Collecting Cancer Data: Bone & Soft Tissue

NAACCR 2015-2016 Webinar Series

Q&A

• Please submit all questions concerning webinar content through the Q&A panel.
• Reminder:
  • If you have participants watching this webinar at your site, please collect their names and emails.
  • We will be distributing a Q&A document in about one week. This document will fully answer questions asked during the webinar and will contain any corrections that we may discover after the webinar.

Fabulous Prizes
Agenda

- Bone
- Epi Moment
- Quiz
- Soft Tissue Sarcomas
- Quiz
- Case Scenarios

Bone

Anatomy
Staging
Treatment

Anatomy - Bone

- Epiphysis
- Metaphysis
- Diaphysis
**Anatomy - Bone**

- Epiphysis
  - Giant-Cell Tumor
- Metaphysis
  - Osteosarcomas
  - Chondrosarcomas
- Diaphysis
  - Ewing's Sarcomas

---

**Pop Quiz**

- Where does most growth occur in the long bones?
  - Epiphyseal line

---

**Laterality**

- Paired
  - C40.0 Long bones of upper limb, scapula and associated joints
  - C40.1 Short bones of upper limb and associated joints
  - C40.2 Long bones of lower limb and associated joints
  - C40.3 Short bones of lower limb and associated joints
  - C41.3 Rib, clavicle and associated joints
  - C41.4 Pelvic bones and associated joints

- Not paired
  - C41.0 Bones of skull and face and associated joints
  - C41.1 Mandible
  - C41.2 Vertebral column
  - C41.3 Sternum
  - C41.4 Sacrum, coccyx, and symphysis pubis
Multiple Primary Rules - Bone and Soft Tissue

- Use Other Sites rules for multiple primaries and histologies
- Use Hematopoietic rules for multiple myeloma

Pop Quiz

A patient presents with pain in his right leg. A CT is done and shows a tumor in the right femur and another in the right tibia. A needle biopsy confirms osteosarcoma.

- How many primaries does this patient have?
  - Osteosarcoma of the femur - C40.2 9180/3
  - Osteosarcoma of the tibia - C40.2 9180/3

Staging

MSTS
Summary Stage
AJCC Stage
Musculoskeletal Tumor Society (MSTS) Staging System

- Low-grade, localized tumors are stage I.
- High-grade, localized tumors are stage II.
- Metastatic tumors (regardless of grade) are stage III.
- Registrars do not code this staging system.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Grade</th>
<th>Tumor</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>G1</td>
<td>T1</td>
<td>M0</td>
</tr>
<tr>
<td>B</td>
<td>G1</td>
<td>T2</td>
<td>M0</td>
</tr>
<tr>
<td>IA</td>
<td>G2</td>
<td>T1</td>
<td>M0</td>
</tr>
<tr>
<td>IB</td>
<td>G2</td>
<td>T2</td>
<td>M0</td>
</tr>
<tr>
<td>II</td>
<td>G1 or G2</td>
<td>T1 or T2</td>
<td>M1</td>
</tr>
</tbody>
</table>

Summary Stage

- In situ is not a valid stage.
- Localized
  - Confined to the cortex of the bone
  - Extension beyond cortex to periosteum (no break in the periosteum)
- Regional by direct extension
  - Extension beyond periosteum to surrounding tissues

Regional lymph nodes
- If no mention of nodes, assume no metastasis
- Distant site(s)/node(s) involved
  - Distant lymph node(s)
  - Extension to skin
  - Further contiguous extension
  - Metastasis

http://www.cancer.org/cancer/osteosarcoma/detailedguide/osteosarcoma-staging

• Applies to all primaries of the bone except:
  • Primary malignant lymphoma
  • Myeloma
• Staging is based on:
  • Grade
  • Tumor size
  • Location of metastasis
• When analyzed stages should be grouped based on site groups
  • Extremities
  • Pelvis
  • Spine

• T Values are Driven by Tumor Size
  • Tumor size ≤ or >8cm
  • Discontinuous tumor in the bone
  • T1-3 (no T4)
• Regional node metastasis is rare
  • Consider N0 (rather than NX) unless clinical node involvement is clearly evident. See note on page 284.
• Distant metastasis most frequently occurs in the lungs

• Grade is included in stage grouping
• Patients with a low grade tumor (G1, G2) have a better prognosis than those with a high grade tumor (G3, G4)
  • Ewing's sarcoma is always G4

