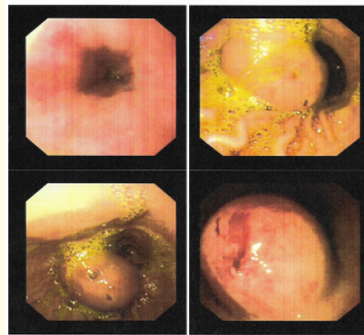
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**Citation:** Alfayez AA, et al. “Encountering a Gastrointestinal Stromal Tumor at Our Institution which is a Rare Entity: A Case Report and Literature Review”. *EC Gastroenterology and Digestive System* 6.9 (2019): 810-814.

**Case Report**

50 years old female, known case of gallstones. Came to our institution electively in 30/12/2017 for open cholecystectomy and excision of gastric mass.

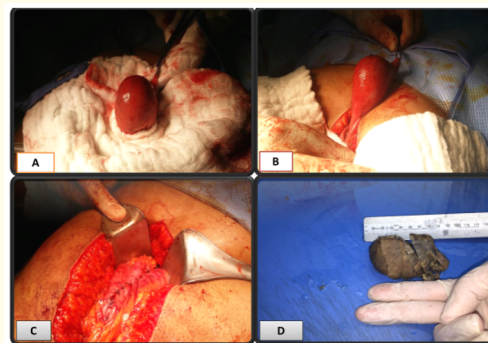
Initially, four days before she came, they did for her an Outpatient CT scan of the abdomen which showed “well defined globular isodense soft tissue lesion seen arising from the entero-superior wall of the stomach related to the inferior margin of the left hepatic lobe without gross evidence of infiltration, measuring about 5 x 4.5 x 4 cm”. So, endoscopy was done for her (Figure 1), biopsy taken which was not conclusive and they Advised for bigger and deeper biopsy to rule out any neoplastic growth. For that we elected to take her for complete excision of the mass.



**Figure 1:** Endoscopical finding, protruding mass is clearly noticed.

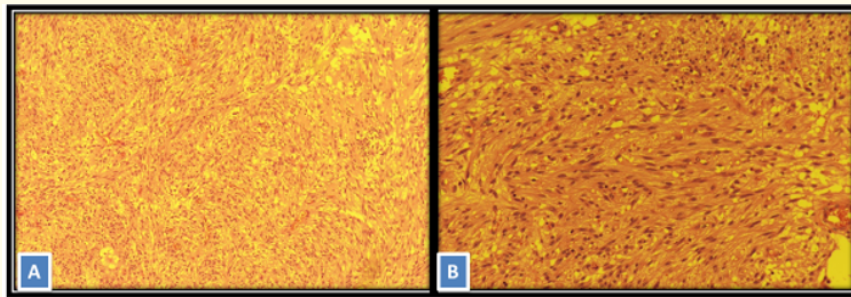
Up on presentation, she gave history of mild on/off abdominal pain in epigastria area, associated with nausea. No history of vomiting and no history of Weight loss. Previously she had surgical history for right inguinal hernia, paraumbilical hernia, appendectomy and tubal ligation. Physical examination revealed a regular mass in epigastric area, with no tenderness, fixed and firm. Pre-operative labs were not conclusive, no tumor markers were done.

Through an open approach, Gastric mass was resected completely along with cholecystectomy and no major complications “Rupture of the capsule or bleeding” was encountered intraoperatively (Figure 2).

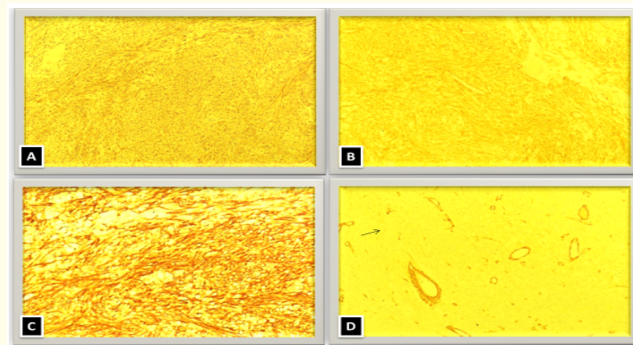


**Figure 2:** Intraoperative finding (A). Another view for the mass before resection (B). Primary repair of the stomach after resecting the mass (C). Gross appearance of the mass measured 5x4x4 cm (D).

