NAACCR 2009-2010 Webinar Series

Collecting Cancer Data:
Soft Tissue Sarcoma, Neuroendocrine Tumors (NET) and Gastrointestinal Stromal Tumors (GIST)

Agenda
• Updates
  • Soft Tissue Sarcoma
    – Overview
    – CSv2
    – MP/H Rules
  • Gastrointestinal Stromal Tumor (GIST)
    – Overview
    – CSv2
  • Neuroendocrine Tumors (NET)
    – Overview
    – CSv2
• Questions from previous webinars

2010 Update
Access to 2010 Information

• “Everything 2010”
  – NAACCR one stop shop for everything 2010
  • EDITS metafile and EDITS software
  • Links to standard setters requirements and manuals
  • CSV2 software products
  • CSV2 coding instructions
  • Hematopoietic and lymphoid neoplasm project
  • Training links

CSV2

• Pre and post treatment CS data fields
  – Implementation delayed until 2012
  – Will be collected only for selected sites
• Next CSV2 version will be released by the end of April

Soft Tissue Sarcoma

Overview
Soft Tissue Sarcoma

- Estimated new cases and deaths from soft tissue sarcoma in the United States in 2009:
  - New cases: 10,660
  - Deaths: 3,820
- Risk factors
  - Radiation
  - Workplace chemicals
  - Genetics

Soft Tissue Sarcoma

- Peripheral Nerves and Autonomic Nervous System
  - C47.0-C47.6, C47.8-C47.9
- Connective, Subcutaneous and Other Soft Tissues
  - C49.0-C49.6, C49.8-C49.9
- Note 1:
  - Laterality must be coded for C47.1-C47.2 and C49.1-C49.2
- Note 2:
  - Soft tissue sarcomas of the heart and mediastinum (C38.0-C38.3 and C38.9) use the Heart, Mediastinum schema

Terminology

- ost(e), oste(o): bone
- chondr(i)o: cartilage, gristle, granule, granular
- lei(o): smooth
- my(o): Of or relating to muscle
- lip(o): fat
- hema or hemo: blood
- angi(o): blood vessel
- fibr(o): fiber
- neur(i), neur(o): Of or pertaining to nerves and the nervous system
Question

- What is the morphology code of pleomorphic leiomyosarcoma arising from the right thigh?

Answer

- Using Rule F in ICD-O-3 (pg. 20)
  - Change 8893/0 (pleomorphic leiomyoma) to 8893/3 for pleomorphic leiomyosarcoma (the malignant form of a leiomyoma).
  
  I & R Team
  46940

ICD-O-3 Coding Guidelines

- Principle Rules
  - Rule 1. Compound morphology diagnoses: Change the order of word roots in a compound term if the term is not listed in ICD-O.
  - Example:
    - Myxofibrosarcoma
      - Assign histology code 8811/3 (fibromyxosarcoma)
Multiple Primary Rules

Other

Multiple Primary Rules-Other

Multiple Tumors

• M8
  — Tumors on both sides of a paired organ are single primary.
• M10
  — Tumors diagnosed more than 1 year apart are multiple primaries.
• M11
  — Tumors with ICD-O-3 topography codes that are different at the second (Cxxx) and/or third characters (Cxx) are multiple primaries.

• M12
  — Tumors with ICD-O-3 topography codes that differ only at the fourth character (Cxx) and are in any one of the following primary sites are multiple primaries.
  • Anus and anal canal (C21_)
  • Bones, joints, and articular cartilage (C40_ - C41_)
  • Peripheral nerves and autonomic nervous system (C47_)
  • Connective subcutaneous and other soft tissues (C49_)
  • Skin (C44_)
Multiple Primary Rules-Other

Multiple Tumors

- M16
  - Abstract as a single primary when one tumor is Sarcoma, NOS (8800) and another is a specific sarcoma.
- M17
  - Tumors with ICD-O-3 histology codes that are different at the first (xxxx), second (xxxx) or third (xxxx) number are multiple primaries.
- Rule M18
  - Tumors that do not meet any of the above criteria are a single primary.