<table>
<thead>
<tr>
<th>AJCC Grade</th>
<th>Terminology</th>
</tr>
</thead>
<tbody>
<tr>
<td>SX</td>
<td>Grade Cannot be assessed</td>
</tr>
<tr>
<td>G1</td>
<td>Well differentiated</td>
</tr>
<tr>
<td>G2</td>
<td>Moderately differentiated</td>
</tr>
<tr>
<td>G3</td>
<td>Poorly differentiated</td>
</tr>
<tr>
<td>G4</td>
<td>Undifferentiated</td>
</tr>
<tr>
<td>Low Grade</td>
<td></td>
</tr>
<tr>
<td>High Grade</td>
<td></td>
</tr>
</tbody>
</table>
### Rules for Classification

- **Clinical**
  - MRI to assess the primary tumor
  - CT to identify distant mets
  - Technetium scintigraphy of the entire skeleton
  - Biopsy to confirm histology and grade
    - Should be done after imaging
- **Pathologic**
  - Resected primary tumor
  - Lymph nodes as appropriate
  - Assessment for distant mets

### Stage Grouping

- Regional lymph node involvement is rare
  - Pathologic stage grouping includes:
    - \( pT \ pN \ pM \ pG \)
    - \( pT \ pN \ cM \ pG \)
    - \( pT \ cN \ cM \ pG \)
  - \( cN \) can be used to calculate the pathologic stage
  - Any \( T \  N1 \) Any \( M \) Any \( G \) is Stage IVB

### Pop Quiz

- A 19 year old white male present with pain in his left forearm.
  - An MRI is done which shows a 10cm bone lesion. The lesion appears to be confined to the bone.
  - A core biopsy confirms high grade osteosarcoma.
  - The patient is treated with chemotherapy.
  - An MRI following chemotherapy shows the tumor has responded and is now 2cm.
  - The tumor is excised and now shows a 2cm poorly differentiated osteosarcoma confined to the cortex of the bone.
Data Items as Coded in Current NAACCR Layout

<table>
<thead>
<tr>
<th>T</th>
<th>N</th>
<th>M</th>
<th>Grade</th>
<th>Stage Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clin</td>
<td>c2</td>
<td>c0</td>
<td>c0</td>
<td>High</td>
</tr>
<tr>
<td>Path</td>
<td>p1</td>
<td>c0</td>
<td>c0</td>
<td>High</td>
</tr>
<tr>
<td>TNM Path Descriptor</td>
<td>4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Summary Stage</td>
<td>1-Localized</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Pop Quiz**

- What if the patient did not have surgery after chemotherapy? What stage would we assign?

Data Items as Coded in Current NAACCR Layout

<table>
<thead>
<tr>
<th>T</th>
<th>N</th>
<th>M</th>
<th>Grade</th>
<th>Stage Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clin</td>
<td>c2</td>
<td>c0</td>
<td>c0</td>
<td>High</td>
</tr>
<tr>
<td>Path</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TNM Path Descriptor</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Summary Stage</td>
<td>3-Localized</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Treatment**
**Diagnostic Workup and Surgery**

- Biopsy – core needle or surgical biopsy
- Surgery
  - Excision (25-26)
  - Limb-sparing resection or Radical Excision (30)
  - Amputation (40-42, 50-54)

**Chemotherapy and Radiation**

- Chemotherapy – Type varies based on type of cancer
- Radiation –
  - IMRT (31)
  - Particle beam (20-30, 40)
  - Stereotactic radiosurgery (41, 42, 43)

**Chondrosarcoma**

- Common in older adults
- Pelvis and Femur
- 85% are of the conventional type
- Intracompartmental vs Extrapartmental
Chondrosarcoma

- Low grade and Intracompartmental
  - Intralesional excision ± Surgical adjuvant
  - Wide excision if resectable
  - Consider Radiation Therapy if unresectable

- High Grade or Clear cell or Extracompartimental
  - Wide excision if resectable
  - Consider Radiation Therapy if unresectable

Chondrosarcoma

- Dedifferentiated
  - 10% of all chondrosarcomas
  - Pelvis bones, femur and humerus
  - Treated as Osteosarcoma

- Mesenchymal
  - 2/3 of cases occur in bone
  - Fast growing tumor
  - Treated as Ewing’s sarcoma

