Getting the Gist of GIST. . .

Radiological Characteristics
of Gastrointestinal Stromal Tumors

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GIST Presentation Goals

• Patient Presentations
• Differential Diagnosis
• Background Information
• Classic Clinical Presentation
• Radiographic Characteristics
• Management Options
• Summary
Our Patient #1

- 67 year old female with several intermittent episodes of dysphagia with solid food. No difficulty with liquids. No GI bleeding.
- PMH: HTN, Hyperlipidemia
- PE: non-contributory
- Upper GI series…
- Endoscopic Ultrasound… (not avail.)
- CT Abdomen…
• Our Patient #1 UGI Series

• normal esophagus, no lower esophageal ring, or hiatal hernia.

• 2.5 x 3.0 cm submucosal mass in the fundus of the stomach.
• Our Patient #1
• CT Abdomen +IV +Oral Contrast

• 2.5 x 2.9 cm mass along the greater curvature in the body of the stomach close to the fundus.
• Mass is hypodense with central ulceration.
• Small area of calcification
• Our Patient #1
• CT Abdomen +IV +Oral Contrast (cont.)

• Mass is also extramural extending to the pancreatic tail and is adjacent to the splenic vein.
Our Patient #2

- 75 year old male with two months of abdominal fullness.
- Visual endoscopy showed a localized 2.9 x 2.7 cm mass at the fundus of the stomach.
- Endoscopic ultrasound with fine needle aspiration with biopsy results suggestive of spindle cell origin or gastrointestinal stromal tumor.
- Scheduled for preoperative abdominal CT scanning prior to laparoscopy and resection.
• Our Patient #2
• CT Abdomen +IV +Oral Contrast (cont.)

• 2.9 x 2.7 cm mass along in the fundus of the stomach.

• Mass is submucosal in origin. Note the outline of the mucosa
Differential Diagnosis

- Gastrointestinal Stromal Tumor
- Other mesenchymal neoplasms such as
  - Leiomyoma
  - Leiomyosarcoma
  - Schwannoma
- Neurofibroma
- Neuroendocrine neoplasms such as a solitary gastric carcinoid
- All may radiographically present similarly to GIST.
The GIST is . . .

- GISTs are nonepithelial mesenchymal tumors. A unique group distinct from smooth muscle and neural tumors such as leiomyomas, leiomyosarcomas and schwannomas.
- 50 years of debate - GIST previously categorized as smooth muscle cell tumors. Until 1983 when electron microscopy and immunohistochemistry showed a lack of smooth muscle cells.
- > 95% of GISTs express the CD-117 receptor which is the tyrosine kinase growth factor receptor, a product of the c-kit protooncogene.
- Mutations in the c-kit gene may result in its uncontrolled constitutive activation causing overexpression of CD-117 receptor.
The GIST is . . .

• The cellular origin of GIST cells is unknown however it is theorized that they arise from primitive stem cells, related to the interstitial cells of Cajal, the intestinal pacemaker cells.
• This theory does not explain phenotypically identical GIST primaries that arise in the omentum and mesentery.
• The frequency of GISTs is 10–20 cases per million.
• Patients with neuro-fibromatosis type 1 have an increased prevalence.
• Usually occur after the age of 50, but may affect a broad age range, and are disproportionately common in men.
Clinical Presentation

• Symptoms include anorexia, weight loss, nausea, vomiting, and abdominal pain.
• Gastrointestinal bleeding 40%
• Abdominal mass 40%
• Abdominal pain 20%
• 25 to 40% of patients present with intestinal obstruction, which may be due to the tumor serving as a lead point for intussusception.
• Most small GISTs remain asymptomatic.
• Large lesions may ulcerate and present with bleeding.
• Small benign GISTs are discovered incidentally during radiologic evaluation or surgery for another condition.
Anatomical Tropism of GIST

- GISTs most frequently occur in the stomach (60-70%)
- Followed by the small intestine (20%-30%)
- anorectum (7%)
- colon, and esophagus (<1%)
Indicators for GIST Malignity

- Advanced Patient Age
- Tumor size: >3cm
- Growth Rate: < 16 month doubling time.
- Presence of Metastases
- Histological/Molecular Markers
- Radiographic Findings
Radiographic Findings of GIST

- Plain film may show a nonspecific soft-tissue mass indented or displacing the gastric air shadow.
Radiographic Findings (cont.)

- UGI Series: lateral view shows a **submucosal mass** that forms **obtuse or right angles** with the gastric wall in profile.
- Usually there is a smooth surface, but there may be ulcerations.
- Rarely it may present as a mucosal polypoid mass.
Radiographic Findings (cont.)

- CT Scan may show…
- An **intramural component**
- As well as an **extragastric extension** into the gastrosplenic ligament
- **Calcification** is a rare finding.

Levy et al. Radiographics 2003; 23:283–304
Radiographic Findings (cont.)

- CT Scan may show...
- **peripheral enhancement** pattern correlating with areas of viable tumor.
- **central low attenuation** corresponds to hemorrhage, necrosis, or cyst formation.
- extragastric **extension** into the gastrohepatic ligament

Levy et al. Radiographics 2003; 23:283–304
Radiographic Findings (cont.)

- CT Scan may show...

- There may be cysts filled with air…

Levy et al. Radiographics 2003; 23:283–304
Radiographic Findings (cont.)

- CT Scan may show...

- … or cysts filled with oral contrast.
Radiographic Findings (cont.)

• If the tumor is very large and its origin is obscured on CT, reformations or MR imaging may be helpful.

• Occasionally the mass may be Homogeneously enhanced.
Management

• Based upon:
  – Confidence in the diagnosis
  – Tumor location, size, presence of metastases
  – Clinical presentation

• Surgical resection is the treatment of choice.

• 5 year survival rate of 34-60%.
New Treatment Option

- Gleevec ® (imatinib mesylate) is a tyrosine kinase inhibiting agent initially FDA approved for treatment of CML.
- can induce dramatic, rapid, clinical benefit for GIST patients with advanced or unresectable metastatic disease.
- Incredibly, appreciable and measurable responses to therapy with Gleevec have been noted as early as 24 hours after initiation of treatment.
- Response to therapy is assessed with CT or PET or PET/CT, the best modality to use is currently under research
Do You Get the GIST?...

• Distinct set of nonepithelial mesenchymal tumors.
• Express CD-117 receptor.
• Stomach is the most common site of GIST.
• GISTs account for 1-3% percent of all stomach neoplasms.
• Radiology, along with pathology, plays an important role in the diagnosis, treatment and prognosis of GIST.
References

- Careweb Clinical Lookup, BIDMC, 2004
- PACS, BIDMC, 2004
Acknowledgments

• Dr. Stien and Dr. Barth
• Dr. Kruskal and Dr. Raptopoulos
• Dr. Lieberman
• Pamela Lepkowski
• Larry Barbaras, Webmaster