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Clinical Image

A Gastrointestinal Stromal Tumor (GIST) at the Gastro-esophageal (GE) Junction in a young female.

Khan Md Nazmus Saqeb[1]*, Mohammad Shamsul Arfin[2]
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Case:

A 35 year old woman presented to the Gastroenterology outdoor with the complaints of dyspepsia for last 2 months. A submucosal gastric mass at the Gastro-esophageal (GE) junction was incidentally identified by endoscopic examination during investigating the cause of dyspepsia. The mass was growing intraluminally without any surface ulceration or any kind of obstruction of the gastric channel. No physical abnormalities were observed. Laboratory tests including hematologic and biochemical analyses revealed no abnormalities. An enhanced computed tomography scan of whole abdomen revealed a solid mass (16 × 18 mm) with a smooth margin & hypervascularity at the cardio-esophageal junction. A fine-needle aspiration was performed, and the pathologic diagnosis of the submucosal tumor was a possible gastric GIST.

Initially a complete endoscopic resection was planned but later this approach was abandoned due to increased risk of bleeding, and the patient was referred to the department of surgery. A complete resection was done there and the specimen was sent for histopathology. The histopathological examination of the resected specimen confirmed the diagnosis of a gastric GIST with negative margins. The tumor was found to be composed of spindle shaped tumor cells. Immunohistochemical analysis revealed that the tumour cells were positive for CD117 and CD34 and negative for SMA & S-100. The mitotic index was less than 5 mitotic figures per 50 high power fields. There were no postoperative complications. The patient was discharged 5 days after the surgery and has been doing well since then. A repeat endoscopy upper GI and a CT scan of whole abdomen is scheduled during the next follow up visit to exclude any kind of recurrence or metastases.

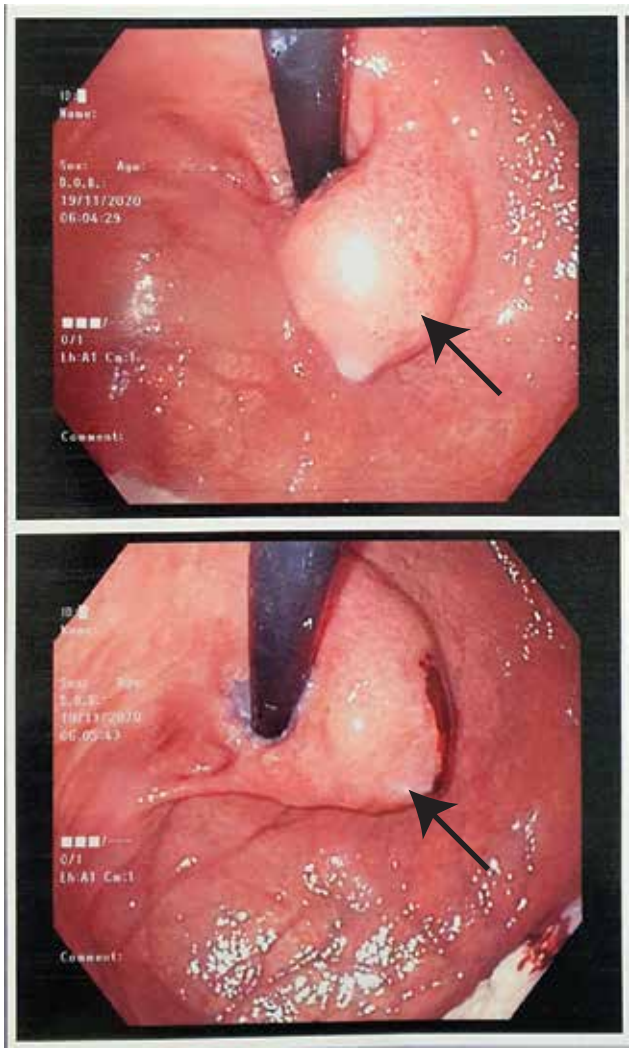

Figure 1: Endoscopy of upper GIT showing a globular submucosal mass with intact surface mucosa at the Gastro-esophageal (GE) junction, suggestive of GIST (black arrow).

Discussion:

GISTs comprise 1% to 3% of all malignant GI tumors; they are the most common mesenchymal tumor of the GI tract.[1] These tumors are derived from the interstitial cells of Cajal and have been shown to harbor gain-of-function mutations in the cell-surface KIT receptor in approximately 90% of cases.[2] GIST develop from the Muscularis propria. They may be acquired or genetic.[3] Most GISTs (60% to 70%) arise in the

1. Assistant Professor, Dept. of Gastrointestinal, Hepatobiliary & Pancreatic diseases, Jahurul Islam Medical College & Hospital, Bajitpur, Kishoreganj, Bangladesh.

2. Chief consultant & Head, Dept. of Gastrointestinal, Hepatobiliary & Pancreatic diseases, Bangladesh Specialized Hospital, Shyamoli, Dhaka, Bangladesh.

***Corresponding Author:**

Dr. Khan Md. Nazmus Saqeb
E-mail: drsaqebk59@gmail.com
Phone: +8801715248062
ORCID ID: https://orcid.org/0000-0002-8080-947X

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stomach; 20% to 30% originate in the small intestine, and less than 10% in the esophagus, colon, and rectum.[1] Gastrointestinal stromal cell tumours (GISTs), arising from the interstitial cells of Cajal are differentiated from other mesenchymal tumours by expression of the *c-kit* proto-oncogene, which encodes a tyrosine kinase receptor.[4]

The clinical presentation of patients with GISTs depends on the anatomic location of the primary lesion, as well as other factors like tumor size and presence or absence of symptomatic metastases. For many GIST patients, initial detection of GISTs may be an incidental finding or result from evaluation of nonspecific symptoms. The most common primary site for GISTs is the stomach. They can be asymptomatic or present with bleeding, pain, or obstruction.[1] These tumours, particularly the smaller lesions of less than 2 cm, are usually benign and asymptomatic, but the larger ones may have malignant potential.[4] Symptoms from GISTs per se are usually noted only after tumors are larger than 5 cm in size or have impinged on a specific anatomic region (e.g., a gastric GIST causing gastric outlet obstruction). Symptoms at presentation may include a palpable abdominal mass or abdominal swelling, abdominal pain, nausea, vomiting, anorexia, and early satiety.[1] Most GIST lesions as seen endoscopically are submucosal rather than mucosal, without overlying ulceration. This explains why many GIST masses may only be visualized on endoscopy as a subtle, smooth protrusion with overtly normal mucosa.[1]

Definitive expert surgery remains the mainstay of treatment for patients with primary localized GISTs (early-stage GISTs).[1] The diagnosis is confirmed by histopathological examination of the resected specimen. The margins of the resected specimen should be carefully examined. The tumor is usually composed of spindle shaped tumor cells.[1] On immunohistochemical analysis the tumour cells usually stain positive for CD117 (95% cases) and CD34 (50% cases) and is negative for SMA & S-100.[1]

Small lesions (< 2 cm) are usually followed by endoscopy, while larger ones require surgical resection. Very large lesions should be treated pre-operatively with imatinib (a tyrosine kinase inhibitor) to reduce their size and make surgery easier. Imatinib can also provide prolonged control of metastatic GISTs.[4] Open surgery is a traditional treatment of GISTs. The indications for laparoscopic surgery have been expanding in recent years.[5]

GISTs rarely involve the regional lymph nodes, and extensive lymph node exploration or resection is rarely indicated. GIST lesions are highly vascularized and often exhibit a fragile pseudo capsule; therefore, surgeons should be careful to minimize the risk of tumour rupture, which subsequently increases the risk of peritoneal dissemination. The margins of resection from the tumour specimen should be carefully oriented and examined, and biopsy samples from several different areas of the tumour should be evaluated by the histopathologist.[1] The management of tumors located near or at the gastro-esophageal (GE) junction poses a particular challenge. The esophagus and vagus nerves are at risk for injury, and the GE junction is at risk for narrowing or dysfunction if the resection is not well-planned.[6] Imatinib has revolutionized the management of locally advanced and metastatic GIST. However, the surgical methods remain the main form of treatment and the only curative one.[7]

Conclusion:

GIST should be kept as a differential in the evaluation of any submucosal mass arising from GIT. Gastric GISTs are the most common one among GISTs arising from GIT. Small GISTs are usually followed by endoscopy, while larger ones require surgical resection.

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