Postoperatively, surgical recovery was smooth. She was extubated and shifted to ICU for extensive care less than 24 hours, with good pain control and DVT prophylaxis, 4 days after she was able to go home, with regular follow up appointment. Post-operative labs were not conclusive apart from low hemoglobin which was expected in her case from 12.6-11.4-10.6-10 g/dl. Histopathology report came as gastrointestinal stromal tumor (GIST) "Benign Cellular Spindle Cell Type" Stage pT2 (Figure 3 and 4). Gallbladder histopathology came to be gallstones.



**Figure 3:** Bland looking spindle cell proliferation in whorls and short intersecting fascicles H&E, low power (A). H&E staining, high power (B).



**Figure 4:** Immunohistochemistry tumor cells are positive with CD117 (A). Positive with DOG1 (B). Strong positive for CD34 (C). Negative for SMA, note the positive internal control in the wall of blood vessels (arrow) (D).

## Discussion

Encountering such a case is not an easy matter in our institution with no oncological center available and less advance specialized equipment. In the stomach, developmental morphology is classified as exogastric (30 - 40%), intramural (29 - 44%), endogastric (18 - 22%) and mixed type (16 - 22%) based on the Skandalakis classification for leiomyomas [3]. Currently GIST represents a distinct entity from other mesenchymal tumors of GI tract. Most GISTs show an activating mutation in either the c-kit or platelet-derived growth factor receptor a (PDGFRA) gene [4]. We had a patient who has tumor resection through open approach and found to have a GIST. The incidence and prevalence of GIST is not well studied in our region with around 4 cases seen till now at our institution.

A retrospective study of 40 Patients proven GIST of the stomach at the Rabin and Kaplan Medical Center between 2004 and 2013. Their symptoms are varying (Figure 5) and 70% of them treated by laparoscopic resection. The median follow-up period was 40 months with 93% disease-free survival [5].

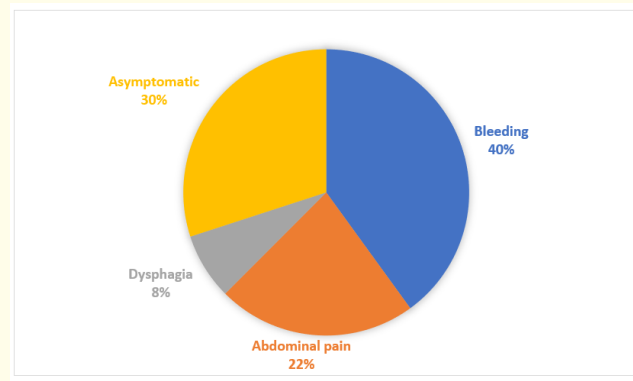


Figure 5: Symptoms of GIST are varying among patients.

The diagnostic approach mainly in literatures was an abdominal CT scan, gastroscopy then followed by pre-operative biopsy [5-7].

Scattered cases raised often in literature, with no clear well-structured guideline explain the management of GIST, most of them supporting dealing with it by surgery either open or laparoscopically depend on tumor size [8] and now a days targeted molecular therapy are promising adjuvant treatment, especially if rupture, recurrence or metastasis is there [9]. Initially, due to small tumor this lady has with low risk classification based on size and mild vague symptoms along with insufficient tissue biopsy taken for diagnosis we took her for entire surgical removal of the mass. She received imatinib for one year as part of effective adjuvant therapy. Till date patient in follow up at our clinic with no recurrence evident by CT. GIST frequently metastasizes to the liver or peritoneum, although nodal metastasis is very rare [10]. Therefore, lymph node dissection is not recommended. Although, GIST has a high morbidity [11,12].

Majority are sporadic, but Familial Gastrointestinal Stromal Tumor Syndrome was described in literature with KIT Exon 11 Mutation [13,14]. Gastrointestinal stromal tumor with synchronous colorectal adenocarcinoma also have been reported [15].

### Conclusion and Learning Points

- Surgical resection is considered to be the best treatment for gastrointestinal stromal tumor (GIST).
- Tumor size, mitotic rate, and anatomic locations are directly related to the potential malignancy, surgical approach, oncological treatment, and recurrence rate.
- Diagnosis of gastric GIST is often incidental to endoscopy and computed tomographic scan.
- The most important technical point is to avoid tumor rupture during removal.
- The most notable symptoms are gastrointestinal bleeding.
- A large tumor size, high mitotic rate, high-risk group, and adjacent organ involvement all contribute to bad outcomes of GISTs.

### Source of Support

None.

### Conflicting Interest

None declared.

### Acknowledgement

I express my sincere respect and gratitude to my colleague at histopathology lab Mr. Bandar Alsaif "Laboratory Technician" who has given his valuable support and cooperation to successfully complete this useful paper.

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Volume 6 Issue 9 September 2019

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