VOLUME-6 | ISSUE-4 | APRIL - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

A CASE OF MALIGNANT GIST OF STOMACH : PEDUNCULATED MOBILE MASS IN RIGHT LUMBAR REGION WITHOUT GI SYMPTOMS

General Surgery

KEYWORDS: STOMACH, GIST, MALIGNANT, LUMBAR MASS

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ABSTRACT

Gastrointestinal stromal tumors are the most common primary non-epithelial tumors of stomach. They are unique owing to origin due to activation of KIT mutation and subsequent expression of KIT proteins. A 45 year male patient presented with mass in right lumbar region since 15 days, with no other positive history. Mass cystic in consistency, well defined margins, smooth surface and moves with respiration. Tomographic scan and fine needle cytology confirmed it as GIST. Intra-operative, pedunculated mass was evident arising from stomach extending upto fat plane of transverse mesocolon. Patient managed with subtotal gastrectomy with Billroth-II procedure. Biopsy report confirmed the diagnosis as GIST tumor with high malignant potential with high mitotic rate with no lymphadenopathy.

GIST, a relatively well circumscribed neoplastic lesion are considered as tumors with uncertain malignant potential.

Introduction

Term 'GIST' coined in 1983 by mazur & clark to describe non-epithelial tumor of GI that lacked Ultrastructural features of smooth muscle cells as well as Immuno histo chemical characteristic of schwann cells[3].

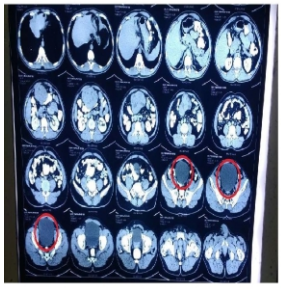
GIST are rare malignancies. Although,they are most common sarcoma of GIT,they represent **only 0.2% of all GI tumors**[1,2]. Arising from **interstitial cells of cajal** ,occurs due to **gain of function mutation of KIT proto-oncogene**.KIT is a Receptor tyrosine kinase,activated when bond to ligand known as steel factor/ stem cells factor.

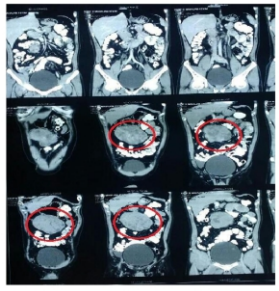
GIST now identified by near universal expression of **CD117 antigen (95%)** In estimated **(5-10%)**KIT staining is faintly or **truly negative**. Approximately **(7%)**of GIST have gain of function mutation of **PGFRA tyrosine kinase receptor**.

Case History

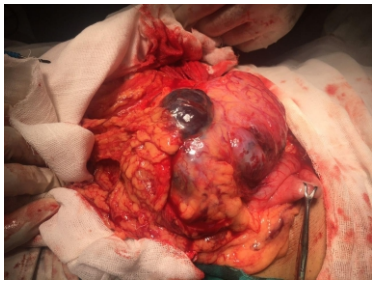
A **45 year** old male patient presented with history of **Mass per abdomen since 15days**, insidious onset, gradually progressive & painless. On examination, Patient well built & nourished with no pallor, icterus or lymphadenopathy. Mass, present in **Right lumbar** region, extending upto umbilical region, Approx **10×8 cm** in size firm consistency, well defined margins, smooth surface, **mobile in all directions** & moves well with respiration. Few other pedunculated swellings viz. Mesenteric cyst, hydatid cyst of mesentry were the differentials. Sonogram revealed hypoechoic lesion with internal vascularity with Calcification, in close approximation to bowel loops. **Cytology revealed spindle cells in fascicles -pointing towards synovial sarcoma with possibility of GIST.CT Scan** revealed a **malignant featuring lesion with loss of fat plane of greater curvature of stomach & transverse colon.** Intra-operative, a pedunculated mass was evident arising from stomach, extending to fat plan of transverse mesocolon. **Patient was managed with Billroth-II procedure with sub-total gastrectomy with omentectomy. Biopsy** report confirmed the diagnosis as **GIST tumor with high malignant potential with high mitotic rate** with no lymphadenopathy with **tumor free resected surgical margins. IHC** suggestive of **CD117 negative** with **vimentin positive.**

