2018 KOPANA Spring Seminar, Vancouver

Critical Review and Appraisal of the Latest AJCC System and/or WHO classification

Bone and Soft Tissue Sarcomas

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AJCC cancer staging manual editions

Edition	Publication year	Effective year
1st	1977	1978
2nd	1983	1984
3rd	1988	1989
4th	1992	1993
5th	1997	1998
6th	2002	2003
7th	2009	2010
8th	2017	2018

Background: 8th edition

- There have been enormous **advances in cancer diagnosis**, **staging,** and **treatment**.

- As a result, we can better **predict the level of cancer risk** and tailor a **more personalized treatment program** for the patient.

- New 8th edition incorporates **newer precision medicine paradigms**, **evidence based medicine,** and **biostatics.**

New features in the 8th edition

- **Levels of evidence** provided for revisions to staging system
- **Imaging** section
- **Risk assessment models** for select cancer sites
- **Recommendations for clinical trial stratification**
- **Prognostic factors**
  - Required for prognostic stage grouping
  - Recommended for clinical care
  - Emerging factors

Levels of evidence

Levels of evidence	Available evidence
I	Consistent results from **multiple, large, well designed**, and **well-conducted** national and international studies
II	At least **one large, well-designed**, and well-conducted study
III	Somewhat **problematic** because of one or more factors, such as the number, size, or quality of individual studies; inconsistency of results across individual studies; appropriateness of the patient population used in one or more studies; or the appropriateness of outcomes used in one or more studies
IV	**Insufficient** because appropriate studies have not yet been performed

How is the stage determined?

- AJCC TNM stage system is based on 4 key pieces of information,
  (1) The extent of the **t**umor (**T**)
  (2) The spread to lymph **n**odes (**N**)
  (3) The spread (**m**etastasis) to distant sites (**M**)
  (4) The **grade** of the cancer

- Once a person's T, N, and M categories have been determined, this information is combined in a process called **stage grouping** to assign an overall stage.

AJCC prognostic stage groups

- **Stage groups** are denoted by Roman numerals from I to IV with increasing extent of disease and generally worsening overall prognosis.
- **Stage I** generally indicates cancers that are smaller or less deeply invasive without regional disease or nodes.
- **Stage II** and **III** defines patients with increasing tumor or nodal extent.
- **Stage IV** identifies those who present with distant metastases (M1) at diagnosis.

Stage classification

Classification	Designation	Details
Clinical	**cTNM** or **TNM**	Used for all patients with cancer identified before treatment
Pathological	**pTNM**	Used for patients if surgery is the first definite therapy
Posttherapy or post neoadjuvant therapy	**ycTNM** and **ypTNM**	Used for patients with systemic and/or radiation therapy before surgery
Recurrence or retreatment	**rTNM**	Used for assigning stage at time of recurrence or progression until treatment is initiated

Histopathologic type

Component of histology	Description
Resource	The **WHO classification of tumors**, published in numerous anatomic site-specific editions, is used most commonly for histopathologic typing.
Histologic codes for staging	Each chapter in AJCC cancer staging manual includes the applicable WHO and ICD-O-3 histopathologic codes. If a specific histology is not listed, the case should not be staged using the AJCC classification in that chapter.

New features in the 8th edition: bone and soft tissue sarcomas

- **New paradigms**
  - Separate staging system based on anatomic site.

- **New chapters/staging systems**
  - Bone: appendicular skeleton/trunk/skull/face, spine, and pelvis
  - Soft tissue sarcoma of the head and neck
  - Soft tissue sarcoma of the trunk and extremities
  - Soft tissue sarcoma of the abdomen and thoracic visceral organs
  - Soft tissue sarcoma of the retroperitoneum
  - Soft tissue sarcoma - unusual histologies and sites

Contents

Primary bone cancers

- Primary bone cancers are **relatively rare**, representing less than 0.2% of all malignancies.

- **Osteosarcoma (35%), chondrosarcoma (30%),** and **Ewing sarcoma (16%)** are the most common forms of primary bone cancer.

- Bone cancers are staged based on the **histologic type, grade, size,** and **localization of the tumor** and **the presence and location of metastases.**

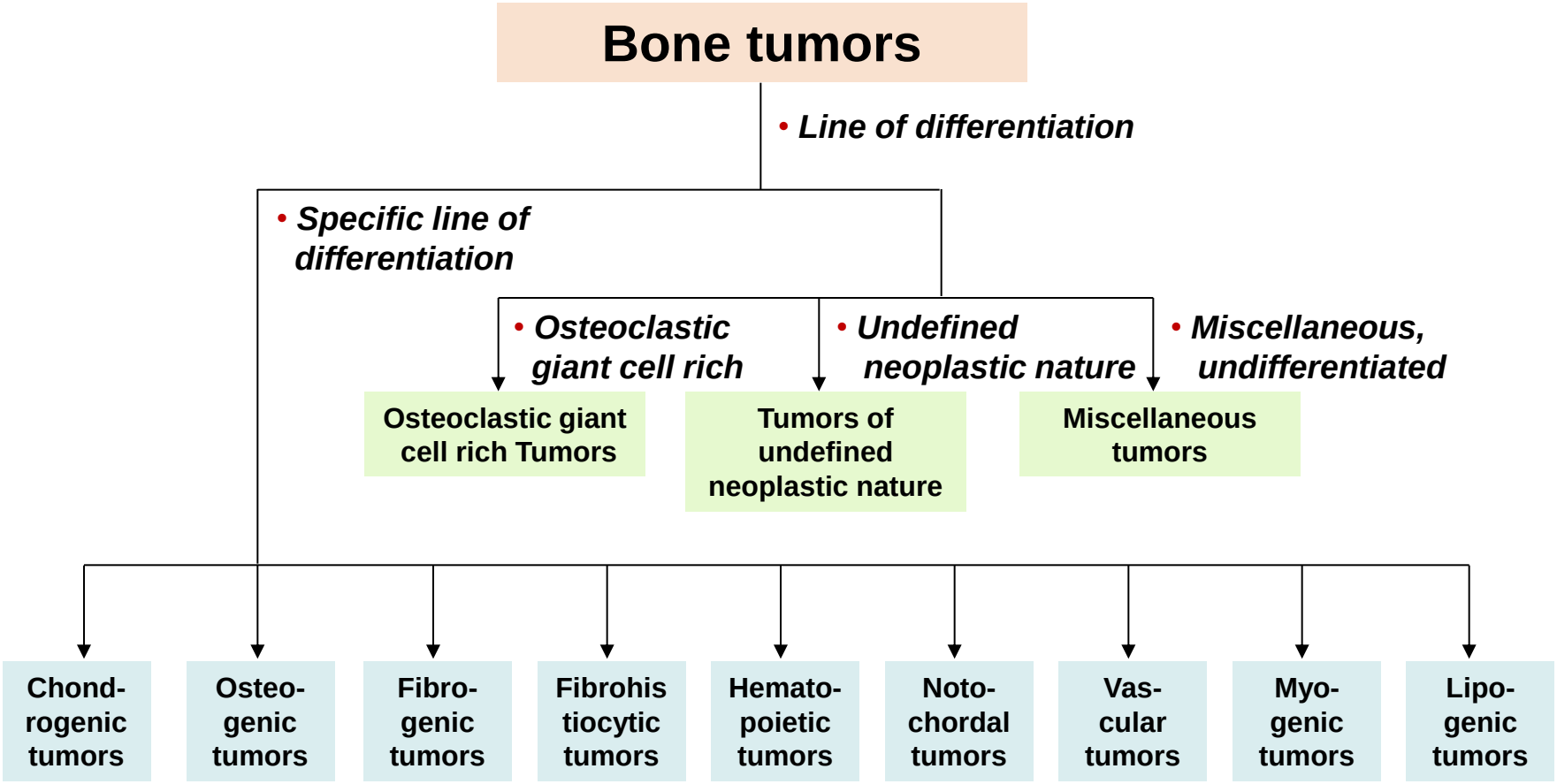
(AJCC 8th ed., 2017)

Primary bone cancers

- **Cancers staged using this staging system**
  - Osteosarcoma, chondrosarcoma, Ewing sarcoma, spindle cell sarcoma, hemangioendothelioma, angiosarcoma, fibrosarcoma/myofibroid sarcoma, chordoma, adamantinoma, and other cancers arising in the bone

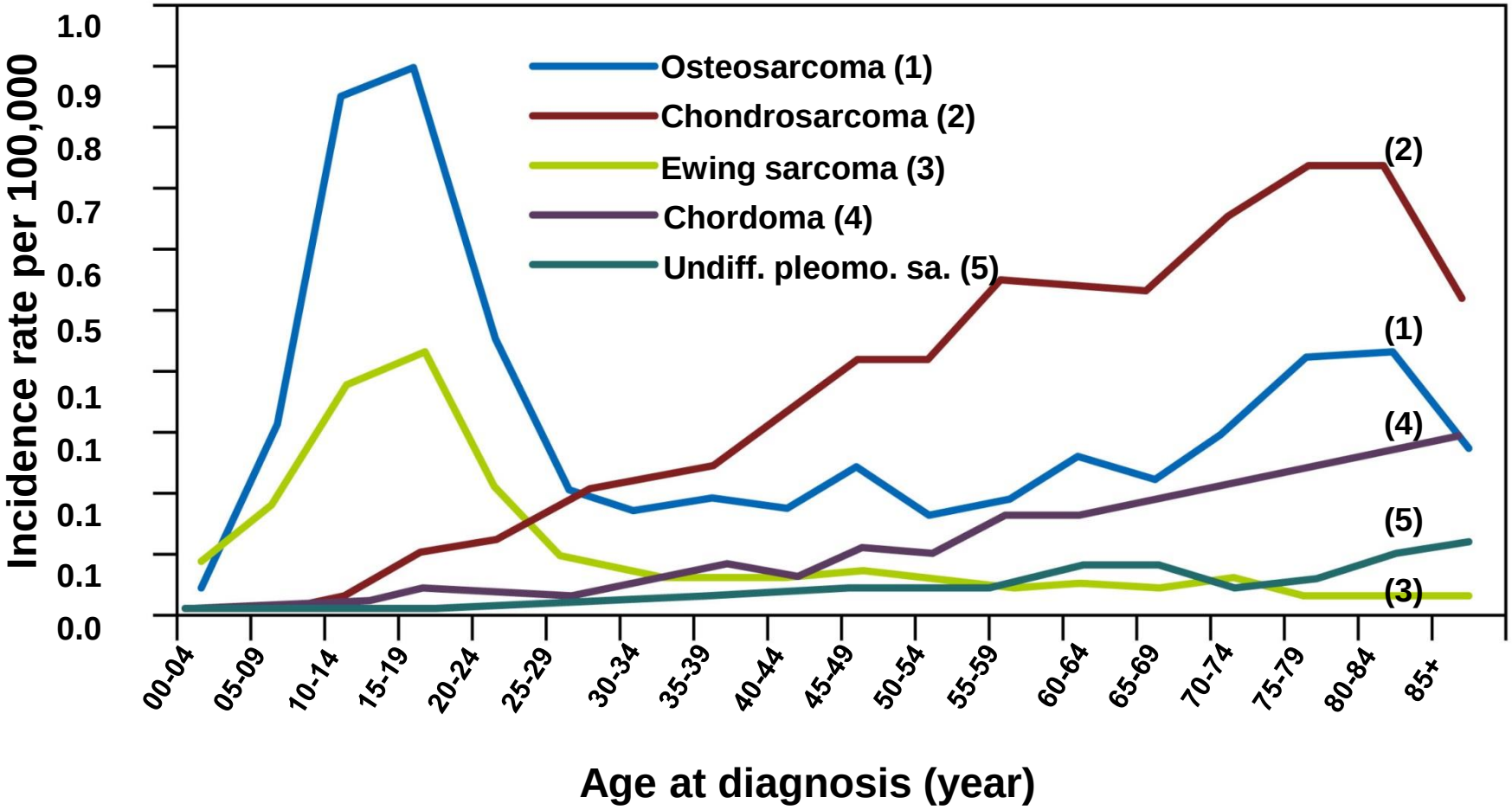
- **Cancers not staged using this staging system**
  - Primary malignant lymphoma
  - Multiple myeloma

WHO classification of bone tumors



(WHO classification, 2013)

Age-specific incidence rates in bone sarcomas



(WHO classification, 2013)

Summary of changes: bone

Change	Details of change
Definitions of AJCC TNM	**Pelvis** and **spine** each have a separate and distinct TNM classification but not a separate stage grouping. Level of evidence: III.
AJCC prognostic stage groups	**Stage III** is reserved for G2 and G3. Level of evidence: III.
Histologic grade (G)	**G4** designation has been eliminated (G1, low grade; G2 and G3, high grade). Level of evidence: III.

Primary sites, regional lymph node, metastatic sites: bone sarcomas

- **Primary sites**

  Site groups of bone sarcomas

  - Appendicular skeleton, trunk, skull, and facial bones
  - Spine
  - Pelvis

- **Regional lymph nodes**

  Regional lymph node metastases from primary bone tumors are extremely rare.

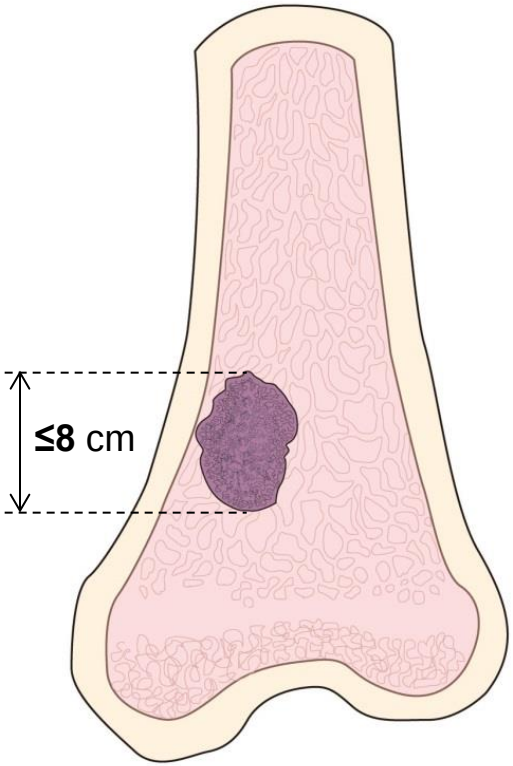
- **Metastatic sites**

  - Pulmonary metastases are the most common site for all bone sarcomas.

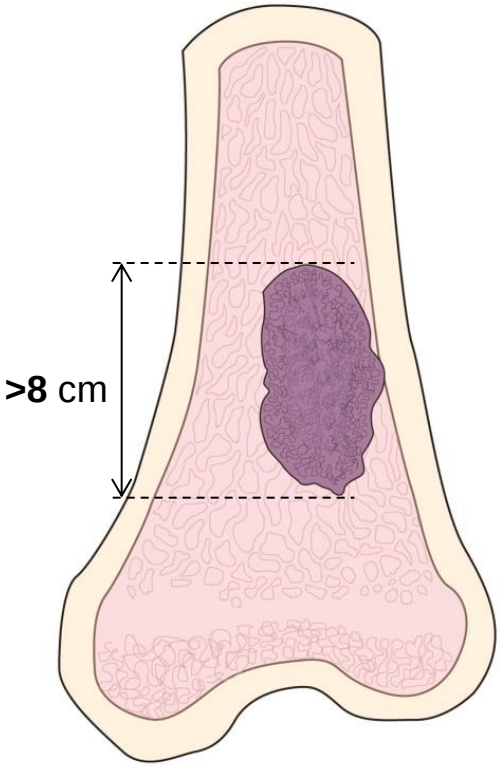
TNM classification: appendicular skeleton, trunk, skull, and facial bone

7th AJCC cancer staging		8th AJCC cancer staging	
T - Primary tumor		**T - Primary tumor**	
TX	Primary tumor cannot be assessed	**TX**	Primary tumor cannot be assessed
T0	No evidence of primary tumor	**T0**	No evidence of primary tumor
T1	Tumor **≤8 cm** in greatest dimension	**T1**	Tumor **≤8** cm in greatest dimension
T2	Tumor **>8 cm** in greatest dimension	**T2**	Tumor **>8** cm in greatest dimension
T3	Discontinuous tumors in the primary bone site	**T3**	Discontinuous tumors in the primary bone site
N - Regional lymph node		**N - Regional lymph node**	
NX	Regional lymph nodes cannot be assessed	**NX**	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis	**N0**	No regional lymph node metastasis
N1	Regional lymph node metastasis	**N1**	Regional lymph node metastasis
M - Distant metastasis		**M - Distant metastasis**	
M0	No distant metastasis	**M0**	No distant metastasis
M1 **M1a** **M1b**	Distant metastasis Lung Other distant metastasis	**M1** **M1a** **M1b**	Regional lymph node metastasis Lung Other distant metastasis

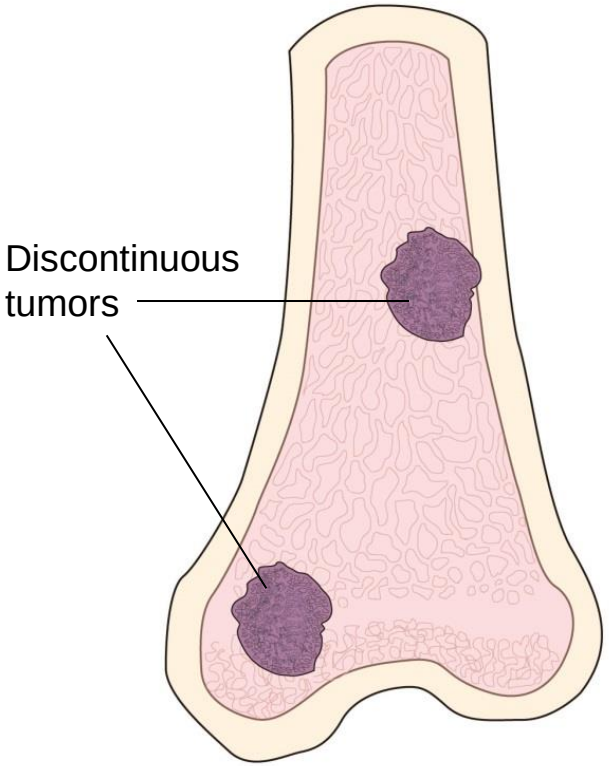
T classification: appendicular skeleton, trunk, skull, and facial bones



T1 is defined as tumor
8 cm or less
in greatest dimension



T2 is defined as tumor
more than 8 cm
in greatest dimension



T3 is defined as
discontinuous tumors in
the primary bone site

(AJCC 8th ed., 2017)

Regional lymph node (N) classification

- Because of the rarity of lymph node involvement in bone sarcomas, the designation **NX** may not be appropriate, and cases should be considered **cN0** unless clinical node involvement is clearly evident.

- For bone and soft tissue sarcoma, **cN0** may be used to assign the pathological stage group - that is, pT1cN0cM0.

- A case in which the lymph nodes were microscopically proven to not contain tumor is categorized as pathologically pN0 (**pN0)**.

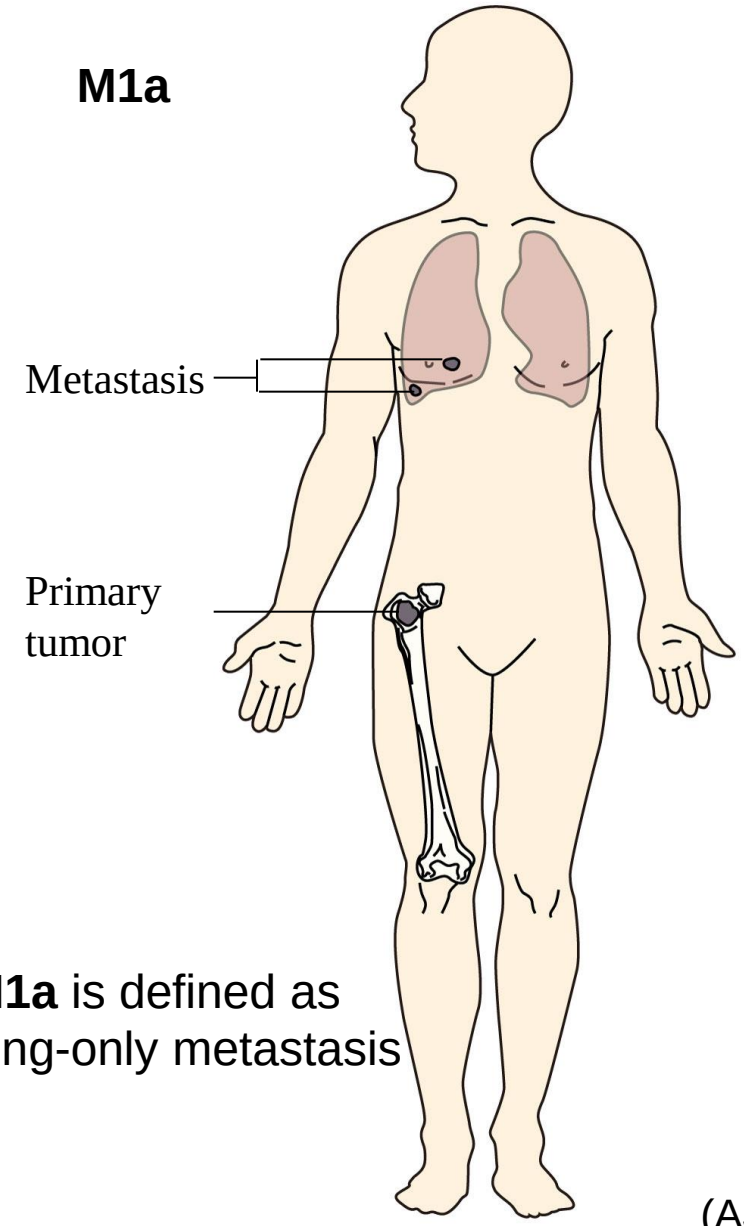
Distant metastasis (M) classification

- If there are no symptoms or signs of distant metastasis, the case is classified as clinically M0 (**cM0**).

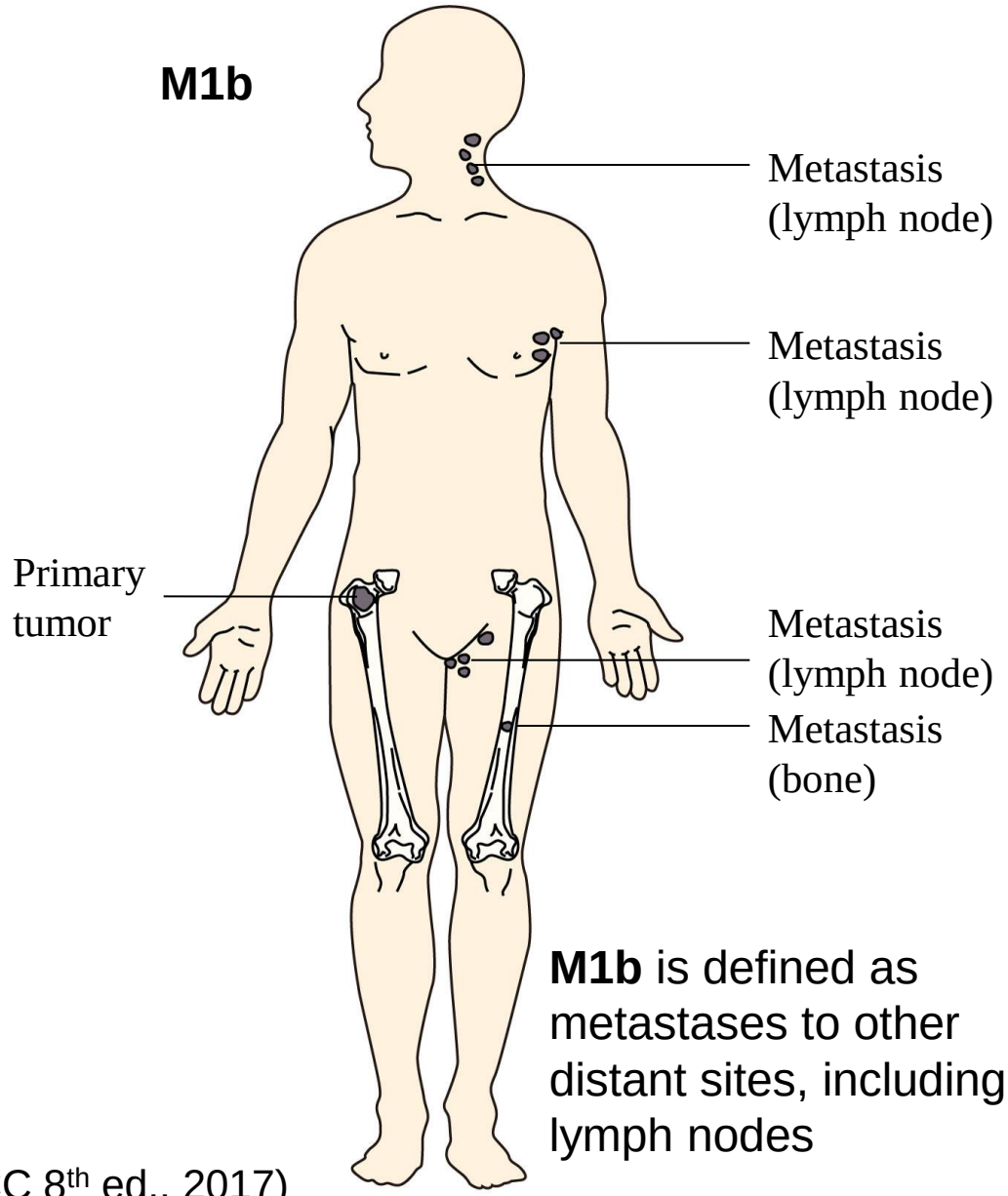
- Patients with clinical evidence of distant metastases by history, physical examination, imaging studies, or invasive procedures, but without microscopic evidence of the presumed distant metastases, are categorized as clinically cM1 (**cM1**).

