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)

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ICD-O-3 Coding Guidelines

- Principle Rules
  - Rule J. *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 (Cxxx) are multiple primaries.

Multiple Primary Rules-Other

Multiple Tumors

- M12
  - Tumors with ICD-O-3 topography codes that differ only at the fourth character (Cxxx) 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

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: SoftTissue

- 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: SoftTissue

- 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: SoftTissue

- 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

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

Localized Tumor
- Code 300: NOS
- Code 302: Stated as T1a
- Code 305: Stated as T2a
- Code 310: Superficial
- Code 312: Stated as T1b

Localized Tumor
- Code 315: Stated as T2b
- Code 320: Deep
- Code 322: Stated as T1 [NOS]
- Code 325: Stated as T2 [NOS]

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

Code	Description
00	No; none
10	Distant lymph node(s)
40	Distant metastases except distant lymph nodes (10)
50	(10) + (40)
60	Distant metastasis, NOS Stated as M1, NOS
99	Unknown

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

SSF1: SoftTissue Grade for Sarcomas

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

SSF2: SoftTissue Neurovascular Invasion

Code	Definition
000	Neurovascular invasion not present/not identified
010	Neurovascular invasion present/identified
888	Obsolete data converted V0200
988	Not applicable
998	No histologic examination of primary site
999	Unknown

SSF3: SoftTissue Bone Invasion

Code	Definition
000	Bone invasion not present/not identified
010	Bone invasion present/identified
888	Obsolete data converted V0200
988	Not applicable
998	No imaging done for bone invasion
999	Unknown

Note 2: Bone invasion based on imaging only

SSF4: SoftTissue Pathologic M1: Source of Pathologic Metastatic Specimen

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

Quiz

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?

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*)
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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.

Curator (I & R Team)

I&R 28619

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
  - PDGFRA 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

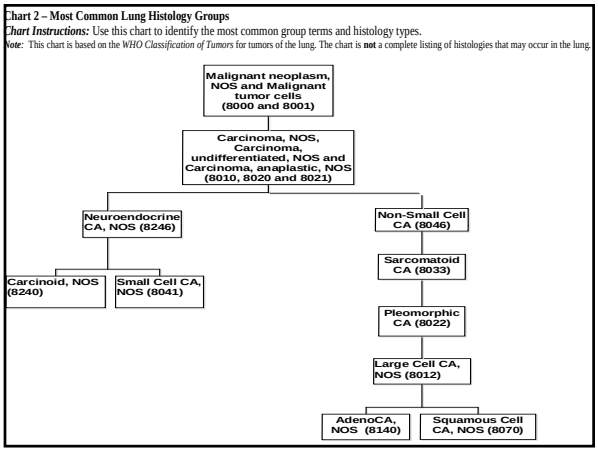
Lung MP/H

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.



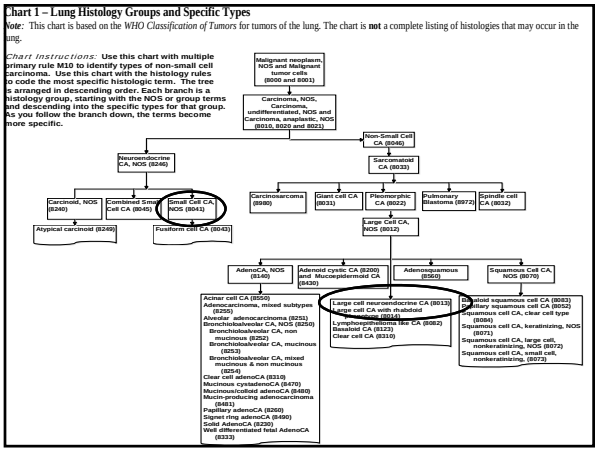
Question

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

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

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