



Agenda

- Overview/Treatment
 - Quiz 1
- Collaborative Stage Data Collection System
 - Quiz 2
- Case Scenarios




OVERVIEW




Key Statistics

- Estimated new cases and deaths from cancer of the **bones and joints** in the United States in 2012:
 - New cases: 2,890
 - Deaths: 1,410



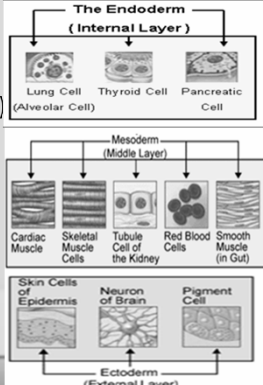
Key Statistics

- Estimated new cases and deaths (adults and children) from soft tissue sarcoma in the United States in 2012:
 - New cases: 11,280.
 - Deaths: 3,900.




Germ Cell Layers

- Ectoderm (outside layer)
- Mesoderm (middle layer)
- Endoderm (Inside layer)



Sarcoma

- A malignancy that arises from cell of mesenchymal origin.
 - Bone
 - Cartilage
 - Fat
 - Connective tissue



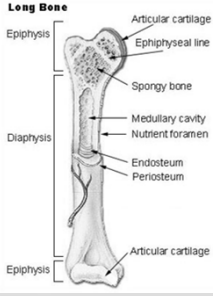
Osteosarcoma

- Usually develops during the period of rapid growth that occurs in adolescence
- Tend to occur in larger bones and in the area of bone with the fastest growth rate
 - Shin
 - Thigh
 - Upper arm
- Metastasizes hematogenously
 - Lung is the most common metastatic site

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Osteosarcoma

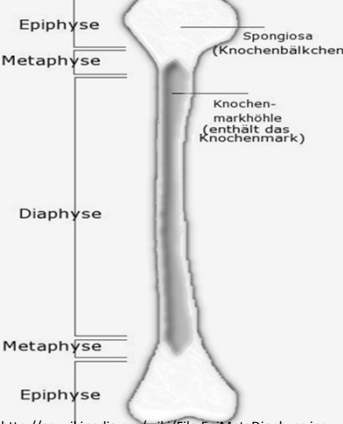
- Intramedullary
 - High grade intramedullary osteosarcoma comprises nearly 80% of all osteosarcoma
- Surface (Periosteum)
- Extra skeletal



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Osteosarcoma


- High grade intramedullary osteosarcoma tends to arise in the metaphyseal areas of the distal femur or proximal tibia



<http://en.wikipedia.org/wiki/File:EpiMetaDiaphyse.jpg>


Osteosarcoma

- Risk factors
 - Trauma
 - Radiation therapy
 - Retinoblastoma




Osteosarcoma-Treatment

<p>High Grade Intramedullary</p> <ul style="list-style-type: none">• Pre-operative chemotherapy• Wide excision• Adjuvant chemotherapy• Patients with unresectable disease may have radiation or chemotherapy as their primary treatment	<p>Low Grade Periosteal</p> <ul style="list-style-type: none">• Pre-operative chemotherapy• Wide excision• Adjuvant chemotherapy for patient with intramedullary and surface disease or high grade disease
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Chondrosarcoma

- Arise from cartilage
- Typically found in middle age and older adults
- May be divided into two categories
 - Tumors arising from previously normal bone preformed from cartilage
 - Secondary or peripheral tumors that arise or develop from preexisting benign cartilage lesions
- Pelvis and femur are the most common primary sites



Chondrosarcoma-Treatment

- Grade and tumor location are key variable for determining grade
 - Wide excision is the preferred treatment
 - Some patients, especially those with high grade disease may benefit from adjuvant radiation therapy
 - Patient with unresectable disease may benefit from radiation therapy

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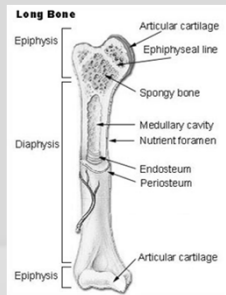
Ewing Sarcoma

- Ewing sarcoma family of tumors
 - Ewing Sarcoma
 - Extra osseous Ewing sarcoma
 - Primitive neuroectodermal tumor (PNET)
 - Primitive neuroectodermal tumor (PNET) of the bone
 - Askin tumor

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Ewing Sarcoma

- More than 50% of patients are adolescence
- Most common sites are:
 - Femur
 - Pelvic bone
 - Bones of the chest wall
- Typically occurs along the diaphysis of the bone
- Nearly 25% present with metastatic disease



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Ewing Sarcoma

- Question
 - What is the histology code for a soft tissue thigh mass that was diagnosed as Ewing sarcoma/PNET, primitive neuroectodermal tumor?

