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

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Bone

Anatomy
Staging
Treatment

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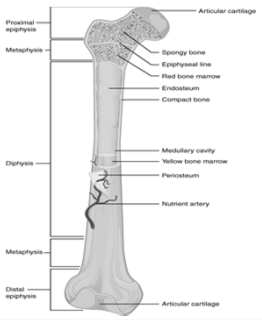
Anatomy - Bone

- Epiphysis
- Metaphysis
- Diaphysis



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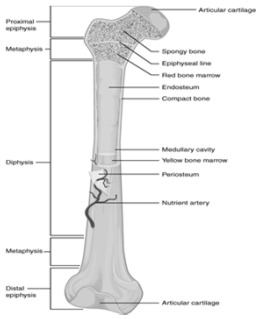
Anatomy - Bone

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



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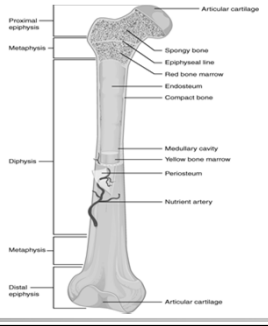
Pop Quiz

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



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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***

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Multiple Primary Rules-Bone and Soft Tissue

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

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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

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Staging

MSTS
Summary Stage
AJCC Stage

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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

Stage	Grade	Tumor	Metastasis
IA	G1	T1	M0
IB	G1	T2	M0
IIA	G2	T1	M0
IIB	G2	T2	M0
III	G1 or G2	T1 orT2	M1

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

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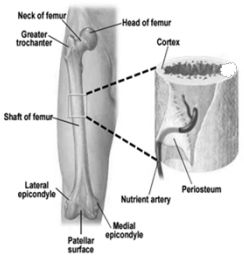
Summary Stage

Page 162

- 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 extenstion
  - Extension beyond periosteum to surrounding tissues



http://seer.cancer.gov/tools/ssm/musculoskel.pdf

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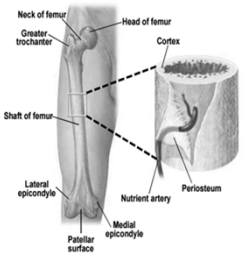
Summary Stage

- 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://seer.cancer.gov/tools/ssm/musculoskel.pdf

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AJCC Staging Bone

Chapter 27 Page 281

- 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

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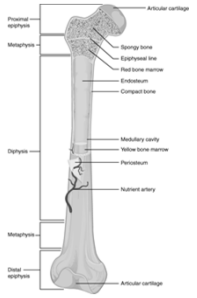
T,N, and M

- 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



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AJCC Grade

- 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

AJCC Grade	Terminology
GX	Grade Cannot be assessed
G1	Well differentiated
G2	Moderately differentiated
G3	Poorly differentiated
G4	Undifferentiated
Low Grade	
High Grade	

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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 imagining
- Pathologic
  - Resected primary tumor
  - Lymph nodes as appropriate
  - Assessment for distant mets

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Stage Grouping

- Regional lymph node involvement is rare
  - Pathologic stage grouping includes:
    - pT pN pM pG
    - pT pN cM pG
    - *pT **cN** cM pG**
    - cT cN pM pG

    **cN can be used to calculate the pathologic stage*
  - *Any T N1 Any M Any G is Stage IVB*

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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.

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Pop Quiz

	Data Items as Coded in Current NAACCR Layout				
	T	**N**	**M**	**Grade**	**Stage Group**
Clin	c2	c0	c0	High	IIB
Path	p1	*c0*	*c0*	High	IIA
TNM Path Descriptor	4				
Summary Stage	1-Localized				

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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				
	T	**N**	**M**	**Grade**	**Stage Group**
Clin	c2	c0	c0	High	IIB
Path					
TNM Path Descriptor	0				
Summary Stage	1-Localized				

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Treatment

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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)

