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…

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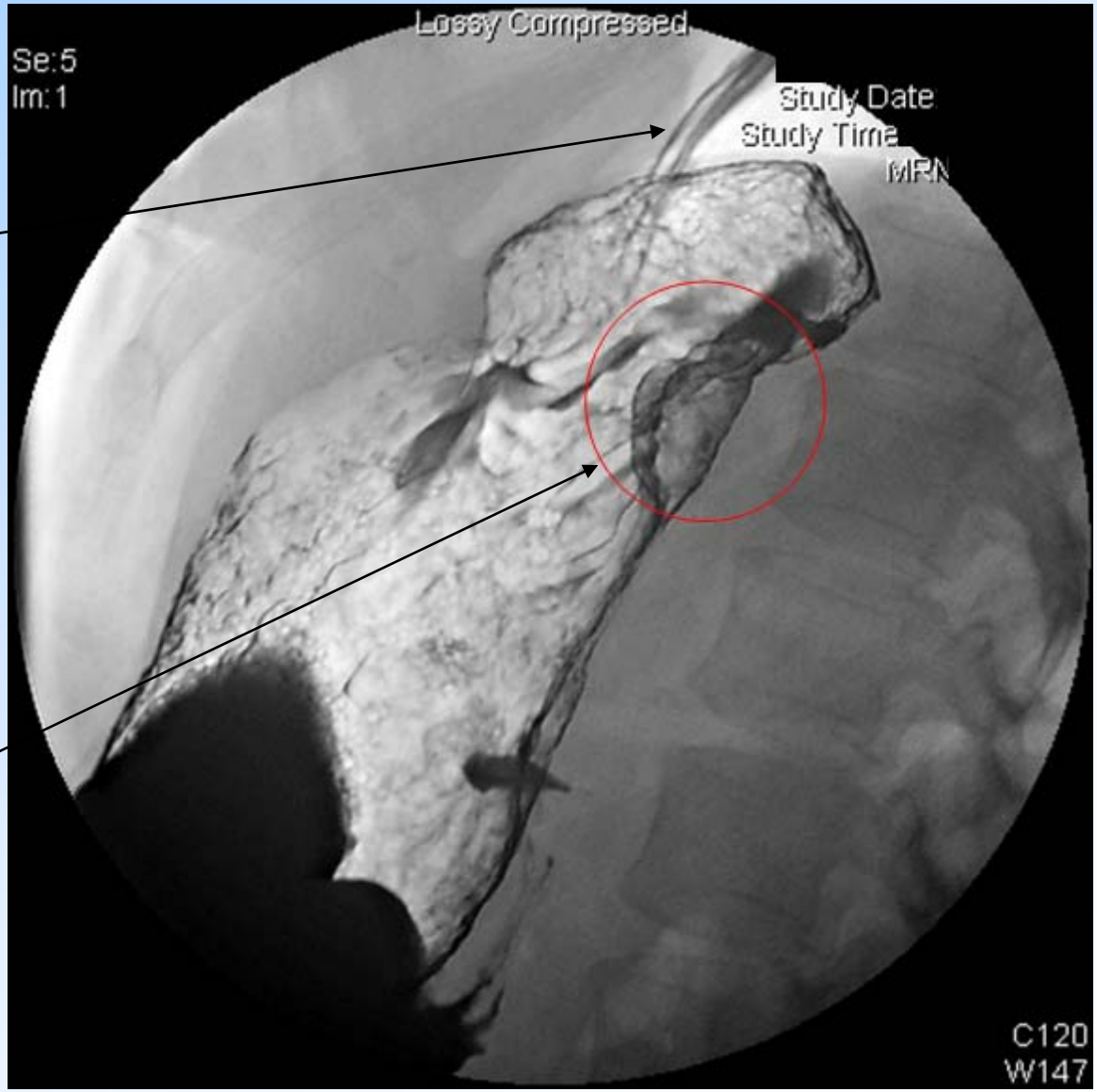
- Our Patient #1 UGI Series

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

- 2.5 x 3.0 cm submucosal mass in the fundus of the stomach.