- Patients in whom there is microscopic evidence confirming distant metastatic disease are categorized as pathologically pM1 (**pM1**).

Distant metastasis (M) classification



M1a is defined as lung-only metastasis



M1b is defined as metastases to other distant sites, including lymph nodes

(AJCC 8th ed., 2017)

Histologic grade: bone sarcomas

7th AJCC cancer staging		8th AJCC cancer staging	
Histologic grade		**Histologic grade**	
GX	Primary tumor cannot be assessed	**GX**	Grade cannot be assessed
G1	Well differentiated, low grade	**G1**	Well differentiated, **low grade**
G2	Moderately differentiated, low grade		
G3	Poorly differentiated, high grade	**G2**	Moderately differentiated, **high grade**
G4	Undifferentiated	**G3**	Poorly differentiated, **high grade**

NOTE: The grading depends on cellularity, cytologic features of tumor cells (pleomorphism), mitotic activity, and necrosis.

Some histologic types determine histologic grade

Grade	Histologic types
Grade 1	• Parosteal osteosarcoma • Grade I chondrosarcoma • Clear cell chondrosarcoma • Low-grade central osteosarcoma
Grade 2	• Periosteal osteosarcoma • Grade II chondrosarcoma • Classic adamantinoma • Chordoma
Grade 3	• Osteosarcoma (conventional, telangiectatic, small cell, secondary, high-grade surface) • Undifferentiated high-grade pleomorphic sarcoma • Ewing sarcoma • Grade III chondrosarcoma • Dedifferentiated chondrosarcoma • Mesenchymal chondrosarcoma • Dedifferentiated chordoma • Malignancy in giant cell tumor

(WHO classification, 2013)

AJCC prognostic stage groups

- **Appendicular skeleton, trunk, skull, and facial bones**

When T is ..	Any N is ..	And M is ..	And grade is ..	Then stage group is ..
T1	N0	M0	G1 or GX	**IA**
T2	N0	M0	G1 or GX	**IB**
T3	N0	M0	G1 or GX	**IB**
T1	N0	M0	G2 or G3	**IIA**
T2	N0	M0	G2 or G3	**IIB**
T3	N0	M0	G2 or G3	**III**
Any T	N0	M1a	Any G	**IVA**
Any T	N1	Any M	Any G	**IVB**
Any T	Any N	M1b	Any G	**IVB**

NOTE: There are no AJCC prognostic stage groupings in **spine and pelvis**.

Case 1: appendicular skeleton

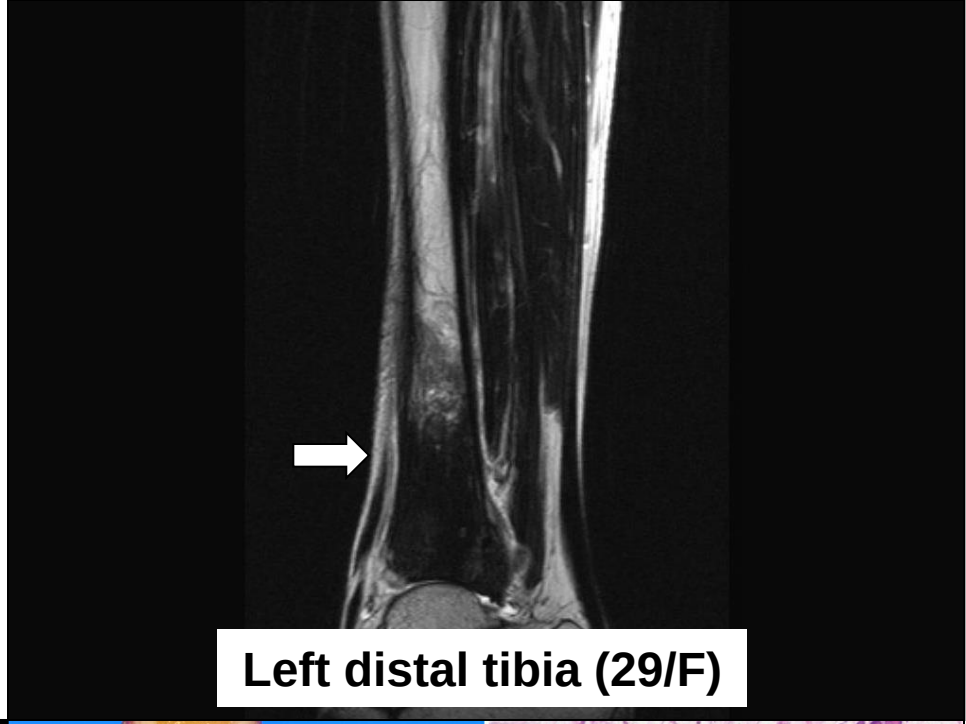
Dx: Conventional osteosarcoma, osteoblastic type

Postneoadjuvant pathological TNM (ypTNM)

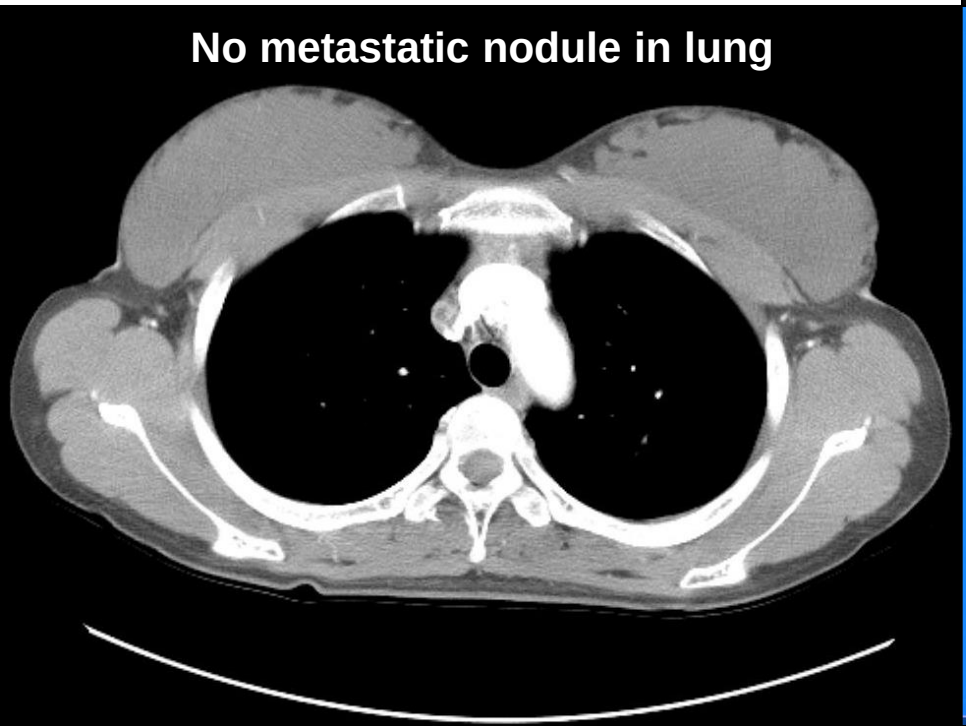
- Primary tumor (T): **ypT1** *(≤8 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade: **G3**

Stage group

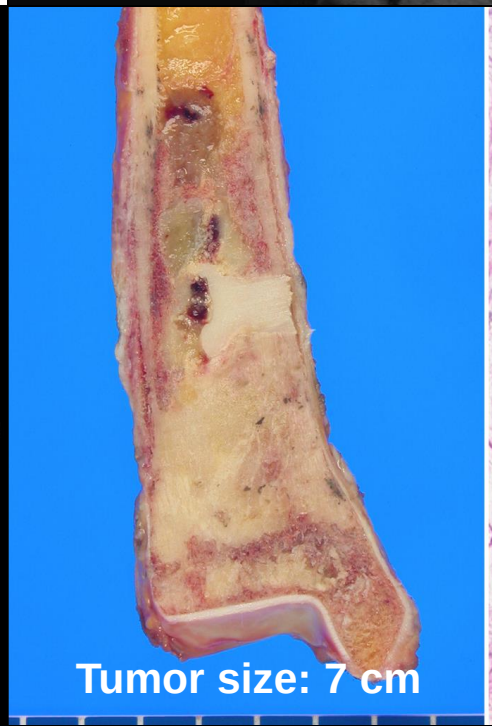
- Stage **IIA**



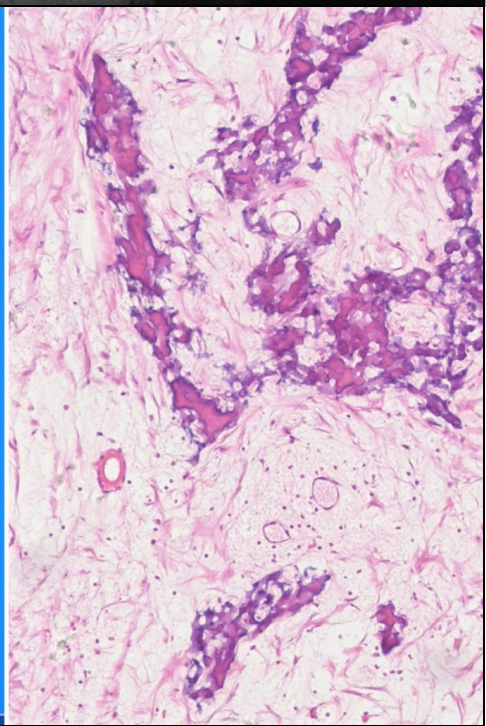
Left distal tibia (29/F)



No metastatic nodule in lung



Tumor size: 7 cm



Case 2: appendicular skeleton

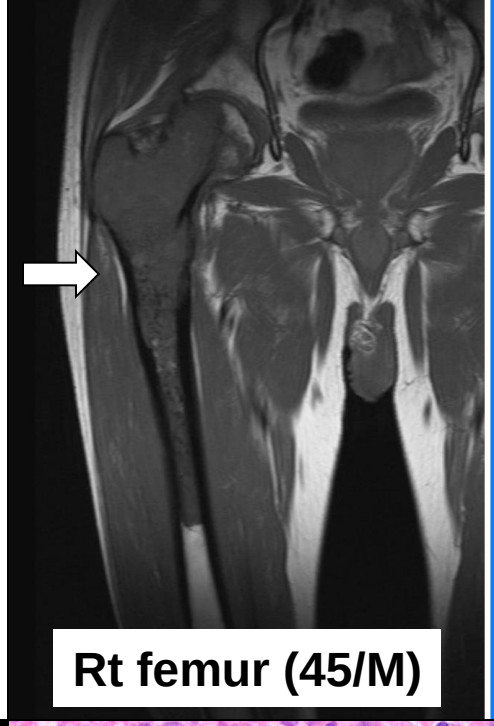
Dx: Dedifferentiated chondrosa.

Pathological TNM (pTNM)

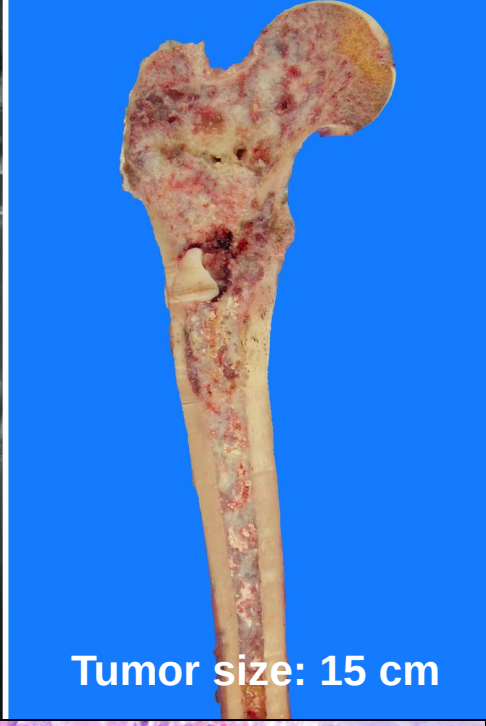
- Primary tumor (T): **pT2** *(>8 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM1a**
- Histologic grade: **G3**

Stage group

- Stage **IVA**

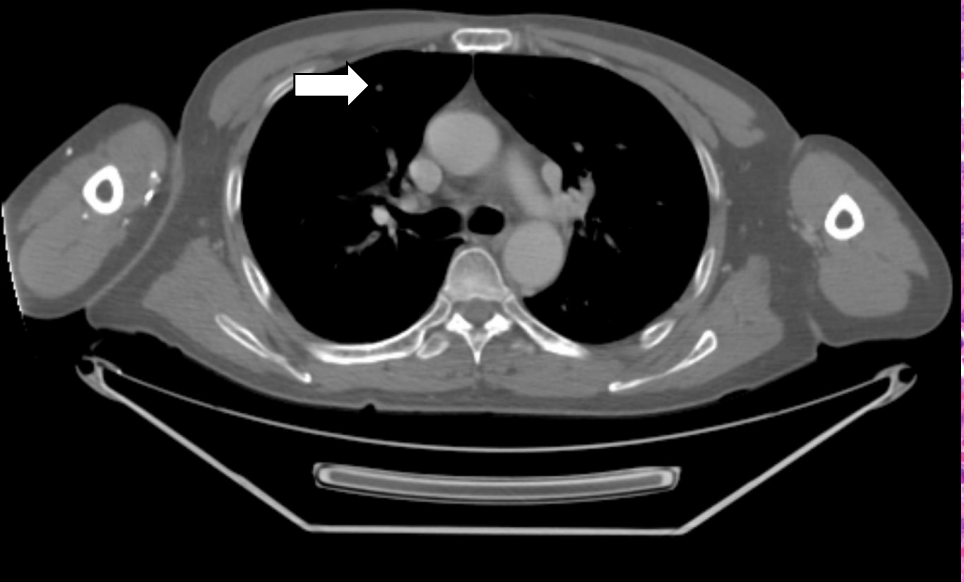


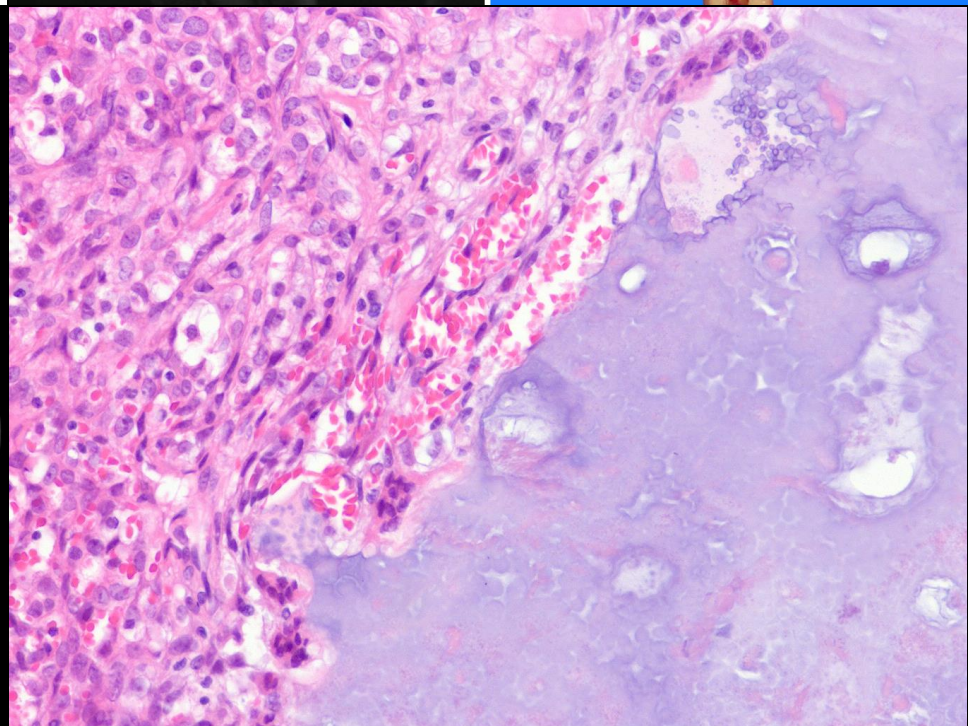
Rt femur (45/M)



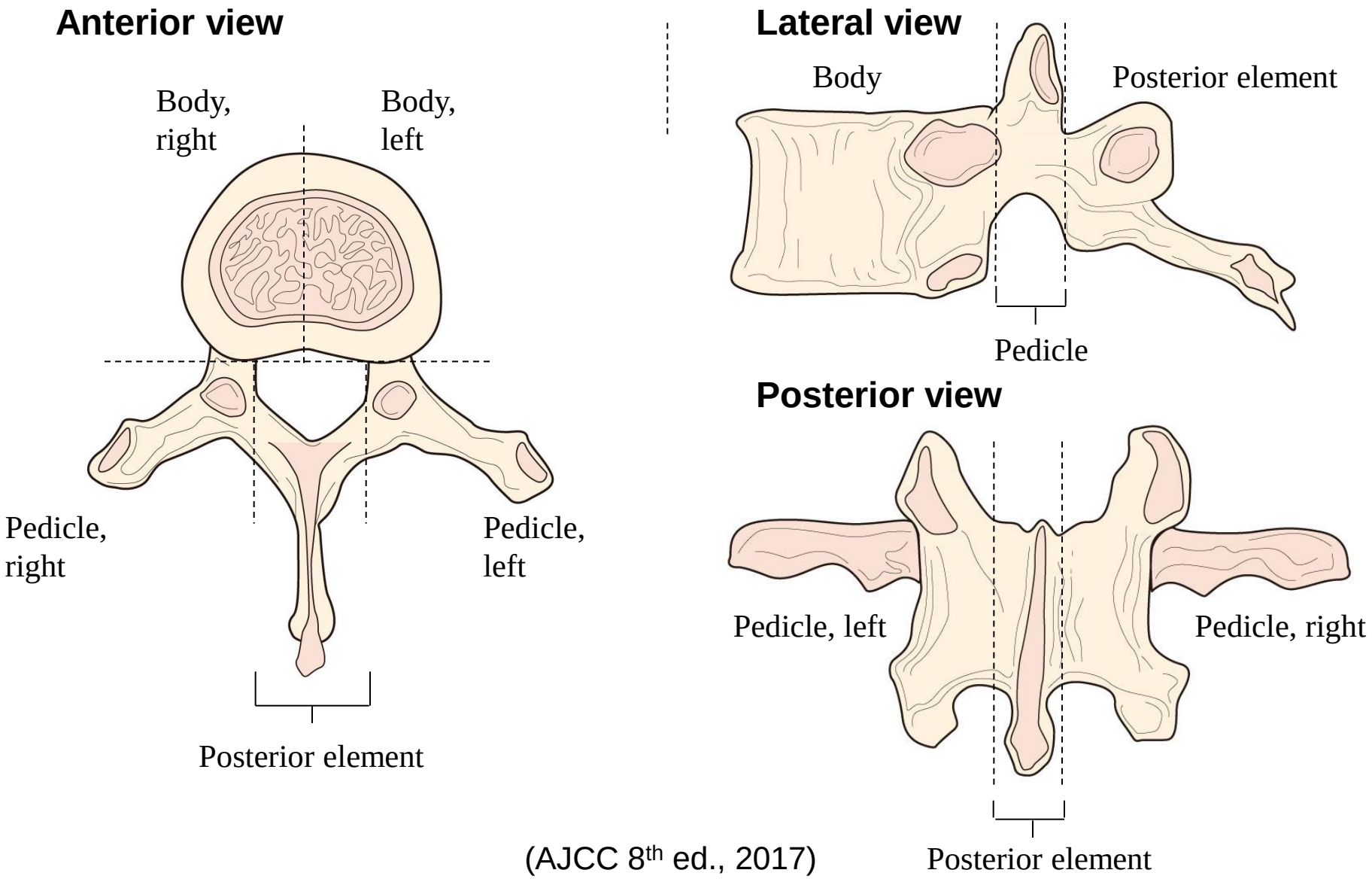
Tumor size: 15 cm

Metastatic nodule in lung





For tumors occurring in the spine, T is now primarily defined by the number of spinal segments involved or involvement of the spinal canal/great vessel.