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Chemotherapy and Radiation

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

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Chondrosarcoma

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

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Chondrosarcoma

- Low grade and Intracompartmental
  - Intralesional excision ± Surgical adjuvant
  - Wide excision if resectable
  - Consider Radiation Therapy if unresectable
- High Grade or Clear cell or Extracompartmental
  - Wide excision if resectable
  - Consider Radiation Therapy if unresectable

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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

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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

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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

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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

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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

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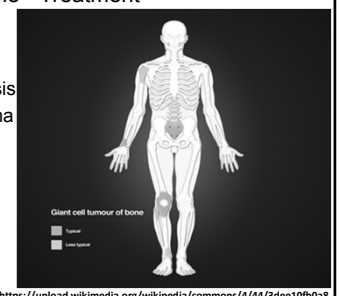
Giant Cell Tumor of the Bone - Treatment

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



https://upload.wikimedia.org/wikipedia/commons/4/44/3dee10fb0a8ad6244470738a8cb382_big_gallery.jpg

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Giant Cell Tumor of the Bone - Treatment

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

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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

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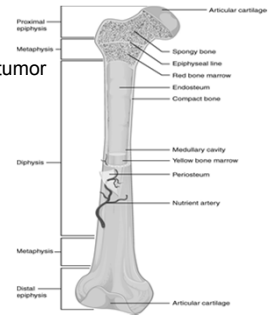
Osteosarcoma

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



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Osteosarcoma

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

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Osteosarcoma

- High-grade: intramedullary + surface
  - Pre-op/Neoadjuvant 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

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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

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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.
  - Plasmacytoma - 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.

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And now a brief pause for...

An Epi Moment

(insert Bones instrumental theme song here)

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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

ICCC

VIII Malignant bone tumors
- VIII(a) Osteosarcomas
- VIII(b) Chondrosarcomas
- VIII(c) Ewing tumor and related sarcomas of bone
- VIII(d) Other specified malignant bone tumors
- VIII(e) Unspecified malignant bone tumors

IX Soft tissue and other extraosseous sarcomas
- IX(a) Rhabdomyosarcomas
- IX(b) Fibrosarcomas, peripheral nerve & other fibrous
- IX(c) Kaposi sarcoma
- IX(d) Other specified soft tissue sarcomas
- IX(e) Unspecified soft tissue sarcomas

Epidemiology of Bone & Soft Tissue Cancers

- 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

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Epidemiology of Bone & Soft Tissue Cancers

- 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

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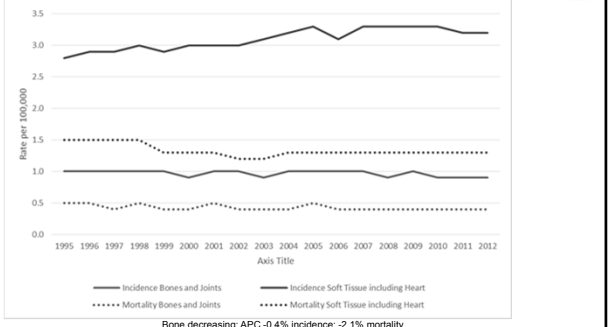
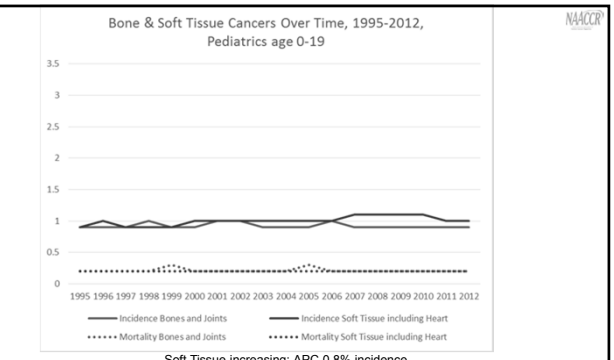


Bone decreasing: APC -0.4% incidence; -2.1% mortality
Soft Tissue increasing: APC1.2% incidence; 0.6% mortality



Soft Tissue increasing: APC 0.8% incidence

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%

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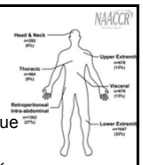
Epidemiology of Soft Tissue Cancers



