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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

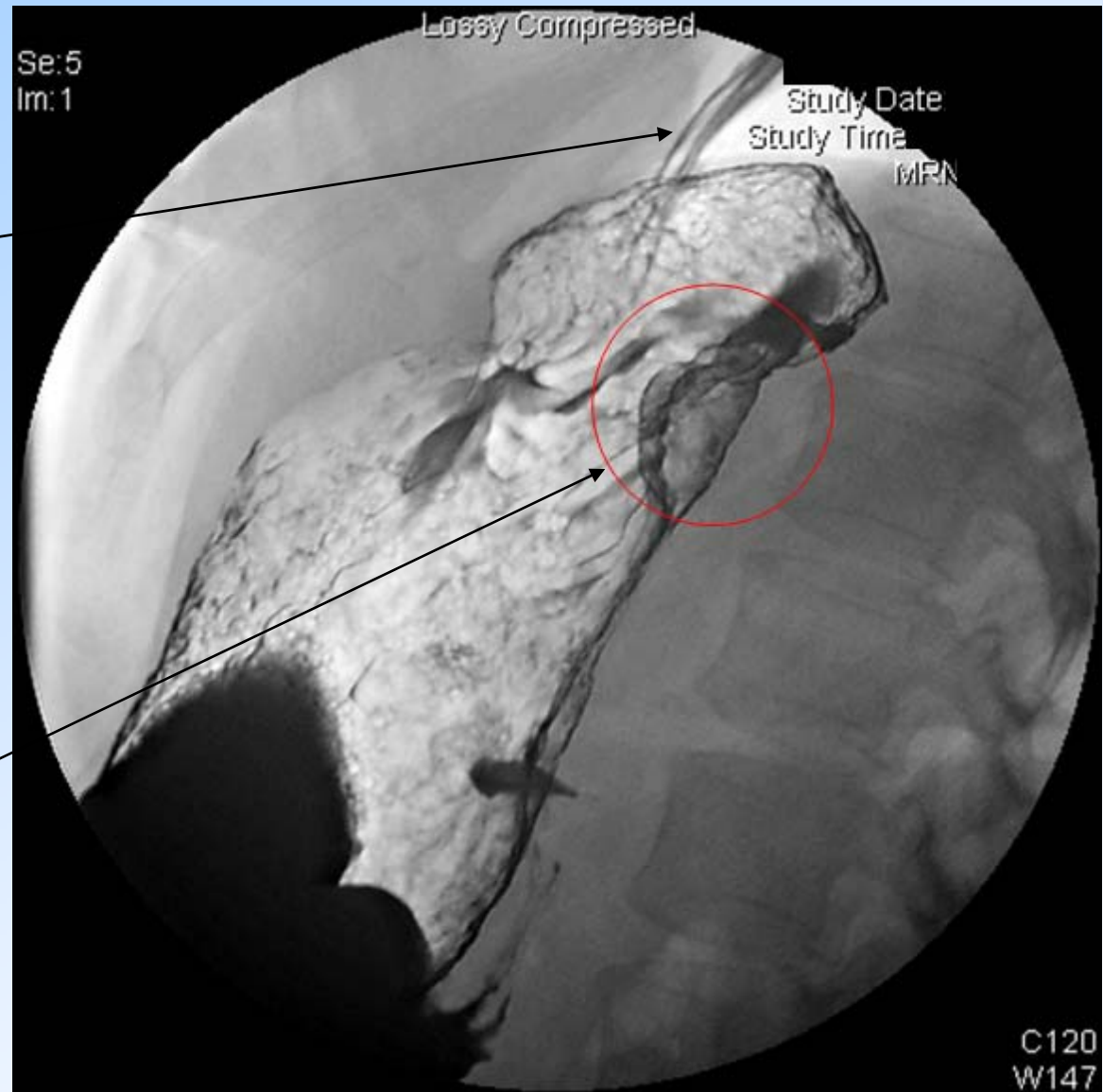
- 67 year old female with several intermittent episodes of dysphagia with solid food. No difficulty with liquids. No GI bleeding.
- PMH: HTN, Hyperlipidemia
- PE: non-contributory
- Upper GI series...
- Endoscopic Ultrasound... (not avail.)
- CT Abdomen...