(AJCC 8th ed., 2017)

Definition of primary tumor (T): spine

T category	T criteria
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Tumor confined to **one** vertebral segment or **two** adjacent vertebral segments
T2	Tumor confined to **three** adjacent vertebral segments
T3	Tumor confined to **four or more** adjacent vertebral segments, or any nonadjacent vertebral segments
T4 **T4a** **T4b**	Extension into the spinal canal or great vessels Extension into the **spinal canal** Evidence of **gross vascular invasion** or **tumor thrombus** in the great vessels

Case 3: spine

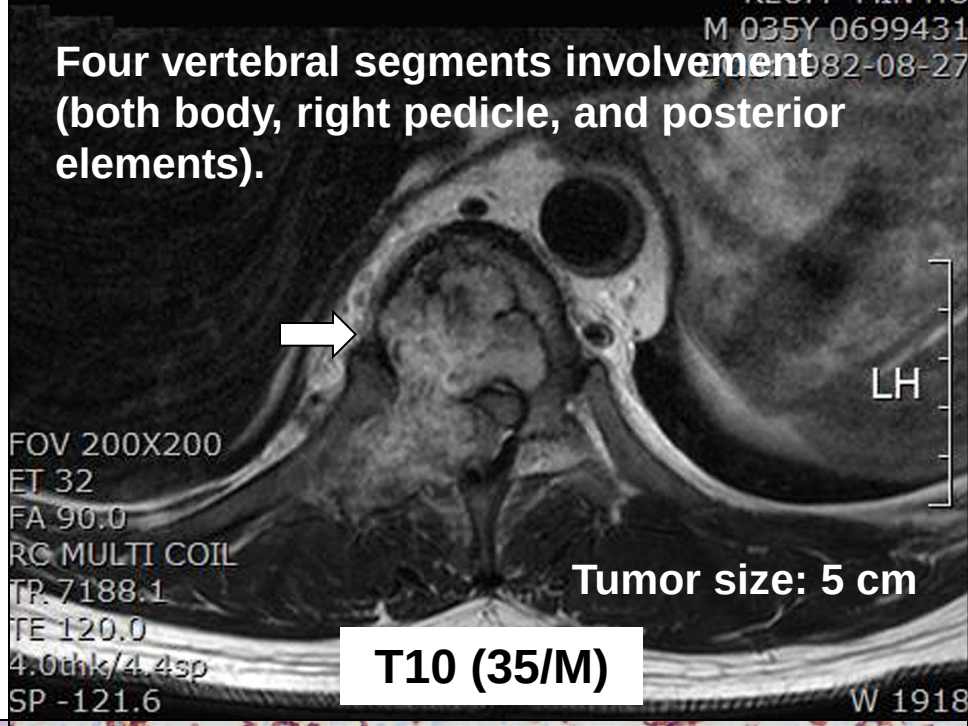
Dx: Epithelioid hemangioendothelioma

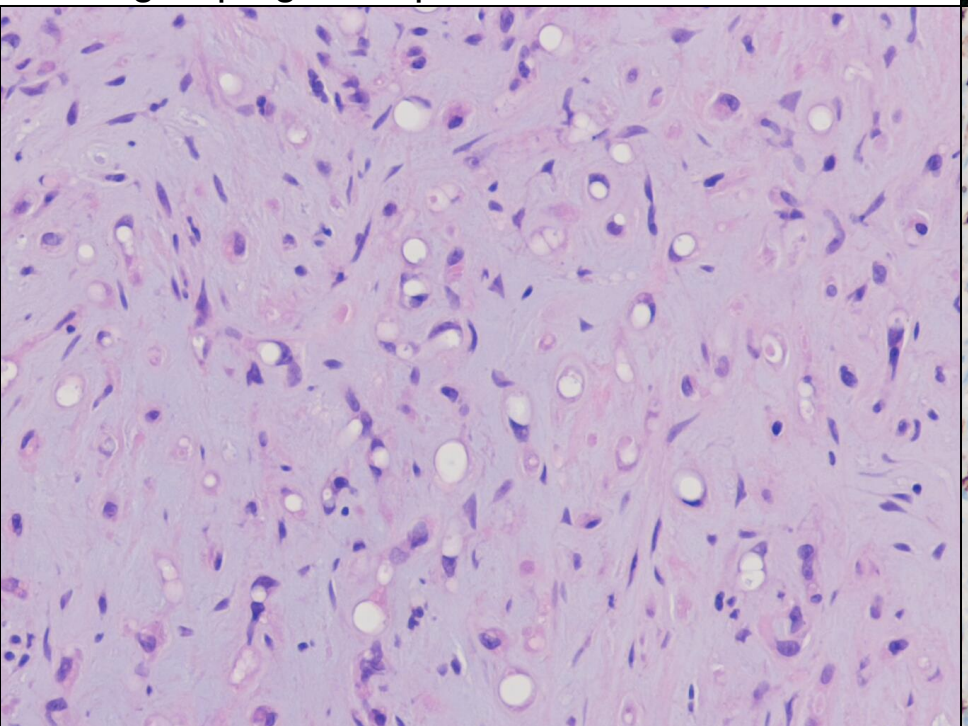
Pathological TNM (pTNM)

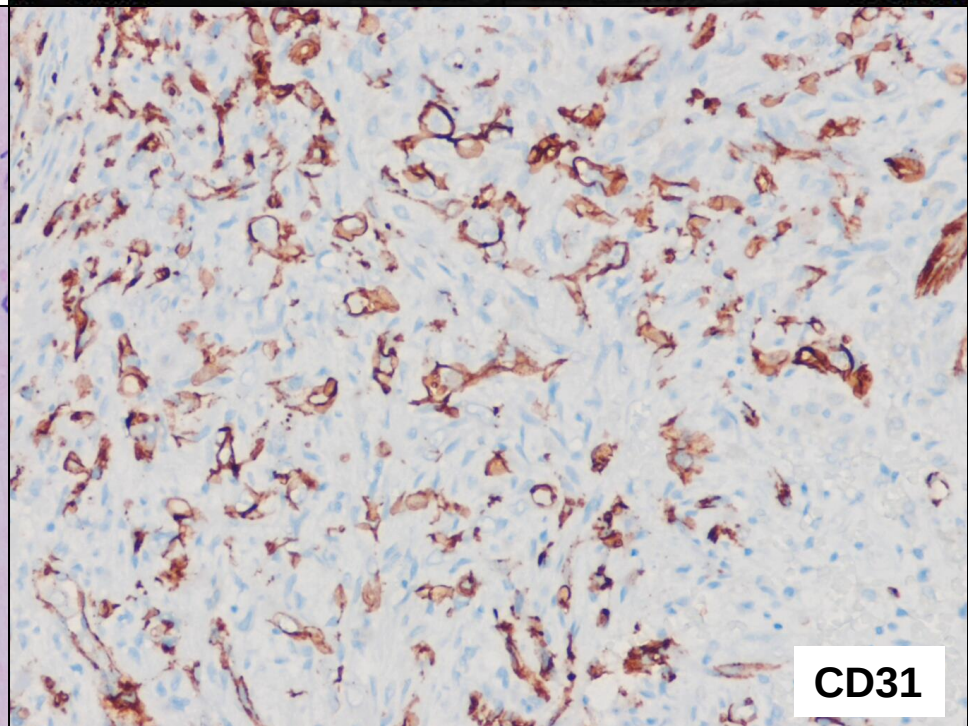
- Primary tumor (T): **pT3** *(involvement of four vertebral segments)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade: **G1**

Stage group*

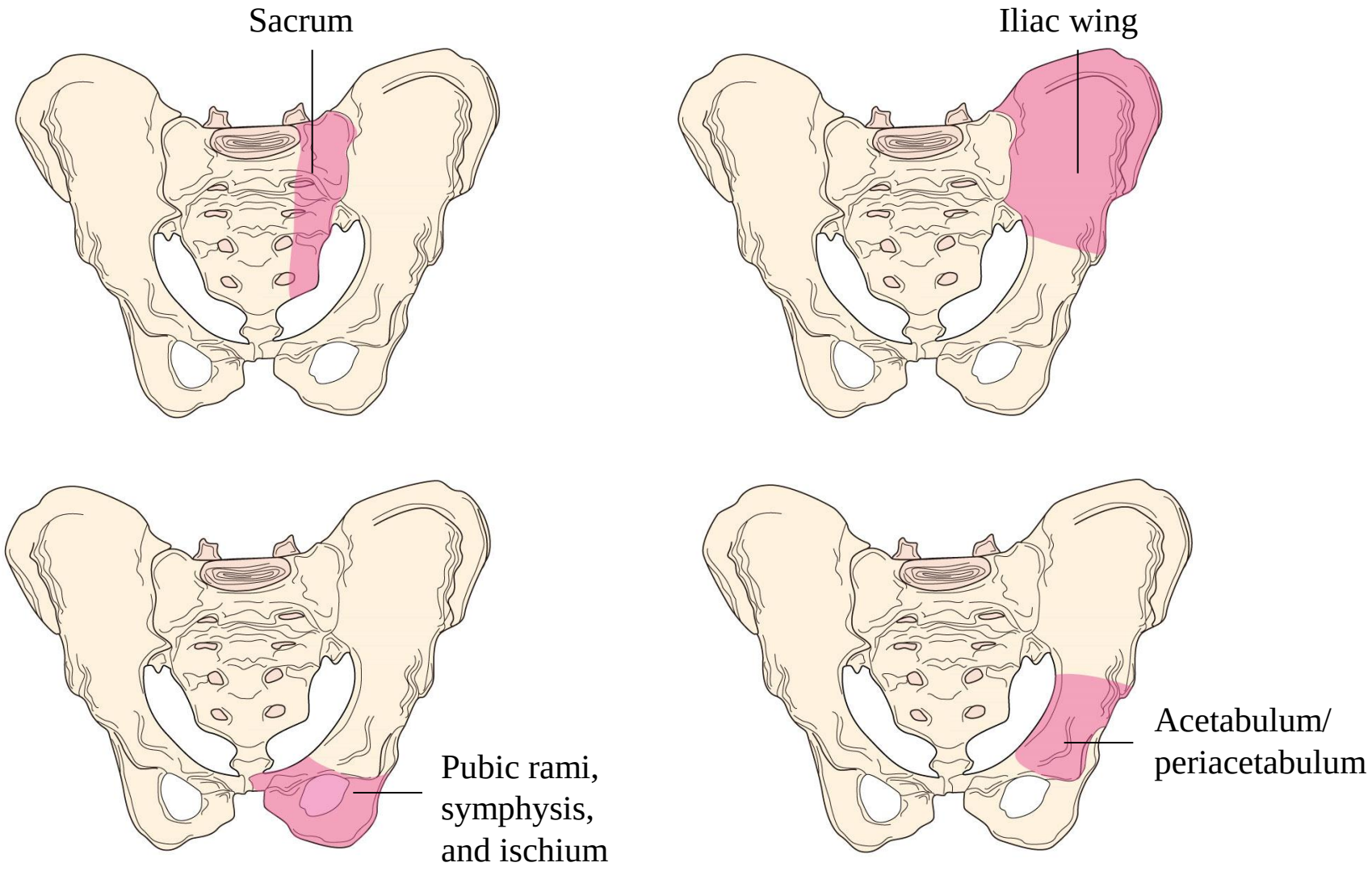
* There are no AJCC prognostic stage groupings for spine sarcomas.







For tumors occurring in the pelvis, T is now defined primarily by the number of pelvic segments involved, the presence of extraosseous extension, or involvement of the pelvic vessels.



(AJCC 8th ed., 2017)

Definition of primary tumor (T): pelvis

T category	T criteria
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1 **T1a** **T1b**	Tumor confined **to one** pelvic segment with no extraosseous extension Tumor **≤8 cm** in greatest dimension Tumor **>8 cm** in greatest dimension
T2 **T2a** **T2b**	Tumor confined to **one** pelvic segment with **extraosseous extension** or **two** segments without extraosseous extension Tumor **≤8 cm** in greatest dimension Tumor **>8 cm** in greatest dimension
T3 **T3a** **T3b**	Tumor spanning **two** pelvic segments **with extraosseous extension** Tumor **≤8 cm** in greatest dimension Tumor **>8 cm** in greatest dimension
T4 **T4a** **T4b**	Tumor spanning **three** pelvic segments or crossing the sacroiliac joint Tumor involves **sacroiliac joint** and extends medial to the **sacral neuroforamen** Tumor encasement of **external iliac vessels** or presence of gross tumor thrombus in **major pelvic vessels**

Case 4: pelvis

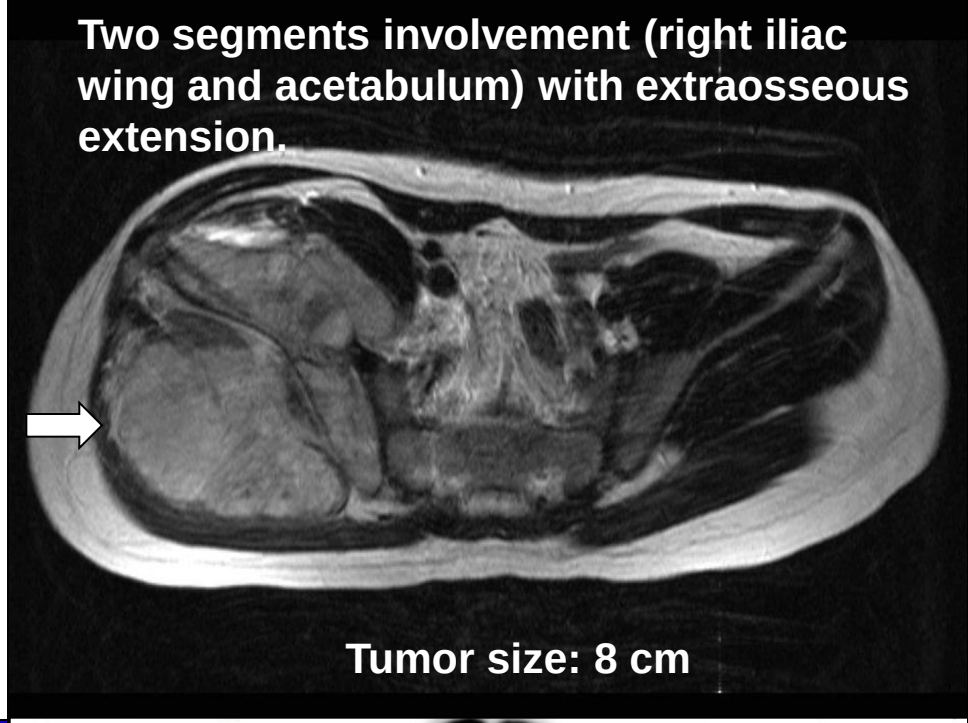
Dx: Ewing sarcoma

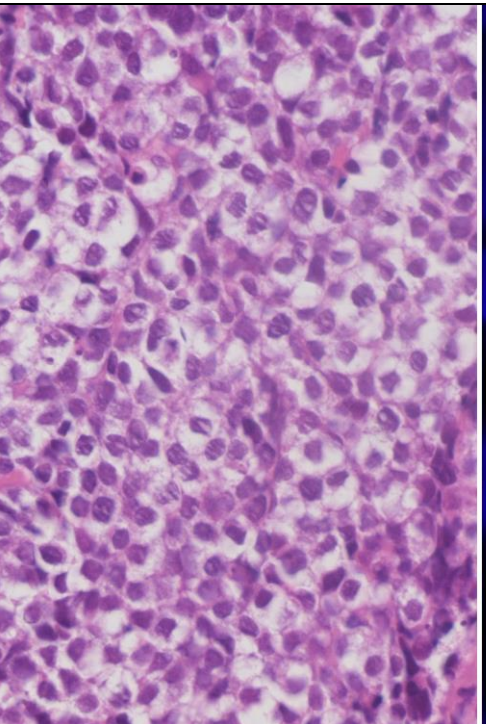
Clinical TNM (cTNM)

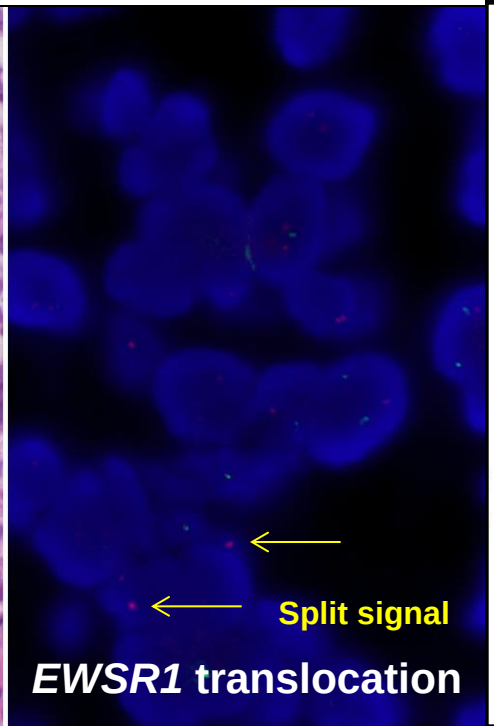
- Primary tumor (T): **pT3a** *(involvement of two pelvic segments with extraosseous extension, ≤ 8 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade: **G3**

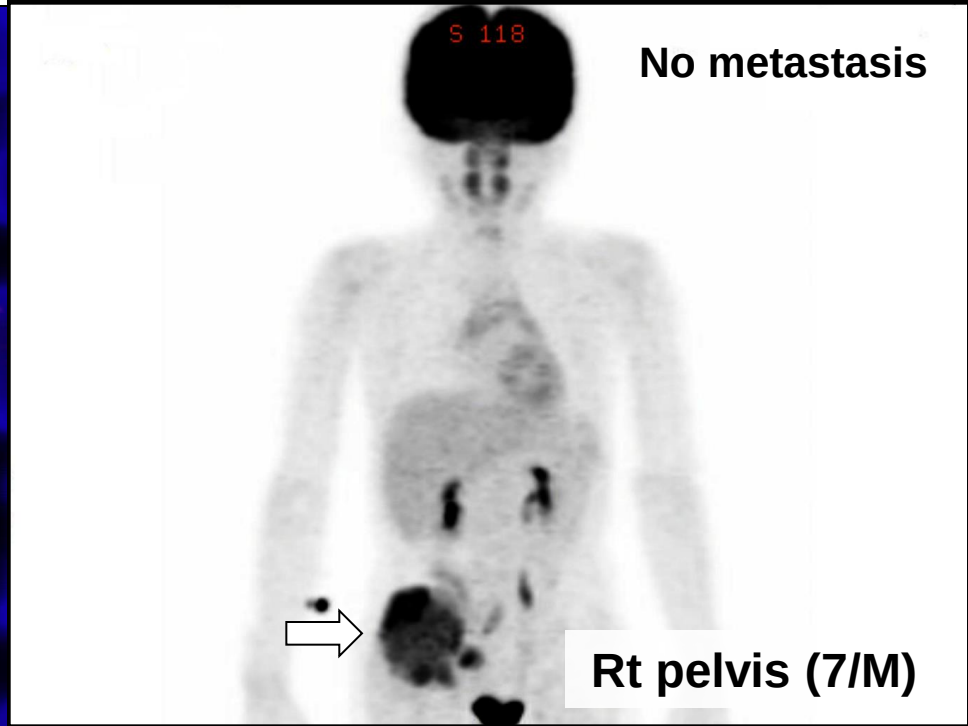
Stage group*

* There are no AJCC prognostic stage groupings for pelvis sarcomas.









Checklist for standard reporting of bone biopsy specimen defined by CAP cancer protocol

Procedure
___ Core needle biopsy
___ Curettage
___ Excisional biopsy
___ Other (specify):
_____
___ Not specified

Tumor Site
___ Appendicular skeleton (specify bone, if known): _____
___ Spine (specify bone, if known):
_____
___ Pelvis (specify bone, if known):
_____
___ Not specified

Tumor Location and Extent (select all that apply)
___ Epiphysis or apophysis
___ Metaphysis
___ Diaphysis
___ Cortex
___ Medullary cavity
___ Surface
___ Tumor involves joint
___ Tumor extension into soft tissue
___ Cannot be determined

Histologic Type (World Health Organization [WHO] classification of bone tumors)
Specify: _____
___ Cannot be determined

Mitotic Rate
Specify: ___ /10 high-power fields (HPF)
(1 HPF x 400 = 0.1734 mm^2; X40 objective; most proliferative area)

Necrosis
___ Not identified
___ Present
Extent: ___%
___ Cannot be determined

Histologic Grade
___ G1: Well differentiated, low grade
___ G2: Moderately differentiated, high grade
___ G3: Poorly differentiated, high grade
___ GX: Cannot be assessed
___ Not applicable

Lymphovascular Invasion
___ Not identified
___ Present
___ Cannot be determined

Additional Pathologic Findings
Specify: _____

Ancillary Studies (required only if applicable)

Immunohistochemistry (specify):
_____
___ Not performed

Cytogenetics (specify): _____
___ Not performed

Molecular Pathology (specify)
_____
___ Not performed

Radiographic Findings
Specify: _____
___ Not available

Comment(s)

* These specify the elements necessary for the pathologists to report the extent and characteristics of cancer specimen (http://www.cap.org).

(College of American Pathologists, Version: Bone 4.0.0.0, June 2017)

Checklist for standard reporting of bone resection specimen defined by CAP cancer protocol (1)

Procedure
___ Intralesional resection
___ Marginal resection
___ Segmental/wide resection
___ Radical resection
___ Other (specify):
_____
___ Not specified

Tumor Site
___ Appendicular skeleton (specify bone, if known): _____
___ Spine (specify bone, if known):
_____
___ Pelvis (specify bone, if known):
_____
___ Not specified

Tumor Location and Extent (select all that apply)
___ Epiphysis or apophysis
___ Metaphysis
___ Diaphysis
___ Cortical
___ Medullary cavity
___ Surface
___ Tumor involves joint
___ Tumor extension into soft tissue
___ Cannot be determined

Tumor Size
Greatest dimension (centimeters): ___ cm
+ Additional dimensions (centimeters):
___ x ___ cm
___ Cannot be determined
___ Multifocal tumor/discontinuous tumor at primary site (skip metastasis)

Mitotic Rate
Specify: ___ /10 high-power fields (HPF)
(1 HPF x 400 = 0.1734 mm^2; X40 objective; most proliferative area)

Necrosis (macroscopic and microscopic)
___ Not Identified
___ Present
Extent: ___%

Histologic Grade
___ G1: Well differentiated, low grade
___ G2: Moderately differentiated, high grade
___ G3: Poorly differentiated, high grade
___ GX: Cannot be assessed
___ Not applicable

Margins
___ Cannot be assessed
___ Uninvolved by sarcoma
Distance of sarcoma from closest margin (centimeters): ___ cm
Specify margin (if known):
_____
___ Involved by sarcoma
Specify margin(s) (if known):
_____

Lymphovascular Invasion
___ Not identified
___ Present
___ Cannot be determined

Regional Lymph Nodes
___ No lymph nodes submitted or found

Lymph Node Examination (required only if lymph nodes are present in the specimen)

Number of Lymph Nodes Involved: _____
___ Number cannot be determined (explain):
_____

Number of Lymph Nodes Examined: _____
___ Number cannot be determined (explain):
_____

(College of American Pathologists, Version: Bone 4.0.0.0, June 2017)

Checklist for standard reporting of bone resection specimen defined by CAP cancer protocol (2)

Pathologic stage classification

TNM Descriptors (required only if applicable) (select all that apply)

___ m (multiple)
___ r (recurrent)
___ y (posttreatment)

Primary Tumor (pT)

Appendicular Skeleton, Trunk, Skull, and Facial Bones

___ pTX:Primary tumor cannot be assessed
___ pT0:No evidence of primary tumor
___ pT1:Tumor ≤8 cm in greatest dimension
___ pT2:Tumor > 8 cm in greatest dimension
___ pT3:Discontinuous tumors in the primary bone site

Spine

___ pTX:Primary tumor cannot be assessed
___ pT0:No evidence of primary tumor
___ pT1:Tumor confined to one vertebral segment or two adjacent vertebral segments
___ pT2:Tumor confined to three adjacent vertebral segments
___ pT3:Tumor confined to four or more adjacent vertebral segments, or any nonadjacent vertebral segments
___ pT4:Extension into the spinal canal or great vessels
___ pT4a:Extension into the spinal canal
___ pT4b:Evidence of gross vascular invasion or tumor thrombus in the great vessels

Pelvis

___ pTX:Primary tumor cannot be assessed
___ pT0:No evidence of primary tumor
___ pT1:Tumor confined to one pelvic segment with no extraosseous extension
___ pT1a:Tumor ≤8 cm in greatest dimension
___ pT1b:Tumor >8 cm in greatest dimension
___ pT2 Tumor confined to one pelvic segment with extraosseous extension or two segments without extraosseous extension
___ pT2a:Tumor ≤8 cm in greatest dimension
___ pT2b:Tumor >8 cm in greatest dimension
___ pT3:Tumor spanning two pelvic segments with extraosseous extension
___ pT3a:Tumor ≤8 cm in greatest dimension
___ pT3b:Tumor >8 cm in greatest dimension
___ pT4:Tumor spanning three pelvic segments or crossing the sacroiliac joint
___ pT4a:Tumor involves sacroiliac joint and extends medial to the sacral neuroforamen
___ pT4b:Tumor encasement of external iliac vessels or presence of gross tumor thrombus in major pelvic vessels

Regional Lymph Nodes (pN)

___ pNX:Regional lymph nodes cannot be assessed
___ pN0:No regional lymph node metastasis
___ pN1:Regional lymph node metastasis

[#] *Note: Because of the rarity of lymph node involvement in bone sarcomas, the designation NX may not be appropriate, and cases should be considered N0 unless clinical node involvement clearly is evident.*

Distant Metastasis (pM) (required only if confirmed pathologically in this case)

___ pM1a:Lung
___ pM1b:Metastasis involving distant sites other than lung
Specify site(s), if known: _____

Additional Pathologic Findings

Specify: _____

Ancillary Studies (required only if applicable)

Immunohistochemistry (specify): _____
___ Not performed

Cytogenetics (specify): _____
___ Not performed

Molecular Pathology (specify): _____
___ Not performed

Radiographic Findings

Specify: _____
___ Not available

Preresection Treatment (select all that apply)

___ No known preresection therapy
___ Chemotherapy performed
___ Radiation therapy performed
___ Therapy performed, type not specified
___ Not specified

Treatment Effect (select all that apply)

___ No known presurgical therapy
___ Not identified
___ Present
Specify percentage of necrotic tumor (compared with pretreatment biopsy, if available): _____%
___ Cannot be determined

Comment(s)

Contents

Summary of changes: soft tissue sarcomas (1)

Change	Details of change
Multiple chapters	A greater emphasis is placed on the **anatomic primary site** of the soft tissue sarcoma, which has implications for local recurrence and metastatic disease. Level of evidence: N/V.
Gastro-intestinal stromal tumor	GIST still has its own staging system and **remains unchanged** but is collected under sarcomas, as these are mesenchymal malignancies. Level of evidence: N/V.
New retro-peritoneal sarcoma staging system	More accurately reflects the **biology of this tumor** site; a validated nomogram may be used to help guide risk assessment in addition to traditional staging categories Level of evidence: I.

Summary of changes: soft tissue sarcomas (2)

Change	Details of change
New head and neck sarcoma staging system	Tumors are recognized at smaller sizes than those at other sites but have **higher risk on a size basis** than those of other sites. Provisional TNM criteria are provided to facilitate prospective data collection. Level of evidence: IV.
New visceral sa. staging system	There are **no superficial tumors** in this anatomic site. Level of evidence: IV.
Definition of primary tumor (T)	A **new size category** reflects the increased risk of metastasis as primary size increases. The **superficial-versus-deep distinction is less important** and has been eliminated. Level of evidence: II.