Single Tumor-Histology

- Rule H11
  - Code the histology when only one histologic type is identified.
- Rule H13
  - Code the most specific histologic term.
    - Sarcoma, NOS (8800) and a more specific sarcoma

Single Tumor-Histology

- Rule H16
  - Code the appropriate combination/mixed code (Table 2) when there are multiple specific histologies or when there is a non-specific histology with multiple specific histologies
    - Any combination of histologies below should be coded to Mixed liposarcoma (8855)
      - Myxoid
      - Round cell
      - Pleomorphic
Collecting Cancer Data: Soft Tissue Sarcoma and GIST

Single Tumor-Histology

- Rule H17
  - Code the histology with the numerically higher ICD-O-3 code.

Treatment

- Low grade early stage (I)
  - Surgery alone
- High grade later stage (II-III)
  - Should consider preoperative chemo or chemoradiation
- Stage IV
  - Chemotherapy/radiation
  - Clinical trial

CSv2

Soft Tissue Sarcoma
CS Tumor Size: Soft Tissue

- Code 995
  - Less than 5 cm, or greater than 4 cm, or “between 4 cm and 5 cm”
  - Stated as T1a or T1b
- Code 996
  - Greater than 5 cm
  - Stated as T2a or T2b

CS Extension: Soft Tissue

- Note 1
  - Defines connective tissue and peripheral nerves and the autonomic nervous system for the soft tissue schema
- Note 2
  - Superficial lesion
    - Located entirely in subcutaneous tissues
- Note 3
  - Deep lesion
    - Located partly or completely within 1 or more muscle groups within the extremity

CS Extension: Soft Tissue

- Note 4
  - Vessels with a name are considered a structure (code 600)
- Note 5
  - Tumors of extremities and trunk ONLY
    - Superficial lesion
      - Does not involve superficial muscular fascia
    - Deep lesion
      - Involves or is beneath superficial fascia
Collecting Cancer Data: Soft Tissue Sarcoma and GIST

CS Extension: SoftTissue

• Note 6
  – All intraperitoneal visceral lesions, retroperitoneal lesions, intrathoracic lesions, and majority of head and neck tumors are deep
  – Assign CS Extension codes 120, 312, 315, 320, 420, 620, 800, 950, or 990 for these sites (C47.0, C47.3-5, C49.0, C49.3-5)

CS Extension: SoftTissue

• Note 7
  – Adjacent connective tissue (code 400)
    • Unnamed tissue immediately surrounding an organ or structure containing the primary cancer

CS Extension: SoftTissue

• Tumor confined to site/tissue of origin
  – Code 100: Invasive tumor, NOS
  – Code 110: Superficial invasive tumor
  – Code 120: Deep invasive tumor
**CS Extension: SoftTissue**

<table>
<thead>
<tr>
<th>Localized Tumor</th>
<th>Localized Tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Code 300: NOS</td>
<td>- Code 315: Stated as T2b</td>
</tr>
<tr>
<td>- Code 302: Stated as T1a</td>
<td>- Code 320: Deep</td>
</tr>
<tr>
<td>- Code 305: Stated as T2a</td>
<td>- Code 322: Stated as T1 [NOS]</td>
</tr>
<tr>
<td>- Code 310: Superficial</td>
<td>- Code 325: Stated as T2 [NOS]</td>
</tr>
<tr>
<td>- Code 312: Stated as T1b</td>
<td></td>
</tr>
</tbody>
</table>

**CS Extension: SoftTissue**

- Adjacent connective tissue
  - Code 400: NOS
  - Code 410: Superficial tumor
  - Code 420: Deep tumor

**CS Extension: SoftTissue**

- Adjacent organs/structures
  - Code 600: NOS
  - Code 610: Superficial tumor
  - Code 620: Deep tumor
CS Lymph Nodes: SoftTissue

- **Note 1**
  - Regional nodes are in vicinity of primary tumor
- **Note 2**
  - Regional node involvement is rare
  - Assume nodes are negative (code 000) when there is no mention of lymph node involvement clinically
- **Note 3**
  - Regional lymph nodes include bilateral or contralateral nodes for head, neck, and trunk primaries ONLY

