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Gastro-intestinal stromal tumors (GIST)

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Abstract
GISTs are the most common sarcomatous tumors of the gastrointestinal tract. They arise from the interstitial cells of Cajal which are the intestinal pacemaker cells controlling gut motility. They have unpredictable biologic behavior. Upper GI endoscopy and CT scan are diagnostic. Radical surgical resection is the mainstay of treatment. This may be accompanied by adjuvant therapy in the form of imatinib to prevent recurrence and improve the survival. Prognosis depends upon the tumor size and the mitotic index.

Keywords: Gastrointestinal, stromal, tumors, management

Introduction
Gastrointestinal stromal tumors are malignancies arising from the mesenchymal tissue. They may originate from any part of the gastrointestinal tract most commonly in the stomach and small intestine. The word GIST was first coined by Mazur and Clark in 1983 [1]. They originate from the Cajal cells which are pacemakers for gut peristalsis and are usually situated in the myenteric plexus of the gut. Various mutations have been associated with GISTs such as mutations of KIT (CD-117) [1, 2]. Majority of GISTs are sporadic, but may be associated with various syndromes.

Etiopathogenesis
GISTs are observed equally in males and females. They commonly present in patients over 50 years of age. They may appear anywhere in the gastrointestinal tract such as in the stomach (40-60%), jejunum and ileum (30%), duodenum (5%) and colon (15%) [2, 3]. They rarely arise in the esophagus and appendix. Extra gastrointestinal sites for GIST's have been reported in the omentum, mesentery, retroperitoneum, gall bladder and rarely in the urinary bladder [4]. The size of the tumors may vary from 1cm to 40cm. Lymph node metastasis are extremely rare. Most common metastasis is to the peritoneum and liver. Size and mitotic index are the best predictive index of metastasis in majority of cases [5]. GIST appear as cystic masses containing large amount of foul-smelling pus or even bowel fistulas in rare cases. Intra-tumoral abscesses is a rare presentation with anecdotal case reports in literature. These tumors are grayish white lesions having whorled appearance on cut surface. Histologically well differentiated smooth muscles are seen.

3 histological types are seen on the basis of cellular appearance
1. Fusiform (77%)
2. Epithelioid (8%)
3. Mixed (15%)

Immunohistochemistry
About 95% GISTs express CD-117 (C-KIT proto-oncogene) while 70-90% express CD-94 (human progenitor cell antigen) [5, 6]. Platelet derived growth factor receptor alpha (PDGFRA mutations) is seen along with C-KIT. C-KIT mutations are usually seen in exon 19 and 11. These tumors may sometimes stain positive for actin (20-30%), S-100 (2-4%) and desmin (2-4%) [5].

Clinical features
Symptomatically GISTs presents with bleeding (hematemesis/malena), abdominal pain,

discomfort and weight loss.
Tumors with intramural growth may also present with signs of obstruction where as those with extramural growth may assume large size to render it palpable.

3 syndromes are linked to GIST [1, 2, 3]

1. Carney triad syndrome comprising of gastric GIST, para-ganglioma and pulmonary chondromas.
2. Carney - Stratakis syndrome comprising of GIST and para ganglioma.
3. Neurofibromatosis type I comprising of multifocal GIST mostly located in the small intestine.

Diagnosis

Diagnosis is best established by upper GI endoscopy, if involving the stomach. For tumors arising lower down in the gut, CT and MRI maybe helpful for assessment of the primary lesion and detection of metastasis [6, 7].

Treatment

Surgery is the mainstay of treatment. Surgical resection should include en-block removal with the pseudo-capsule intact. Tumors greater than 2 cms have a higher chance of metastasis [6, 7].
The aim of surgery is to achieve an R0 resection. For smaller tumors wedge resection is sufficient but for larger tumors a wider extend of resection is indicated. No lymph node resection is necessary [7].
Adjuvant therapy with imatinib (Tyrosine kinase receptor inhibitor) is used to prevent recurrence following surgery in unresectable cases [8].
It is effective in patients who have mutations in exon 11 of the KIT gene. Patients having mutations in exon 9 of the KIT gene may also respond to imatinib but in higher doses [8]. Patients devoid of mutations do not respond to imatinib. For imatinib refractory GISTs, sunitinib (Tyrosine kinase inhibitor) is useful as the drug targets multiple kinases such as VEGF, EDGFR, PDGFRA, KIT and FLT3 [9]. If facilities for PET scan are available then pre-operative screening for GIST as well as detecting early response to imatinib can be done.
Follow up for GIST is essential. Low risk patients may require an abdominal CT or MRI every 6-12 months for the first 5 years. High risk patients may require follow up with CT/MRI every 3-6 months for the first 5 years during adjuvant therapy. After completion of adjuvant therapy, follow up every 3 months for the first 2 years and subsequently every 6 months for next 5 years and thereafter annually for additional 5 years is advisable [8, 9, 10].

Conclusion

GISTs are malignancies arising from the mesenchymal tissues. Typical symptoms and associations should raise the suspicion of a GIST especially in the stomach and small intestine.
Upper GI endoscopy and CT scan are diagnostic. Surgical resection with adjuvant therapy helps in improving the survival. Inoperable, recurrent or metastatic cases may be treated only by adjuvant therapy.

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