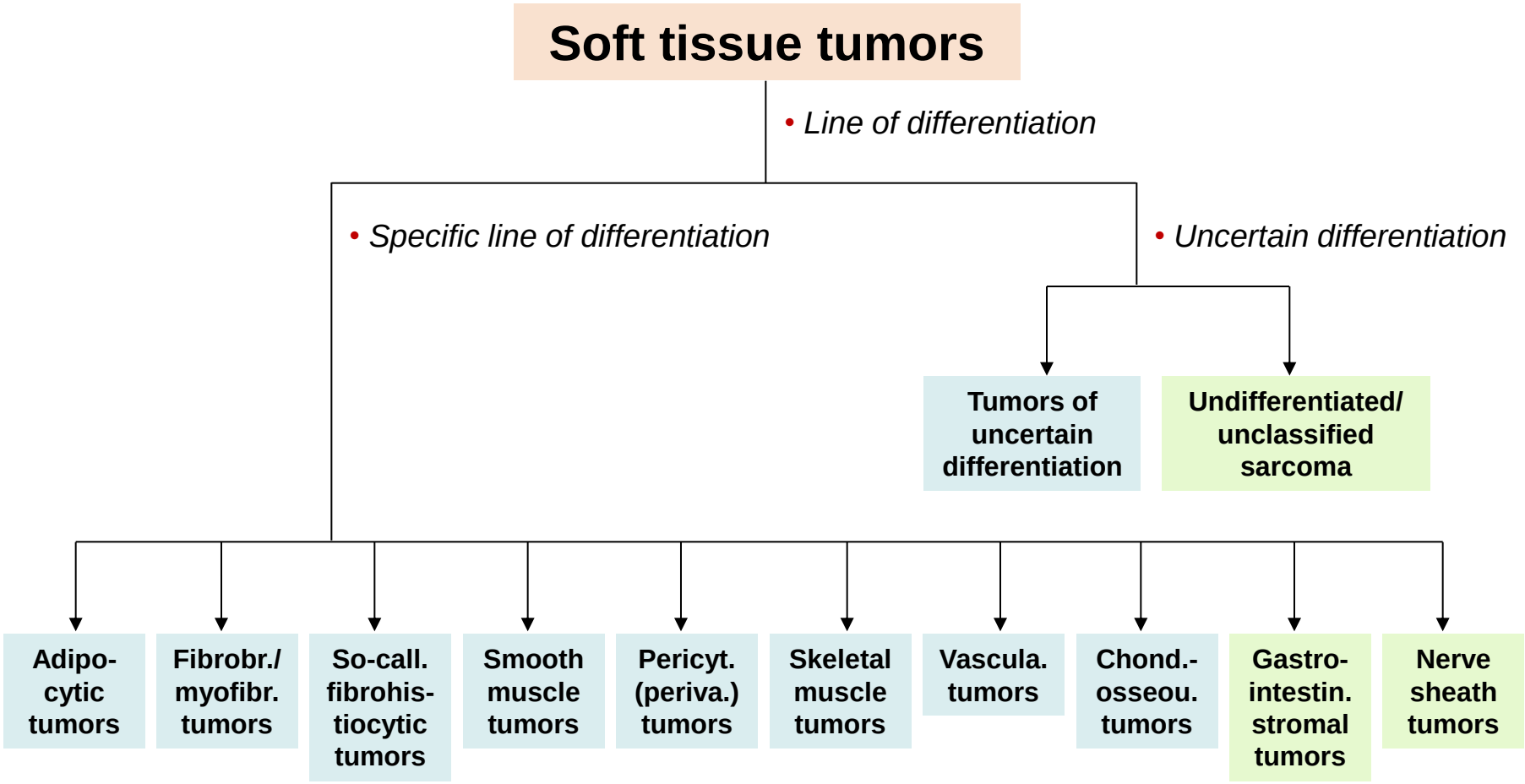
Summary of changes: soft tissue sarcomas (3)

Change	Details of change
Definition of regional lymph node (N)	**N1 disease** behaves similarly between stage III and stage IV disease and is captured as **stage IV** disease for simplicity. Level of evidence: II.
Unusual sites and histologies	Guidance is provided regarding **some unique histologies** and their biological behavior. Some sarcomas metastasize early, but patients may live with metastatic disease far longer than with other sarcoma histologies. Level of evidence: N/A.

Anatomic primary sites: soft tissue sarcomas

Anatomic primary sites	
(1) Extremity and trunk (Extremity: 40-50% of all sarcomas) (Trunk: 10% of all sarcomas) **(2) Retroperitoneum** (15% of all sarcomas)	Outcomes are **well characterized** and good predictive models exist for recurrence based on staging data.
(3) Head and neck (10% of all sarcomas) **(4) Abdomen and thoracic visceral sites** (15-20% of all sarcomas)	The available data are **more limited**, and the criteria presented herein will serve as a starting point and research tool for refining risk for these anatomic sites in future editions.

WHO classification of soft tissue tumors



(WHO classification, 2013)

Histologic grade: soft tissue sarcomas

- Soft tissue sarcomas constitute a family of **more than 50 different subtypes** of cancer, as well as lesions that are locally aggressive and only infrequently or never metastasize.

- **Histologic subtype**, **grade**, and **tumor size** are essential for staging system.

- The French Federation of Cancer Centers Sarcoma Group (**FNCLCC**) system is preferred over NCI system because of its ease of use/reproducibility and slightly superior performance.

Histopathological parameters in FNCLCC grading system

Tumor differentiation	
Score 1	Sarcomas closely resembling normal adult mesenchymal tissue and potentially difficult to distinguish from the counterpart benign tumor (e.g., well-differentiated liposarcoma, well-differentiated leiomyosarcoma)
Score 2	Sarcomas for which histologic typing is certain (e.g., myxoid liposa., myxofibrosa.)
Score 3	Embryonal and undifferentiated sa., synovial sa., sarcomas of doubtful type
Mitotic count	
Score 1	0-9 mitoses/10 HPF (high power fields)
Score 2	10-19 mitoses/10 HPF
Score 3	≥20 mitoses/10 HPF
Tumor necrosis	
Score 0	No necrosis
Score 1	<50% tumor necrosis
Score 2	≥50% tumor necrosis
Histologic grade (tumor differentiation + mitotic count + tumor necrosis)	
Grade 1	Total score 2,3
Grade 2	Total score 4,5
Grade 3	Total score 6,7,8

Low grade (= FNCLCC grade 1), **high grade** (= FNCLCC grade 2 and 3)

Tumor differentiation score according to histological type

Histologic type	Tumor differentiation
• Well differentiated liposarcoma • Well differentiated leiomyosarcoma	**Score 1**
• Myxoid liposarcoma • Conventional fibrosarcoma • Myxofibrosarcoma • Conventional MPNST • Conventional leiomyosarcoma • Conventional angiosarcoma	**Score 2**
• High grade myxoid (round cell) liposarcoma • Pleomorphic/dedifferentiated liposarcoma • Poorly differentiated/pleomorphic leiomyosarcoma • Poorly differentiated/epithelioid angiosarcoma • Poorly differentiated MPNST • Malignant Triton tumor • Synovial sarcoma • Rhabdomyosarcoma • Mesenchymal chondrosarcoma • Extraskeletal osteosarcoma • Extraskeletal Ewing sarcoma • Epithelioid sarcoma • Clear cell sarcoma • Alveolar soft part sarcoma • Malignant rhabdoid tumor • Undifferentiated (spindle cell and pleomorphic) sarcoma	**Score 3**

Lymph node metastasis: soft tissue sarcomas

- Involvement of regional lymph nodes by soft tissue sarcomas is uncommon in adults.

- Specific histologies in which regional lymph node metastatic disease is most commonly observed include **alveolar rhabdomyosarcoma, embryonal rhabdomyosarcoma, epithelioid sarcoma,** and **angiosarcoma.**

(AJCC 8th ed., 2017)

Metastatic sites: soft tissue sarcomas

- Metastatic sites for soft tissue sarcoma often depend on the original site of the primary lesion.

- For example, the most common site of metastatic disease for patients with **extremity sarcoma** is the **lung,** whereas **retroperitoneal and gastrointestinal sarcomas** often have **liver** as the first site of metastasis.

(AJCC 8th ed., 2017)

Contents

Soft tissue sarcomas: head and neck (1)

- **Sarcomas of the head and neck** comprise about 10% of soft tissue sarcomas overall and < 1% of head & neck malignancies.

- Four most **common groups**
  1. Neck (include. larynx/pharynx): liposa., MPNST, synovial sa.
  2. Scalp and facial skin: angiosa., DFSP
  3. Sinonasal tract: MPNST, angiosa., myxofibrosa., rhabdomyosa.
  4. Oral cavity: leiomyosa., rhabdomyosa.

- **Pediatric** population
  - Embryonal and alveolar rhabdomysoarcoma

Soft tissue sarcomas: head and neck (2)

- **Cancers staged using this staging system**
  - Soft tissue sarcomas of the head and neck

- **Cancers not staged using this staging system**
  - Sarcoma of orbit
  - Embryonal and alveolar rhabdomyosarcoma
  - Cutaneous angiosarcoma
  - Kaposi sarcoma
  - Dermatofibrosarcoma protuberans

Summary of changes: head and neck

Change	Details of change
New classification	This **classification** is being **introduced** for the first time because the previous classification developed for sarcomas elsewhere is not suited to this anatomic region. Level of evidence: N/A.
Definition of primary tumor (T)	A **new set** of **T** categories (**T1-T4**) has been created. Traditional T1 and T2 according to the 5 cm-breakpoint for soft tissue sarcoma have been eliminated in the head and neck. Level of evidence: IV.
Definition of regional lymph node (N)	Follow criteria used for extremity and trunk lesions. Level of evidence: IV.
Histologic grade (G)	Follow criteria used for extremity and trunk lesions Level of evidence: IV.

Definition of primary tumor (T): head and neck

7th AJCC cancer staging		8th AJCC cancer staging	
T - Primary tumor		**T - Primary tumor**	
TX	Primary tumor cannot be assessed	**TX**	Primary tumor cannot be assessed
T0	No evidence of primary tumor		
T1	Tumor **5 cm or less** in greatest dimension	**T1**	Tumor **≤2 cm**
T1a	Superficial tumor	**T2**	Tumor **>2** to **4 cm**
T1b	Deep tumor	**T3**	Tumor **>4 cm**
T2	Tumor **more than 5 cm** in greatest dimension	**T4**	Tumor with **invasion of adjoining structures**
T2a	Superficial tumor	**T4a**	Tumor with orbital invasion, skull base/dural invasion, invasion of central compartment viscera, involvement of facial skeleton, or invasion of pterygoid muscles
T2b	Deep tumor	**T4b**	Tumor with brain parenchymal invasion, carotid artery encasement, prevertebral muscle invasion, or central nervous system involvement via perineural spread

AJCC prognostic stage groups: head and neck

- This is a new classification that needs data collection before defining a stage grouping for head and neck sarcomas.

(AJCC 8th ed., 2017)

Case 5: head & neck

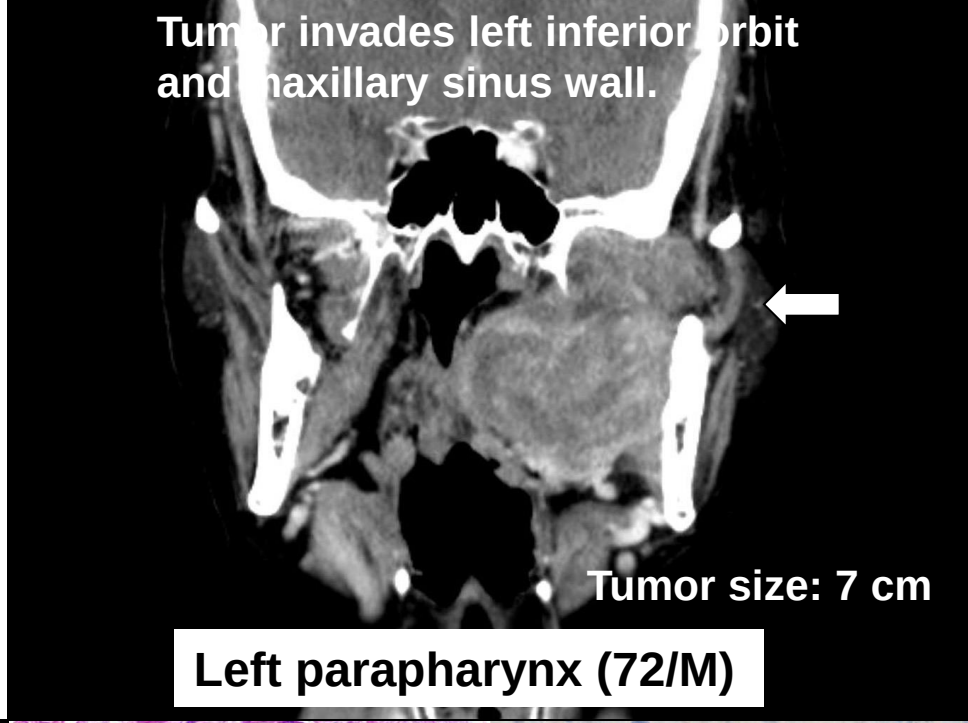
Dx: Dedif. solitary fibrous tumor

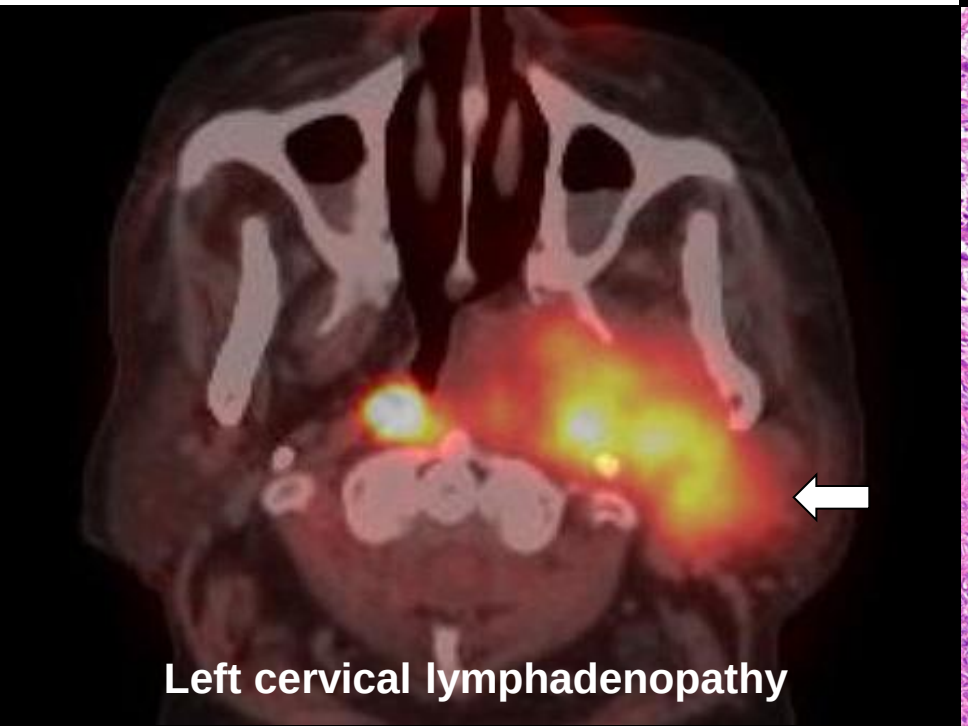
Pathological TNM (pTNM)

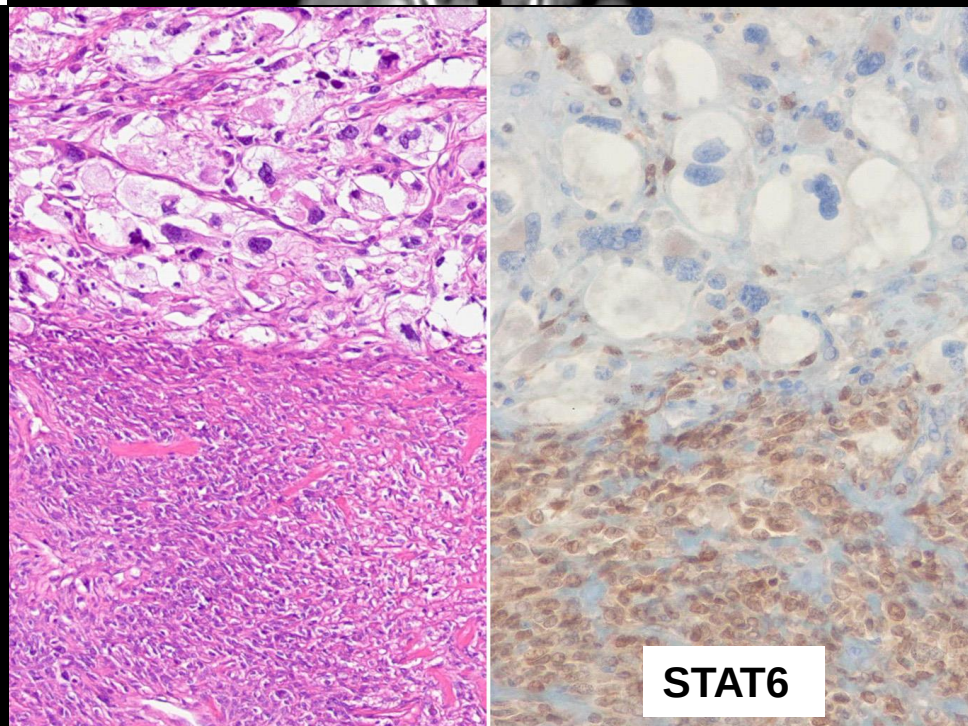
- Primary tumor (T): **pT4a** *(invasion of adjoining structures: orbit)*
- Regional lymph node (N): **cN1**
- Distant metastasis (M): **cM0**
- Histologic grade: **G3**

Stage group*

* Needs data collection before defining stage grouping for head & neck sarcoma.







Contents

Soft tissue sarcomas: trunk and extremities

- About **40-50%** of soft tissue sarcomas occur in the extremities, and about **10%** occurs in the trunk.
- **Tumor size** and **histologic grade** are essential for soft tissue sarcoma staging
- This staging system applies to all extremity and trunk soft tissue sarcomas **except desmoid tumors** and **Kaposi sarcoma**.
- The breast is a trunk site and often is collected as a separate anatomic site.

(AJCC 8th ed., 2017)

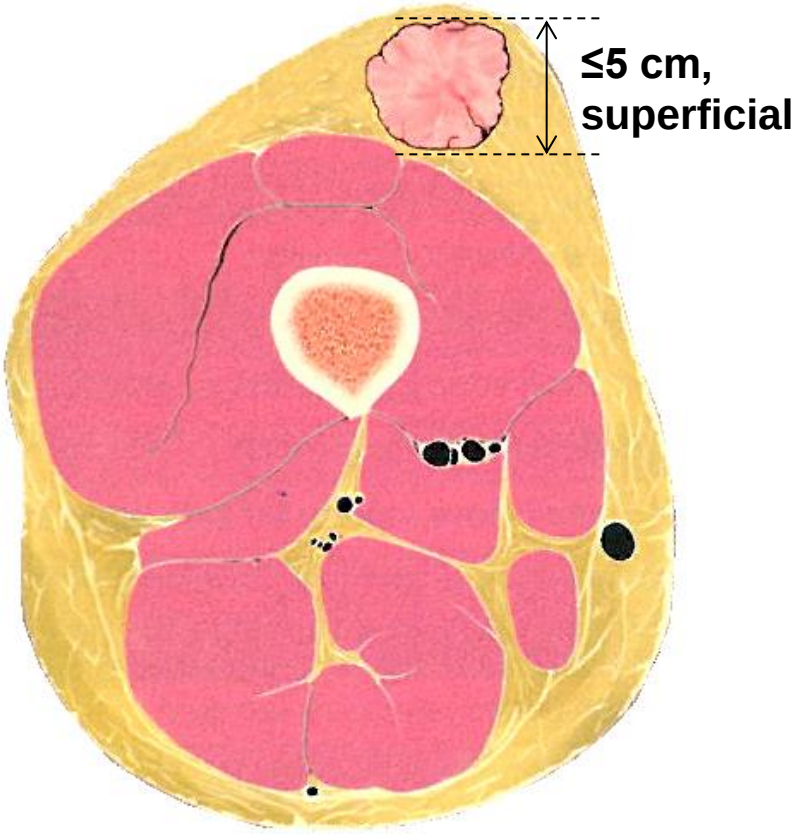
Summary of changes: trunk and extremities

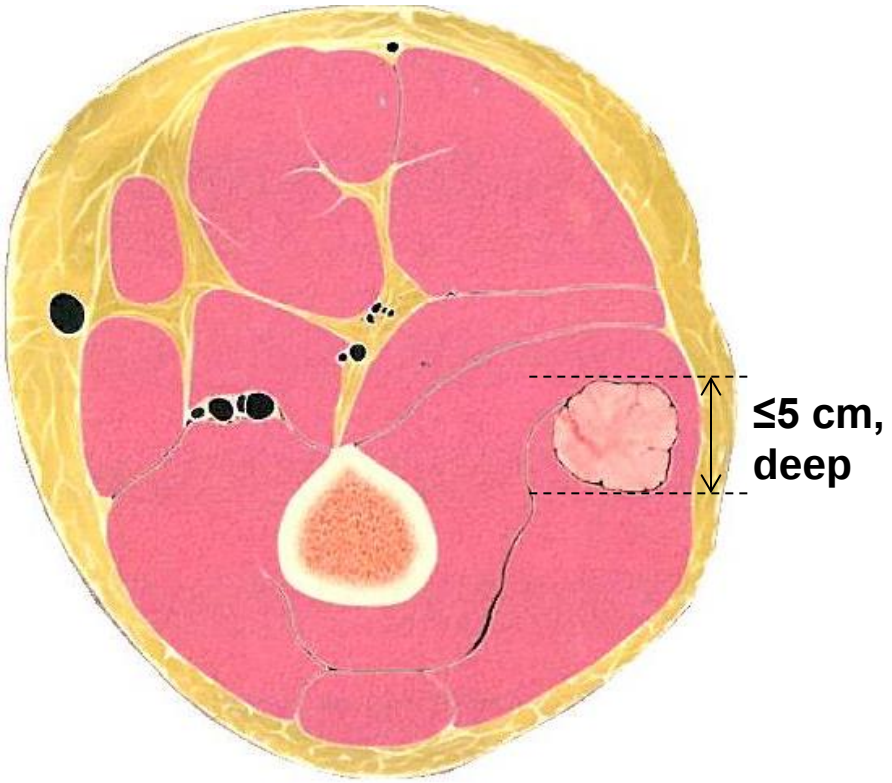
Change	Details of change
New chapter	This classification is being introduced for the first time. Level of evidence: N/A.
Definition of primary tumor (T)	**Superficial and deep location has been removed as part of T criteria**. T categories have been increased from two to four. **T1** remains as tumor **5 cm** or less in greatest dimension. **T2** is now tumor more than **5 cm** and less than or equal to **10 cm** in greatest dimension. **T3** is newly categorized as tumor more than **10 cm** and less than or equal to **15 cm** in greatest dimension. **T4** is a new category defined as tumor more than **15 cm** in greatest dimension. Level of evidence: II.
AJCC prognostic stage groups	AJCC prognostic stage groups have been changed. Level of evidence: N/A.