CS Lymph Nodes: SoftTissue

- **Code 100**
  - Regional lymph nodes by primary site
- **Code 120**
  - Submental nodes for neck primary only
- **Code 150**
  - Neck primary only: code 100 + code 120

CS Mets at DX: SoftTissue

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>00</td>
<td>No; none</td>
</tr>
<tr>
<td>10</td>
<td>Distant lymph node(s)</td>
</tr>
<tr>
<td>40</td>
<td>Distant metastases except distant lymph nodes (10)</td>
</tr>
<tr>
<td>50</td>
<td>(10) + (40)</td>
</tr>
<tr>
<td>60</td>
<td>Distant metastasis, NOS</td>
</tr>
<tr>
<td></td>
<td>Stated as M1, NOS</td>
</tr>
<tr>
<td>99</td>
<td>Unknown</td>
</tr>
</tbody>
</table>
SSF1: SoftTissue Grade for Sarcomas

- Three grade sarcoma grading systems
  - National Cancer Institute (NCI) system
  - French Federation of Cancer Centers Sarcoma Group (FNCLCC)
    - AJCC 7th Edition preferred sarcoma grading system

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Mapping of Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>010</td>
<td>Grade 1 of 3</td>
<td>1</td>
</tr>
<tr>
<td>020</td>
<td>Grade 2 of 3</td>
<td>2</td>
</tr>
<tr>
<td>030</td>
<td>Grade 3 of 3</td>
<td>3</td>
</tr>
<tr>
<td>100</td>
<td>Low grade [NOS]</td>
<td>1</td>
</tr>
<tr>
<td>200</td>
<td>High grade [NOS]</td>
<td>3</td>
</tr>
<tr>
<td>888</td>
<td>Obsolete data converted V0200</td>
<td>9</td>
</tr>
<tr>
<td>988</td>
<td>Not applicable</td>
<td>9</td>
</tr>
<tr>
<td>998</td>
<td>No histologic examination of primary site</td>
<td>9</td>
</tr>
<tr>
<td>999</td>
<td>Unknown</td>
<td>9</td>
</tr>
</tbody>
</table>

SSF2: SoftTissue Neurovascular Invasion

<table>
<thead>
<tr>
<th>Code</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>000</td>
<td>Neurovascular invasion not present/not identified</td>
</tr>
<tr>
<td>010</td>
<td>Neurovascular invasion present/identified</td>
</tr>
<tr>
<td>888</td>
<td>Obsolete data converted V0200</td>
</tr>
<tr>
<td>988</td>
<td>Not applicable</td>
</tr>
<tr>
<td>998</td>
<td>No histologic examination of primary site</td>
</tr>
<tr>
<td>999</td>
<td>Unknown</td>
</tr>
</tbody>
</table>
### SSF3: SoftTissue

**Bone Invasion**

<table>
<thead>
<tr>
<th>Code</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>000</td>
<td>Bone invasion not present/not identified</td>
</tr>
<tr>
<td>010</td>
<td>Bone invasion present/identified</td>
</tr>
<tr>
<td>888</td>
<td>Obsolete data converted V0200</td>
</tr>
<tr>
<td>988</td>
<td>Not applicable</td>
</tr>
<tr>
<td>998</td>
<td>No imaging done for bone invasion</td>
</tr>
<tr>
<td>999</td>
<td>Unknown</td>
</tr>
</tbody>
</table>

**Note 2:** Bone invasion based on imaging only

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### SSF4: SoftTissue

**Pathologic M1: Source of Pathologic Metastatic Specimen**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>000</td>
<td>No pathological mets at diagnosis identified</td>
</tr>
<tr>
<td>010</td>
<td>Liver mets present/identified</td>
</tr>
<tr>
<td>020</td>
<td>Lung mets present/identified</td>
</tr>
<tr>
<td>030</td>
<td>Brain mets present/identified</td>
</tr>
<tr>
<td>040</td>
<td>Bone mets present/identified</td>
</tr>
<tr>
<td>050</td>
<td>Other mets present/identified</td>
</tr>
<tr>
<td>060</td>
<td>Combination of code 100-500</td>
</tr>
<tr>
<td>888</td>
<td>Obsolete data converted V0200</td>
</tr>
<tr>
<td>988</td>
<td>Not applicable</td>
</tr>
<tr>
<td>998</td>
<td>No microscopic examination of metastatic</td>
</tr>
<tr>
<td>999</td>
<td>Unknown</td>
</tr>
</tbody>
</table>

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**Quiz**

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Gastrointestinal Stromal Tumors (GIST)

Overview

Gastrointestinal Stromal Tumors (GIST)

- What is the difference between GIST, NOS and a malignant GIST?
  - GIST, NOS  8936/1
  - Malignant GIST  8936/3

Question

- Path report states: Gastrointestinal stromal tumor, intermediate malignant potential, gastric fundic polyp. What is the behavior code?
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Answer

- As you can see in the ICD-O-3 book, GIST behavior can range from /0, /1 or /3. Question the pathologist as to which description best fits this diagnosis; if that is not possible, we must use GIST, NOS and code 8936/1.

  (I & R Team)

46868

GIST Histology

- Stromal sarcoma, NOS 8935/3
- Gastrointestinal stromal sarcoma 8936/3
  - Gastrointestinal stromal tumor, malignant
  - GIST Malignant

Question

- Gastrointestinal stromal tumor, epithelioid cell type with myxoid pattern...what is the morphology code?
Answer

- GIST, epithelioid and myxoid may be a specific subtype, but currently, there is no ICD-O-3 code to differentiate this subtype.
  - Follow Other Rule H11 and code the only type found (8936).
  - Review with the pathologist to determine if the tumor is malignant to determine the 5th digit.

Multiple Primary and Histology Rules

- Use the site where the tumor originated to determine MPH chapter to use.

GIST Multiple Primary Rules

- Stomach Other
- Small intestine Other
- Esophagus Other
- Large intestine Colon
- Rectum Other
- Other (very rare) Other
  - Peritoneum, mesentery, omentum, liver, pancreas, ovaries, uterus, prostate
Staging

AJCC 7th Edition TNM Stage

- Two staging forms
  - Gastric GIST
  - Small Intestine GIST
    - Esophagus
    - Colorectal
    - Peritoneum

Question

- Pathologists have used tumor size and mitotic activity to determine whether GISTS were benign or malignant.
- The 7th Edition AJCC Manual uses criteria for Stage 1 GIST which would otherwise be considered benign.
- Could you clarify if we are to go by staging criteria to determine if a GIST is reportable?
Answer

• ...Staging does NOT determine reportability or the histology behavior codes.
  (I & R Team)
  46803

AJCC 7th Edition TNM Stage

• Same T,N and M values for both forms
  – T1 Tumor 2cm or less
  – T2 Tumor more than 2cm but not more than 5cm
  – T3 Tumor more than 5cm but not more than 10cm
  – T4 Tumor more than 10cm in greatest dimension

Appendix, Colon, and Rectum GIST SSF’s

• SSF 1
  – OBSOLETE (Pre-Operative Carcinoembryonic Antigen (CEA))
• SSF 2
  – OBSOLETE (Clinical Assessment of Regional Lymph Nodes)
Appendix, Colon, and Rectum GIST SSF’s

*Required by CoC and SEER
- SSF 11*
  - Mitotic Count
- SSF 12*
  - KIT Gene Immunohistochemistry (IHC)

Appendix, Colon, and Rectum GIST SSF’s

Not required by CoC and SEER
- SSF 13
  - KIT Gene Mutations
- SSF 14
  - PDGFRα Gene Mutation
- SSF 15
  - Tumor Multiplicity

Esophagus, Stomach and Small Intestine GIST

Required by CoC and SEER
- SSF 6
  - Mitotic Count
- SSF 7
  - KIT Gene Immunohistochemistry (IHC)
Esophagus, Stomach and Small Intestine GIST
Not required by CoC and SEER
• SSF 8
  – KIT Gene Mutations
• SSF 9
  – PDGFRA Gene Mutation
• SSF 10
  – Tumor Multiplicity

Peritoneum GIST
Required by CoC and SEER
• SSF 5
  – Mitotic Count
• SSF 6
  – KIT Gene Immunohistochemistry (IHC)
• SSF 10
  – Location of Primary Tumor

Peritoneum GIST
Not required by CoC and SEER
• SSF 7
  – KIT Gene Mutations
• SSF 8
  – PDGFRA Gene Mutation
• SSF 9
  – Tumor Multiplicity
CSv2 GIST

Quiz

Neuroendocrine Tumor (NET)

Overview
Multiple Primary and Histology Rules

- Colon
- Other
- Lung

Colon MP/H

- Neuroendocrine carcinoma (8246): Neuroendocrine carcinoma is a group of carcinomas that include typical carcinoid tumor (8240), atypical carcinoid tumor (8249).

Colon MP/H

- Rule H8
  - Code 8240 (carcinoid tumor, NOS) when the diagnosis is neuroendocrine carcinoma (8246) and carcinoid tumor (8240).
- Rule H9
  - Code 8244 (composite carcinoid) when the diagnosis is adenocarcinoma and carcinoid tumor.
- Rule H10
  - Code 8245 (adenocarcinoid) when the diagnosis is exactly "adenocarcinoid."
Question

- How is histology coded for an appendix primary when the final pathology report states “mucinous adenocarcinoma arising in goblet cell carcinoid”?

Answer

- We follow the Colon Rules until Rule H9 adenocarcinoma and carcinoid, code 8244.

Curator (I & R Team)
27032

Other MP/H

- Stomach
- small intestine
- Pancreas
- Thyroid gland
- Adrenal gland
- Thymus
- Heart
- Other sites that develop carcinoids and small cell carcinomas
- Skin
**Lung MP/H**

Equivalent terms
- Low grade neuroendocrine carcinoma, carcinoid

**Definitions**
- Neuroendocrine carcinoma (8246): Neuroendocrine carcinoma is a group of carcinomas that include typical carcinoid tumor and small cell carcinoma. Code the specific histology when given. Code neuroendocrine carcinoma, NOS (8246) when no specific histology is documented.

**Question**
- What is the histology for large cell neuroendocrine carcinoma with areas of small cell carcinoma of right upper lobe lung and why?
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Answer

• Large cell neuroendocrine carcinoma (8013/3) and small cell carcinoma, NOS (8041/3) are on different branches of the histology tree.
• Large cell neuroendocrine is on the non-small cell branch and small cell is on the small cell branch.
• Go to rule H7 and code the higher numeric histology (small cell carcinoma 8041/3).

Curator (I & R Team), 24947

Required SSF's
**Ampulla NET**

Required by the CoC and SEER
- SSF 5
  - Serum Chromogranin A (CgA) Lab Value
- SSF 6
  - Urinary 5-HIAA Lab Value Level

Not Required by the CoC and SEER
- SSF 4
  - Mitotic Count

**Colon & Rectum NET**

Required by CoC and SEER
- SSF 2
  - Clinical Assessment of Regional Lymph Nodes
- SSF 16
  - Serum Chromogranin A (CgA) Lab Value
- SSF 17
  - Urinary 5-HIAA Lab Value Level

Not Required by CoC and SEER
- SSF 11
  - Mitotic Count

**Small Intestine & Stomach NET**

Required by CoC and SEER
- SSF 1 (Stomach only)
  - Clinical Assessment of Regional Lymph Nodes
- SSF 11
  - Serum Chromogranin A (CgA) Lab Value
- SSF 12
  - Urinary 5-HIAA Lab Value Level

Not Required by CoC and SEER
- SSF 6
  - Mitotic Count
Collecting Cancer Data: Soft Tissue Sarcoma and GIST

CSv2

Questions?

Quiz
Q&A

• Answers to questions from previous webinars

Thank You!

• Using Geographic Information System Mapping for Spatial Analysis
  – Presented by the NAACCR GIS committee
  – May 6, 2010