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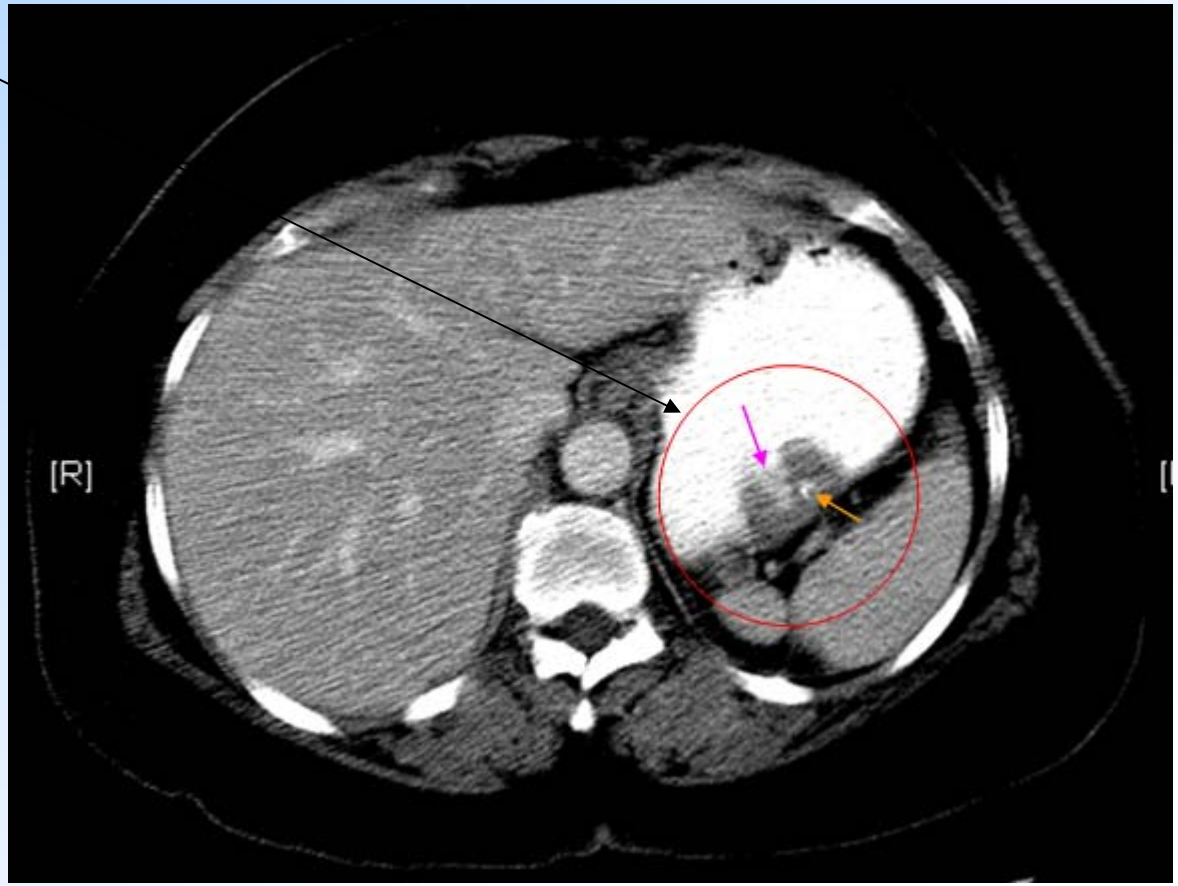
- Our Patient #1
- CT Abdomen +IV +Oral Contrast