Definition of primary tumor (T): trunk and extremities

7th AJCC cancer staging		8th AJCC cancer staging	
T - Primary tumor		**T - Primary tumor**	
TX	Primary tumor cannot be assessed	**TX**	Primary tumor cannot be assessed
T0	No evidence of primary tumor		
T1	Tumor **5 cm or less** in greatest dimension	**T1**	Tumor 5 cm or less in greatest dimension
T1a	Superficial tumor		
T1b	Deep tumor		
T2	Tumor **more than 5 cm** in greatest dimension	**T2**	Tumor more than **5 cm** and less than or equal to **10 cm** in greatest dimension
T2a	Superficial tumor	**T3**	Tumor more than **10 cm** and less than or equal to **15 cm** in greatest dimension
T2b	Deep tumor	**T4**	Tumor more than **15 cm** in greatest dimension

T classification: comparison of 7th and 8th edition

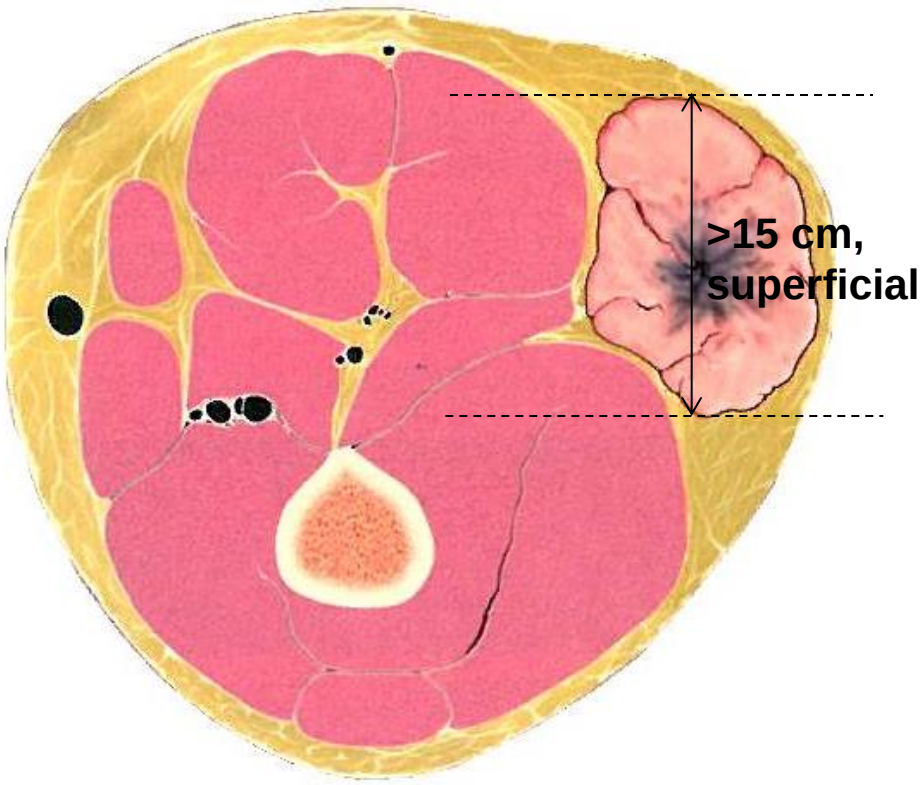


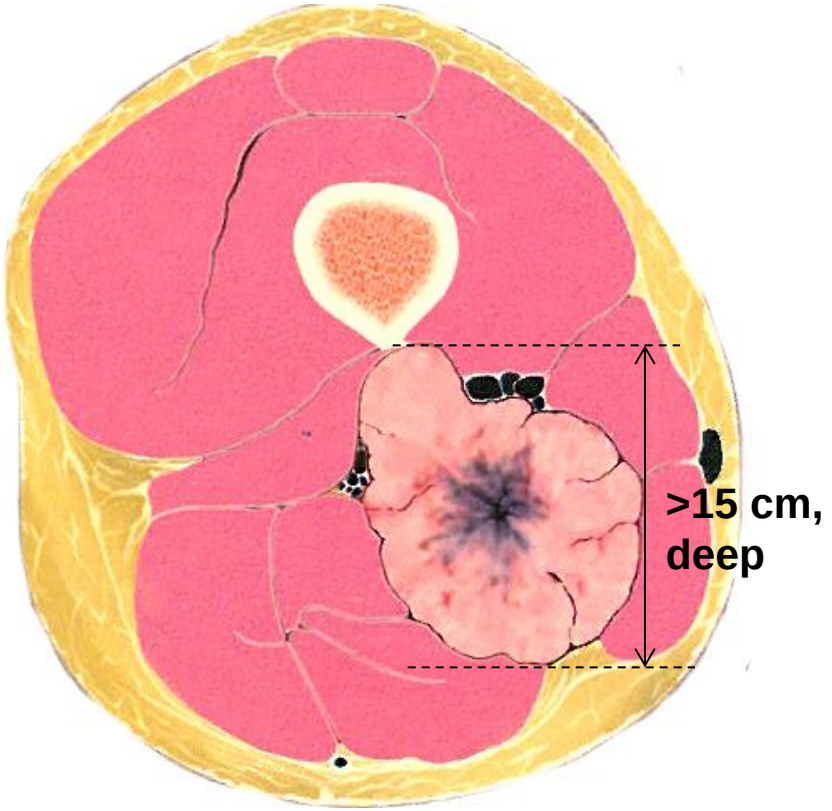


7th edition	8th edition
T1a: Tumor ≤5 cm is located **superficial** to the fascia	**T1:** Tumor ≤5 cm in greatest dimension

7th edition	8th edition
T1b: Tumor ≤5 cm is located **deep** to fascia	**T1:** Tumor ≤5 cm in greatest dimension

T classification: comparison of 7th and 8th edition





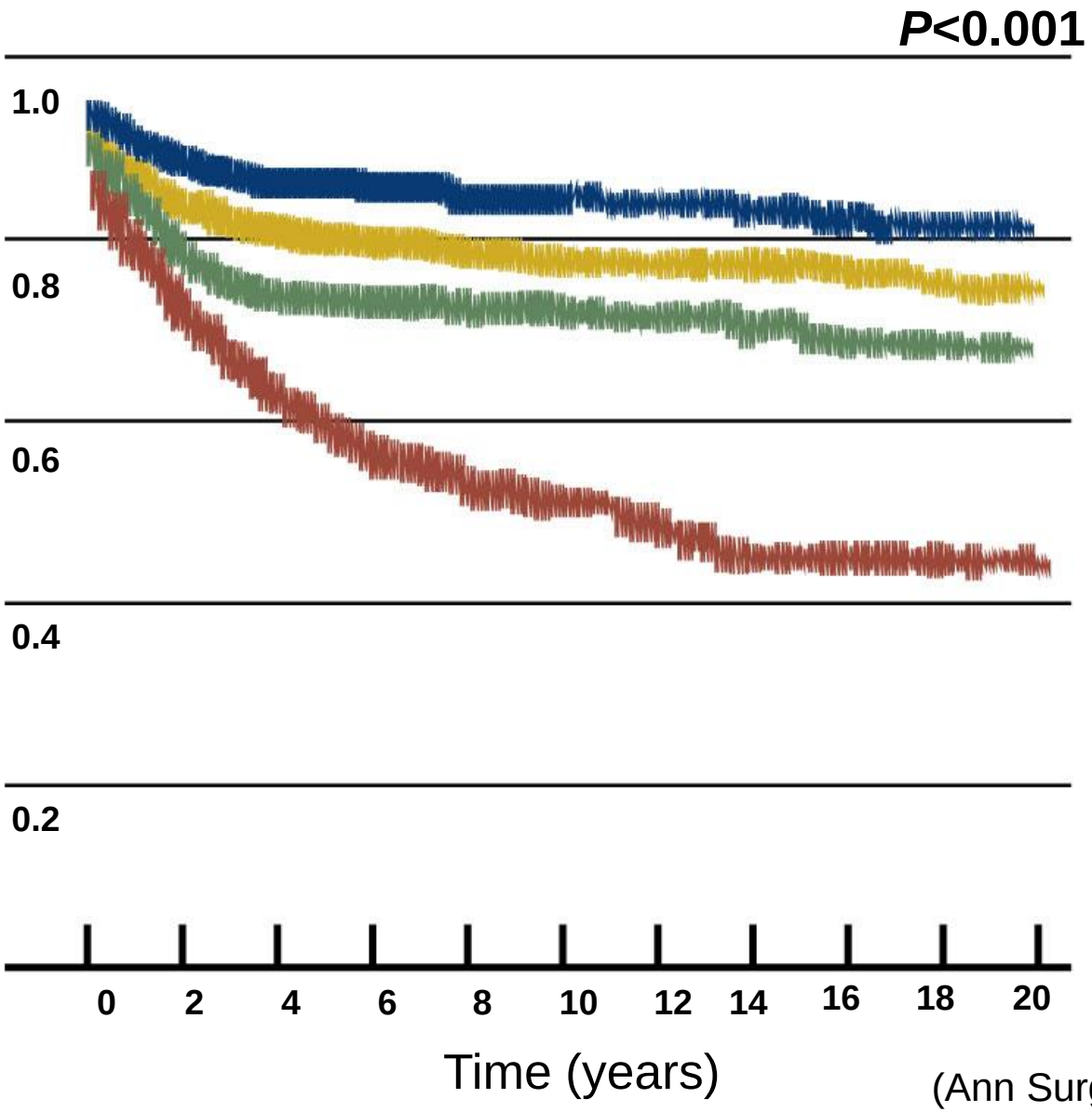
7th edition	8th edition
T2a: Tumor >5 cm is located **superficia**l to fascia	**T4:** Tumor >15 cm in greatest dimension

7th edition	8th edition
T2b: Tumor >5 cm is located **deep** to fascia	**T4:** Tumor >15 cm in greatest dimension

AJCC prognostic stage groups: trunk and extremities

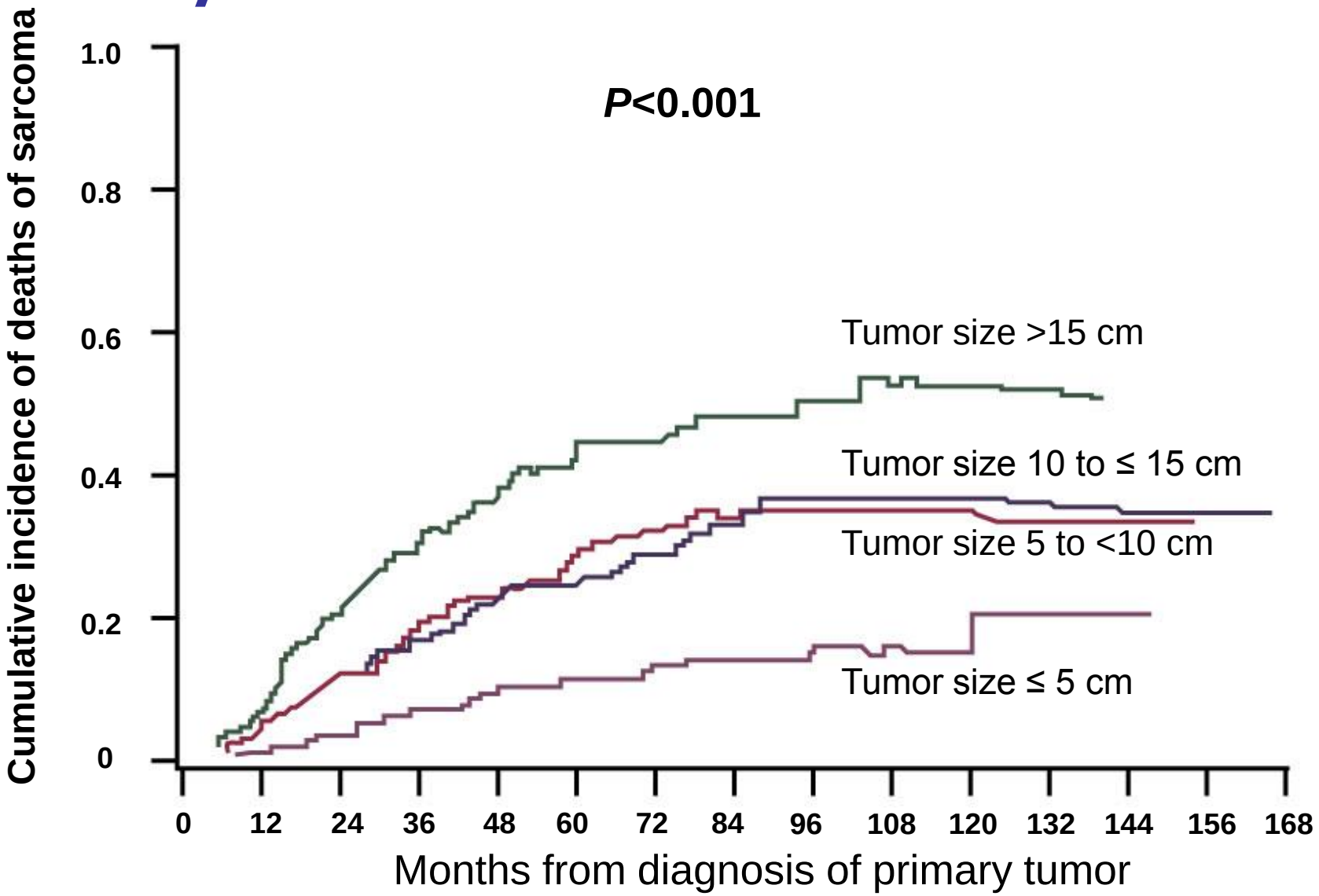
When T is ..	Any N is ..	And M is ..	And grade is ..	Then stage group is ..
T1	N0	M0	G1 or GX	**IA**
T2, T3, T4	N0	M0	G1 or GX	**IB**
T1	N0	M0	G2 or G3	**II**
T2	N0	M0	G2 or G3	**IIIA**
T3, T4	N0	M0	G2 or G3	**IIIB**
Any T	N1	M0	Any G	**IV**
Any T	Any N	M1	Any G	**IV**

Local disease-free survival by size category: trunk and extremities



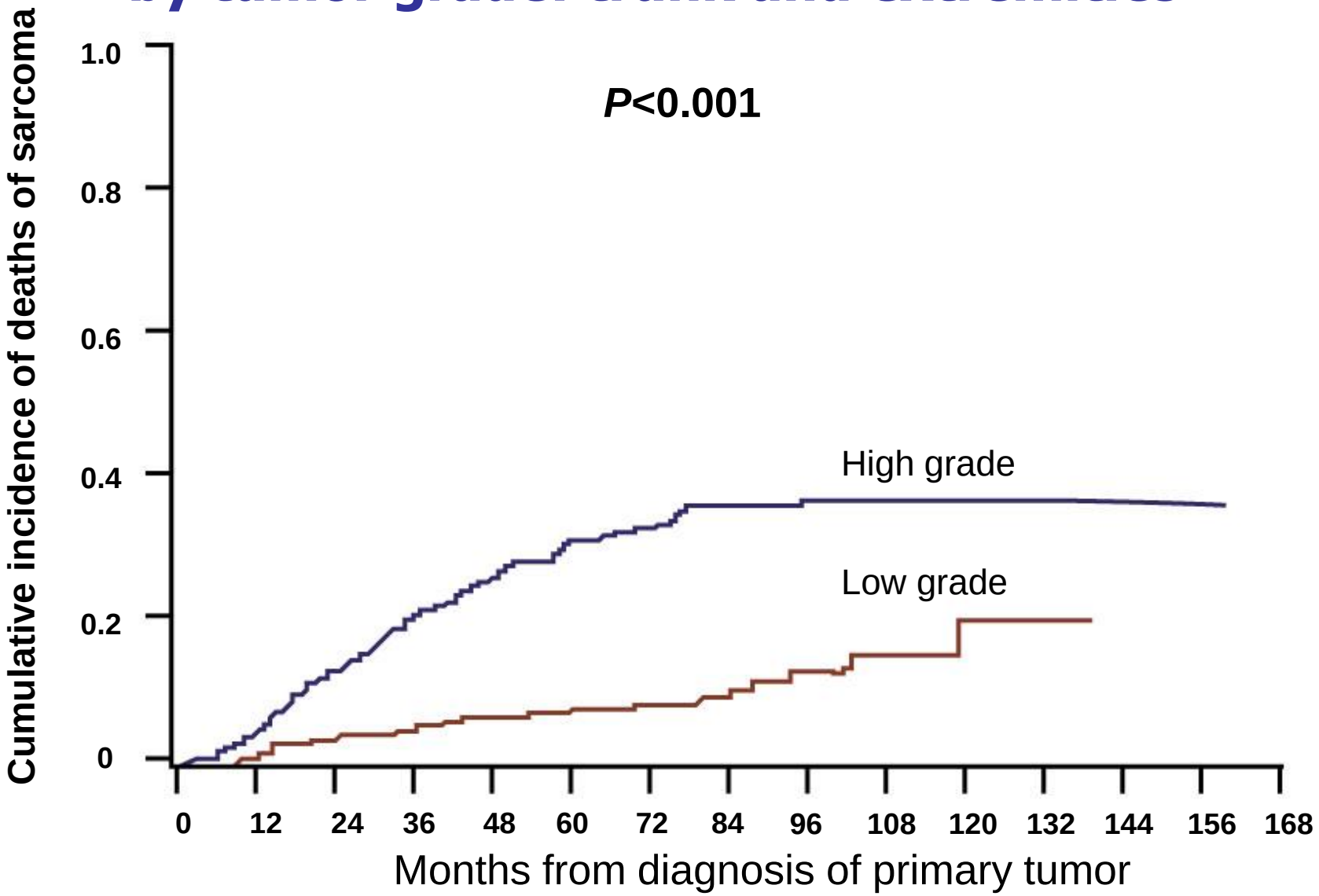
(Ann Surg Oncol 2013;20:3377-83)

Cumulative incidence of sarcoma-related deaths by tumor size: trunk and extremities



(Ann Surg Oncol 2008;15:2739-48)

Cumulative incidence of sarcoma-related deaths by tumor grade: trunk and extremities



(Ann Surg Oncol 2008;15:2739-48)

Case 6: trunk & extremities

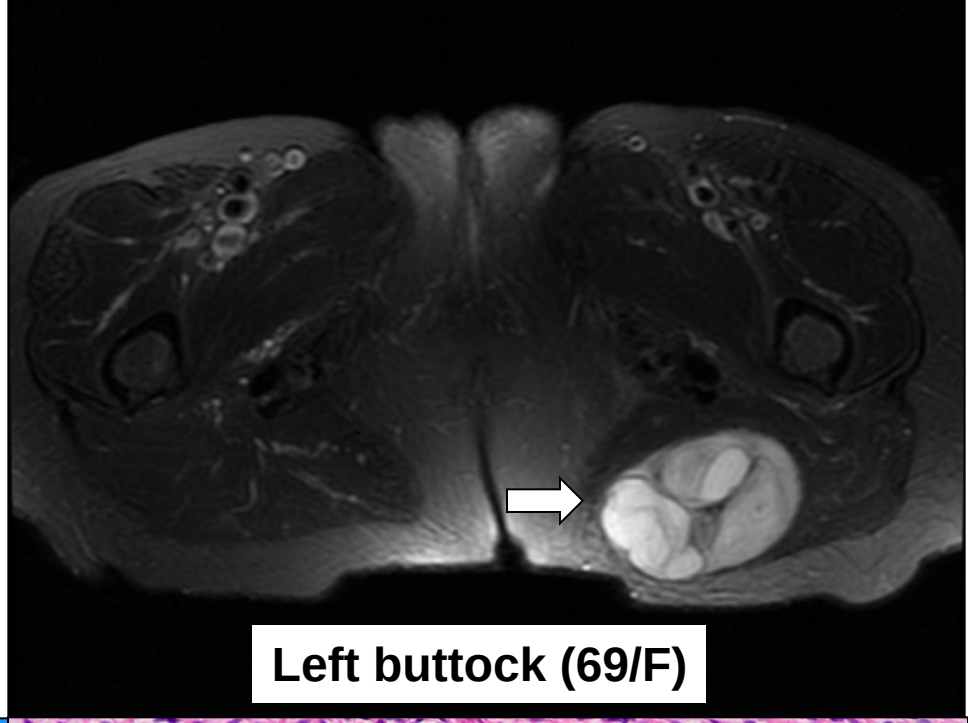
Dx: Myxofibrosarcoma

Pathological TNM

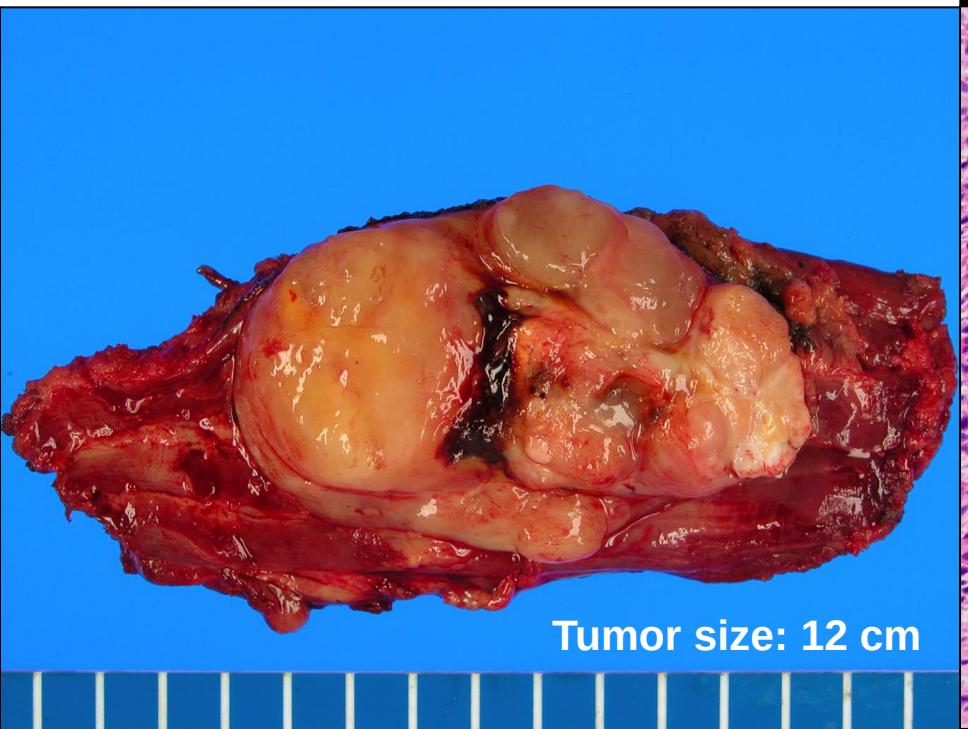
- Primary tumor (T): **pT3** *(>10 - ≤15 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade **3**

Stage group

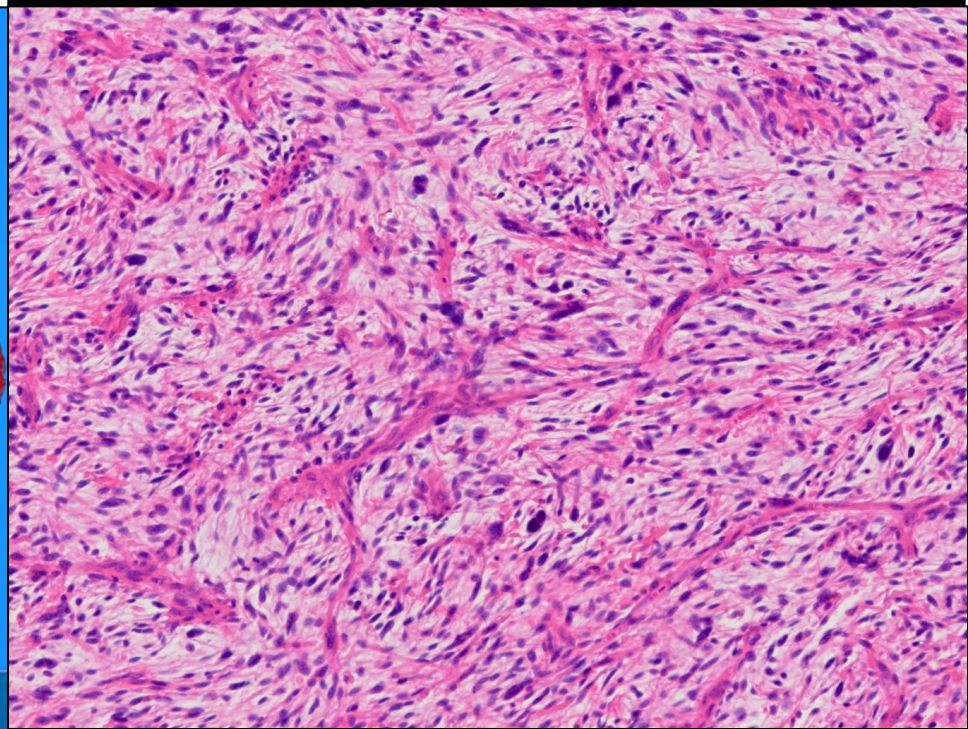
- Stage **IIIB**



Left buttock (69/F)