• 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.



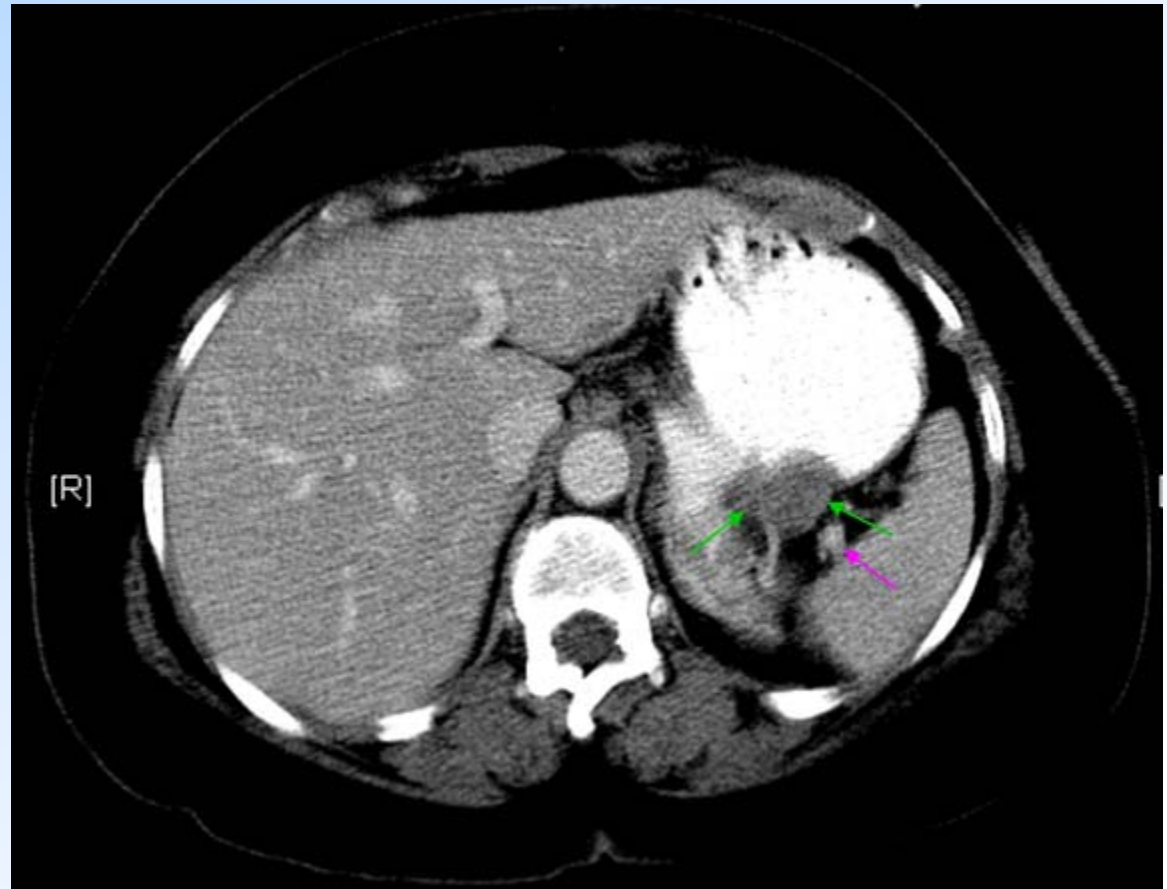


- Our Patient #1
- CT Abdomen +IV +Oral Contrast
- 2.5 x 2.9 cm **mass** along the greater curvature in the body of the stomach close to the fundus.
- Mass is hypodense with **central ulceration**.
- Small area of **calcification**





