Giant Gastric Gastrointestinal Stromal Tumor Encasing Splenic Hilum - A Case Report

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Abstract
Gastrointestinal stromal tumour (GIST) is a rare tumour of GI tract (0.2%). GIST arise from Interstitial cell of Cajal which intercalates between smooth muscle cells and intramural neuron. Stomach is the most common site followed by small bowel, rectum and colon. Commonly presents as abdominal pain, GI bleed, Intestinal obstruction and perforation. In spite of the impressive advances in the diagnosis and target therapy for GIST, surgery remains the standard treatment. We report a case of GIST originating from stomach in a 53 year male who presented with the mass abdomen and weight loss with emphasis on its going through surgical resection in our Institute.

Keywords: GIST, Stomach, GIANT.

Introduction
Gastrointestinal Stromal Tumour (GIST) is a rare disease. It is the most common mesenchymal tumour of the GI tract around 0.2% of all GI tumour(1). It arise from Interstitial cell of Cajal (intestinal pace maker cell). Originally GIST is believed to be arising from smooth muscle of GI tract (formerly termed as leiomyoma and leiomyosarcoma)(2). It may be sporadic or familial. Mostly arise from stomach then from small bowel, rectum and colon(3). 95% GIST express c-kit (CD117) mutation and 5% express mutation of platelet derived growth factor alpha (PDGF-A). Clinically presents as abdominal mass, pain abdomen, weight loss, Intestinal obstruction and perforation. Diagnosis mainly done by CT and MRI of abdomen. Surgery is the main mode of treatment in resectable cases. Imatinib Mesylate a tyrosine kinase inhibitor is much beneficial as an adjuvant therapy to surgery and in unresectable cases.

Case Report
A 53 years old male got admitted in our Department with chief complaints of Pain in upper abdomen for 6 months, a swelling in left upper abdomen for 4 months. Pain was insidious in onset, dull aching in nature, not radiating or shifting to any other site, aggravated by taking food, not associated with nausea, vomiting.
Following this he noticed a small lemon sized swelling over left upper abdomen, gradually increasing in size to attain the present size, which was associated with decrease in appetite and weight loss of around 6-8kgs in last 2 months. There was no H/O fever, bleeding P/R, black stool, altered bowel habit, yellowish discolouration of urine or sclera. On clinical examination patient was of average body build, mild pallor, no icterus, no edema, no lymphadenopathy. On per abdominal examination there was a lump palpable over left hypochondrium extending toward epigastric region of size 14 X 12 cm with smooth surface, irregular margin, upper pole can be reached, firm in consistency, moving with respiration, mobile side to side. On head rising test, swelling was found to be intra-abdominal. Shifting dullness and fluid thrill both were absent.

CT of abdomen revealed a large approx. 16 X 12X 9 cm sized centrally necrotic mass lesion in left upper quadrant, in left sub phrenic region closely related to body of stomach, indenting the body of pancreas with mild adjacent fat stranding. Fat planes with the adjacent structure appear to be maintained with no significant regional lymphadenopathy- appearance suggestive of neoplastic pathology most likely GIST arising from body of stomach.

On Histopathological study of the tumour- Micro section showed interlacing bundles of spindle and epitheloid cells with hyper cellularity, moderate nuclear atypia with mitosis >10 /50 HPF. At places showing nuclear palisading suggestive of malignant GIST. Immunohistochemistry was positive for CD117. We referred him for oncology consultation. Presently he is on Tab Imatinib (400)- 1 tab OD ,Tab Iron, folic acid- 1 tab OD. At present patient is asymptomatic and is on regular follow up.

Discussion
Golden and Stout in 1941: described the mesenchymal tumors arising in bowel as tumors arising from smooth muscle cells: leiomyoblastoma, leiomyoma and leiomyosarcoma. Term GIST was 1st used by Mazur and Clark in 1983. In 1998 Japanese research workers (Hirota et al)
discovered KIT mutations in GIST that possibly distinguish GIST from other tumor. GIST is a GI tract associated stromal (mesenchymal) neoplasm with activating mutations in c-KIT (CD117) or PDGF-A, whose line of differentiation recapitulates the interstitial cells of Cajal and has broad spectrum of biological behavior. Most common benign nonepithelial tumor of the GI tract. Oesophagus: 5%, Stomach: 50-70%, SI: 25-40%, Duodenum – 10-20 %, Jejunum – 27-37%, Ileum: 27-53%, Colorectal: 10%, Extra - gastrointestinal GIST : 6.7% (5). DOG 1 (discovered on GIST 1): 87-97.8%, CD117 up to 95%, Protein kinase C theta – 96%, Heavy caldesmon -80%, CD 34 -70%, Nestin – non specific (positive in schwannoma, leiomyosarcoma and melanoma), Smooth muscle actin 20-30%, S100 – 5%(5). Often asymptomatic, especially when small - Symptoms nonspecific — GI bleeding (53%), Abdominal pain (32%), Pulpable mass (13%). Other symptoms may include- Early satiety, Fatigue from anemia, rarely obstruction. The number of mitotic figures present can be used to histologically grade GISTs. In general, GISTs with less than 1 mitotic figure per 50 high-powered fields (HPFs) are correlated with benign behavior. A finding of 1-5 mitoses per 10 HPFs suggests potential malignancy. A finding of more than 5 per 10 HPFs indicates malignancy. A finding of more than 10 per 10 HPFs denotes high-grade malignancy(6,8).

Initial patient workup should include- Complete hemograms, LFT, RFT, CT of abdomen and pelvis with oral IV contrast. Selective use of tissue biopsy- Cytology/pathology for spindle cell morphology, CD117(c-KIT), Comorbidty assessment. Surgery optimal for resectable GIST- Goals of surgery, Complete gross resection, Negative microscopic margins (R0 resection). If recurrence develops after surgery, disease is usually not curable(7). Imatinib mesylate is approved for treatment of GIST resistant to or intolerant of imatinib- Inhibits multiple receptor tyrosine kinases, Antitumor and anti-angiogenic activities in preclinical studies. It can be used for metastatic or unresectable cases, as an adjuvant or neoadjuvant therapy.

**Conclusion**

Surgery is the first-line treatment for patients with resectable GISTs – Up to 50% patients have recurrence after complete resection. Tyrosine kinase inhibitor imatinib now is the standard treatment for unresectable or metastatic or advanced GIST. Because of specific therapy, accurate preoperative diagnosis is very useful and can help to plan surgery in resectable lesions or alternative therapy in unresectable or metastatic lesions.

**References**