Tumor size: 12 cm



Case 7: trunk & extremities

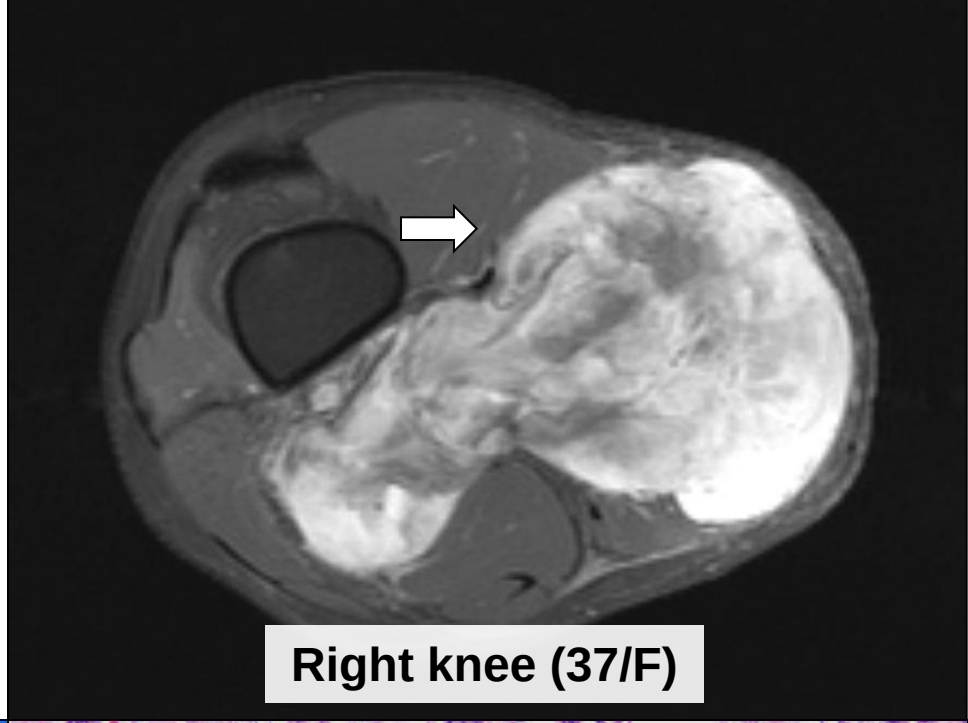
Dx: Myxoid liposarcoma

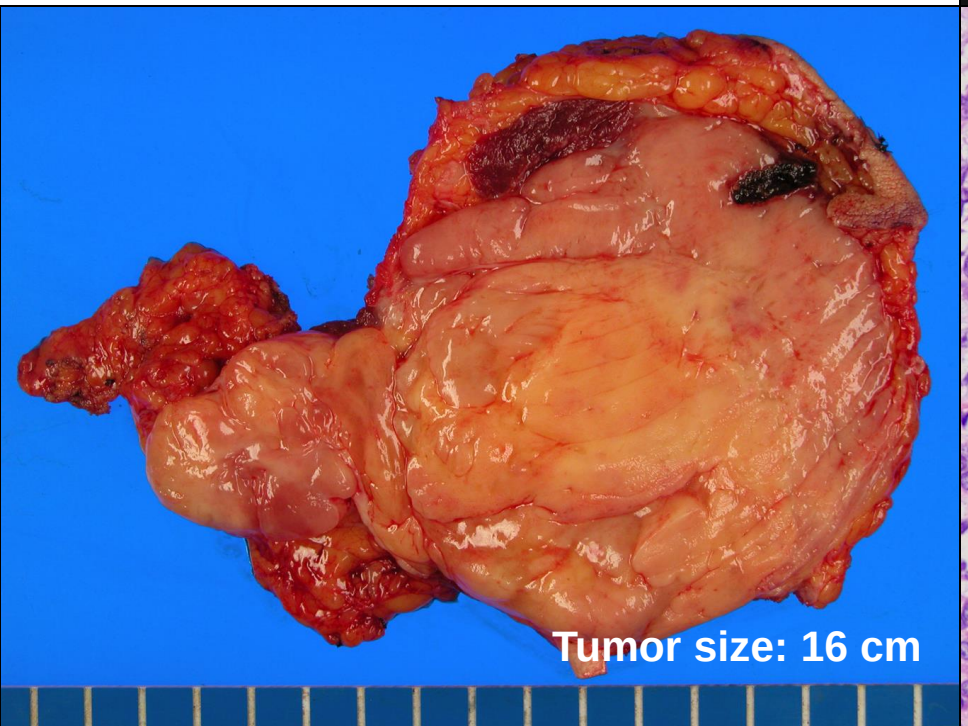
Pathological TNM

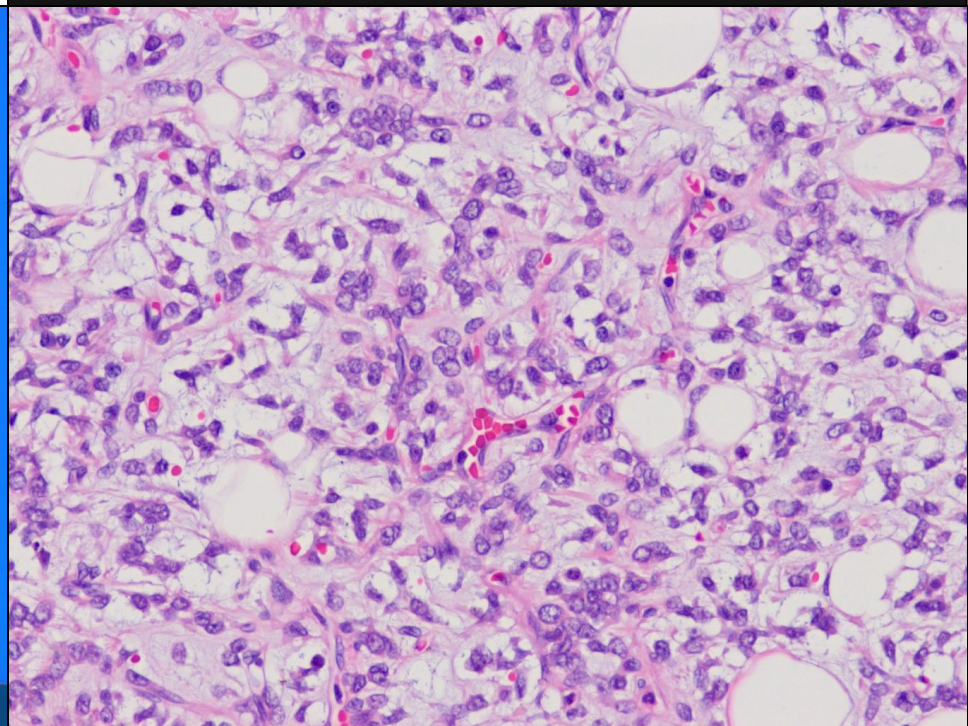
- Primary tumor (T): **pT4** *(>15 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade **3**

Stage group

- Stage **IIIB**







Contents

Soft tissue sarcomas: abdomen and thoracic visceral organs

- Sarcomas of the abdominal and thoracic viscera represent a varied and **heterogeneous** group of mesenchymal neoplasms.

- **Traditional staging algorithms** have not been able to reliably prognosticate this cohort of sarcomas.

- This chapter provides a brief introduction to this unique group of sarcomas and proposes a **new T classification** system for purposes of future data collection and potential development of a specific visceral sarcoma staging system.

(AJCC 8th ed., 2017)

Soft tissue sarcomas: abdomen and thoracic visceral organs

- **Cancers staged using this staging system**
  - Soft tissue sarcomas of the abdominal and thoracic visceral organs
- **Cancers not staged using this staging system**
  - Desmoplastic small round cell tumor
  - Epithelioid hemangioendothelioma
  - Inflammatory myofibroblastic tumor
  - Perivascular epithelioid cell tumor (PEComa)
  - Solitary fibrous tumor
  - Gastrointestinal stromal tumor (sarcoma)
  - Leiomyosarcoma, uterine and retroperitoneal

(AJCC 8th ed., 2017)

Summary of changes: abdomen and thoracic visceral organs

Change	Details of change
New staging system	A new staging system for abdominal and thoracic visceral sarcomas is introduced. Level of evidence: IV.
Definition of primary tumor (T)	A **new** designation for **T** category is proposed. The designation of **deep versus superficial sarcoma** does not make sense for these anatomic sites and is deleted. Level of evidence: IV.

(AJCC 8th ed., 2017)

Definition of primary tumor (T): abdomen and thoracic visceral organs

7th AJCC cancer staging		8th AJCC cancer staging	
T - Primary tumor		**T - Primary tumor**	
TX	Primary tumor cannot be assessed	**TX**	Primary tumor cannot be assessed
T0	No evidence of primary tumor		
T1	Tumor **5 cm or less** than in greatest dimension	**T1**	Organ confined
		T2	Tumor extension into tissue beyond organ
T1a	Superficial tumor	**T2a**	Invades serosa, visceral peritoneum
T1b	Deep tumor	**T2b**	Extension beyond serosa (mesentery)
T2	Tumor **more than 5 cm** in greatest dimension	**T3**	Invades another organ
		T4	Multifocal involvement
T2a	Superficial tumor	**T4a**	Multifocal (2 sites)
T2b	Deep tumor	**T4b**	Multifocal (3-5 sites)
		T4c	Multifocal (>5 sites)

Metastatic disease vs. multifocality

- There are no criteria by which to declare multifocality vs metastatic disease.

- Generally, **dominant lesion with small implants** elsewhere should be considered metastatic disease, whereas lesions **without a dominant primary site** can be considered multifocal.

(AJCC 8th ed., 2017)

Case 8: abdomen & thoracic visceral organ

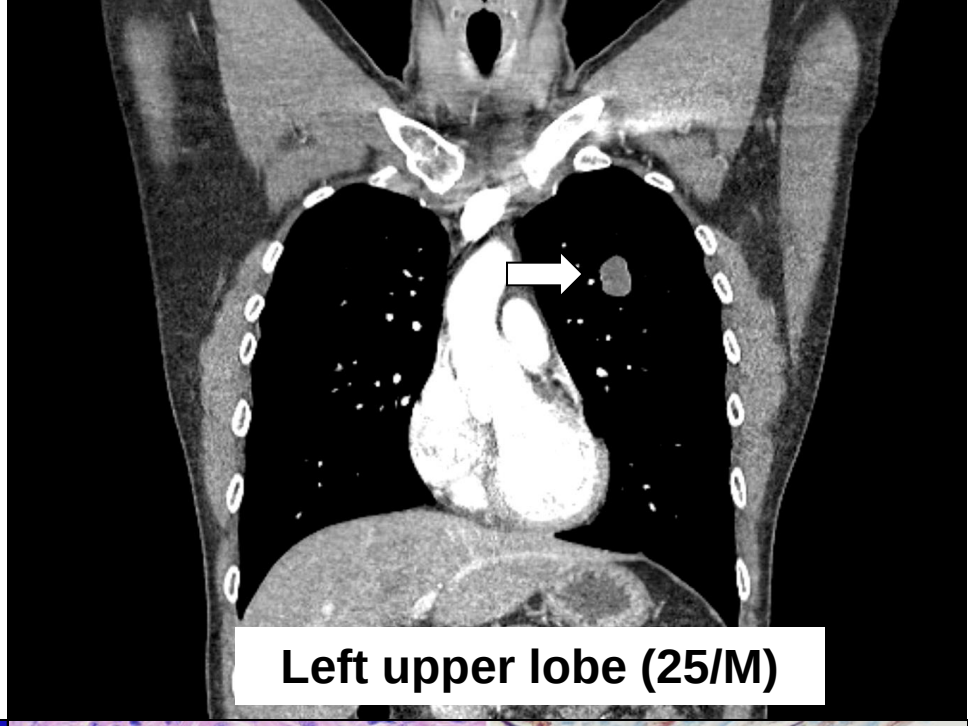
Dx: Leiomyosarcoma

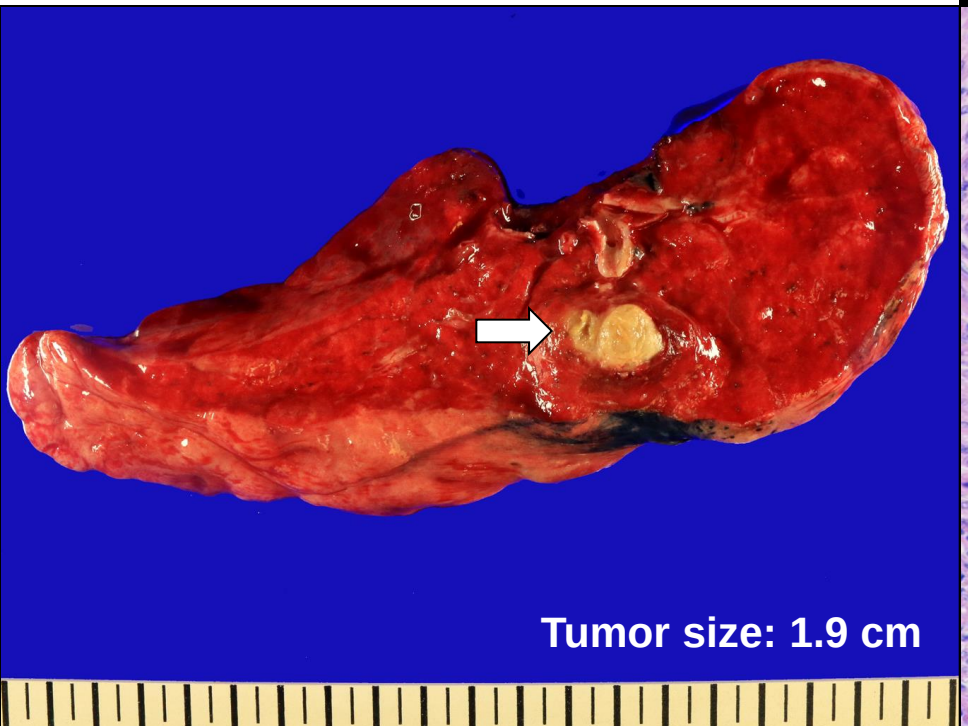
Pathological TNM

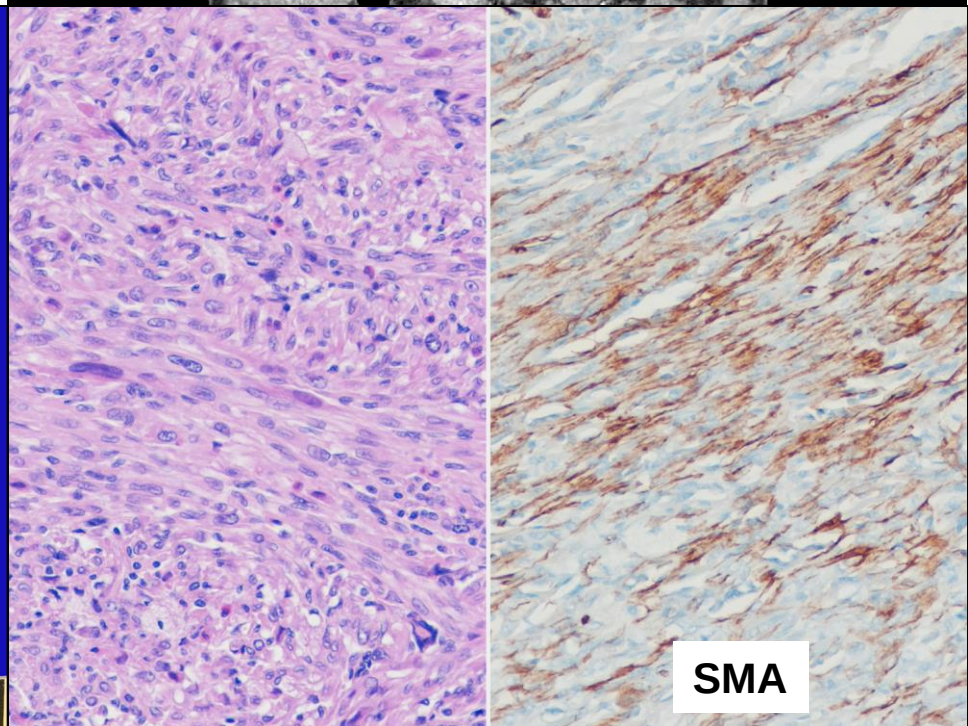
- Primary tumor (T): **pT1** *(organ confined)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade **1**

Stage group

* There is no recommended AJCC prognostic stage group at this time.







Contents

Soft tissue sarcoma: GIST

- **Cancers staged using this staging system**
  - Gastrointestinal stromal tumor (GIST)

- **Cancers not staged using this staging system**
  - Pediatric GIST
  - Familial GIST (germline mutant *KIT* or *PDGFRA*)
  - Syndromic GIST

(AJCC 8th ed., 2017)

Summary of changes: GIST

- There are **no changes** to this staging system.

(AJCC 8th ed., 2017)

TNM classification: GIST

T category	**T criteria**
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Tumor 2 cm or less
T2	Tumor more than 2 cm but not more than 5 cm
T3	Tumor more than 5 cm but not more than 10 cm
T4	Tumor more than 10 cm in greatest dimension
N category	**N criteria**
N0	No regional lymph node metastases or unknwon lymph node status
N1	Regional lymph node metastasis
M category	**M criteria**
M0	No distant metastasis
M1	Distant metastasis
Mitotic rate	**Definition**
Low	5 or fewer mitoses per 5 mm^2, or per 50 HPF
High	Over 5 mitoses per 5 mm^2, or per 50 HPF

GIST: risk assessment
NIH consensus classification system, 2002

Risk category	Size	Mitotic count
Very low risk	<2 cm	≤5/50 HPF
Low risk	2-5 cm	≤5/50 HPF
Intermediate risk	≤5 cm >5-10 cm	6-10/50 HPF ≤5/50 HPF
High risk	>5 cm >10 cm Any size	>5/50 HPF Any mitotic rate >10/50 HPF

(Hum Pathol 2002;33:459-65)

AJCC prognostic stage groups: gastric and omental GIST

When T is ..	Any N is ..	And M is ..	And mitotic rate is ..	Then stage group is ..
T1 or T2	N0	M0	Low	**IA**
T3	N0	M0	Low	**IB**
T1	N0	M0	High	**II**
T2	N0	M0	High	**II**
T4	N0	M0	Low	**II**
T3	N0	M0	High	**IIIA**
T4	N0	M0	High	**IIIB**
Any T	N1	M0	Any rate	**IV**
Any T	Any N	M1	Any rate	**IV**

AJCC prognostic stage groups: small intestine, esophageal, colorectal, mesenteric, peritoneal GIST

When T is ..	Any N is ..	And M is ..	And mitotic rate is ..	Then stage group is ..
T1 or T2	N0	M0	Low	**I**
T3	N0	M0	Low	**II**
T1	N0	M0	High	**IIIA**
T4	N0	M0	Low	**IIIA**
T2	N0	M0	High	**IIIB**
T3	N0	M0	High	**IIIB**
T4	N0	M0	High	**IIIB**
Any T	N1	M0	Any rate	**IV**
Any T	Any N	M1	Any rate	**IV**

Case 9: GIST

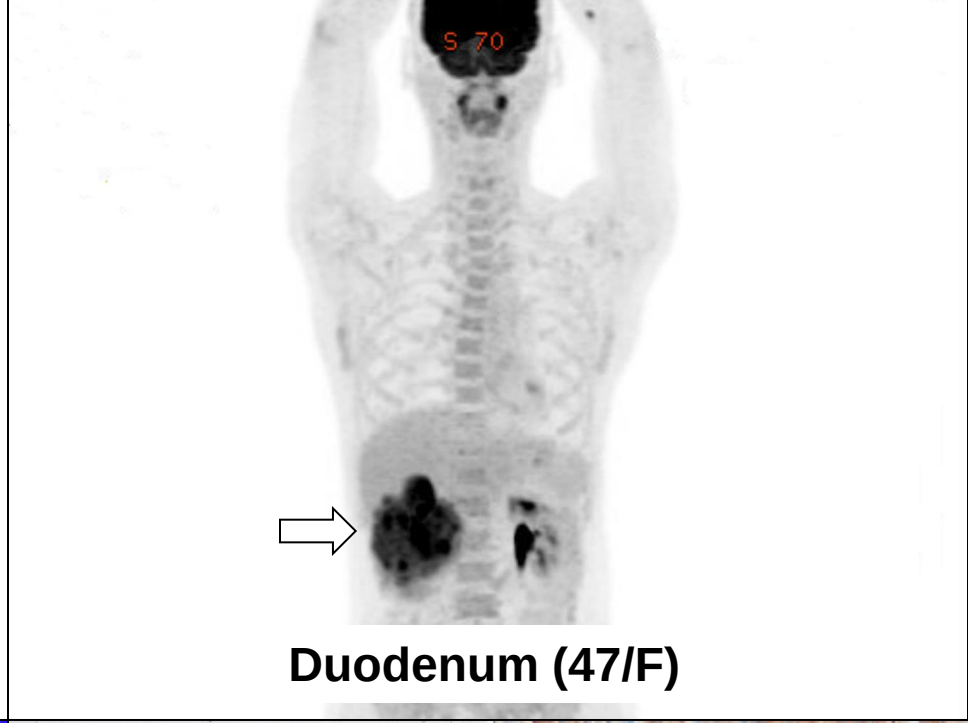
Dx: GIST, high risk

Pathological TNM (pTNM)

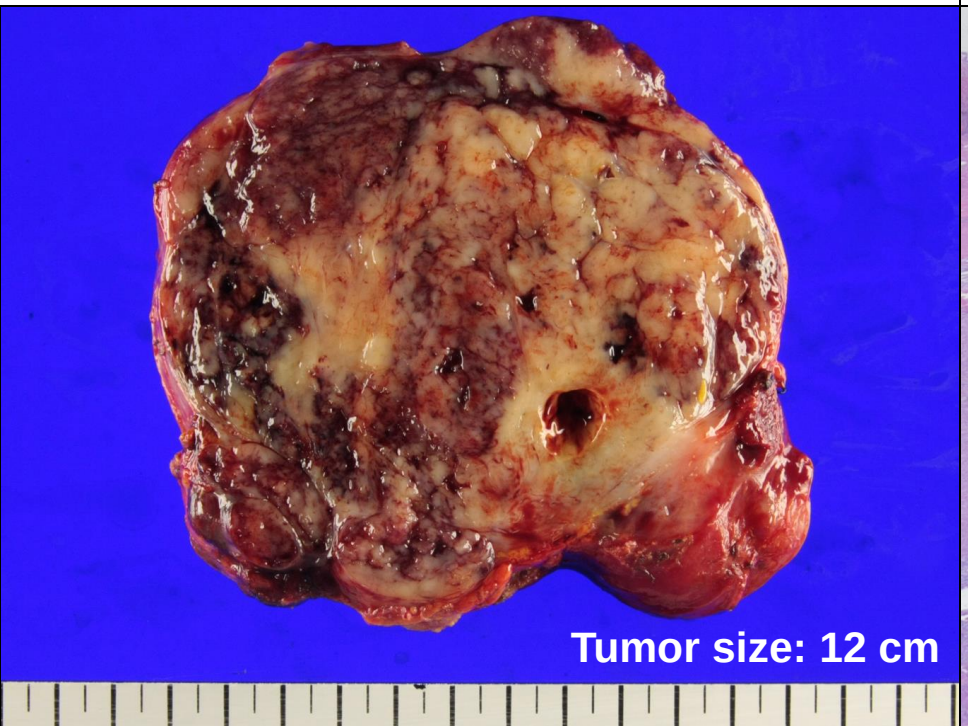
- Primary tumor (T): **pT4** *(>10 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Mitotic rate: **low** (5/50 HPFs)

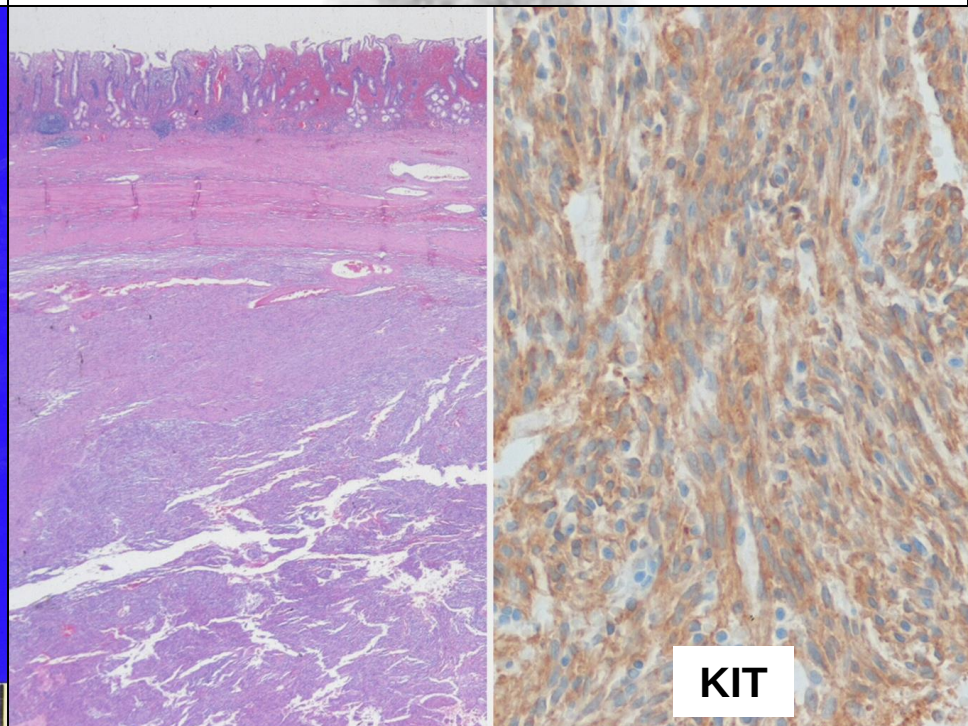
Stage group

- Stage **IIIA**



Duodenum (47/F)





Contents

- Introduction
- Bone
- **Soft tissue sarcoma**
  - Head and neck
  - Trunk and extremities
  - Abdomen and thoracic visceral organs
  - Gastrointestinal stromal tumor (GIST)
  - Retroperitoneum
  - Unusual histologies and sites
- Summary

Most common and less common sarcomas arising in the retroperitoneum

Most common sarcomas	Less common sarcomas
• Well differentiated liposarcoma • Dedifferentiated liposarcoma • Leiomyosarcoma	• Pleomorphic liposarcoma • Undifferentiated pleomorphic sa. • MPNST • Solitary fibrous tumor (malignant)

Summary of changes: retroperitoneum (1)

Change	Details of change
Definition of primary tumor (T)	Retroperitoneal sarcomas use the **same revised tumor size** (T) classification for extremity and trunk sarcomas. Level of Evidence: II. **Superficial** and **deep location** has been removed as part of T criteria. Level of Evidence: II. **T categories** have been increased from two to four. **T1** remains as tumor **5 cm** or less in greatest dimension. **T2** is now tumor more than **5 cm** and less than or equal to **10 cm** in greatest dimension. **T3** is newly categorized as tumor more than **10 cm** and less than or equal to **15 cm** in greatest dimension. **T4** is a new category defined as tumor more than **15 cm** in greatest dimension. Level of Evidence: II.