- 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

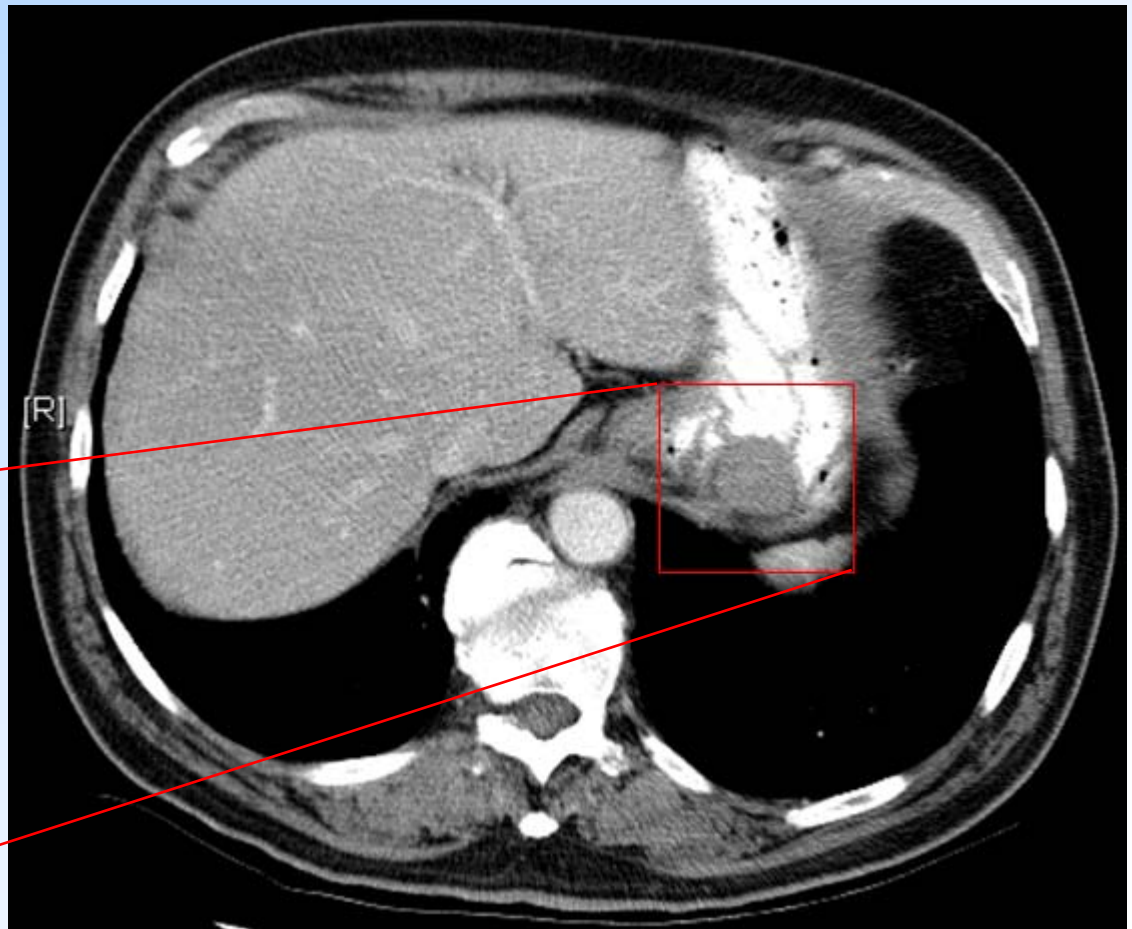
- 75 year old male with two months of abdominal fullness.
- Visual endoscopy showed a localized 2.9 x 2.7 cm mass at the fundus of the stomach.
- Endoscopic ultrasound with fine needle aspiration with biopsy results suggestive of spindle cell origin or gastrointestinal stromal tumor.
- Scheduled for 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



2x



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%)