- 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

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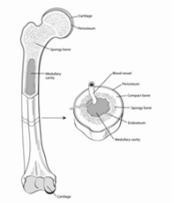
Risk Factors for Bone Cancers



- 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

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Risk Factors for Soft Tissue Cancers

- Largely unknown
- Medical treatment (secondary cancer)
  - Radiation
    - Lag time about 10-15 year after tx
    - Tx has improved, more precise and lower dose
  - Damaged lymph system
    - Lymphedema due to surgery or radiation
- 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)

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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

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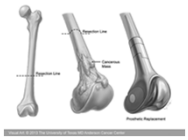
Bone Cancers—Treatment & Survival



- Treatment
  - Surgery
  - Chemotherapy
    - Not effective for chondrosarcoma
  - Radiation
    - Generally for chondrosarcoma, also palliative
  - Cryosurgery (liquid nitrogen)
- 5-year relative survival 70% (adults & peds)
  - 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

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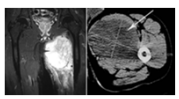
Diagnosing Soft Tissue Cancers

- 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
  - Ultrasound, PET
  - Biopsy generally guided by MRI



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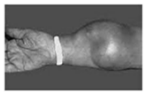
Soft Tissue Cancers—Treatment & Survival

- 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



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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

Childhood and adolescent cancer statistics, 2014 in CA: A Cancer Journal for Clinicians

http://onlinelibrary.wiley.com/doi/10.3322/caac.21219/full

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Quiz 1

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Soft Tissue Sarcomas

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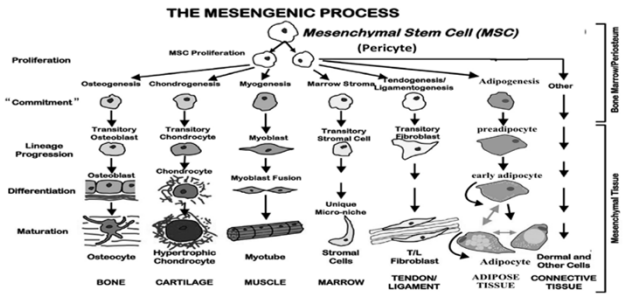
Anatomy – Soft Tissue



DiMarino AM, Caplan AI and Bonfield TL (2013) Mesenchymal stem cells in tissue repair. Front. Immunol. 4:201. doi: 10.3389/fimmu.2013.00201

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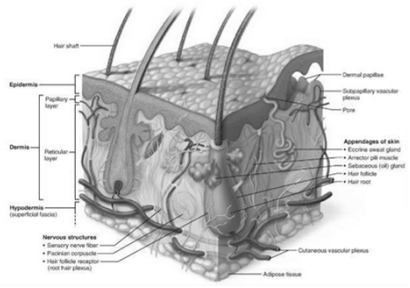
Anatomy – Soft Tissue



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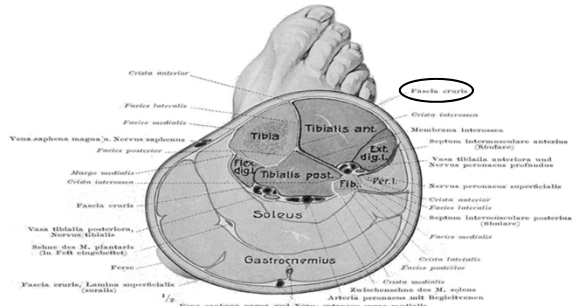
Anatomy – Soft Tissue



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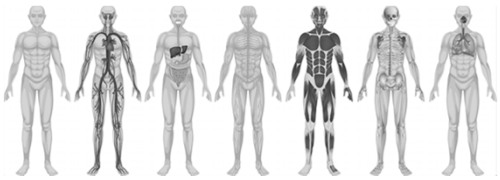
Anatomy – Soft Tissue



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Staging

Summary Stage
AJCC Stage

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Summary Stage

- Based on where sarcoma arises
  - PERIPHERAL NERVES AND AUTONOMIC NERVOUS SYSTEM; CONNECTIVE, SUBCUTANEOUS, AND OTHER SOFT TISSUES
    - C47.0-C47.6, 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

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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.