Summary of changes: retroperitoneum (2)

Change	Details of change
Risk assessment models	The retroperitoneum poses particular **challenges to staging**, especially in the context of resectable retroperitoneal sarcoma (AJCC stage I-III). These difficulties are particularly apparent in using the AJCC staging system to counsel patients regarding prognosis in that **most resectable retroperitoneal sarcomas** present as large lesions (T2) without any metastasis (N0M0). In light of this relative lack of prognostic discrimination of the AJCC soft tissue sarcoma staging system, a **prognostic nomogram** is now included as a means to assess prognosis more accurately for patients bearing retroperitoneal soft tissue sarcoma. Level of Evidence: I

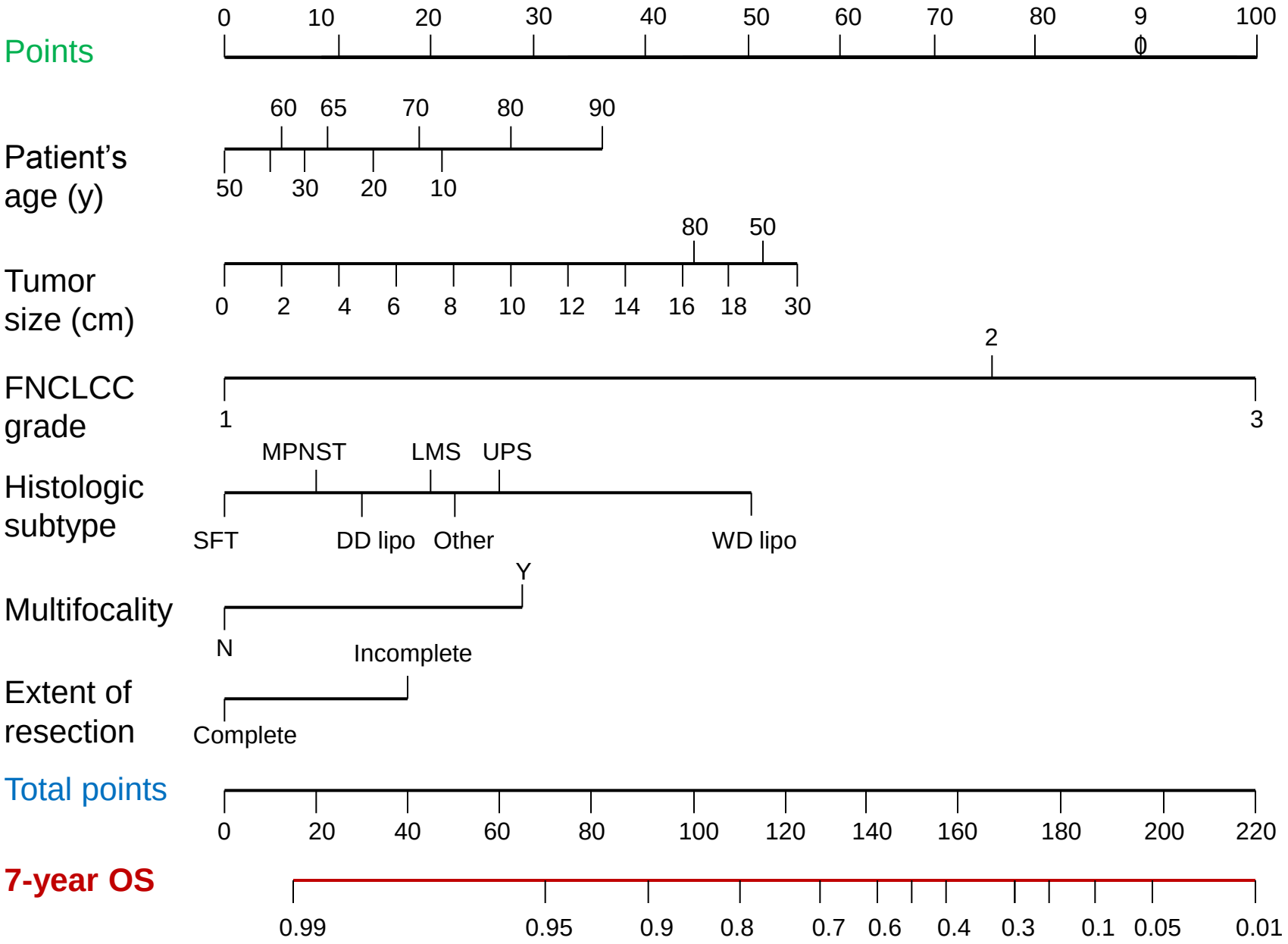
Definition of primary tumor (T): retroperitoneum

7th AJCC cancer staging		8th AJCC cancer staging	
T - Primary tumor		**T - Primary tumor**	
TX	Primary tumor cannot be assessed	**TX**	Primary tumor cannot be assessed
T0	No evidence of primary tumor	**T0**	No evidence of primary tumor
T1	Tumor **5 cm or less** than in greatest dimension	**T1**	Tumor **5 cm** or less in greatest dimension
T1a	Superficial tumor	**T2**	Tumor more than **5 cm** and less than or equal to **10 cm** in greatest dimension
T1b	Deep tumor		
T2	Tumor **more than 5 cm** in greatest dimension	**T3**	Tumor more than **10 cm** and less than or equal to **15 cm** in greatest dimension
T2a	Superficial tumor		
T2b	Deep tumor	**T4**	Tumor more than **15 cm** in greatest dimension

AJCC prognostic stage groups: retroperitoneum

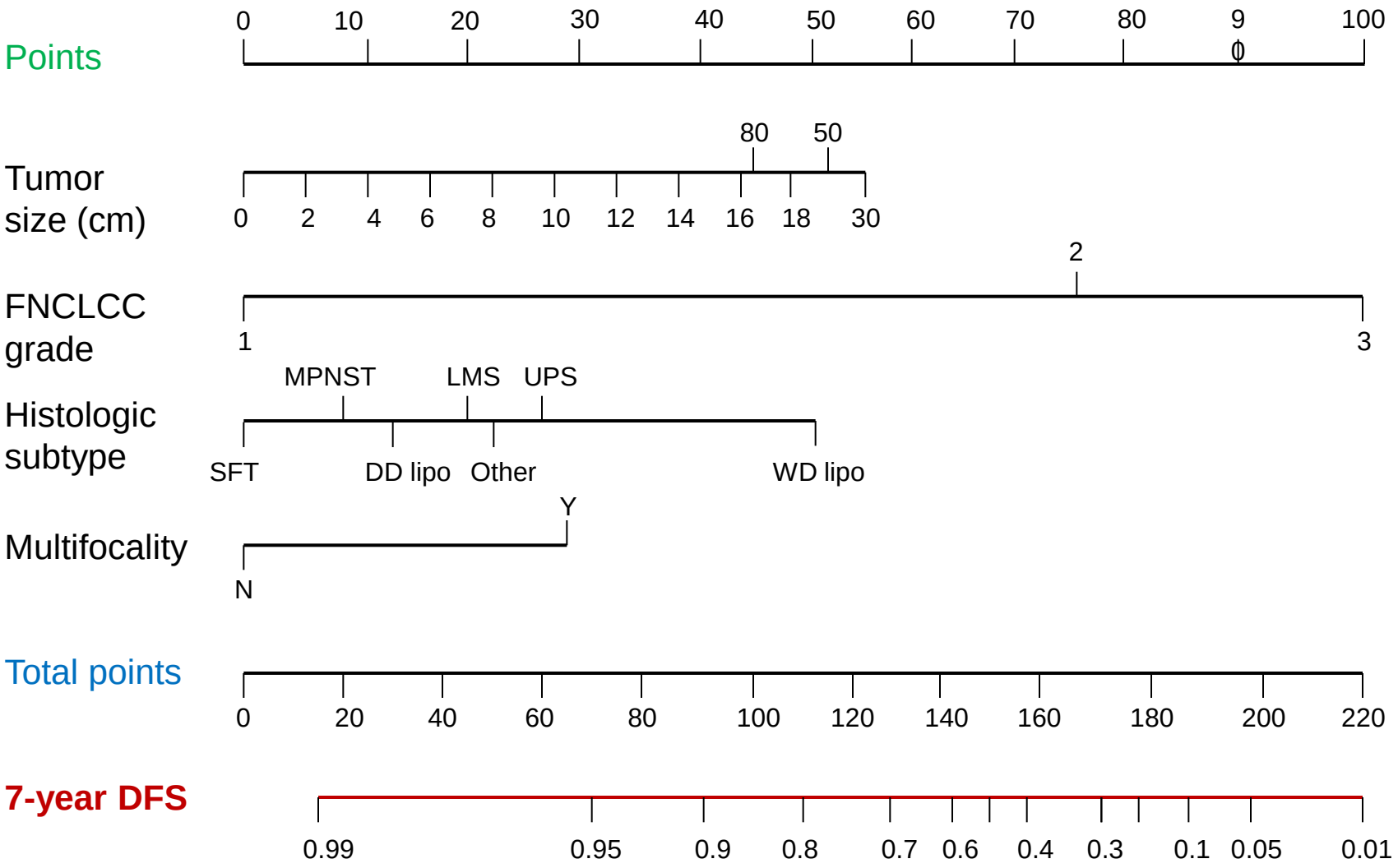
When T is ..	Any N is ..	And M is ..	And grade is ..	Then stage group is ..
T1	N0	M0	G1, GX	**IA**
T2, T3, T4	N0	M0	G1, GX	**IB**
T1	N0	M0	G2, G3	**II**
T2	N0	M0	G2, G3	**IIIA**
T3, T4	N0	M0	G2, G3	**IIIB**
Any T	N1	M0	Any G	**IIIB**
Any T	Any N	M1	Any Ge	**IV**

Nomogram for 7-year overall survival: retroperitoneal sarcoma



(J Clin Oncol 2013;31:1649-55)

Nomogram for 7-year disease free survival: retroperitoneal sarcoma



(J Clin Oncol 2013;31:1649-55)

Case 10: retroperitoneum

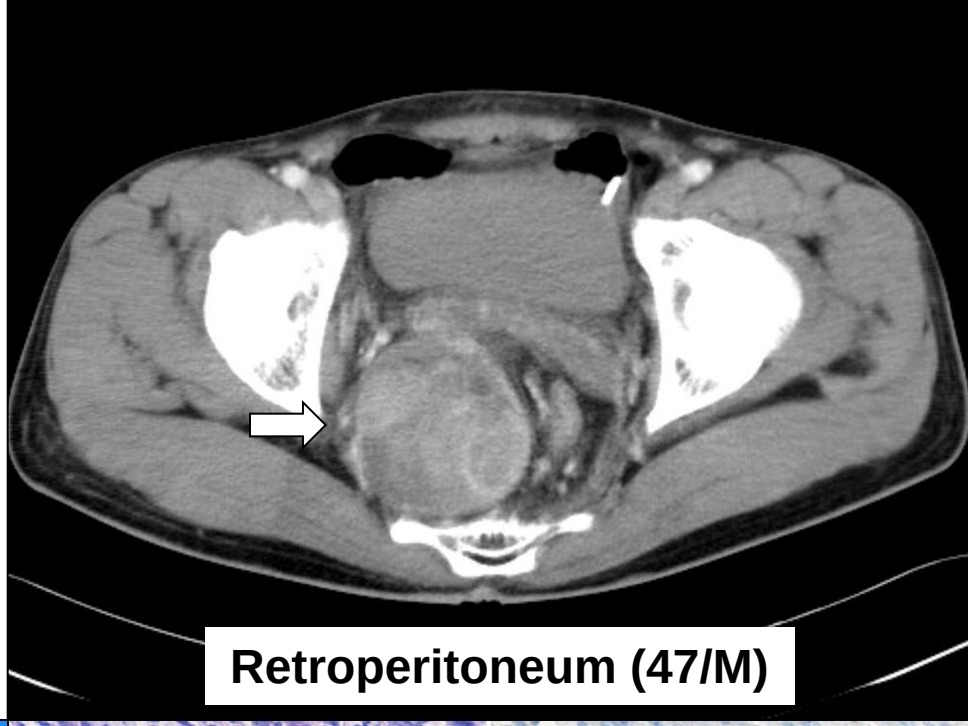
Dx: MPNST

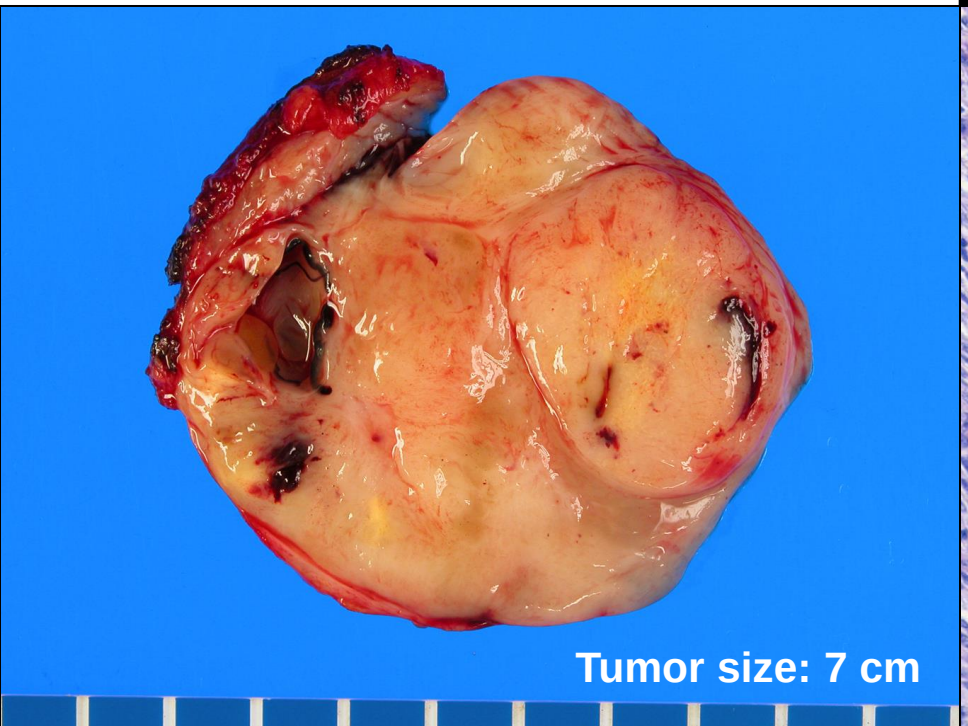
Pathological TNM (pTNM)

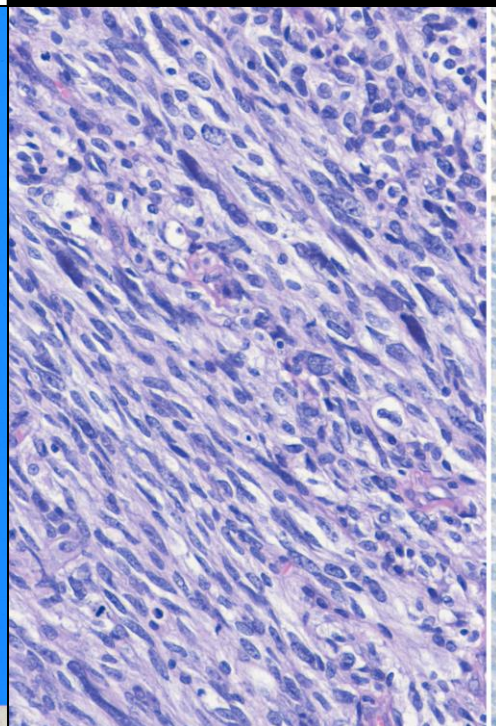
- Primary tumor (T): **pT2** *(>5 - ≤10 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade: **G2**

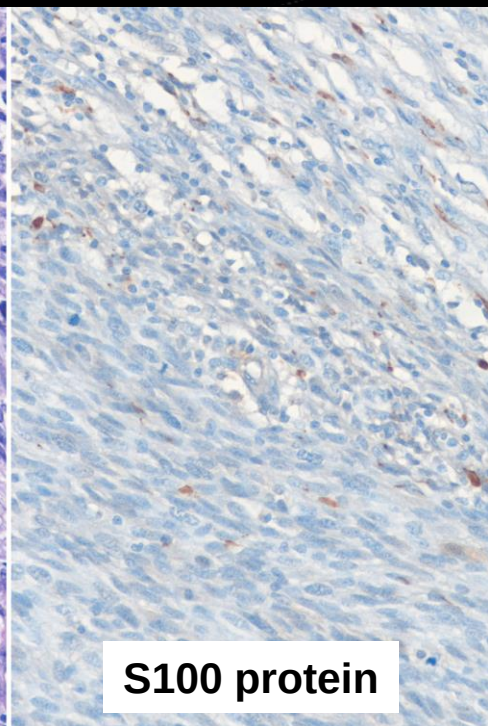
Stage group

- Stage **IIIA**









Case 11: retroperitoneum

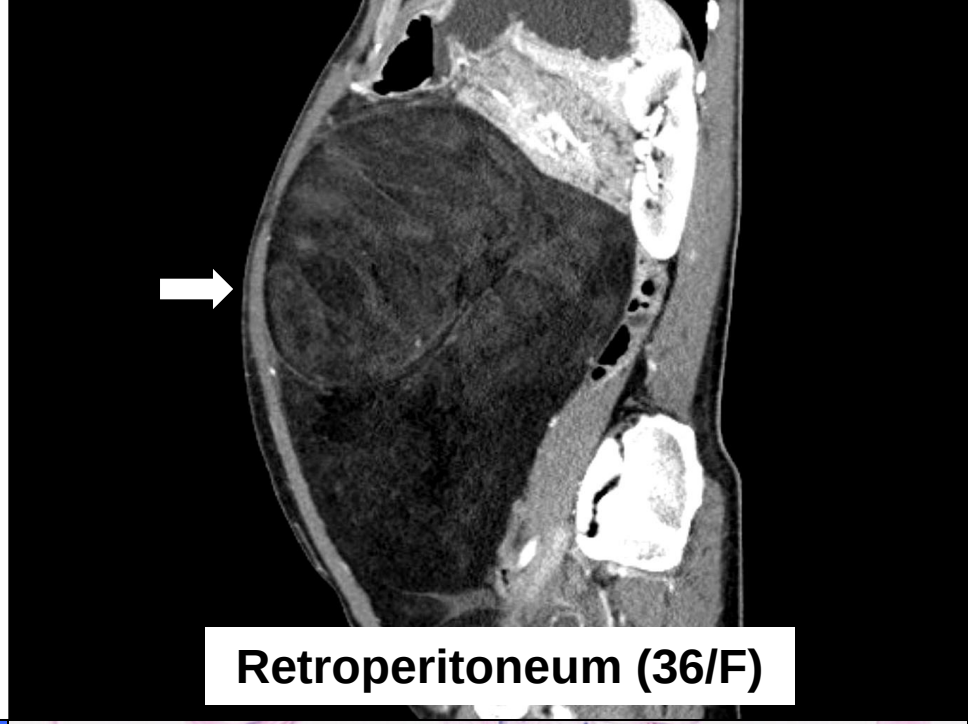
Dx: Well differen. liposarcoma

Pathological TNM (pTNM)

- Primary tumor (T): **pT4** *(>15 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade: **G1**

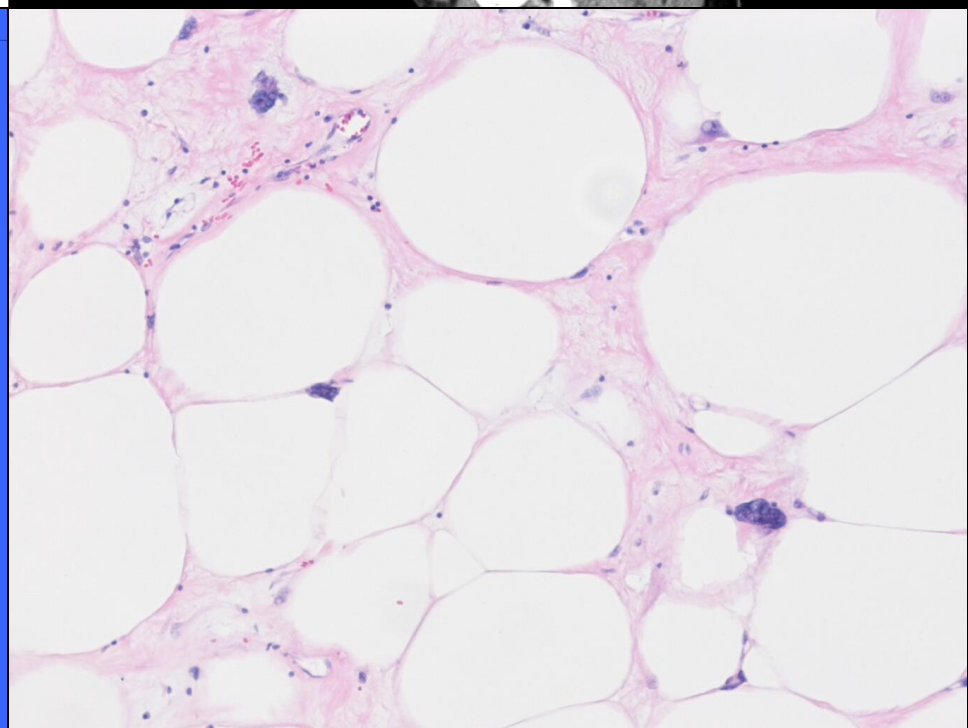
Stage group

- Stage **IB**



Retroperitoneum (36/F)

Tumor size: 45 cm



Contents

Soft tissue sarcoma - unusual histologies and sites: difficult to stage or present unique features

- **Diagnoses discussed in this chapter**
  - Alveolar soft part sarcoma
  - Angiosarcoma
  - Desmoplastic small round cell tumor
  - Epithelioid hemangioendothelioma
  - Extraskeletal myxoid chondrosarcoma
  - Inflammatory myofibroblastic tumor
  - Kaposi sarcoma
  - Osteosarcoma of soft tissue
  - Phyllodes tumor
  - Rhabdomyosarcoma
  - Solitary fibrous tumor

Soft tissue sarcoma - unusual histologies and sites: no risk or extremely low risk of metastasis

- **Diagnoses not staged using this staging system**
  - Desmoid tumor (deep fibromatosis)
  - Superficial fibromatosis
  - Lipofibromatosis
  - Giant cell fibroblastoma
  - Plexiform fibrohistiocytic tumor
  - Giant cell tumor of soft tissue
  - Kaposiform hemangioendothelioma
  - Hemosiderotic fibrolipomatous tumor
  - Atypical fibroxanthoma
  - Angiomatoid fibrous histiocytoma
  - Pleomorphic hyalinizing angiectatic tumor

Summary of changes: soft tissue sarcoma - unusual histologies and sites

Change	Details of change
New chapter	Given the difficulty of classifying more than 70 different cancers using a single staging system, this chapter discusses key histologies that are **troublesome regarding their staging**. Reference is made to other sections or chapters in which these diagnoses are addressed in more detail. Level of evidence: N/A.