Pre-operative CT :





Intra-operative:



Discussion

Primary GIST can arise throughout GI tract but **most common in stomach(40-70%)** small bowel(20-40%) & colo-rectal(5-15%) with least common in esophagus(5%)[3]. Most common in 40-60 years of age group with equal incidence in males & females. A few GIST are familial, caused by germline mutation of KIT. **Many GIST are asymptomatic or may present as mass lesion** or vague abdominal pain & **can grow to large size before producing symptoms.**

GIST can be highly vascular & bleeding is one of the most common presenting symptoms.Tumor is typically soft & friable & can cause life threatening hemorrhage. **CT scans** are critical to determine the **anatomic extent of GIST** & to assist with operative planning. **PET scan** to assess **Imatinib response** is recommended to guide therapeutic dosing or timing of surgery with a baseline pre treatment scan followed by repeat scan after 2-4 weeks of drug therapy.

Tumors >5 cm, increased Mitotic Activity while once arising from **small bowel** indicate **poor prognosis.**

	Roux-En-Y	Loop
	gastro-jejunostomy	gastro-jejunostomy
Functional	**More**	**Less**
Anastomosis		
Anastomotic Leak	Less	More
Time Duration	More	Less
Skill & Learning	Longer	Shorter
Curve		

CLASSIFICATION OF PRIMARY GASTROINTESTINAL STROMAL TUMORS BY RISK OF METASTASIS

RISK CATEGORY	SIZE	MITOTIC COUNT
Very low	<2cm	<5 per 50 HFPs
Low	2-5cm	<5 per 50 HFPs
Intermediates	<5cm	6-10 per 50 HFPs
	5-10 cm	<5 per HFPs
High	>5cm	>5 per HFPs

Conclusion

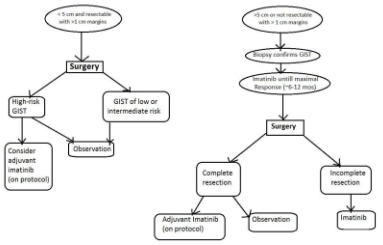
GIST are histologically heterogeneous group of mesenchymal tumors, showing CD117 positivity regardless of the site.These are most common primary Non-epithelial tumor of stomach & small bowel. **These are smooth muscle tumors of uncertain malignant potential.** Between **15-50% of GIST presenting with overtly metastatic disease**,most common metastatic sites are liver & peritoneum[3,4].

GIST almost never metastasize to regional lymph nodes but can invade adjacent organs. **Surgery remains the standard therapy for all Non-metastatic tumors** with reported **resectability rate** for localized primary GIST is **70-80%. Endoscopic submucosal recession** is preferred for **pedunculated GIST**and **tumor size less than 5 cm. Subtotal gastrectomy** is advocated for **tumors** arising from **body of stomach or pyloric region**, while **total gastrectomy** for tumors **more than 5 cm.** Surgical tumor **Resection with Approx 1 cm** modest margins is **standard goal.Regional lymphadenectomy** or intra-operative **incisional biopsy is not recommended.**

If lesion involves adjacent organs, en block resection is considered to avoid tumor spillage. In a **study by Singer** and associates **5 years recurrence free survival (76 +/-9%)** per patient **with clear surgical margins**, while (15+/-8%) with grossly or microscopically positive margins.The typical sites of **tumor recurrence post surgery** are **local resection bed**, liver and peritoneum.

Post operative administration of imatinib mesylate improves disease control. In **series of patient treated with imatinib after resection bumming and associates reported recurrence free interval of 7 to 13months.** In absence of adjuvant therapy, approximately 50% of Patient's receiving potentially curative surgery develop either local recurrence or metastatic disease within 5 years, yielding **5 years survival rate of 40-55%. Surgery** in form of **Debulking** has important role **in palliation** of patient's **recurrent or Metastatic** disease. **Radiation** has very **limited role** in management of the GIST. The advent of effective chemotherapy-**Imatinib Mesylate** being **drug of choice**, has limited the role of radiation therapy, & **achieved localized tumor shrinkage**, also **haulting** the **disease progression.**

SOLITARY INTRAABDOMINAL MASS SUGGESTING SUBMUCOSAL ORIGIN



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