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Ewing Sarcoma

- Answer
 - The histologies stated for this case are Ewing sarcoma (9260) and PNET, primitive neuroectodermal tumor,(9364).
 - Use the Other Site Rules, starting with H8.
 - Stop at H17 and assign the higher histology code -- 9364/3 (Peripheral neuroectodermal tumor).

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Ewing Sarcoma-Treatment

- Multi-agent chemotherapy prior to surgery
 - VAC (Vincristine, doxorubicin, and cyclophosphamide)
 - VAC/IE (Vincristine, doxorubicin and cyclophosphamide alternating with Ifosfamide and etoposide)
 - VIDE (vincristine, ifosfamide, doxorubicin, and etoposide)
- Re-staging
- Wide excision
- Adjuvant chemotherapy +/- Radiation

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Primary Site

- Paired sites
 - C400 Long bones of upper limb, scapula, and associated joints
 - C401 Short bones of upper limb and associated joints
 - C402 Long bones of lower limb and associated joints
 - C403 Short bones of lower limb and associated joints
 - C413 Rib, clavicle (excluding sternum)
 - C414 Pelvic bones (excluding sacrum, coccyx, symphysis pubis)

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Soft Tissue Sarcoma

<p>Arise in mesodermal tissues</p> <ul style="list-style-type: none"> • Muscle (musculo, my, myo) • Fat (adip or adipo or lip or lipo) • Blood vessels (angi or angio) • Nerves (neuri or neuro) • Tendons (tendin or tendino) • Joint linings (synovial lining) 	<p>Occur in soft tissues of the</p> <ul style="list-style-type: none"> • Arms and legs (50%) • Trunk and retroperitoneum (40%) • Head and neck (10%)
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Primary Site

- Paired sites
 - C471 Peripheral nerves and autonomic nervous system of upper limb and shoulder
 - C472 Peripheral nerves and autonomic nervous system of the lower limb and hip
 - C491 Connective, subcutaneous, and other soft tissues of upper limb and shoulder
 - C492 Connective, subcutaneous, and other soft tissues of the lower limb and hip

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Soft Tissue Sarcoma

- More than 50 different histologic subtypes
 - Malignant fibrous histiocytoma (MFH)
 - Gastrointestinal stromal tumor (GIST)
 - Liposarcoma
 - Leiomyosarcoma
 - Synovial sarcoma
 - Malignant peripheral nerve sheath tumors




Soft Tissue Sarcoma

- Superficial
 - Lesions located entirely in the subcutaneous tissues without any degree of extension through the muscular fascia or into underlying muscle.
- Deep
 - Deep lesions are located partly or completely within one or more muscle groups within the extremity.
 - Deep tumors may extend through the muscular fascia into the subcutaneous tissues or even to the skin but the critical criterion is location of any portion of the tumor within the muscular components of the extremity.




Soft Tissue Sarcoma

- According to AJCC, "All intraperitoneal visceral lesions, retroperitoneal lesions, and intrathoracic lesions, and the majority of head and neck tumors are considered deep."