- 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



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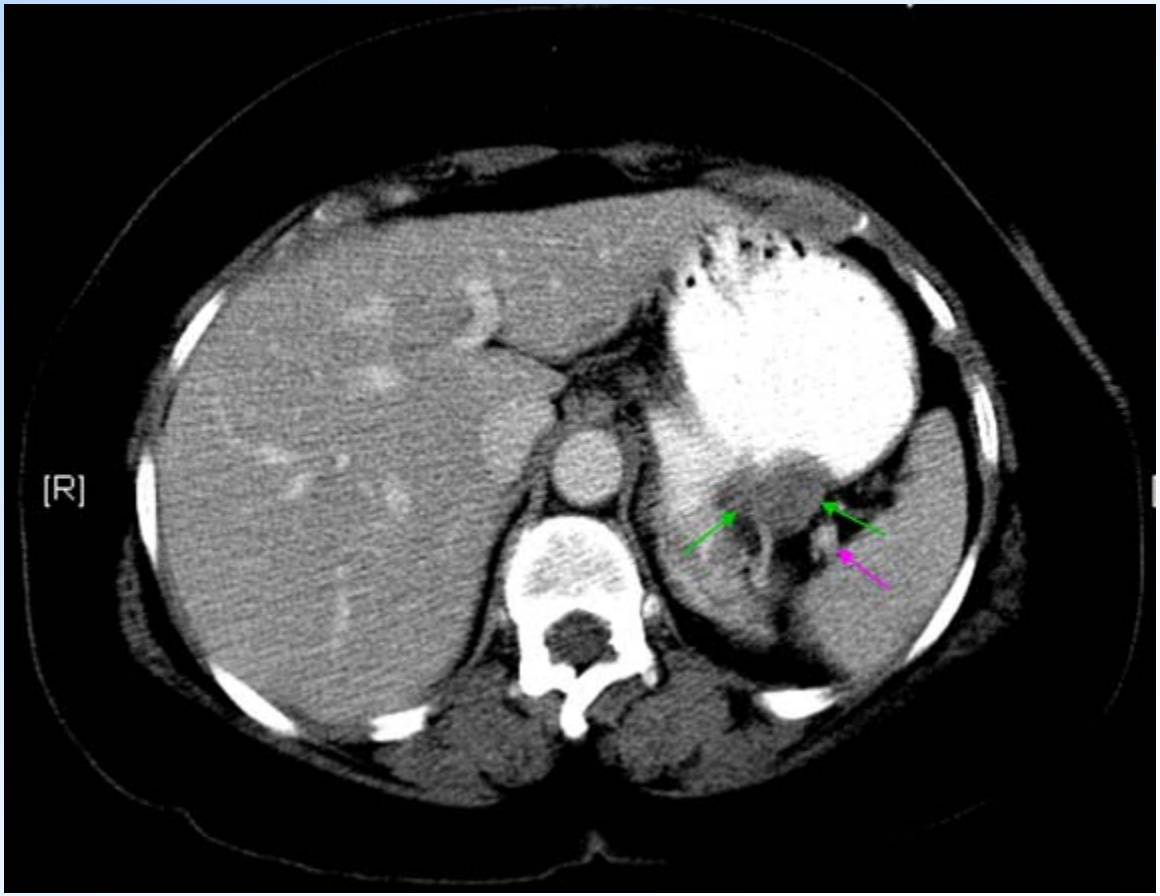
- Our Patient #1
- CT Abdomen +IV +Oral Contrast (cont.)

- 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.