Chordoma - Histology

- Conventional
  - Classical – most common

- Chondroid
  - 5% - 15%
  - features of both chordoma and chondrosarcoma

- Dedifferentiated
  - 2% - 8%
  - Features of high-grade pleomorphic spindle cell soft tissue sarcoma
**Chordoma - Treatment**

- Sacrococcygeal or mobile spine
  - If resectable
    - Wide resection ± Radiation Therapy; adjuvant treatment
  - If unresectable
    - Consider Radiation Therapy
- Skull base/Clival
  - If resectable
    - Intralesional excision ± Radiation Therapy
  - If unresectable
    - Consider Radiation Therapy
- Dedifferentiated – Treated as Soft tissue Sarcoma

**Ewing's Sarcoma**

- Primitive Neuroectodermal tumor (PNET) of bone
  - Children and adults under 25 yrs
- Askin's tumor
  - PNET of soft tissue of chest wall
- Extraosseous Ewing's Sarcoma
  - Common primary sites: Trunk, Extremity, Head and neck, Retroperitoneum

**Ewing's Sarcoma - Treatment**

- Multi-agent Chemotherapy
  - 12 weeks prior to local therapy
- Response to Chemo (Restage)
  - Definitive Radiation Therapy and chemo
  - Wide excision
  - Amputation
- Progression of Disease
  - Consider Radiation Therapy and surgery
Giant Cell Tumor of the Bone - Treatment

- Malignancy approx. 2% of cases
- Slow to develop
- Lung most common site of metastasis
- 1-3% transform to malignant sarcoma

Giant Cell Tumor of the Bone - Treatment

- **Localized Disease**
  - Resectable
    - Excision
  - Resectable - Unacceptable morbidity and/or Unresectable axial lesions
    - Scleral embolization
    - Denosumab
    - IFN or PEG IFN
    - Radiation Therapy

Giant Cell Tumor of the Bone - Treatment

- **Metastatic Disease**
  - Resectable
    - For primary lesion treat as you would localized disease
    - Consider excision of metastatic sites
  - Unresectable
    - Denosumab
    - IFN or PEG IFN
    - Radiation Therapy
    - Observation
Osteosarcoma

- Most common malignant bone tumor
- 20 years old
- 3 main subtypes
  - Intramedullary 80%
  - Surface 5%
  - Extraskeletal

Osteosarcoma

- Low-grade: intramedullary + surface
  - Wide Excision
    - High grade → Chemo
- Periosteal osteosarcoma
  - Consider chemotherapy
  - Wide Excision
    - High grade → Chemo

Osteosarcoma

- High-grade: intramedullary + surface
  - Pre-op/Necadjuvant Chemo (restage)
  - Unresectable – Radiation Therapy or Chemo
  - Resectable – Wide Excision
    - Positive Margins
      - Chemo or surgical resection ± Radiation Therapy
    - Surgical resection ± Radiation Therapy or change chemo
    - Negative Margins
      - Chemo
      - Change of chemo
Osteosarcoma

- Metastatic disease at presentation
  - Resectable (pulmonary, visceral or skeletal metastases)
    - Chemo
    - Metastasectomy
    - Same Treatment as high grade osteosarcoma
- Unresectable
  - Chemo
  - Radiation Therapy
- Extraskeletal osteosarcoma – treat as Soft Tissue Sarcoma

Myeloma

- Accounts for more than 40% of bone tumors
- Hematopoietic disease
  - Multiple myeloma - most common form: More than 90 percent of people with myeloma have this type. Multiple myeloma affects several different areas of the body.
  - Plasmacloma - only one site of myeloma cells evident in the body, such as a tumor in the bone, skin, muscle, or lung.
  - Localized myeloma - found in one site with exposure to neighboring sites.
  - Extramedullary myeloma - involvement of tissue other than the marrow, such as the skin, muscles or lungs.

And now a brief pause for...