Soft Tissue Sarcoma-Treatment

- Extremities, trunk, head and neck
 - Stage I
 - Wide excision of tumor
 - Radiation may be done if margins are within 1cm and the intact facial plane was not remove
 - Stage II-III
 - May have neo adjuvant RT or chemotherapy
 - Wide excision of tumor and may have radical lymphadenectomy
 - Post operative chemotherapy if surgical margins are positive




Soft Tissue Sarcoma-Treatment

- Extremities, trunk, head and neck
 - Stage IV-limited metastasis
 - Same as stage II and III disease
 - May have metastasectomy
 - Stage IV-disseminated metastasis
 - Watchful waiting
 - Palliative radiation, surgery, or chemotherapy




Soft Tissue Sarcoma-Treatment

- Retroperitoneal/Intra-abdominal Soft Tissue Sarcoma
 - Pre-operative radiation in select patients
 - Intra operative radiation with or without external beam radiation
 - Electron beam
 - HDR Brachytherapy
 - Surgical resection
 - Postoperative radiation therapy




Rhabdomyosarcoma

- More common among children than adults
 - Accounts for 50% of soft tissue sarcomas in children
 - Three subtypes
 - Embryonal
 - Alveolar
 - Pleomorphic




Rhabdomyosarcoma

- Favorable prognostic factors
 - Age 1-9
 - Site of origin
 - Tumor less than 5cm
 - Localized
 - Histopathologic subtype




Rhabdomyosarcoma-Treatment

- Multimodality treatment
 - Surgery
 - Radiation
 - Chemotherapy



Kaposi Sarcoma

- Classic Kaposi sarcoma
- African Kaposi sarcoma
- Immunosuppressive treatment-related Kaposi sarcoma
- Epidemic Kaposi Sarcoma



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Kaposi Sarcoma-Multiple Primary Rules

- Rule M5
 - Kaposi sarcoma (any site or sites) is always a single primary.

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Kaposi Sarcoma-Treatment

- Radiation
- Surgery
 - Local excision
 - Electrodesiccation
 - Cryosurgery
- Chemotherapy
 - Liposomal chemotherapy (doxorubicin)
- BRM
 - Interferon

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Grade

Bone Sarcoma <ul style="list-style-type: none">• Usually a four grade system<ul style="list-style-type: none">– G1 Well differentiated– G2 Moderately differentiated– G3 Poorly differentiated– G4 Undifferentiated• Low Grade-2• High grade-4• Grade is included in AJCC stage	Soft Tissue Sarcoma <ul style="list-style-type: none">• Various grading systems<ul style="list-style-type: none">– Four grade– Low/High– French (FNCLCC)<ul style="list-style-type: none">• Three grade system based on:<ul style="list-style-type: none">– Differentiation– Mitotic rate– Extent of necrosis– NIH• Grade is included in AJCC stage
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Multiple Primary Rules

- Rule M8
 - Tumors on both sides (right and left) of a site listed in Table 1 are multiple primaries.
- Rule M10
 - Tumors diagnosed more than one (1) year apart are multiple primaries.
- Rule M11
 - Tumors with ICD-O-3 topography codes that are different at the second (Cxxx) and/or third characters (Cxxx) are multiple primaries.

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Multiple Primary Rules

- Rule 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.
 - Bones, joints, and articular cartilage (C40_ - C41_)
 - Peripheral nerves and autonomic nervous system (C47_)
 - Connective subcutaneous and other soft tissues (C49_)

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Multiple Primary Rules

- Rule 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.

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Multiple Primary Rules

Question

- Does a prior clinical diagnosis of a metastatic deposit for a previously diagnosed sarcoma have priority if the diagnosis on a subsequent resection (18 months later) indicates it is also a sarcoma but does not state it represents metastasis from the original sarcoma primary?

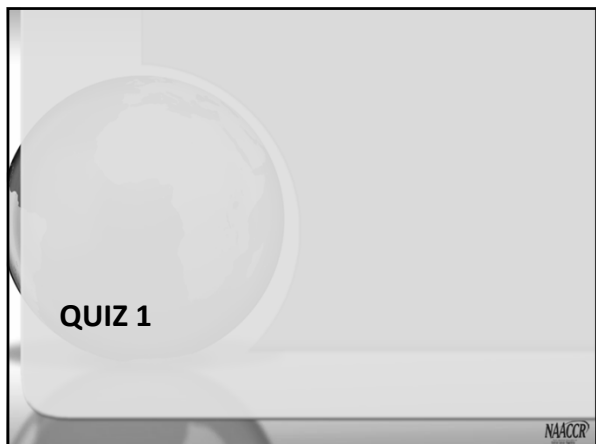
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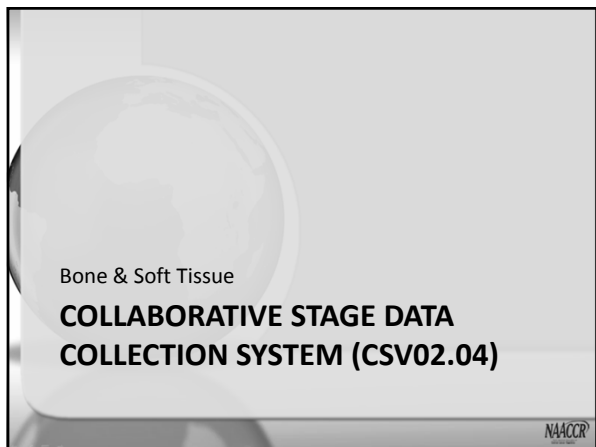
Multiple Primary Rules-Other