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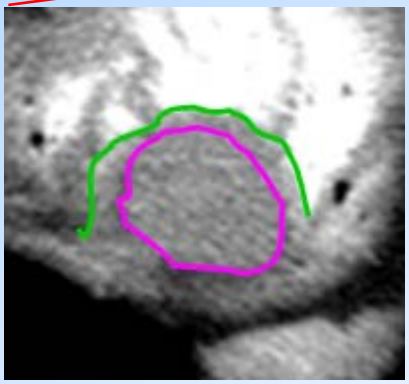
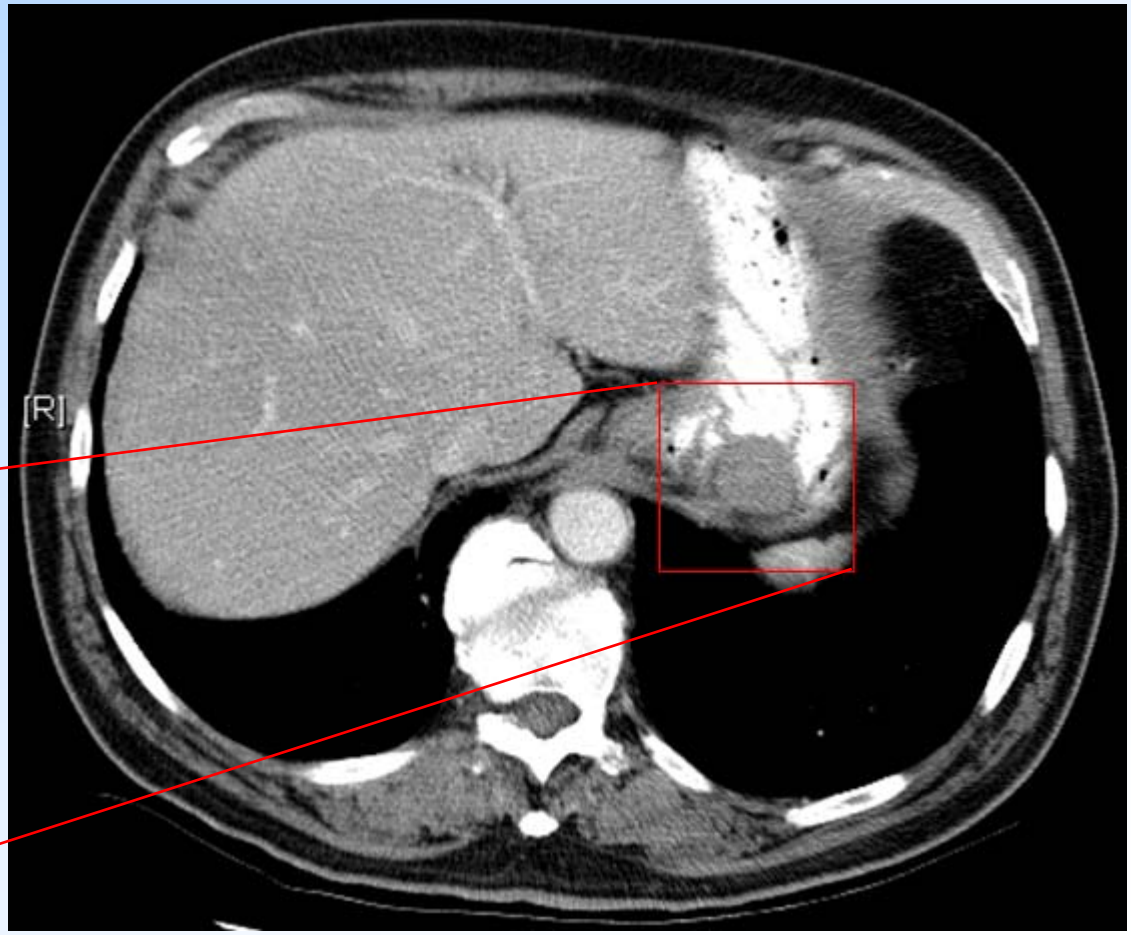
- Our Patient #2
- CT Abdomen +IV +Oral Contrast (cont.)

• 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.

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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.

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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%)

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Indicators for GIST Malignity

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

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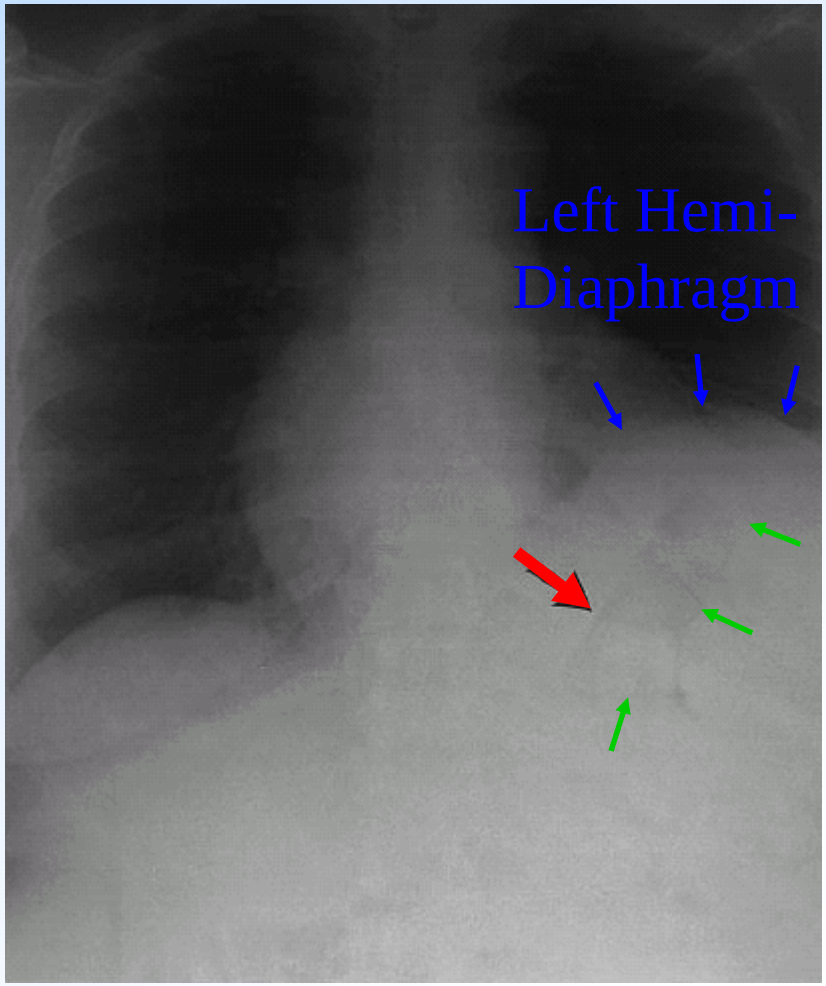
Radiographic Findings of GIST

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



Levy et al. Radiographics 2003; 23:283–304

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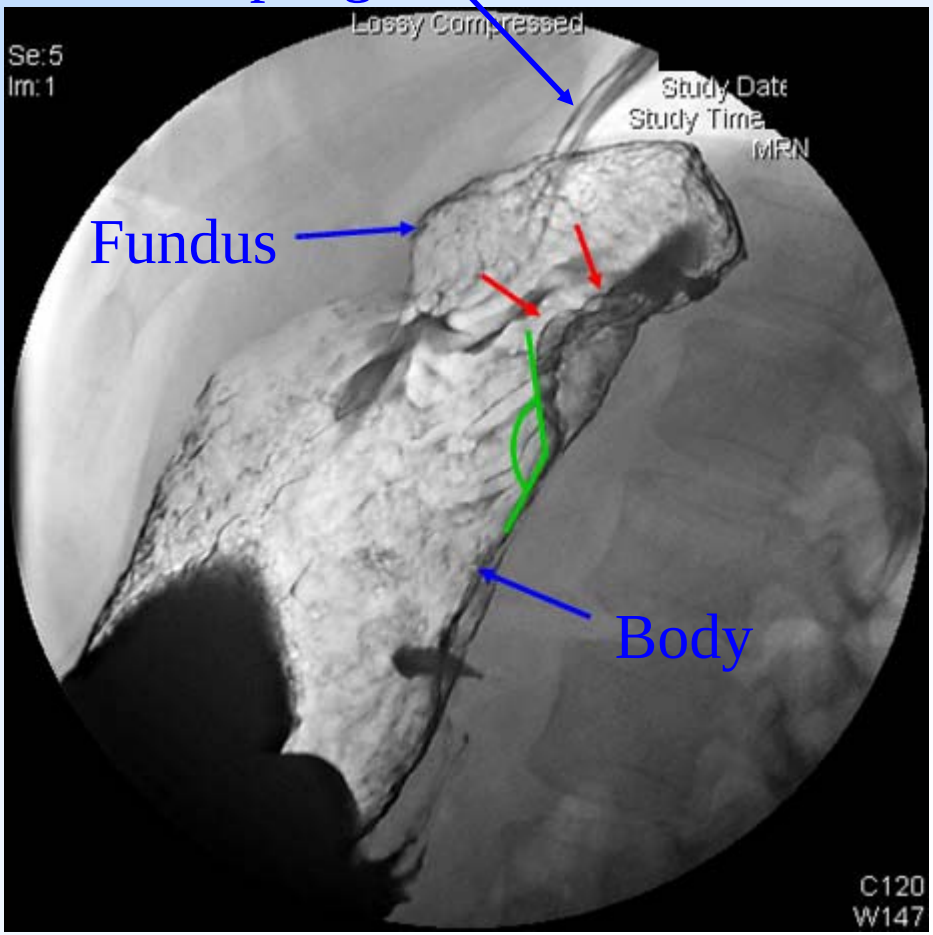
Radiographic Findings (cont.)