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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

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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

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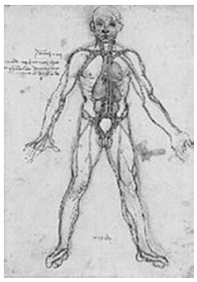
Site Groups for Soft Tissue Sarcomas

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



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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

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Grade

- FNLCC 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

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SoftTissue

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: Record the grade from any three-grade sarcoma grading system the pathologist uses prior to neoadjuvant treatment. Do not code terms such as "well differentiated" or "poorly differentiated" in this field.
- Note 3: In some cases, especially for needle biopsies, grade may be specified only as "low grade" or "high grade". Use code 100, which maps to G1, or 200, which maps to G3. Codes 010-030 take priority over codes 100 and 200.
- Note 4: The mapping of grade as shown in this table is used in the derivation of AJCC 7 staging.

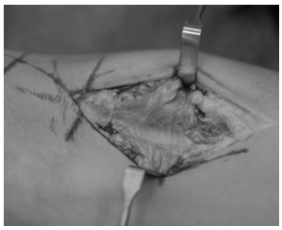
Code	Description	Mapping of Grade
010	Specified as Grade 1 [of 3]	1
020	Specified as Grade 2 [of 3]	2
030	Specified as Grade 3 [of 3]	3
100	Grade stated as low grade, NOS	1
200	Grade stated as high grade, NOS	3

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



http://www.handtoelbow.com/radial-nerve-and-posterior-interosseous-nerve-decompression/

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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

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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

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Pop Quiz

- 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.

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Pop Quiz

- How do we stage this case?

	Data Items as Coded in Current NAACCR Layout				
	T	**N**	**M**	**Grade**	**Stage Group**
Clin	c1b	c0	c0	X	IA
Path	p1b	*c0*	*c0*	G2	IIA
TNM Path Descriptor	0				
Summary Stage	1-Localized				

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Gastrointestinal Stromal Tumor (GIST)

- Summary Stage based on location of tumor
  - Stomach-stomach schema (pg 74)
- AJCC Chapter 16 Gastrointestinal Stromal Tumor (pg 175)

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GIST-AJCC Staging

- 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

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CS Site-Specific Factor 6
Mitotic Count

- Note 1: The mitotic rate, the count of mitoses (also termed "mitotic figures") per 50 high-power fields (HPF), reflects the potential aggressiveness or prognosis of gastrointestinal stromal tumors (GISTs) and is used alone to determine their histologic grade (low or high). The mitotic rate is also a factor in assigning the AJCC 7 anatomic stage/prognostic group. This site-specific factor presumes the denominator of 50 HPF or its equivalent, so just the numerator (the mitotic count) is coded here. For other schemas in which mitotic count is collected, the denominator may vary.
- Note 2: A HPF usually has a magnification objective of 40 (a 40x field). As described in the AJCC chapter on GIST, 50 HPF are equivalent to viewing a total area of 5 square millimeters (mm) at 40x magnification.
- Note 3: Record mitotic count, to the nearest tenth of a mitosis, as documented in the pathology report. For example, a mitotic count of 6/50 HPF, or 6 per 5 square mm, would be coded 060.
- Note 4: Code the specific mitotic count only per 50 HPF or 5 square mm; assume the denominator is 50 HPF or 5 square mm if not specified.
- Note 5: Use code 996 for a description of mitoses, NOS, or if the mitotic count is expressed with a specific denominator other than 50 HPF or 5 square mm.