An Epi Moment

(insert Bones instrumental theme song here)
Epidemiology of Bone & Soft Tissue Cancers

- Rare cancers, often 2nd primary, sarcomas
- Analyzed as 2 groups
  - Bone & joints
  - Soft tissue including heart
- Important pediatric cancers

<table>
<thead>
<tr>
<th>Code</th>
<th>Tumor Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>VIII</td>
<td>Malignant bone tumors</td>
</tr>
<tr>
<td>VIII(a)</td>
<td>Osteosarcoma</td>
</tr>
<tr>
<td>VIII(c)</td>
<td>Chondrosarcoma</td>
</tr>
<tr>
<td>VIII(d)</td>
<td>Ewing tumor and related sarcomas of bone</td>
</tr>
<tr>
<td>VIII(e)</td>
<td>Other specified malignant bone tumors</td>
</tr>
<tr>
<td>IX</td>
<td>Soft tissue and other extramusculosarcomas</td>
</tr>
<tr>
<td>IX(a)</td>
<td>Rhabdomyosarcoma</td>
</tr>
<tr>
<td>IX(b)</td>
<td>Fibrosarcoma, peripheral nerve &amp; other fibrous</td>
</tr>
<tr>
<td>IX(c)</td>
<td>Kaposi sarcoma</td>
</tr>
<tr>
<td>IX(d)</td>
<td>Other specified soft tissue sarcomas</td>
</tr>
<tr>
<td>IX(e)</td>
<td>Unclassified soft tissue sarcoma</td>
</tr>
</tbody>
</table>

Incidence per 100,000 2008-2012
- Bone – 0.9 total, 1.1 males, 0.8 females
  - 1.0 white, 0.8 black, 0.6 API & AIAN
- Soft Tissue – 3.3 total, 3.9 male, 2.8 females
  - 2.3 API, 2.1 AIAN

Mortality per 100,000 2008-2012
- Bone – 0.4 total, 0.5 males, 0.3 females
- Soft Tissue – 1.3 total, 1.5 male, 1.2 females
  - Slightly higher among blacks 1.5

Pediatric Cancers, age 0-19

Incidence per 100,000 2008-2012
- Bone – equivalent to all age incidence by sex & race
- Soft Tissue – lower, 1.2 total
  - Same sex & race rankings as adults

Mortality per 100,000 2008-2012
- Lower, 0.2 for bone & soft tissue
  - No difference by sex or race
**Epidemiology of Bone Cancers**

- Primary bone cancer < 0.2% of all cancers
  - Benign more common than malignant (not collected; rarely life-threatening)
- Common 2nd primary
- Common metastatic site
  - Breast, prostate, lung (identified by bone pain)
  - Incurable but “treatable”—most commonly with bisphosphonates to relieve pain and reduce risk of fractures (IV)
- Adults:
  - 40% chondrosarcomas, 28% osteosarcomas, 10% chordomas, Ewing tumors 8%, malignant fibrous histiocytoma/fibrosarcoma 4%
- Pediatrics:
  - Osteosarcoma 56%, Ewing tumors 34%, chondrosarcoma 6%
50 types of soft tissue sarcomas
- Benign tumors more common than benign bone (lipomas)
- Muscle, tendons, fat, lymph & blood vessels, nerves, and tissue around joints
  - >50% arm/leg, about 20% abdomen, 10% trunk, 10% head/neck
- Rhabdomyosarcoma most common in pediatrics (<10 yo)
  - Skeletal muscles
- Less common in adults & also less treatable due to location
- Kaposi sarcoma (analyzed as separate category)
  - AIDS-related, Mediterranean (older adults), African (herpesvirus)
- GIST (gastrointestinal stromal tumors)
  - Rare but more common in adults aged 50+
- Uterine sarcomas—5% of uterine cancers

Epidemiology of Soft Tissue Cancers

Largely unknown

Genetic conditions
- Osteosarcomas
  - Li-Fraumeni and Rothmund-Thomson syndrome
- Pediatric genetic retinoblastoma (secondary cancer)
- Paget disease

Medical treatment (secondary cancer, pediatric)
- Radiation
  - Lag time about 10-15 year after tx
  - Tx has improved, more precise and lower dose
- Cancer drugs, Bone marrow transplant

Risk Factors for Bone Cancers

Largely unknown

Genetic conditions
- Neurofibromatosis
- Li-Fraumeni, Gardner, Gorlin, or Werner syndromes
- Pediatric genetic retinoblastoma (secondary cancer)
- Chemical exposures (possible)
  - Dioxin and other herbicides (farm worker exposure)

Risk Factors for Soft Tissue Cancers
**Diagnosing Bone Cancers**

- No screening tests (no *in situ*)
  - Pain most common
  - Swelling & Fractures associated with bone pain
  - Numbness/tingling/weakness if tumor presses on nerves
  - Weight-loss & fatigue
- Diagnosed initially by X-ray
  - CT scan used for staging
  - MRI & Radionuclide bone scans used to look for mets
  - Biopsy to determine primary cancer or mets
- Blood tests not useful in pediatrics