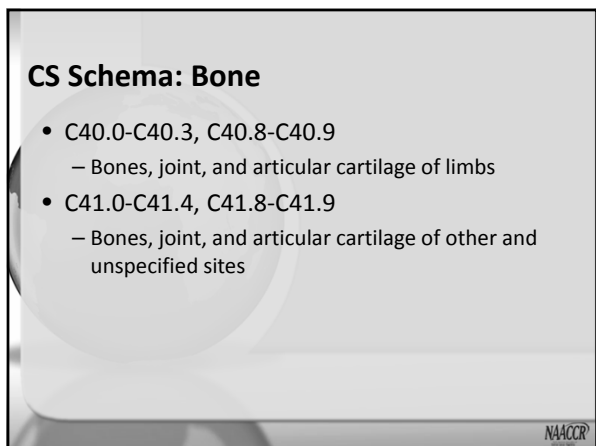
- For cases diagnosed 2007-2013, this is a single primary per Rule M1.
 - According to our expert pathologist, "if multiple solid tissue tumors are present (sarcomas), then almost always there is one primary and the rest are metastases. There are infrequent occasions of multifocal liposarcoma or osteosarcoma occurring, but the patient would be treated as a patient with metastatic disease."

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CS Tumor Size: Bone

- T1 and T2 categories based on tumor size
- Code 995
 - Less than 8 cm or between 5 cm and 8 cm
 - Stated as T1 [NOS] with no other information on size
- Code 996
 - Greater than 8 cm
 - Stated as T2 [NOS] with no other information on size




CS Extension: Bone

- T1 and T2 categories based on tumor size
 - For CS Extension codes 100-800, T category is based on value of CS Tumor Size
- Cortex: Outer shell of bone that provides its strength
 - Code 100: Invasive tumor confined to cortex of bone
- Periosteum: Fibrous membrane covering of bone that contains blood vessels and nerves
 - Code 200: Extension beyond cortex to periosteum



CS Extension: Bone

- Skip metastasis
 - Discontinuous metastasis in same bone as primary tumor
 - Assign CS Extension code 820
 - Discontinuous metastasis in an adjacent bone
 - Assign appropriate code in CS Mets at DX



Pop Quiz

- MRI: Malignant neoplasm confined to right femur less than 8 cm in size with skip metastasis in right femur.
- Biopsy of right femur lesion: High-grade osteosarcoma.
- Treatment summary: Patient had neoadjuvant chemotherapy followed by wide excision of right femur. Pathology report documented 4.2 cm osteosarcoma confined to the cortex without metastasis.

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Pop Quiz

- What is the code for CS Tumor Size?
 - 042
 - 996: Described as "less than 8 cm" or "between 5 cm and 8 cm"; Stated as T1 NOS with no other information on size
- What is the code for CS Extension?
 - 100: Invasive tumor confined to cortex of bone
 - 300: Localized NOS
 - 820: Discontinuous tumors in the primary bone site

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CS Lymph Nodes: Bone

- Regional node involvement is rare
- Assign code 000 (no regional lymph node involvement) if no mention of node involvement
- Assign code 999 (unknown) only when no information is available on patient's extent of disease