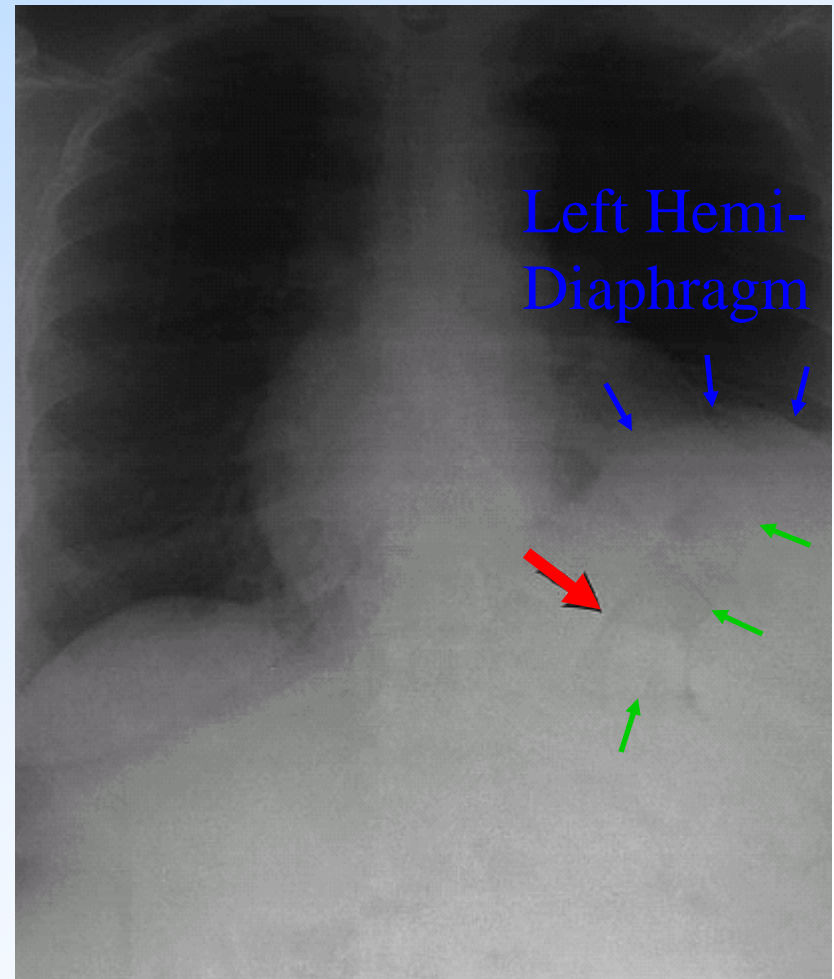
Indicators for GIST Malignancy

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



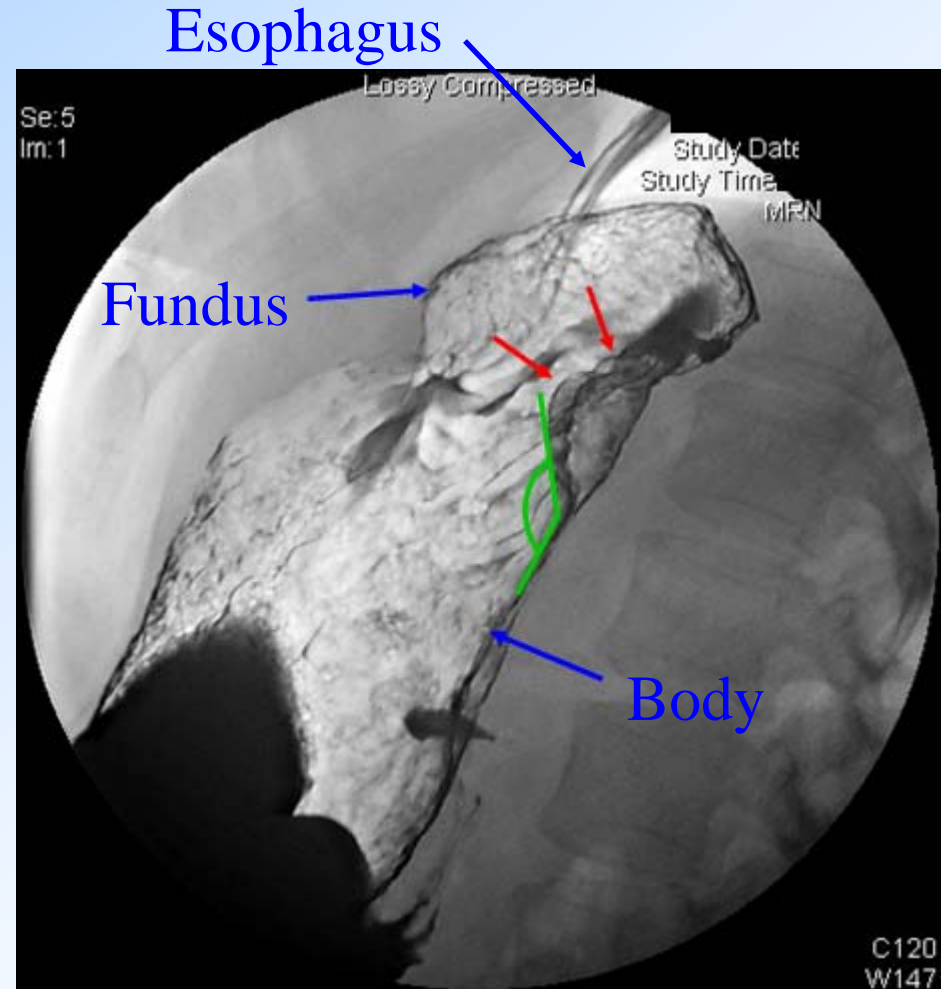
Radiographic Findings of GIST

- Plain film may show a nonspecific soft-tissue mass **indenting** or displacing the **gastric air shadow**.



Radiographic Findings (cont.)

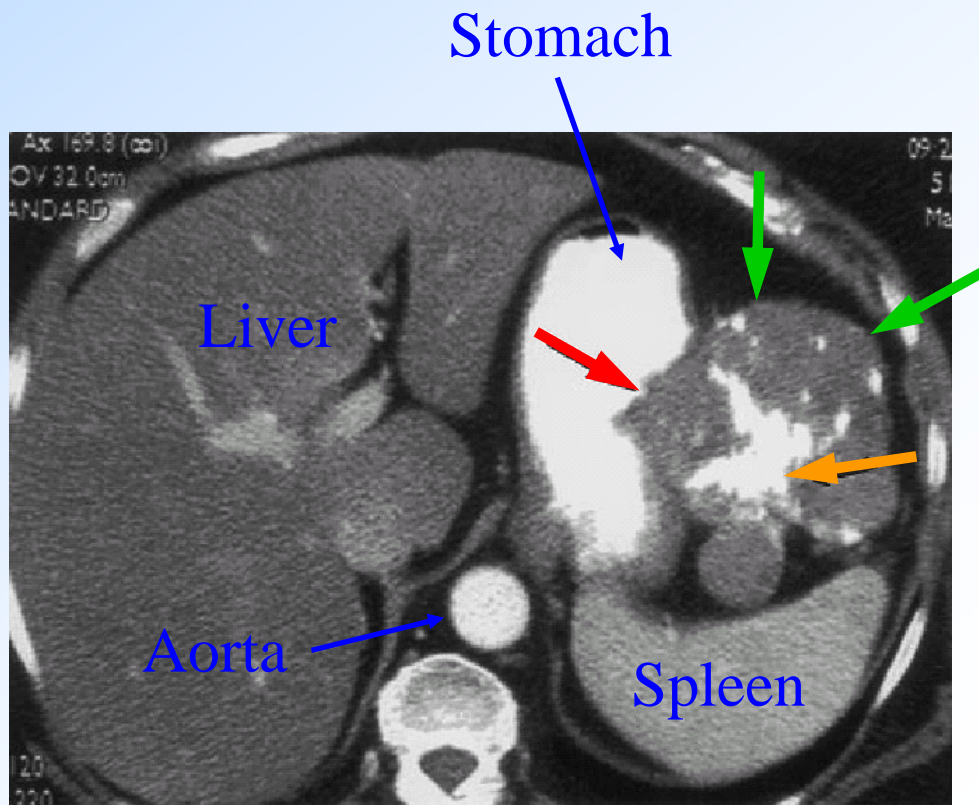
- UGI Series: lateral view shows a **submucosal mass** that forms **obtuse or right angles** with the gastric wall in profile
- Usually there is a smooth surface, but there may be ulcerations
- Rarely it may present as a mucosal polypoid mass.



Radiographic Findings (cont.)

- CT Scan may show...

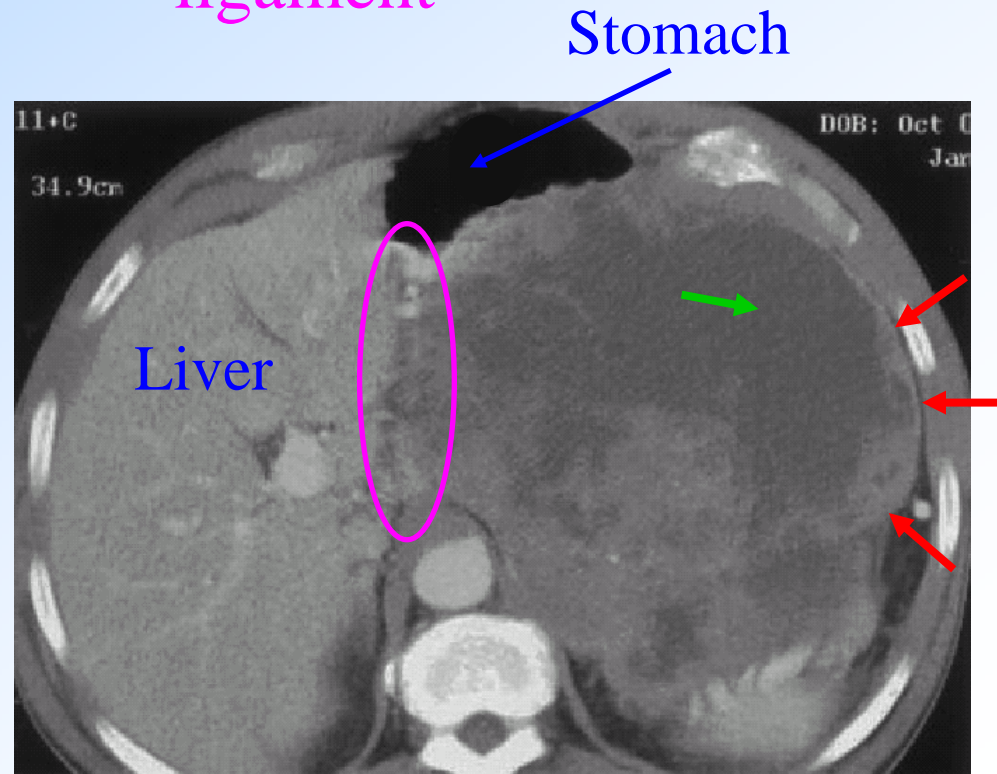
- An **intramural component**
- As well as an **extragastric extension** into the gastrosplenic ligament
- **Calcification** is a rare finding.



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.

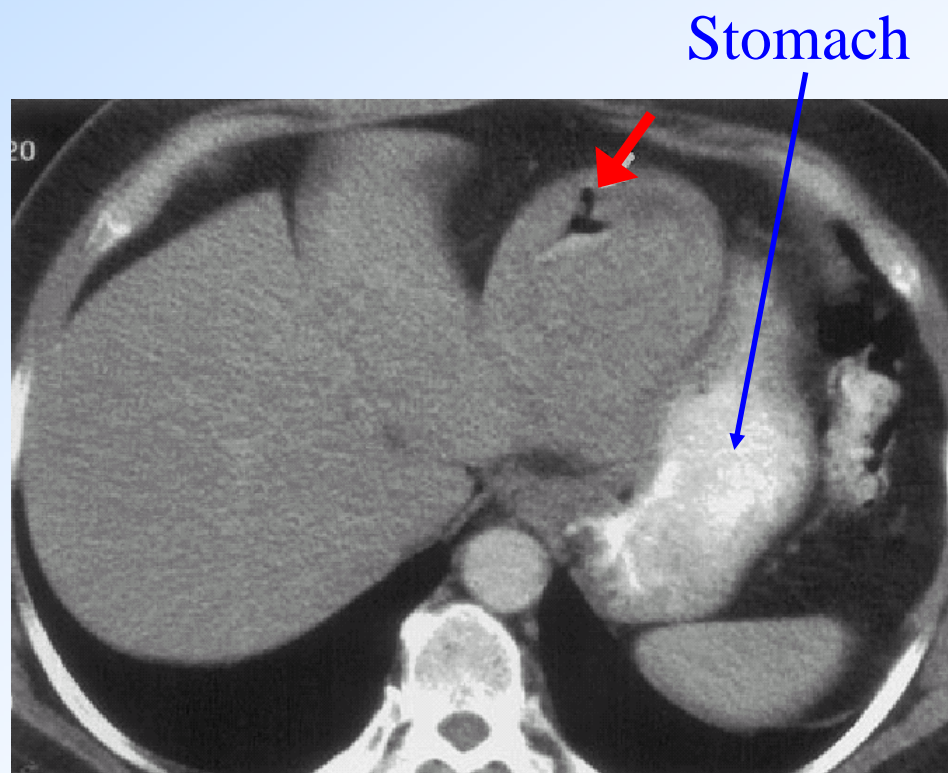
- extragastric **extension** into the gastrohepatic ligament





Radiographic Findings (cont.)

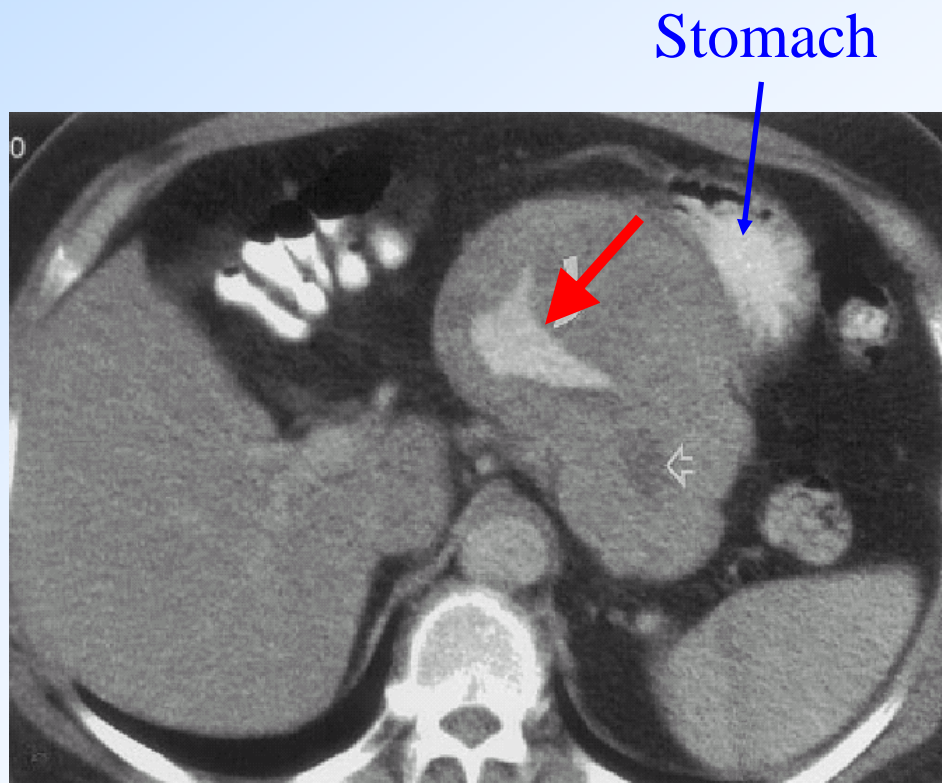
- CT Scan may show...
- There may be **cysts filled with air**...





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

- Bralow et al. Gastrointestinal stromal tumors and sarcomas of the gastrointestinal tract. Up To Date Online, version 12.2, April 2004.
- Burkill et al. Malignant Gastrointestinal Stromal Tumor: Distribution, Imaging Features, and Pattern of Metastatic Spread. *Radiology* 2003; 226:527–532.
- Careweb Clinical Lookup, BIDMC, 2004
- Chun et al. Gastrointestinal leiomyoma and leiomyosarcoma: CT differentiation. *J Comput Assist Tomogr* 1998; 22:69.
- Levy et al. Gastrointestinal Stromal Tumors: Radiologic Features with Pathologic Correlation. *Radiographics* 2003; 23:283–304.
- PACS, BIDMC, 2004



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