**Bone Cancers—Treatment & Survival**

- Treatment
  - Surgery
  - Chemotherapy
    - Not effective for chondrosarcoma
  - Radiation
    - Generally for chondrosarcoma, also palliative
  - Cryosurgery (liquid nitrogen)
- 5-year relative survival 70% (adults & pedis)
  - Chondrosarcomas (adults) 80%
  - Osteosarcomas 60-80% if local; 15-30% if mets
  - Ewing Tumors 70% if local; 15-30% if mets
  - 2008-2012 19% distant

**Diagnosing Soft Tissue Cancers**

- No screening tests (no *in situ*)
  - Lump or swelling
    - Particularly for rhabdomyosarcomas which leads to early diagnosis
  - Often asymptomatic until advanced stage
    - pressing on nearby nerves or blockage/bleeding of stomach or bowels (abdominal pain, blood in stool or vomit)
    - GIST generally diagnosed late
  - Genetic testing for those with family history may be useful
- Diagnosed by imaging
  - MRI main imaging tool
  - X-ray (bone involvement)
  - CT substituted for MRI if metal implants
  - Ultrason, PET
  - Biopsy generally guided by MRI
Soft Tissue Cancers—Treatment & Survival

• Treatment
  • More effective among children
  • Surgery—limb sparing
  • Radiation—also palliative
  • Chemotherapy
    • Isolated limb perfusion
    • Targeted Therapy
      • Votrient—for advance patients after chemo, Gleevec—for GIST

• 5-year relative survival
  • Local 83% (56% diagnosed local)
  • Regional 54% (20% diagnosed regional)
  • Distant 16% (15% diagnosed distant)
  • Most who survive 5 years are "cured" soft

CURRENT CINA Research

Recent Publications:
ACS Facts & Figures 2014, Special Section: Cancer in Children & Adolescents
http://www.cancer.org/acs/groups/content/@research/documents/webcontent/acspc-041787.pdf

Quiz 1
Soft Tissue Sarcomas

Anatomy – Soft Tissue

THE MESENGENIC PROCESS

Mesenchymal Stem Cell (MSC) (Pericyte)

Proliferation

"Commitment"

Lineage differentiation

"Differentiation"

Cytokine

Neural crest derivative cells

Bone

Cartilage

Muscle

Adipose

Connective tissue

Bone

Cartilage

Muscle

Adipose

Connective tissue

Anatomy – Soft Tissue

2015-2016 NAACCR Webinar Series
Summary Stage

- Based on where sarcoma arises
  - PERIPHERAL NERVES AND AUTONOMIC NERVOUS SYSTEM; CONNECTIVE, SUBCUTANEOUS, AND OTHER SOFT TISSUES
    - C47.0-C47.9, C47.8-C47.9, C49.0-C49.6, C49.8-C49.9
  - RETROPERITONEUM AND PERITONEUM
    - C48.0-C48.2, C48.8
  - HEART, MEDIASTINUM
    - C38.0-C38.3, C38.8
  - For any other sites use the schema for that chapter
    - E.g. for a breast sarcoma use the breast schema

Summary Stage

- Code 0 is not applicable for this site
  - 1 Localized
    - Invasive tumor confined to the site/tissue of origin
  - 2 Regional by direction extension only
    - Adjacent tissue
    - Unnamed tissues that immediately surround an organ or structure containing a primary cancer.
    - Adjacent organs
    - Organs are anatomic structures with specific physiologic functions other than (or in addition to) support and storage.
    - Adjacent structures
    - Connective tissues large enough to be given a specific name would be considered adjacent structures.

Summary Stage

- 3 Regional Lymph Nodes
  - Regional lymph nodes are listed in the manual by primary site.
    - Arm/shoulder:
      - Axillary
      - Epitrochlear for hand/forearm
      - Spinal accessory (posterior cervical) for shoulder
  - 7 Distant sites/lymph nodes
    - Distant lymph nodes
    - Further contiguous extension
    - Metastasis
AJCC Chapter 28 Soft Tissue Sarcoma

- Applies to all soft tissue sarcomas except
  - Kaposi Sarcoma
  - GIST (Chapter 16 Gastrointestinal Stromal Tumor)
  - Fibromatosis
  - Infantile fibrosarcoma
- Sarcoma’s arising from the following sites are not “optimally” staged by this system
  - Sarcomas arising in dura mater
  - Sarcomas arising in parenchymal organs
  - Sarcomas arising in visceral hollow organs

Site Groups for Soft Tissue Sarcomas

- Head and Neck
- Extremity and superficial trunk
- Gastrointestinal
- Genitourinary
- Visceral retroperitoneum
- Gynecologic
- Breast
- Lung, pleura, mediastinum
- Other

Rules for Classification

- Clinical Staging
  - Based on imaging and clinical evaluation prior to any treatment.
    - Tumor size can be measured clinically or radiographically (MRI or CT)
    - Evaluation for metastasis should be based on imaging. Most likely spot for distant metastasis is lungs.
- Pathologic Staging
  - Based on resection of the primary tumor and clinical/radiologic evaluation for regional and distant metastasis.
    - Tumor size can be based on imaging if an accurate tumor size cannot be obtained from the resected specimen
    - Grade should be based on tumor obtained prior to neoadjuvant treatment
Grade

- FNCLCC grade is based on three parameters
  - Differentiation
  - May be histologically specific (Table 28.1 pg 294)
  - Not all histologies will be assigned a differentiation
  - Mitotic activity
  - Extent of necrosis
- Stage grouping is strongly influenced by grade

Soft Tissue

CS Site-Specific Factor 1
Grade for Sarcomas

- Note 1: Comprehensive grading of soft tissue sarcomas is strongly correlated with disease specific survival and incorporates differentiation, mitotic rate, and extent of necrosis. The grading system of the French Federation of Cancer Centers Sarcoma Group (FNCLCC) is preferred system.
- Note 2: Report the grade from any three grade sarcoma grading system the pathologist uses prior to neoadjuvant treatment. Do not code forms such as “well differentiated” or “poorly differentiated” in this field.
- Note 3: In some cases, especially for musculoaponeurotic tumors, grade may be specified only as “low grade” or “high grade”. Use code 159, which maps to C1, or 290, which maps to C02. Codes 015-033 take priority over codes 159 and 290.
- Note 4: The mapping of grade as shown in this table is used in the derivation of AJCC staging.

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Mapped Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>015</td>
<td>Specified as Grade 1</td>
<td>1</td>
</tr>
<tr>
<td>020</td>
<td>Specified as Grade 2</td>
<td>2</td>
</tr>
<tr>
<td>025</td>
<td>Specified as Grade 3</td>
<td>3</td>
</tr>
<tr>
<td>159</td>
<td>Grade stated as low</td>
<td>1</td>
</tr>
<tr>
<td>290</td>
<td>Grade stated as high</td>
<td>3</td>
</tr>
</tbody>
</table>

Primary Tumor

- T1 and T2 are based on tumor size
  - Tumors 5cm or less are T1
  - Tumors more than 5cm are T2
- T1 and T2 are subdivided into “a” and “b”.
  - “a” indicates tumor is superficial
  - “b” indicates tumor is deep
- T3 and T4 are not defined
Superficial vs Deep

- Superficial "a"
  - Located entirely in the subcutaneous tissues without any degree of extension through muscular fascia or into underlying muscle
- Deep "b"
  - Tumor arising within subcutaneous tissue with invasion into or through the superficial fascia
  - Tumor entirely beneath the superficial fascia
  - Tumor arising beneath the deep fascia with invasion into or through the superficial fascia

Metastasis

- Regional lymph nodes
  - Regional lymph node metastasis is uncommon in adults
- Distant metastasis
  - Most common sites vary based on location of sarcoma
    - Lung is most common site for sarcomas of the extremities (arms and legs)
    - Liver is most common site for sarcomas of the retroperitoneum and GI tract

Stage Grouping

- Stage I and II are based on grade
  - GX or G1 are stage I
  - G2 or G3 are stage II or higher
- Lymph node involvement is Stage III or higher
A 24 year old white female presents with a mass in her left calf that had been present for several months and had been getting larger.
• A core needle biopsy was done that showed well differentiated fibrosarcoma.
• An MRI and CT were done that showed a 3cm tumor arising in the gastrocnemius muscle. No indication of metastasis. Tumor confined to musculature.
• The tumor was excised and the pathologist confirmed a 3cm well differentiated fibrosarcoma grade 2. The tumor approached but did not invade the fascia.

How do we stage this case?

<table>
<thead>
<tr>
<th>Data Items as Coded in Current NAACCR Layout</th>
</tr>
</thead>
<tbody>
<tr>
<td>T</td>
</tr>
<tr>
<td>Clin</td>
</tr>
<tr>
<td>Path</td>
</tr>
<tr>
<td>TNM Path Descriptor</td>
</tr>
<tr>
<td>Summary Stage</td>
</tr>
</tbody>
</table>

Gastrointestinal Stromal Tumor (GIST)

• Summary Stage based on location of tumor
  • Stomach-stomach schema (pg 74)
  • AJCC Chapter 16 Gastrointestinal Stromal Tumor (pg 175)
Follow rules for classification for peripheral soft tissue tumors

Primary tumor assessment is based on tumor size
- 2cm or less
- >2 but not more than 5cm
- >5 but not more than 10
- >10

Regional node metastasis is very rare
- NX should not be used

Distant metastasis
- Usually intraabdominal
- Bone, soft tissue, skin
- Lung metastasis is very rare

Mitotic Rate
- Mitotic rate strongly influences stage group
  - SSF 6

Mitotic Count

<table>
<thead>
<tr>
<th>Code</th>
<th>Mitotic Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>000</td>
<td>0.0 mitoses per 50 high power fields (HPF)</td>
</tr>
<tr>
<td>001-009</td>
<td>0.01-0.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>010-099</td>
<td>1.0-9.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>100-299</td>
<td>10.0-99.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>300-599</td>
<td>100.0-499.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>600-999</td>
<td>500.0-999.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>1000-4999</td>
<td>1000.0-4999.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>5000-9999</td>
<td>5000.0-9999.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>10000-49999</td>
<td>10000.0-49999.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>50000-99999</td>
<td>50000.0-99999.9 mitoses per 50 HPF</td>
</tr>
<tr>
<td>100000-</td>
<td>100000.0 or more mitoses per 50 HPF</td>
</tr>
</tbody>
</table>

Mitotic rate strongly influences stage group

Stage grouping is different for tumors arising in the stomach and tumors arising in the small intestine
- See tables 16.1 and 16.2
- Tumors arising in sites other than stomach or small intestine should be grouped based on Small Intestine stage group.
Table 1. Risk Stratification of Primary GIST by Mitotic Index, Tumor Size, and Tumor Location

<table>
<thead>
<tr>
<th>Mitotic Index, %</th>
<th>Size, cm</th>
<th>Site and Risk of Progressive Disease (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-5 per 50</td>
<td>Low (0)</td>
<td>Low (6)</td>
</tr>
<tr>
<td>5-15 per 50</td>
<td>Low (10)</td>
<td>Low (80)</td>
</tr>
<tr>
<td>&gt;15 per 50</td>
<td>Moderate (10%)</td>
<td>High (90)</td>
</tr>
<tr>
<td>&gt;10</td>
<td>Low (60)</td>
<td>Low (60)</td>
</tr>
<tr>
<td>1-10</td>
<td>Moderate (10%)</td>
<td>High (60)</td>
</tr>
<tr>
<td>&gt;10</td>
<td>Low (60)</td>
<td>Low (60)</td>
</tr>
</tbody>
</table>

NAACCR Webinar Series 28

http://www.cancer.gov/types/soft-tissue-sarcoma/hp/gist-treatment-pdq#link/_22_toc

Pop Quiz

• A pathology report comes across your desk and the final diagnosis is GIST NOS. Is this case reportable?
  • Not unless it is a reportable by agreement case or your state registry requires you to report the case.

• A physician completes a staging form for the patient above. Is the case reportable?
  • Same answer as above.

Pop Quiz

• If you decide to abstract this case reportable by agreement or to fulfill a state reporting requirement, what sequence number would you assign (assuming no previous reportable primaries)?
  • 60

• If the patient returns a year later and the physician refers to the GIST as malignant, what is the date of diagnosis and sequence?
  • The date of diagnosis is the date the physician refers to the tumor as malignant.
  • The exception would be if the physician or pathologist specifically state that the tumor was actually malignant at the time of the original diagnosis. In that case it would be the date of the original diagnosis.
Treatment

- Surgery
  - Excision (25-26)
  - Limb Sparing (30)
  - Amputation (40-42; 50-54)
- Radiation Therapy
  - Brachytherapy (50-54)
  - Intraoperative Radiation Therapy
  - Intensity-modulated Radiation Therapy (31)
- Chemotherapy

Extremity/superficial Trunk, Head/Neck

- Unique Histologies
  - Desmoid Tumors
  - Ewing's Sarcoma (extraosseus)
  - Gastrointestinal Stromal Tumors (GISTs)
  - Rhabdomyosarcoma
**Bone and Soft Tissue**

---

**Extremity/superficial Trunk, Head/Neck**

- Stage IA or IB
  - Surgery with adequate margins
  - Failure to obtain appropriate margins
  - Re-section or Observation (Stage IA)
  - Consider Radiation Therapy (Stage IA or IB)

---

**Extremity/superficial Trunk, Head/Neck**

- Stage II, III; Resectable, Positive Functional Outcomes
  - Stage IIA
    - Pre-op Radiation Therapy → Surgery
    - Surgery → Radiation Therapy
    - Surgery
  - Stage IIB, III
    - Pre-op Radiation Therapy or Radiation Therapy + Adjuvant Chemo
    - Pre-op Radiation Therapy/Chemoradiation → Surgery → Consider Radiation Therapy boost + Adjuvant Chemo
    - Pre-op Chemo → Surgery → Radiation Therapy/Radiation Therapy + Adjuvant Chemo

---

**Extremity/superficial Trunk, Head/Neck**

- Stage II, III, Resectable, Negative Functional Outcomes; Unresectable
  - Radiation Therapy
  - Chemoradiation
  - Chemotherapy
  - Regional limb therapy
**Extremity/superficial Trunk, Head/Neck**

- **Synchronous Stage IV**
  - Single organ and limited tumor bulk
  - **Primary Treatment**
    - Consider metastasectomy ± preop or post op chemo ± Radiation Therapy
    - Ablation
    - Embolization
    - Stereotactic body radiation therapy (SBRT)
    - Observation
  - **Disseminated Mets**
    - Palliative Treatment

**Retroperitoneal/Intra-Abdominal**

- **Resectable**
  - Biopsy done
    - GIST or Desmoid Tumors - treat as such
    - Other sarcoma
      - Surgery
      - Pre-op Therapy: Radiation Therapy or Chemo → Surgery
  - No Biopsy or nondiagnostic
    - Surgery ± Intraoperative Radiation Therapy (IORT)
      - GIST or Desmoid tumors → treat as such
      - Other sarcoma: depends on surgical margins
**Retroperitoneal/Intra-Abdominal**

- Unresectable or Stage IV
  - Biopsy
    - Attempt to shrink tumor
    - Palliative care only

**Gastrointestinal Stromal Tumors**

- Resectable disease
  - Surgery followed by Imatinib
- Resectable risk of significant morbidity or unresectable
  - Neoadjuvant Imatinib followed by reassessment for possible surgery

**Desmoid Tumors**

- Resectable
  - Observation
  - Treatment
    - Surgery
    - Radiation Therapy and/or Systemic therapy
**Desmoid Tumors**

- Unresectable or Unacceptably morbid
  - Definitive Radiation Therapy
  - Systemic therapy
  - Radical Surgery – considered if other treatments fail
  - Observation

**Rhabdomyosarcoma**

- Pleomorphic
  - Treat like soft tissue sarcoma
- Non – Pleomorphic
  - Alveolar
  - Embryonal

**Kaposi Sarcoma**

- Kaposi’s sarcoma (KS) is a tumor caused by Human herpesvirus 8 (HHV8)
- AJCC Staging is not used for Kaposi sarcoma. Patients may be assigned a Summary Stage.
Kaposi Sarcoma
• Radiation
• Surgery
  • Local excision
  • Electrodesication
  • Cryosurgery
• Chemotherapy
  • Liposomal chemotherapy (doxorubicin)
• BRM
  • Interferon

Quiz 2 & Case Scenarios

Questions?
Coming Up…

• Collecting Cancer Data: Breast  
  • 2/4/16
• Abstracting and Coding Boot Camp: Cancer Case Scenarios  
  • 3/3/16

And the winners are…

CE Certificate Quiz/Survey

• Phrase
  • Subcutaneous
• Link
Thank you!

Jim Hofferkamp, CTR
jhofferkamp@naaccr.org
217 698 0800 x 5

Angela Martin, CTR
amartin@naaccr.org
217 698 0800 x 9