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CS Mets at DX: Bone

- Code 30
 - Distant metastasis to lung only; Stated as M1a with no other information on distant metastasis
- Code 35
 - Distant lymph nodes
- Code 40
 - Distant metastasis except distant lymph nodes or "lung only"; Distant metastasis to lung plus other sites except distant lymph nodes; Carcinomatosis
 - Includes skip metastasis to adjacent bone
- Code 50
 - (30 or 40) + 35




Pop Quiz

- MRI: Malignant neoplasm confined to left humerus, size between 3 and 4 cm, with skip metastasis in left humerus.
- CT chest: 3 metastatic lesions in left lung; right lung clear.
- Biopsy of left humerus: High-grade chondroblastic osteosarcoma.
- Oncology: Patient to begin neoadjuvant chemotherapy with Cisplatin and Doxorubicin next week. Wide excision of humerus tumor and metastatic lesions to follow completion of neoadjuvant chemotherapy.




Pop Quiz

- What is the code for CS Mets at DX?
 - 00: No distant metastasis
 - 30: Distant metastasis to lung only
 - 40: Distant metastasis except distant lymph node(s) or "lung only"; Distant metastasis to lung plus other sites except distant lymph nodes; Carcinomatosis
 - 60: Distant metastasis NOS




SSF1: Tumor Size- 2nd Largest Dimension

- Code the 2nd largest tumor dimension in mm
 - Pathologic tumor size from resected specimen is priority
 - Code clinical size if no resection
 - Assign code 999 (unknown) if only 1 tumor dimension is documented




SSF2: Tumor Size- 3rd Largest Dimension

- Code the 3rd largest tumor dimension in mm
 - Pathologic tumor size from resected specimen is priority
 - Code clinical size if no resection
 - Assign code 999 (unknown) if only 1 tumor dimension is documented



Pop Quiz

- MRI: Malignant neoplasm of the pelvis greater than 8 cm in size.
- Biopsy of right pelvic lesion: High-grade osteosarcoma.
- Treatment summary: Patient had multi-agent neoadjuvant chemotherapy followed by wide excision of pelvic lesion. Pathology report documented 4.2 cm x 2.1 cm x 3.2 cm osteosarcoma confined to the cortex.



Pop Quiz

- What is the code for CS Tumor Size?
 - 042
 - 997: Described as "greater than 8 cm"
- What is the code for SSF1 (Tumor Size- 2nd Largest Dimension)?
 - 032
 - 999: Unknown
- What is the code for SSF2 (Tumor Size- 3rd Largest Dimension)?
 - 021
 - 999: Unknown

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SSF3: Percent Necrosis Post Neoadjuvant Chemotherapy

- Code exact percentage of tumor necrosis post neoadjuvant chemotherapy
- Assign code 998 if:
 - No surgical resection of primary tumor
 - No histologic examination of primary tumor
 - No neoadjuvant chemotherapy

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SSF4: Resected Pulmonary Metastasis

- Record the exact number of pulmonary metastases found at initial diagnosis that were resected
- Assign code 000 if no lung metastases resected

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CS Schemas: Soft Tissue Sarcomas

- HeartMediastinum
 - C38.0-C38.3, C38.8
- SoftTissue
 - C47.0-C47.6, C47.8-C47.9
 - Peripheral nerves and autonomic nervous system
 - C49.0-C49.6, C49.8-C49.9
 - Connective, subcutaneous, and other soft tissues
- Retroperitoneum
 - C48.0
- Peritoneum
 - C48.1-C48.2, C48.8




CS Schemas: Soft Tissue Sarcomas

- *Excluded*
 - Kaposi's sarcoma
 - Gastrointestinal stromal tumors (GIST)
 - Fibromatosis (desmoid tumor)
 - Sarcoma arising in dura mater, brain, parenchymatous organs, or hollow viscera



CS Schemas: Soft Tissue Sarcoma

- *Question*
 - Patient has synovial sarcoma of the breast. Should the primary site be coded to C50.9 (breast NOS) or C49.3 (soft tissues of thorax)? Primary site determines if CS breast schema or CS soft tissue schema is used.



