# Contemporary Management of Gastro-Intestinal Stromal Tumors

Marcus Tan, MBBS, FACS Assistant Professor, Division of Surgical Oncology & Endocrine Surgery Vanderbilt University Medical Center

American Foregut Society, September 25

## No Disclosures

# Overview

Work-up of the subepithelial gastric mass

**Essential features of GIST** 

Prognosis and risk-stratification

**Decision-making** 

**Operative approaches** 

Post-operative considerations

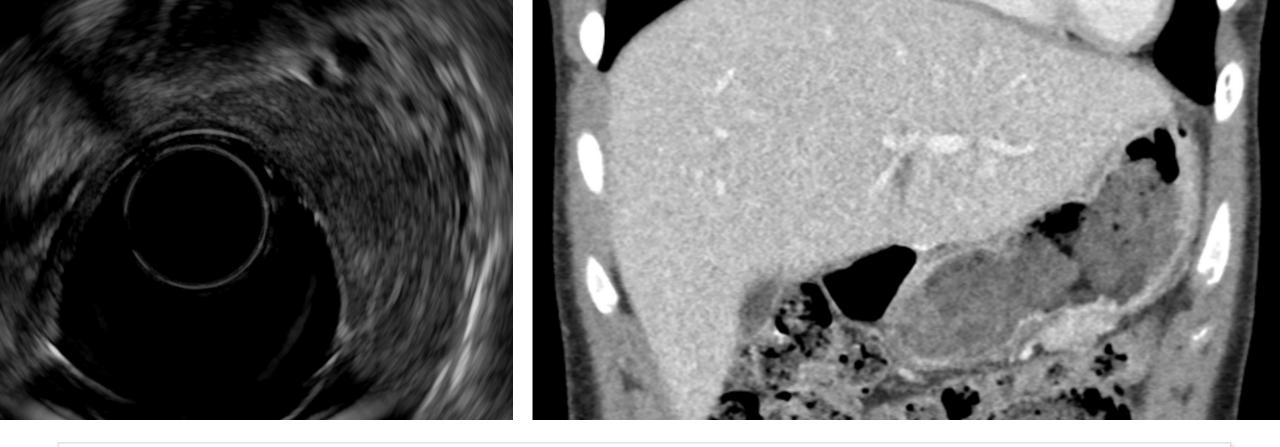
#### Subepithelial Gastric Mass

#### Differential diagnoses

- Gastro-Intestinal Stromal Tumor (GIST)
- Leiomyoma / leiomyosarcoma
- Schwannoma
- Desmoid



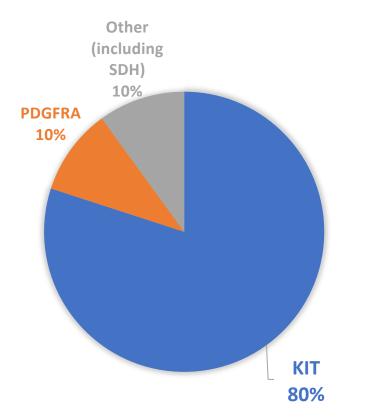




#### EUS FNA: spindle cells

- GIST: CD117, DOG1
- Leiomyomas: SMA
- Schwannomas: S100

#### DISTRIBUTION OF GENE MUTATIONS



# GIST: Essential Features

- most common mesenchymal (nonepithelial) neoplasm of the gut.
- originate from the interstitial cells of Cajal
- rare (7-8 cases / million / year), 1-2% of all GI cancers

## Prognosis & Risk Stratification

- Tumor Size (>5cm, >10cm)
- Mitotic Index (>5, >10 mitoses per 50 HPFs)
- Location (Stomach = most favorable)

## **Decision-Making**

#### Standard: Complete gross resection

#### Exceptions

- Gastric GISTs <2cm, without high-risk features</li>
  >observe
- Where operation would be highly morbid or require multi-visceral resection
  - Proximal stomach (? Esophagectomy)
    - Must determine distance to GE junction
  - Proximal duodenum (? Whipple)
    - Must determine distance to ampulla
  - Neoadjuvant imatinib

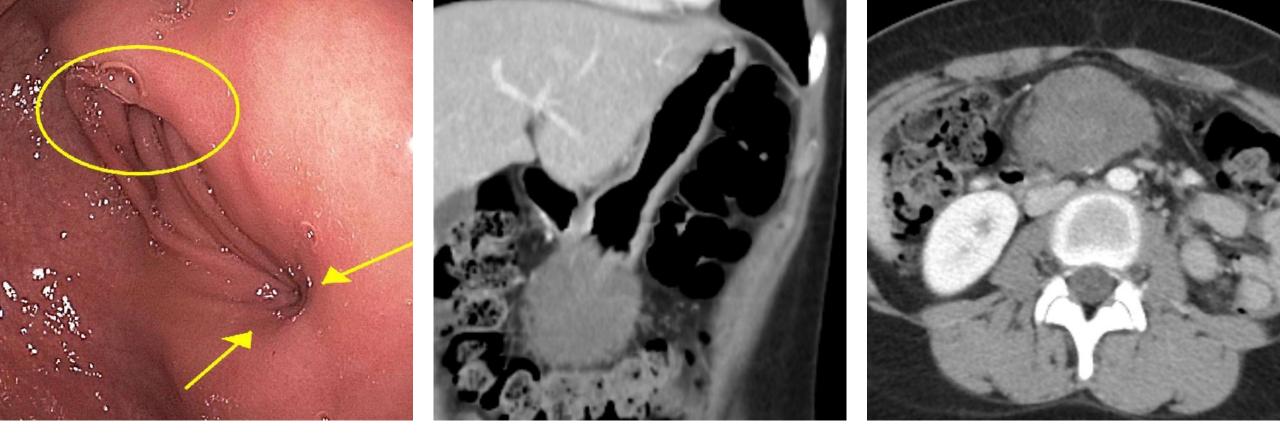
## Operative Approaches (1)

- Distal greater curve resection
  - Laparoscopic approach
  - 34 Fr gastric lavage tube
  - Stapled resection
  - Frozen section
- Surgical Pathology
  - 5cm GIST, 0 mitoses
  - PDGFRA mutation
- Post-op plan:
  - Annual surveillance
  - No adjuvant imatinib

#### Operative Approaches (2)

- Proximal greater curve resection
  - Laparoscopic approach
  - 50 Fr Maloney dilator
  - Stapled resection
  - Frozen section
- Surgical Pathology
  - Multifocal GIST: 9.4cm with 3 x 0.5cm tumors
  - 3 mitoses / 50 HPF
  - KIT mutation
- Post-op:
  - Discussed at weekly sarcoma conference
  - Adjuvant imatinib, 3 yrs

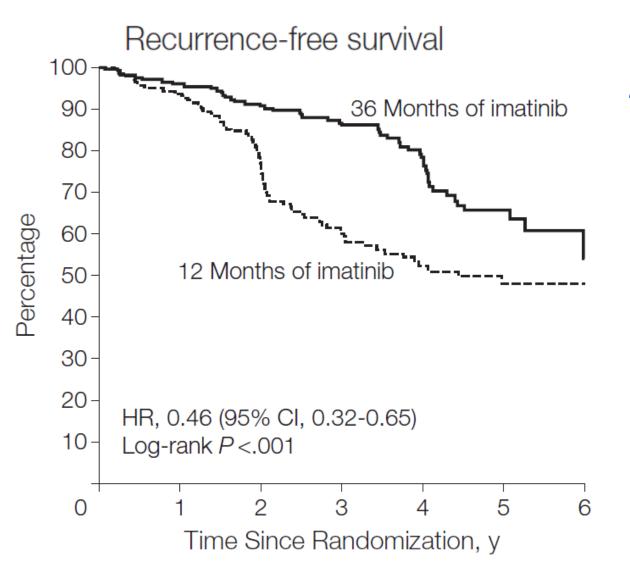




# EUS FNA: Spindle cells

- Laparoscopy: densely adherent, converted to open: mass adherent to posterior wall of stomach, transverse colon, SMA -> deemed unresectable; additional core biopsies obtained.
- Final surgical pathology: desmoid-type fibromatosis

## Post-operative Considerations (1)



# Adjuvant Imatinib for

high-risk disease:

- Tumor size >10cm
- Mitotic count >10 mitoses / 50 HPF
- Tumor size >5cm AND mitoses >5
- Tumor rupture
- 5 year RFS 66% vs 48%
  - (HR 0.46, 95% CI 0.32-0.65)

## Post-operative Considerations (2)

#### **Surveillance** for resected GIST:

• Clinical review and CT abdomen / pelvis q6 months for 3 years, then annually

#### **Resection of Metastases**

- If liver-only metastases, then resection combined with imatinib = best opportunity for long-term disease control
- Pre-operative imatinib ~ 6 months

# Summary: Contemporary Management of GIST

- EUS biopsy usually necessary
- Stage with contrasted CT
- Tumor size, mitotic rate and organ location are prognostic
- Complete resection = mainstay of therapy
- High-risk disease benefits from 3 years of adjuvant imatinib.

### Questions?

#### Modified NIH risk stratification criteria for GIST with rupture included

Risk category	Tumor size (cm)	Mitotic index (per 50 HPFs)	Primary tumor site
Very low risk	<2.0	≤5	Any
Low risk	2.1 to 5.0	≤5	Any
Intermediate risk	2.1 to 5.0	>5	Gastric
	<5.0	6 to 10	Any
	5.1 to 10.0	≤5	Gastric
High risk	Any	Any	Tumor rupture
	>10 cm	Any	Any
	Any	>10	Any
	>5.0	>5	Any
	2.1 to 5.0	>5	Nongastric
	5.1 to 10.0	≤5	Nongastric

NIH: National Institutes of Health; GIST: gastrointestinal stromal tumor; HPF: high power fields.

Reproduced from: Joensuu H. Risk stratification of patients diagnosed with gastrointestinal stromal tumor. Hum Pathol 2008; 39:1411. Table used with the permission of Elsevier Inc. All rights reserved.