Code	Description
000	0.0 mitoses per 50 high-power fields (HPF) (40x fields) 0.0 mitoses per 5 square millimeters (mm) Mitoses absent No mitoses present
001-008	0.1-0.8 mitoses per 50 HPF (40x field) 0.1-0.8 mitoses per 5 square mm
009	0.9 mitoses per 50 HPF (40x fields) 0.9 mitoses per 5 square mm Stated as less than 1 mitosis per 50 HPF (40x fields) Stated as less than 1 mitosis per 5 square mm
010-100	1 - 10 mitoses per 50 HPF (40x fields) 1 - 10 mitoses per 5 square mm
110	11 or more mitoses per 50 HPF (40x fields) 11 or more mitoses per 5 square mm

GIST Stage Grouping (pg 177)

- 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.

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

Table 1. Risk Stratification of Primary GIST by Mitotic Index, Tumor Size, and Tumor Location[a]

Mitotic Index, hpf	Size, cm	Site and Risk of Progressive Disease (%)			
		Gastric	Duodenum	Jejunum/Ileum	Rectum
≤5 per 50	≤2	None (0)	None (0)	None (0)	None (0)
	>2 ≤5	Very low (1.9)	Low (4.3)	Low (8.3)	Low (8.5)
	>5 ≤10	Low (3.6)	Moderate (24)	(Insufficient data)	(Insufficient data)
	>10	Moderate (10)	High (52)	High (34)	High (57)
>5 per 50	≤2	None	High[b]	(Insufficient data)	High (54)
	>2 ≤5	Moderate (16)	High (73)	High (50)	High (52)
	>5 ≤10	High (55)	High (85)	(Insufficient data)	(Insufficient data)
	>10	High (86)	High (90)	High (86)	High (7)

GIST = gastrointestinal stromal tumors; hpf = high-power field, assessed from an area that on initial screen appears to have the highest mitotic activity.

[a]Annual review of pathology by ANNUAL REVIEWS, INC. Reproduced with permission of ANNUAL REVIEWS, INC., in the format Internet posting via Copyright Clearance Center.[2]

[b]Small numbers of cases.

References

2. Corless CL, Heinrich MC: Molecular pathobiology of gastrointestinal stromal sarcomas. Annu Rev Pathol 3: 557-86, 2008. [PUBMED Abstract]

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.

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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 date the physician refers to the tumor as malignant. Sequence would be 00.
  - 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.

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Treatment

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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

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Extremity/superficial Trunk, Head/Neck

- Unique Histologies
  - Desmoid Tumors
  - Ewing's Sarcoma (extraossesus)
  - Gastrointestinal Stromal Tumors (GISTs)
  - Rhabdomyosarcoma

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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)

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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
    - Surgery → 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

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Extremity/superficial Trunk, Head/Neck

- Stage II, III, Resectable, Negative Functional Outcomes; Unresectable
  - Radiation Therapy
  - Chemoradiation
  - Chemotherapy
  - Regional limb therapy

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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

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Retroperitoneal/Intra-Abdominal

- Resectable
  - Biopsy done
    - GIST or Desmoid Tumors - treat as such
    - Other sarcoma
      - Surgery
      - Pre-op Therapy: Radiation Therapy or Chemo → Surgery

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Retroperitoneal/Intra-Abdominal

- Resectable
  - No Biopsy or nondiagnostic
    - Surgery ± Intraoperative Radiation Therapy (IORT)
      - GIST or Desmoid tumors – treat as such
      - Other sarcoma: depends on surgical margins

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Retroperitoneal/Intra-Abdominal

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

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Gastrointestinal Stromal Tumors

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

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Desmoid Tumors

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

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Desmoid Tumors

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

97

Rhabdomyosarcoma

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

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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
  - Electrodesiccation
  - Cryosurgery
- Chemotherapy
  - Liposomal chemotherapy (doxorubicin)
- BRM
  - Interferon

Quiz 2 & Case Scenarios

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Questions?

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Coming Up…

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

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And the winners are…





CE Certificate Quiz/Survey

- Phrase
  Subcutaneous
- Link
  - http://www.surveygizmo.com/s3/2519180/Bone-and-Soft-Tissue

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Thank you!

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