CS Tumor Size: Soft Tissue

- T1 and T2 categories based on tumor size
- Code 995
 - Less than 5 cm OR greater than 4 cm OR between 4 cm and 5 cm
 - Stated as T1a or T1b or T1 [NOS] with no other information on size
- Code 996
 - Greater than 5 cm
 - Stated as T2a or T2b or T2 [NOS] with no other information on size

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CS Extension: Soft Tissue

- Superficial tumor
 - Located entirely in subcutaneous tissues without extension through muscular fascia or into muscle
 - For extremities and trunk ONLY
 - Tumor does not involve superficial muscular fascia
- Codes
 - 110: Superficial tumor confined to site/tissue of origin
 - 310: Superficial: localized tumor NOS
 - 410: Superficial tumor involving adjacent connective tissue

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CS Extension: Soft Tissue

- Deep tumor
 - Located partly or completely within 1 or more muscle groups
 - For extremities and trunk ONLY
 - Tumor involves or is beneath superficial fascia
 - Intraperitoneal visceral lesions, retroperitoneal lesions, intrathoracic lesions, and majority of head and neck tumors (C47.0, C47.3-C47.5, C49.0, C49.3-C49.5)

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CS Extension: Soft Tissue

- Deep Tumor
 - Codes
 - 120: Deep tumor confined to site/tissue of origin
 - 320: Deep: localized tumor NOS
 - 420: Deep tumor involving adjacent connective tissue
 - 620: Deep tumor involving adjacent organs/structures including bone/cartilage

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CS Extension: Soft Tissue

- T1 and T2 categories based on tumor size
 - For CS Extension codes 100-800, T category is based on value of CS Tumor Size
 - Stated as T categories with no other information on extension
 - 302: Stated as T1a with no other info on extension
 - 305: Stated as T2a with no other info on extension
 - 312: Stated as T1b with no other info on extension
 - 315: Stated as T2b with no other info on extension
 - 322: Stated as T1 with no other info on extension
 - 325: Stated as T2 with no other info on extension

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CS Extension: Soft Tissue

- Other codes
 - 100: Tumor confined to site/tissue of origin NOS
 - 300: Localized NOS
 - 400: Adjacent connective tissue
 - Unnamed tissues surrounding organ or structure containing primary cancer
 - 600: Adjacent organs/structures including bone/cartilage
 - Includes named vessels

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Pop Quiz

- Wide excision of right forearm lesion: Spindle cell sarcoma of forearm connective tissue extending into muscle; 3 cm tumor size; FNCLCC grade 1; pT1b.




Pop Quiz

- What is the code for CS Extension?
 - 100: Invasive tumor confined to site/tissue of origin, NOS
 - 110: Superficial invasive tumor confined to site/tissue of origin
 - 120: Deep tumor confined to site/tissue of origin
 - 312: Stated as T1b with no other information on extension




CS Lymph Nodes: Soft Tissue

- Regional node involvement by soft tissue sarcoma is rare
- Assign code 000 (no regional lymph node involvement) if no mention of node involvement
- Assign code 999 (unknown) only when no information is available on patient's extent of disease




CS Mets at DX: Soft Tissue

- Record metastasis to distant sites and/or lymph nodes at time of diagnosis
- Lung is most common metastatic site for extremity sarcoma




SSF1: Grade for Sarcoma

- Record grade from any 3-grade system for sarcoma
 - French Federation of Cancer Centers Sarcoma Group (FNCLCC) is preferred grading system
 - Mitotic activity, extent of necrosis, and differentiation
- Code grade from path report
 - Codes for grade 1, 2, or 3 (010-030) take precedence over low grade (100) or high grade (200)
 - Do not code well differentiated, poorly differentiated or similar terminology



Pop Quiz

- Biopsy of soft tissue mass right side of abdomen: High grade sarcoma
- Wide excision of abdominal lesion: Sarcoma of abdominal connective tissue and muscle; 5.8 cm tumor size; FNCLCC grade 2.
- What is the code for SSF1 (Grade for sarcoma)?
 - 020: Specified as Grade 2 [of 3]
 - 200: Grade stated as high grade NOS



SSF2: Neurovascular Invasion

- Code the absence or presence of neurovascular invasion as documented on path report
 - May be called blood vessel invasion/involvement, vascular invasion/involvement, involvement/invasion of nerve, involvement/invasion of neurovascular bundle

