

CASE REPORT

DUODENAL TUMOR IN GASTROINTESTINAL STROMAL TUMORS: A RARE CASE

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ABSTRACT

Gastrointestinal stromal tumors are commonly developed in gastrointestinal tract but in duodenum is less than 3% of all gastrointestinal neoplasm. The early diagnostic criteria are very difficult in these tumors in worldwide and being a challenge for medical team due to difficult anatomical structure of duodenum. The study's aim is to know the incident rate of duodenal gastrointestinal stromal tumors. The average age of these tumors is more than 40 years but in the current case, 19-years old female patient present with complaint of melena and anemia. Her family history was negative. She was diagnosed by scope test of oral gastric duodenum with biopsy, abdominal x-rays, computerized tomography scan and histopathology. The patient was treated by Classic Whipple's procedure. According to the oncologist, patient has low risk of duodenal tumor from gastrointestinal stromal tumor. These tumors have no need to treat with adjuvant therapy: chemotherapy (Glavic) and radiation therapy after surgical resection.

Keywords: Gastrointestinal Stromal Tumors; Duodenum; Neoplasm; Hemoglobin.

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INTRODUCTION

The term gastrointestinal stromal tumor was initially invented in 1983 by Mazur and Clark. When Hirota and colleagues started working in 1998 on mutation in the KIT proto-oncogene, GIST came into recognition. "GISTs are thought to be originated from interstitial cells of Cajal (ICC) or ICC precursor cells, and are characterized by activating mutations in the KIT (CD117) and platelet-derived growth factor receptor alpha (PDGFRα) proto- oncogenes in 85-95% of all cases. The clinical presentation and tumor biology of GISTs are widely variable, with several advances being made over the past two decades in the understanding of GIST tumor biology and pathophysiology¹."

CASE REPORT

This is a recent case of a duodenal GIST of gastric origin. A 19-year-old female visited hospital with the complaint of melena since 2 months, weakness and 1 episode of hematemesis. On investigations, her hemoglobin was 8.3g/dl. Abdominal CT scan showed circumferential thickening of 2nd part of duodenum causing narrowing of lumen. Oral

Gastric duodenal scope showed the polypoidal growth in Duodenum part1 and 2 junction.

The histopathology report revealed as immune/ histochemical stains: (cluster of differentiation) CD34=Positive in spindle tumor cell, discovered on gastrointestinal stromal tumors protein1 (DOG1) = Positive in spindle tumor cell, Cytokeratins = Negative in spindle tumor cell confirmed GIST. Patient visited gastroenterologist along with the reports of Complete Blood Counts, Urea and electrolytes, Liver Function Tests and Computerized Tomography staging, CT chest, abdomen and pelvis with contrast showed a mass like lesion with peripheral enhancement and central hypo density along the first part of duodenum. Collectively measuring 2.8 x 2.6 cm, closely abutting the head of pancreas without any frank infiltration, Pylorus not involved. Other abdominal viscera were unremarkable.

Patient had a history of laparoscopic surgery for hemorrhagic left ovarian cyst year back. According to patient; no cyst fluid cytology or biopsy done at that time. Her father is asthmatic. Patient looks pallor on examination and treated as pyloric one

week ago, she had a single episode of hematemesis associated with dyspepsia, post-prandial fullness, nausea and the bile stained vomiting, dark colored stools and epigastric pain. She presented with fatigue, malaise, and weight loss, she is also complaining of vomiting, loss of appetite, dyspepsia and altered bowel habit, however on any issue with head, ear, eyes, neck, throat and skin. No any abnormal finding examined on cardiovascular, respiratory, musculoskeletal, genitourinary and neurological system.

Oro gastro duodenoscopy was performed and biopsy sent for histopathology, which gives impression of 3cm ulcerated lesion at duodenum part-1 and duodenum part-2 Junction on the posterior wall. Patient's reports were discussed in GI conference and decided for surgical evaluation on priority. Patient admitted for elective surgery (Whipple's procedure), informed consent obtained, and 3unit bloods were in hand along with latest investigation, post Classic Whipple's procedure. Patient shifted to Intensive Care Unit with postoperative management.

Three samples of surgical procedure "Classic Whipple procedure with pancreatic jejunostomy" obtained. Histopathology shows the following results; specimen#1: One lymph node (LN), free of tumor. Specimen#2: chronic Cholecystitis, no malignancy evidence. However, in Specimen#3 (Whipple resection): Gastrointestinal stromal tumor 2.5 cm, with low risk. All margins are free of tumor.

Pharmacological intervention: antibiotics, analgesia, antiemetic, anticoagulant, hormonal therapy, proton pump inhibitors and aluminum complex. Patient was discharged from hospital in stable condition. Nursing intervention: provided specific care such as pain management, wound care, infection prevention, diet, drains handling, electrolytes replacement, and early mobilization to prevent from deep vein thrombosis and physiotherapy. Patient was appointed with medical oncologist for further plan. According to the oncologist, patient has no need of adjuvant Imatinib (low risk GIST) on the impression of following results: Size 2.5 cm, Mitotic rate 4/5 mm², LN reactive and patient recall after 3 months with the report of CBC.

DISCUSSION

Small bowel tumors are particularly indefinable and pretense a unique challenge for a physician across the world. Small intestine has two types of classification benign and malignant tumors, which are neuroendocrine, epithelial, lymphoproliferative, mesenchymal, and metastatic lesions. Gastrointestinal stromal tumor is the type of mesenchymal tumor and primary tumors of duodenum are uncommon and rarely diagnosed

before surgery. Up till now, it account for >3% of all gastrointestinal cancers and geographically small intestine malignancies are higher in Moari of new Zealand and ethnic Hawaiian and low in India, Romania and other part of Eastern Europe¹.

One study was conducted in national level in Karachi, which indicates 15 cases out of 1 million in USA and 11 cases out of 1 million in Northern Europe. The incidents of GIST in Pakistan are still unknown as large studies have not been conducted².

The pathogenesis of specific type of GIST is analogous to helicobacter pylori in the stomach; have association including chronic infection stimulation, which may irritate mucosal layer or smooth muscles; immune mediated reaction of mitotic change lead to malignant changes, which can lead to GIST³. Moreover the other causes include hereditary, celiac disease, Cohn's disease, fatty diet, <40years, tobacco, alcohol and smoking. These patients presented with abdominal pain, anorexia, vomiting, melena, weight loss and hematemesis. Diagnostic studies are endoscopic procedures, CT scan, x-ray, MRI and pathological test³. The treatment options like surgery, adjuvant chemotherapy, and radiotherapy may improve the survival rate³.

Gastrointestinal stromal tumor (GIST) is the most common type of gastrointestinal mesenchymal tumor. Universally, GIST is a well-defined lesion involved of sheets or fascicles of uniform spindle or epithelioid cells. It is situated in the muscular wall of the GI tract, specifically in the stomach, also located in adenocarcinoma of gastric and gastric schwannoma. Usually Gastrointestinal stromal tumors are classified as low risk tumors; these apathetic lesions have low ability to metastasis. Immunohistochemical testing is positive for c-kit, cluster of differentiation (CD34) and Discovered for GIST-1 (DOG1), and staining results are variable for smooth muscle actin S-100 protein, and cytokeratin. The gold standard of treatment is surgical resection⁴.

The incident rate of duodenal gastrointestinal stromal tumor is more common after 40 years as well as male gender is more prone. Usually low risk tumors have symptom of melena, GI bleeding and anemia. Anatomically, duodenal GIST is located in the duodenal muscles' layers and may go into submucosal layer and lamina propria. Duodenal GIST is mostly present on ascending duodenum followed by horizontal duodenum⁵. As per literature, this patient was diagnosed at the age of 19 years, which is rare tumor, and her CT showed a mass like lesion with peripheral enhancement and central hypo density along the first part of duodenum, collectively measuring 2.8 x 2.6 cm, closely abutting the head of pancreas without any frank infiltration, Pylorus not involved. Other abdominal viscera were

unremarkable. Bowel loops are not dilated. No retroperitoneal lymphadenopathy. In the pelvis, urinary bladder outlines normally. A mild free fluid is seen in the endometrial cavity. Bilateral adnexal cysts are seen; index left adnexal cyst measures 24 mm. No pelvic sidewall or inguinal lymphadenopathy. No abdominopelvic free fluid. No suspicious pulmonary parenchymal nodularity. Thyroid gland enhances normally. No supraclavicular, axillary or mediastinal lymphadenopathy. Central airways are patent. No endobronchial lesion. Central pulmonary vasculature enhances normally without any filling defect. No pleural or pericardial effusion.

Duodenal Gastrointestinal stromal tumors developed due to continued irritation of mucosal layer of Gastrointestinal Tract, which leads to polypoid of small intestine on any region and cell mutation occurred, which increased the cell growth and may cause intussusception. Intussusception associated with common symptom of nausea and vomiting, GI bleeding, abdominal pain and melena. Endure irritant stimulation may develop the lesion on duodenal gastrointestinal stromal tumor⁶.

These factors have been related to increase the risk of malignancy in small bowel such as tobacco, alcohol, refined sugar and carbohydrates, red meat or smoked food, however, frequent intake of coffee, fish, fruit, and vegetables have been seen to reduce the risk of cancer. Some predisposing factors demonstrated the small bowel tumor. Such as inflammatory bowel disease, Crohn's disease, coeliac disease and hereditary syndromes: familial adenomatous polyposis, Lynch syndrome⁷.

Clinical manifestation of gastrointestinal stromal tumors is Melena, GI bleeding, vomiting and dyspepsia caused by pressure necrosis and ulceration of the overlying mucosa. Hardly, the patient present with complaint of bowel obstruction or tumor rupture with hemoperitoneum. An exact diagnosis is initiated on a combination of complete medical history, thorough physical examination, and imaging modalities. First diagnostic tool is abdominal X-ray, which shows the symptoms of obstruction. Barium studies of upper gastrointestinal tract show stacked coin or coiled spring sign due to edematous mucosal folds. However, endoscopy and abdominal computed tomography (CT) scans are also helpful to diagnose the primary gastrointestinal stromal tumor⁸.

Approximately 5% duodenal gastrointestinal stromal tumors are considered surgical resection. However, 30% of primary tumors of duodenal gastrointestinal stromal tumors are account to very difficult to diagnose whether a duodenal gastrointestinal stromal tumor is benign or malignant. Moreover, the anatomical location of duodenal gastrointestinal stromal tumor makes it more complex and unique in nature, which leads to challenging surgical

interventions⁹. The size of tumor, extent of disease and location of tumor indicate the treatment procedure type. Basic curative treatment of duodenal gastrointestinal stromal tumor is complete surgical resection of all margins¹⁰.

As we discussed, the incident rate of duodenal gastrointestinal stromal tumors is rare in duodenum part 1 and duodenum part 2. The 15 patients diagnosed out of 1 million in USA, male gender is at high risk instead of female and median age of this tumor is more than 40 years. The smooth muscles layers of gastrointestinal tract involve in mutation of oncogene are associated with polypoid and may cause obstruction or intussusception. Duodenum gastrointestinal stromal tumor mostly present with abdominal pain, nausea and vomiting, melena and anemia. The primary cause of gastrointestinal stromal tumor is unknown but other predisposing factors increase the risk of neoplasm as age, hereditary, food and alcohol. The importance of diagnostic approach at initial stage is curable. Surgical resection of duodenal gastrointestinal stromal tumor is the first line treatment with adjuvant therapy, chemotherapy and radiation therapy.

CONCLUSION

In conclusion, the rate of duodenal GIST is <3% as per data. It is uncommon in D1 and D2 part of duodenum. Secondly, male is at high risk rather than female, these patients mostly present with the complaint of abdominal pain, nausea and vomiting, melena and anemia. The etiology of GIST is unknown but age, hereditary, food and alcohol can increase the risk of malignancy. Early detection is highly recommended in GIST, moreover surgical removal of duodenal GIST and adjuvant therapy showed positive outcomes.

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CONFLICT OF INTEREST

There was no conflict of interest among the authors.

ETHICS APPROVAL

The study approval was sort from the Shaukat Khanam Memorial Cancer Hospital and Research Center Institutional Review Board (IRB Number is EX-29-07-19-02).

PATIENTS CONSENT

Informed consent was taken from the patient of the study.

AUTHORS CONTRIBUTION

SH conceived the idea, did bench work, AN searched this case, compiles data and wrote the manuscript, AA helped in designing of the case report and facilitated, JA supervised the case report and finalized the manuscript.

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