- 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.



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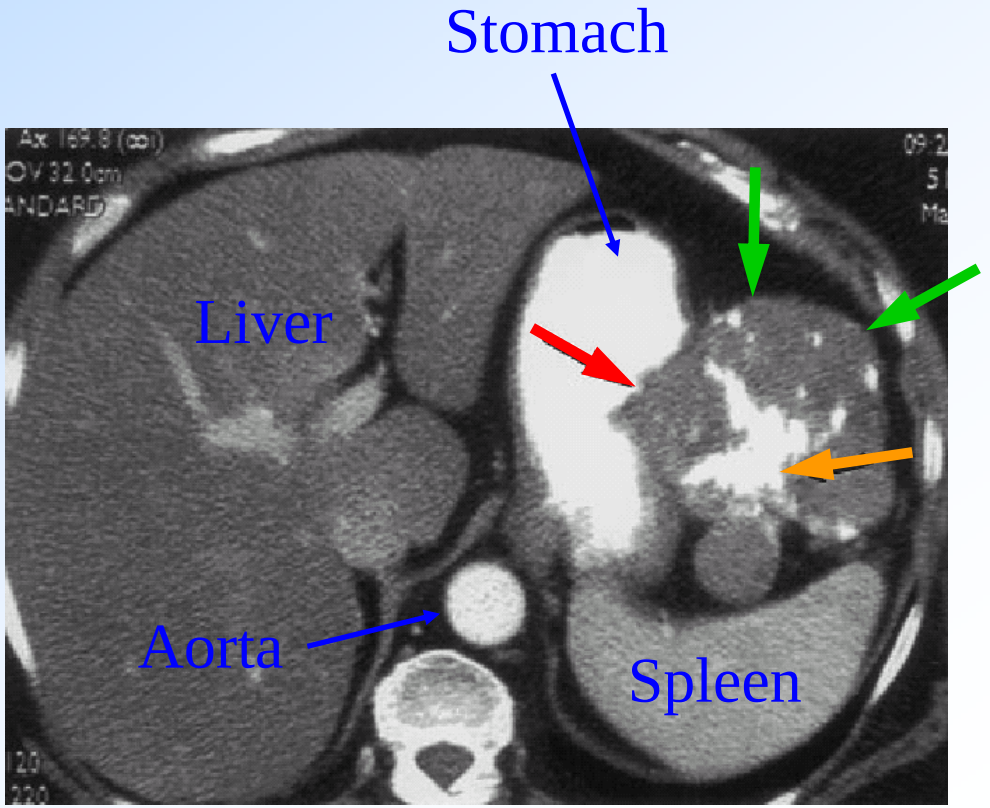
Radiographic Findings (cont.)

- CT Scan may show…

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



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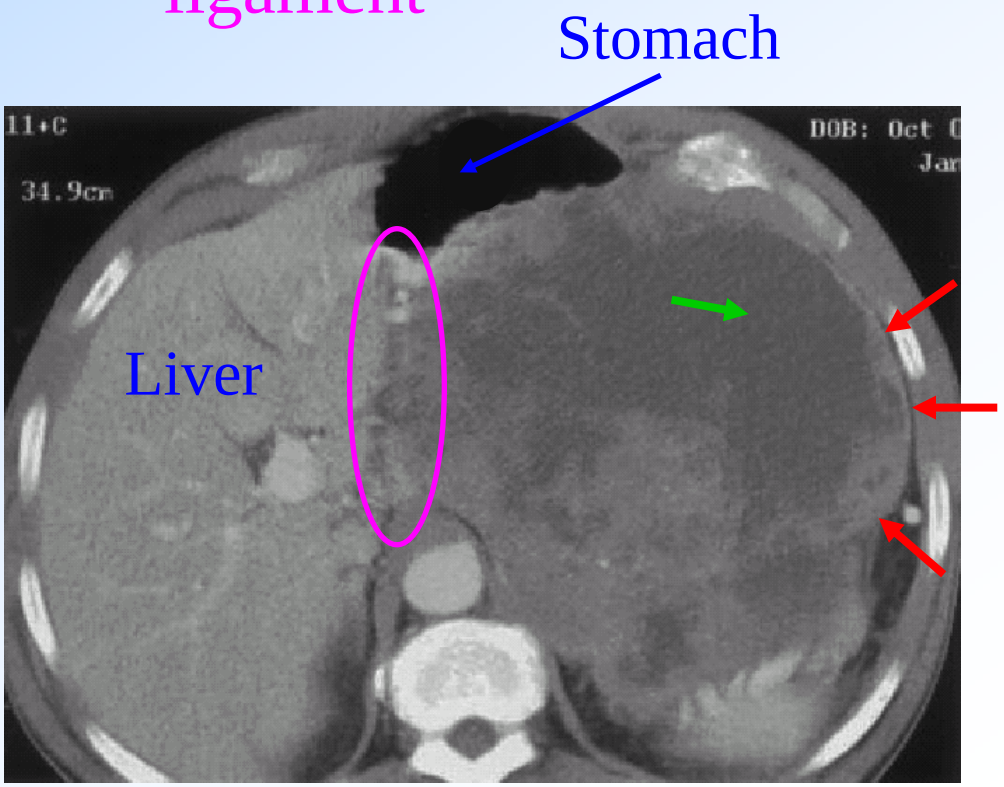
Radiographic Findings (cont.)

- 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



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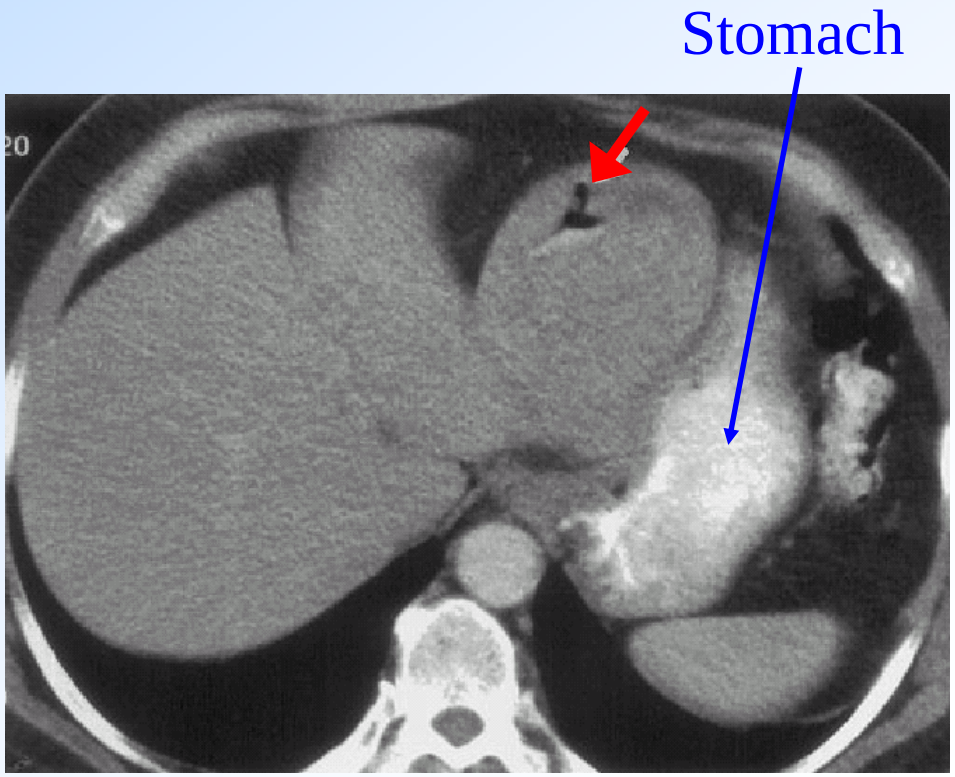
Radiographic Findings (cont.)

- CT Scan may show…
- There may be cysts filled with air…



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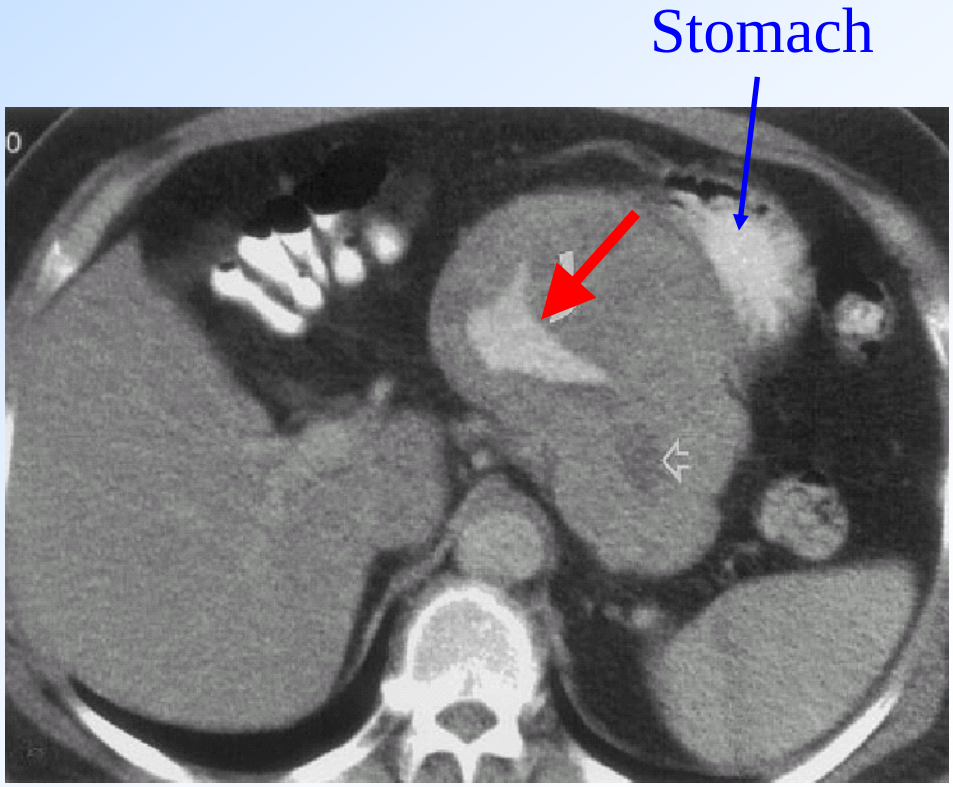
Radiographic Findings (cont.)

- CT Scan may show…
- … or cysts filled with oral contrast.



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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

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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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