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SSF3: Bone Invasion

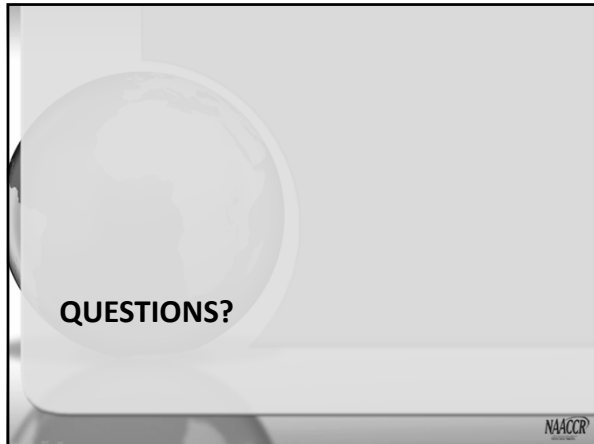
- Code the absence or presence of direct tumor extension from the primary sarcoma into adjacent bone
 - Information based on imaging techniques
 - Assign code 998 if no imaging for bone invasion was done

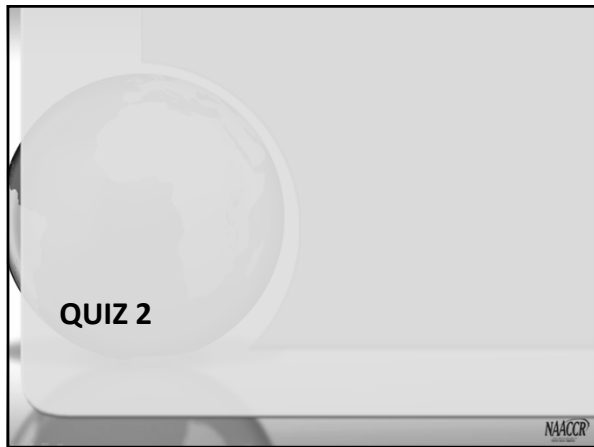
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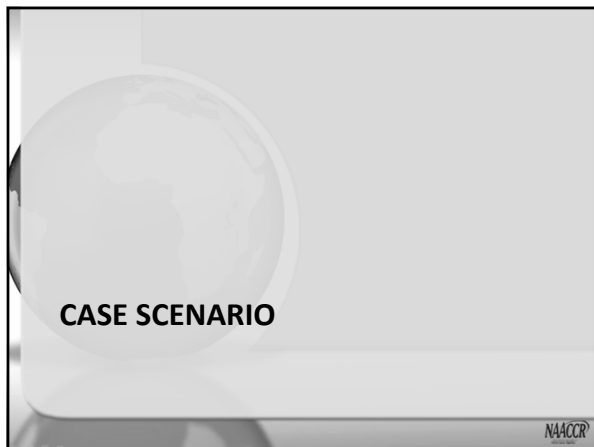
SSF4: Pathologic M1 - Source of Pathologic Metastatic Specimen

- Code the type of distant metastasis identified microscopically at the time of diagnosis
 - Do not include distant metastasis identified only by imaging or other clinical methods
 - Assign code 998 (no microscopic examination of metastatic site/no clinical evidence of metastasis and/or only clinical evidence of metastasis)
- Do not code disease progression or distant recurrence

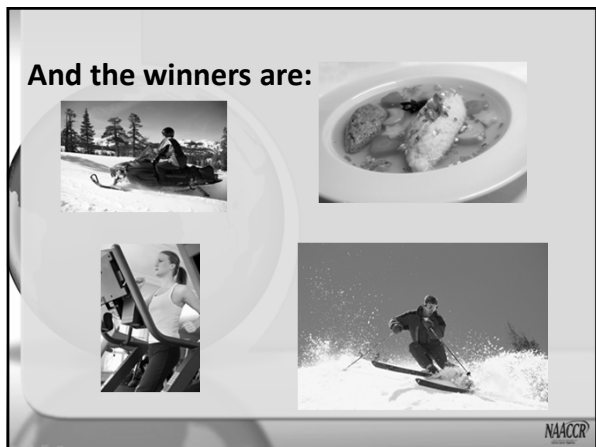
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And the winners are:

