What the GIST?

Gastrointestinal Stromal Tumors
and
Reportability Standards

Cheryl Moody, BA, CTR
California Cancer Registry
Public Health Institute

Overview

- Background
- Recoding Audits Results
- Clinician Perspective
- Standard Setters Perspective
- Options/Conclusion

Definition

- **Gastrointestinal Stromal tumor:**
  - **One of the most common mesenchymal tumors of the gastrointestinal tract**
  - **Per AJCC 7th Edition:**
    - **The designation of GIST refers to a specific tumor type that is generally immunohistochemically KIT-positive and is driven by KIT or PDGFRA (platelet–derived growth factor receptor alpha) activating mutations.**

BACKGROUND

- **GIST, NOS (not stated to be malignant or benign)**
  - **ICD-0-3 Behavior = /1**
- **Reportability criteria**
- **SEER POC study 2010 – identified issue**

Recoding Audit Results

- GIST tumors:
  - 2,908 GIST cases in the central database
  - 320 GIST cases recoded
  - 40 cases from each regional registry
    - 8 regions
- Recoded behavior based on text documentation
- Discrepancy identified if original and auditor recode did not match
- Peer Review method (2 auditors)

Results

- 320 total cases recoded
- 211 (or 65.9%) GIST Malignant
  - Reportable per SEER's coding standard
- 109 (or 34.1%) GIST Borderline
  - Non-reportable per SEER's coding standard

Analysis

- **Auditor issues:**
  - **Excellent text documentation describes:**
    - **Tumor size and extent**
    - **Invasion**
    - **Mitotic Rate**
    - **Chemotherapy administered**
    - **Terminology such as “high malignant potential” or “malignant risk”**

Audit Documentation Examples

- **Invasion of adjacent tissues and/or organs (through muscularis propria & abuts serosa; invades pericolic fat tissue; extends into pancreas; involves visceral peritoneum)**
- **Omental caking**
- **Malignant Risk per path report based on 17.5 cm tumor size**
- **High malignant potential; omentum inv by tumor**
- **High mitotic activity > 10/50 HPF; high risk for aggressive behavior moderate malignant potential**
- **CD 117 positive; KIT exon 11 positive**

Clinician's Perspective

- Three Pathologists contacted
  - All three concur:
  - For GIST tumors,
    - Behavior is not categorized as benign or malignant
    - National consensus to categorize GIST tumors using a standard table
      - Tumor Size
      - Mitotic Count

Clinical Perspective

- **Dr. Lawrence Weiss, Chief Pathologist, City of Hope National Medical Center, Duarte, CA**

  "We do not speak of GISTs as benign or malignant anymore (if we don't have to). The most recognized (of the various classification systems) is the NIH system. Very low risk equates to benign, low risk equates to uncertain malignant potential, intermediate risk equates to low-grade malignant, and high risk equates to high-grade malignant. As you can see, it all depends on the size and the mitotic count as to which category a particular tumor falls in."

Clinical Perspective

Dan Phan, MD, Pathologist, Diagnostic Pathology Medical Group (DMPG), Sacramento, CA

"Criteria for classifying GIST are based on size and mitosis. We use a national risk classification system (NCI). We use this rather than the terms "benign vs. malignant".

Alexander Borowsky, MD, Pathologist, University of California Davis Medical Center, Sacramento, CA

"Is there such a thing as "benign" GIST? I think I would not use the term. To me they are all malignant, but can be divided into high and low grades (both considered "malignant")".

Risk Stratification Criteria (per Dr. Phan)

Risk Stratification of Primary GIST by Mitotic Index, Size and Site

Tumor Parameters	Risk for Progressive Disease* (%), Based on Site of Origin				
Mitotic Rate	Size	Stomach	Jejunum/Ileum	Duodenum	Rectum
< 5 per 50 HPF	< 2 cm	None (0%)	None (0%)	None (0%)	None (0%)
	>2, < 5 cm	Very low (1.9%)	Low (4.3%)	Low (8.3%)	Low (8.5%)
	>5, < 10 cm	Low (3.6%)	Moderate (24%)	Insufficient data	Insufficient Data
	> 10 cm	Moderate (10%)	High (52%)	High (34%)	High (57%)
> 5 per 50 HPF	< 2 cm	None	High+	Insufficient Data	High (54%)
	>2, < 5 cm	Moderate (16%)	High (73%)	High (50%)	High (52%)
	>5, < 10 cm	High (55%)	High (85%)	Insufficient Data	Insufficient Data
	> 10 cm	High (86%)	High (90%)	High (86%)	High (71%)

Data are based on long-term follow up of 1055 gastric, 629 small intestinal, 144 duodenal, and 111 rectal GISTs

*Defined as metastasis or tumor-related death

+Denotes small number of cases

Abbreviations: GIST: Gastrointestinal stromal tumors; HPF: high-power field

Adapted from: Miettinen M, Lasota J. Gastrointestinal tumors:pathology and prognosis at different sites, (Sem Dian Pathol 2006;23;70-83)

Risk Criteria (per Dr. Weiss)

Risk Criteria	Size	Mitotic count Per 50 high power fields
Very Low risk	<2	<5
Low risk	≥2 <5	<5
Intermediate risk	<5	5-10
High risk	≥5 >10 Any size	≥5 Any mitotic count >10

Support for Clinician's Perspective

- **AJCC 7th – GIST Schema**
- **ICD-0-3 – Histology codes specific to GIST tumors**
- **Collaborative Stage GIST Schemas**
  - **Appendix, Colon, Esophagus, Peritoneum, Rectum, Small Intestine and Stomach**

Support for Clinical Perspective

- **Collaborative Stage - Site Specific Factors**
  - **KIT**
  - **PDGFRA**
  - **Tumor Multiplicity**
  - **Mitotic Count**
  - **Location of Primary Tumor**

Standard Setter's Perspective

- **PER SEER SINQ 20091021**

"According to the current reportability criteria, malignant GIST (8936/3) is reportable to SEER.

GIST coded to 8936/0 or 8936/1 is not reportable.

If your pathologist will not indicate "malignant" or "benign," code 8936/1 applies according to ICD-O-3 and, therefore, these are not reportable to SEER."

(See also SEER SINQ: 20021151 and 2010014)

Standard Setter's Perspective

- **Per CoC Inquiry & Response 48098 (8/6/2010):**

  "Gastrointestinal Stromal Tumors (GIST), NOS have the histology code 8936/1 and are not reportable.

  If the pathologist confirms that this is malignant (8936/3) the case is reportable."
  (*I & R Team*)

Support for Standard Setter's Perspective

- Consistency and accuracy are key to quality data
- Past experience with unclear coding instructions
  - CIN III, dysplasia vs in situ
  - Muddy data
- Obligation to Researchers to provide consistent and accurate data

Disconnect

There appears to be a disconnect

between standard setters reportability perspective

And

Clinicians perspective in regards to GIST tumors

Dilemma

What to do with GIST, NOS cases?

Options

- **Include GIST, NOS in the central registry database – because based on clinical practice there may be no definitive statement of benign or malignant behavior with regard to GIST tumors**
  - **Classify as reportable by agreement**
  - **Assign benign sequence number**
  - **Assign borderline behavior**
  - **Use appropriate CS schema and codes**

or

- **Exclude GIST, NOS from the central registry database – because current coding standards state that without a definitive statement of benign or malignant GIST, the tumor is considered "borderline" and therefore, non-reportable**

Recommendation/Conclusions

- **Including GIST, NOS in the Central Registry database allows for the option of later reclassifying as clinical standards evolve**
- **Valuable data is retained**
- **A more accurate accounting of the incidence of GIST will be available to researchers**

Bottom line

GIST - Keep ‘em!

Questions?

Contact Information

Cheryl Moody, BA, CTR
Quality Control Specialist
California Cancer Registry
1825 Bell Street, Ste 102
Sacramento, CA 95825
(916) 779-0311
cmoody@ccr.ca.gov