Soft tissue sarcomas: unusual histologies and sites

Histology	Anatomic site	Clinical features	Implication for staging
Angiosarcoma	Head and neck	May present with satellite lesions; radiation-associated may present as multifocal disease	Record size of largest lesion according to multifocality guidelines.
Clear cell sarcoma	Joint tendons, aponeurosis, small bowel		
Desmoplastic small round cell tumor	Peritoneum	Typically presents as multiple masses	If possible, record size of largest lesion according to multifocality guidelines
Epithelioid hemangioendo-thelioma	Liver, lung, pleura, rarely elsewhere	Typically presents as multiple masses	If possible, record size of largest lesion according to multifocality guidelines
Epithelioid sarcoma, proximal type	Shoulder girdle, hip musculatures	May present as multifocal disease; lymph node involvement common	If possible, record size of largest lesion according to multifocality guidelines

Case 12: Unusual histologies

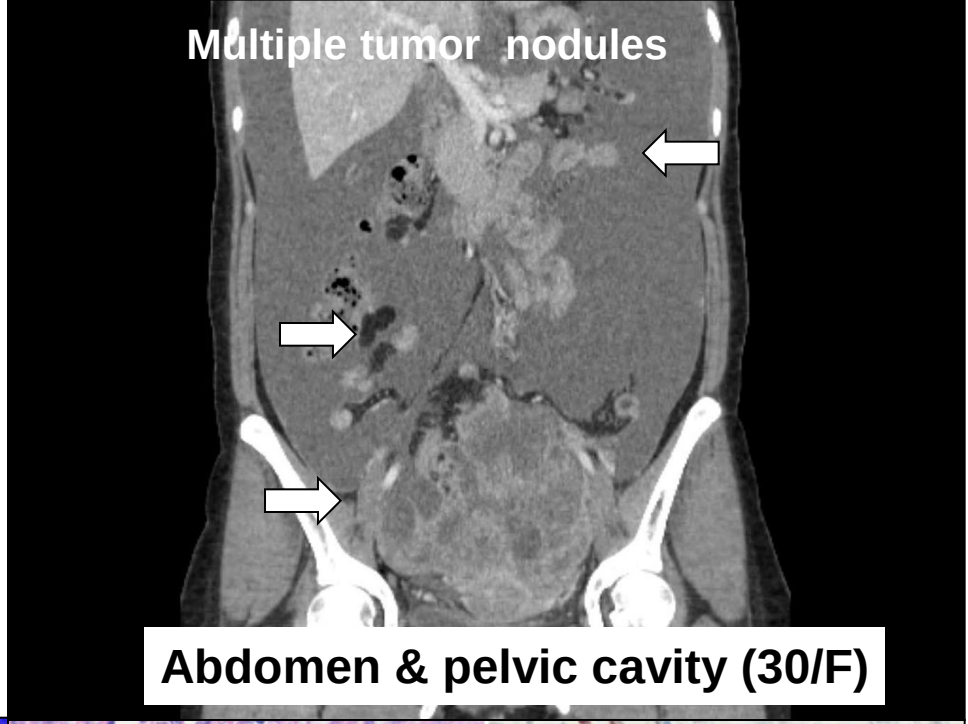
Dx: Desmoplastic small round cell tumor

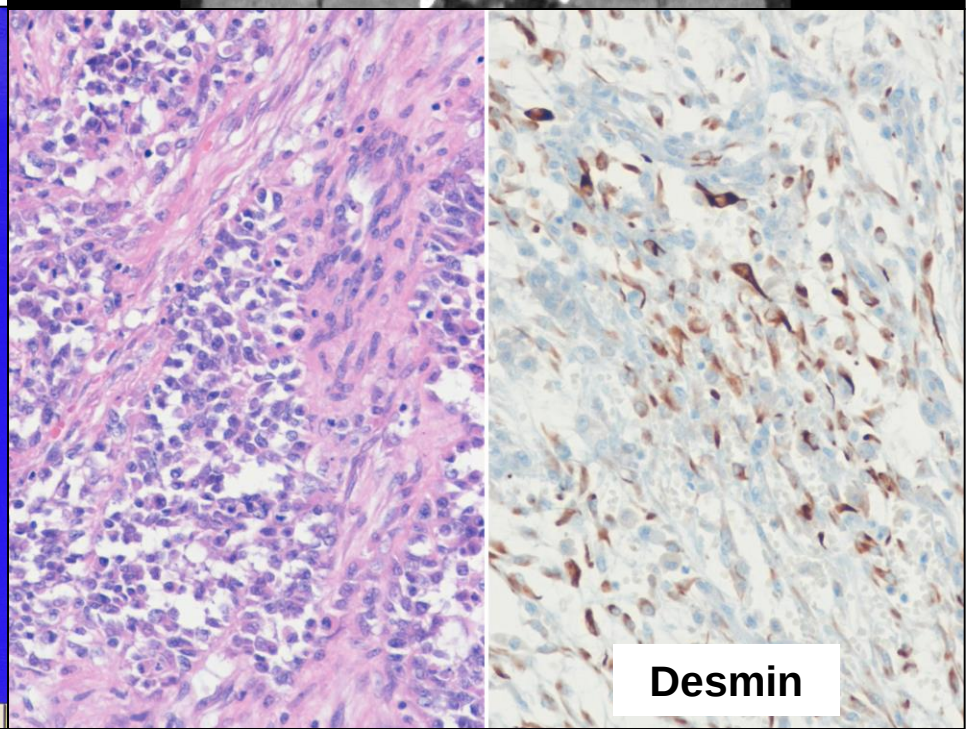
Pathological TNM (pTNM)

- Primary tumor (T): **pT4** (**m**) *(multifocal involvement by according to AJCC)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **cM0**
- Histologic grade: **G3**

Stage group

* There is no currently validated staging system. Several staging methods have been proposed based on the size ad number of lesion.





Case 13: Unusual histologies

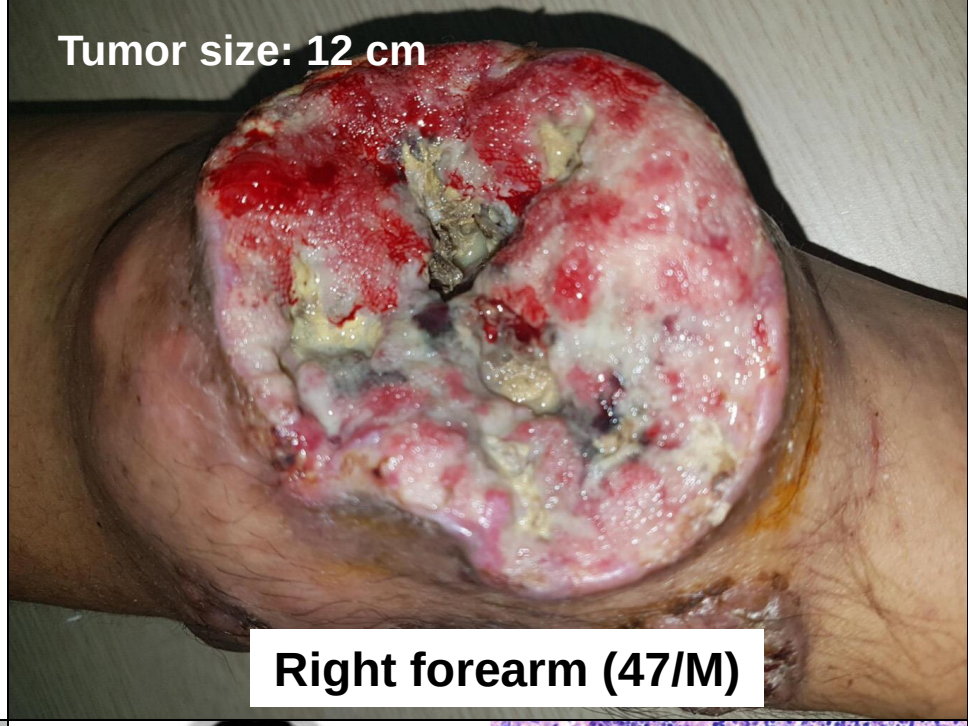
Dx: Epithelioid sarcoma, proximal type

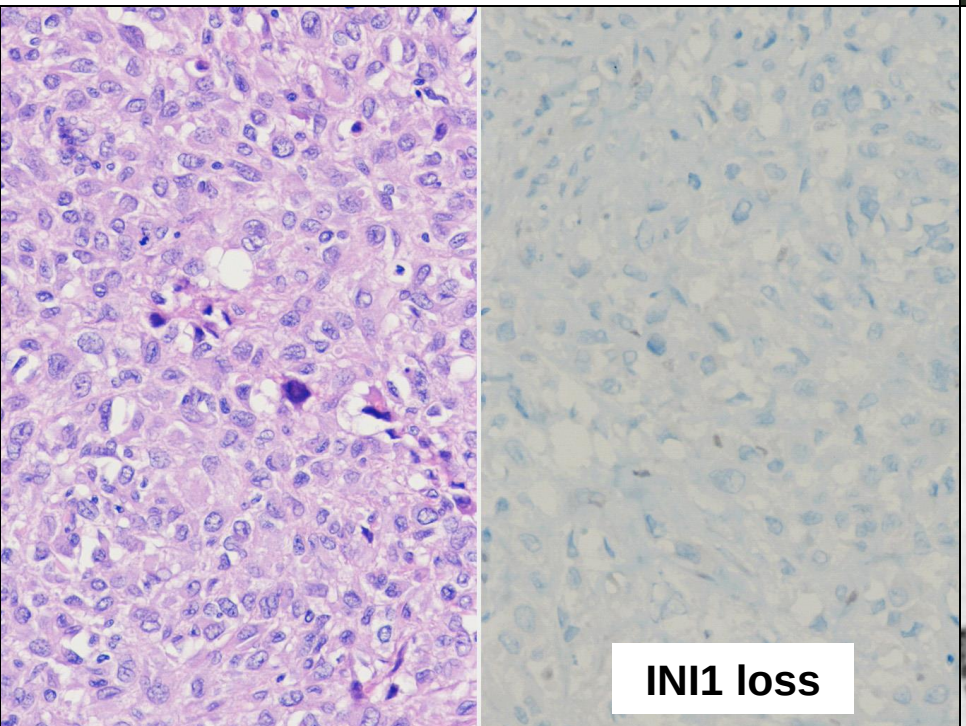
Pathological TNM (pTNM)

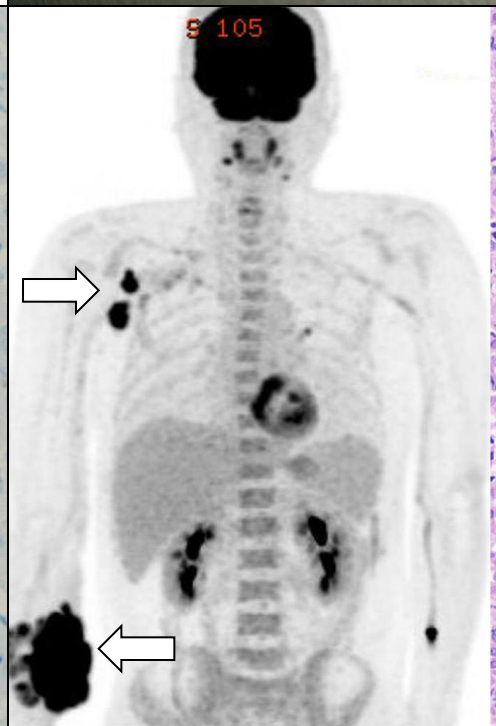
- Primary tumor (T): **pT3** *(>10 - ≤15 cm)*
- Regional lymph node (N): **pN1**
- Distant metastasis (M): **cM0**
- Histologic grade: **G3**

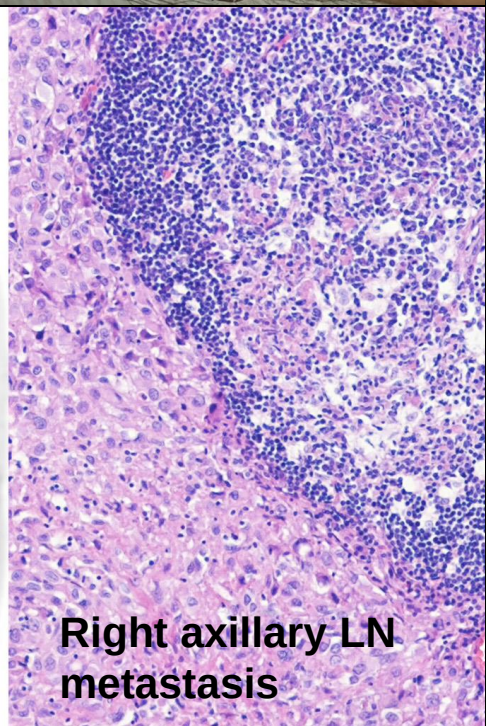
Stage group

- Stage **IV**









Case 14: Unusual histologies

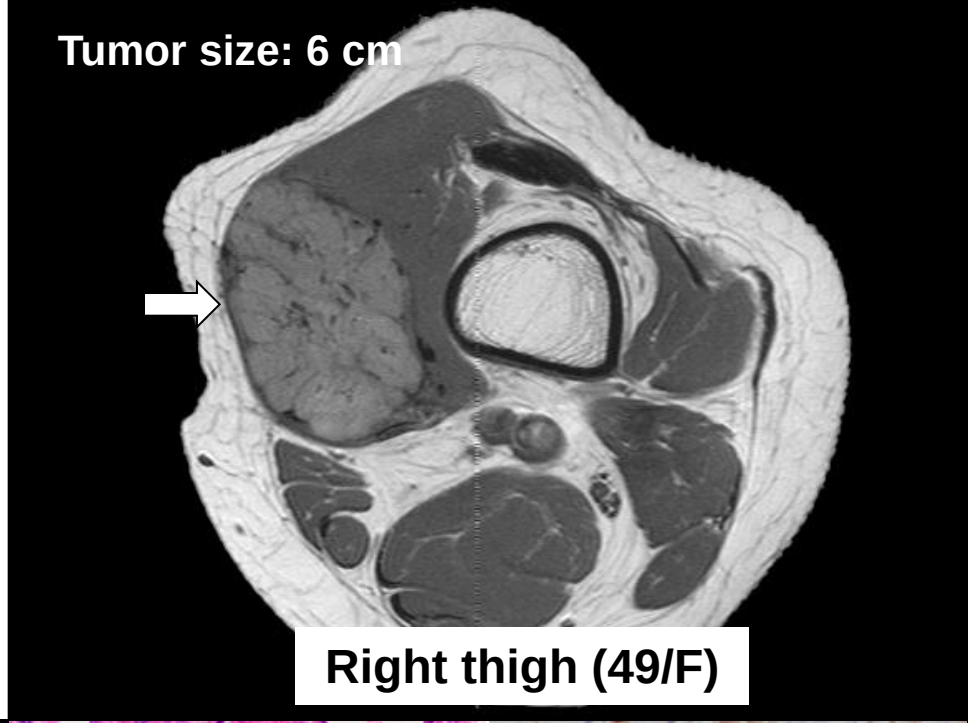
Dx: Alveolar soft part sarcoma

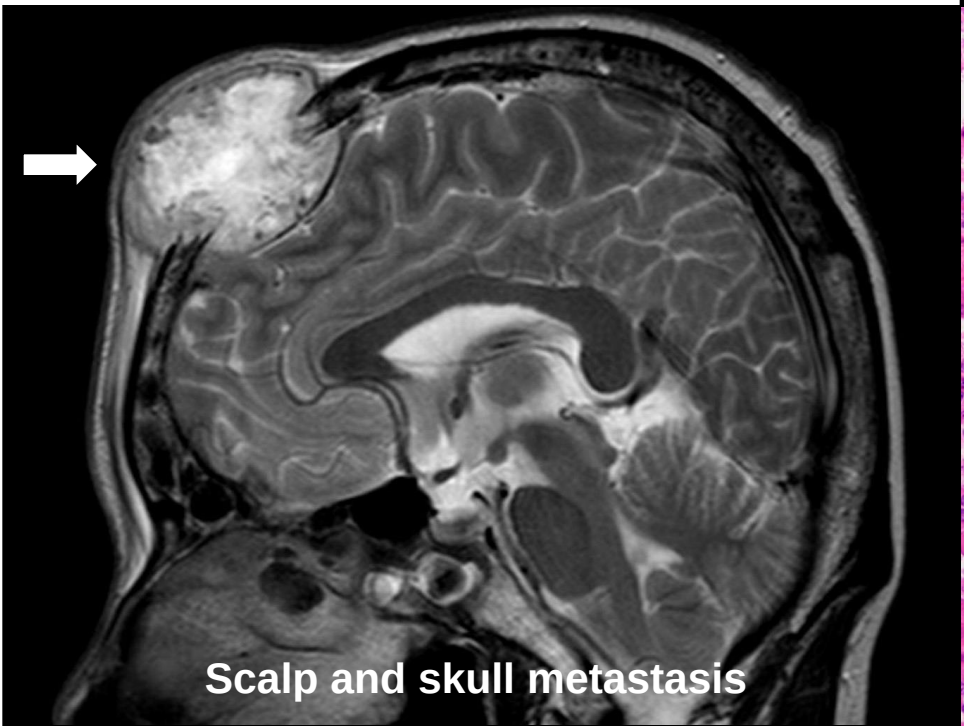
Pathological TNM (pTNM)

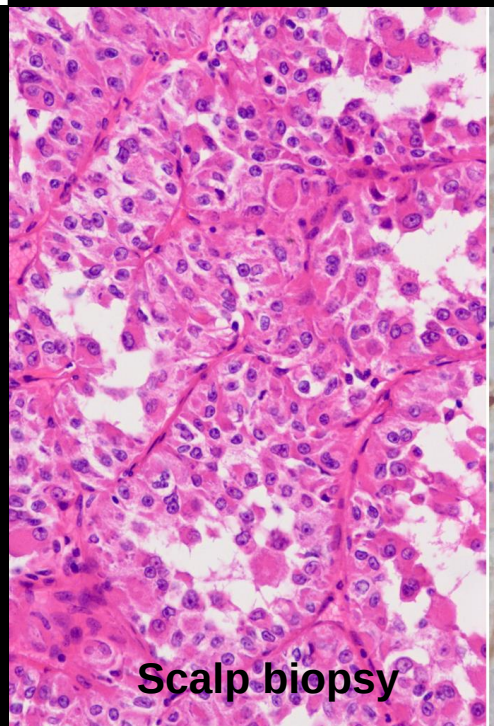
- Primary tumor (T): **pT2** *(>5 - ≤10 cm)*
- Regional lymph node (N): **cN0**
- Distant metastasis (M): **pM1**
- Histologic grade: **G3**

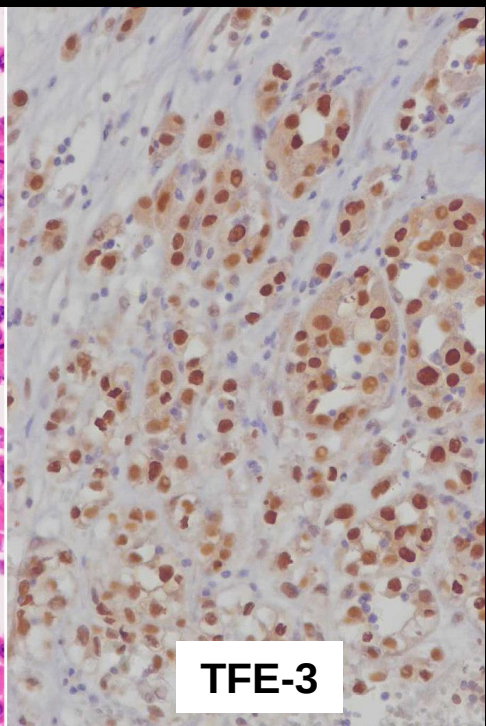
Stage group

- Stage **IV**









Checklist for standard reporting of soft tissue biopsy specimen recommended by CAP cancer protocol

Procedure
___ Core needle biopsy
___ Incisional biopsy
___ Excisional biopsy
___ Other (specify): _____
___ Not specified

Tumor Site
___ Head and neck (specify site, if known): _____
___ Trunk and extremities (specify site, if known): _____
___ Abdominal visceral organ(s) (specify site, if known): _____
___ Thoracic visceral organ(s) (specify site, if known): _____
___ Retroperitoneum (specify site, if known): _____
___ Orbit (specify site, if known): _____
___ Not specified

Histologic Type (World Health Organization [WHO] classification of soft tissue tumors)
Specify: _____
___ Cannot be determined

Mitotic Rate
Specify: ___ /10 high-power fields (HPF)
(1 HPF x 400 = 0.1734 mm^2; X40 objective; most proliferative area)

Necrosis
___ Not identified
___ Present
Extent: ___%
___ Cannot be determined

Histologic Grade (French Federation of Cancer Centers Sarcoma Group [FNCLCC])
___ Grade 1
___ Grade 2
___ Grade 3
___ Ungraded sarcoma
___ Cannot be assessed

Margins (for excisional biopsy only)
___ Cannot be assessed
___ Uninvolved by sarcoma
Distance of sarcoma from closest margin (centimeters): ___ cm
Specify margin: _____
Specify other close (<2.0 cm) margin(s): _____
___ Involved by sarcoma
Specify margin (s): _____

Lymphovascular Invasion
___ Not identified
___ Present
___ Cannot be determined

Additional Pathologic Findings
Specify: _____

Ancillary Studies (required only if applicable)

Immunohistochemistry (specify): _____
___ Not performed

Cytogenetics (specify): _____
___ Not performed

Molecular Pathology (specify):_____
___ Not performed

Prebiopsy Treatment (select all that apply)
___ No known prebiopsy therapy
___ Chemotherapy performed
___ Radiation therapy performed
___ Therapy performed, type not specified
___ Not specified

Treatment Effect
___ No known prebiopsy therapy
___ Not identified
___ Present
Specify percentage of viable tumor: ____%
___ Cannot be determined

Comment(s)

(College of American Pathologists, Version: Soft tissue 4.0.0.0, June 2017)

Checklist for standard reporting of soft tissue resection specimen defined by CAP cancer protocol (1)

Procedure
___ Intralesional resection
___ Marginal resection
___ Wide resection
___ Radical resection
___ Other (specify): _____
___ Not specified

Tumor Site
___ Head and neck (specify site, if known):
_____
___ Trunk and extremities (specify site, if known):
_____
___ Abdominal visceral organs (specify site, if known): _____
___ Thoracic visceral organs (specify site, if known): _____
___ Retroperitoneum (specify, if known):
_____
___ Orbit (specify site, if known): _____
___ Not specified

Tumor Size
Greatest dimension (centimeters): ___ cm
Additional dimensions (centimeters): ___ x ___ cm
___ Cannot be determined (explain):

Histologic Type (World Health Organization [WHO] classification of soft tissue tumors)
Specify: _____
___ Cannot be determined

Mitotic Rate
Specify: ___ /10 high-power fields (HPF)
(1 HPF x 400 = 0.1734 mm^2; X40 objective; most proliferative area)

Necrosis (macroscopic or microscopic)
___ Not identified
___ Present
Extent: ____%

Histologic Grade (French Federation of Cancer Centers Sarcoma Group [FNCLCC])
___ Grade 1
___ Grade 2
___ Grade 3
___ Ungraded sarcoma
___ Cannot be assessed

Margins
___ Cannot be assessed
___ Uninvolved by sarcoma
Distance of sarcoma from closest margin (centimeters): ___ cm
Specify closest margin: _____
Specify other close (less than 2.0 centimeters) margin (s) (if applicable):
_____
___ Involved by sarcoma
Specify margin (s): _____

Lymphovascular Invasion
___ Not identified
___ Present
___ Cannot be determined

Regional Lymph Nodes
___ No lymph nodes submitted or found

Lymph Node Examination (required only if lymph nodes present in specimen)

Number of Lymph Nodes Involved: _____
___ Number cannot be determined (explain):
_____

Number of Lymph Nodes Examined: _____
___ Number cannot be determined (explain):
_____

(College of American Pathologists, Version: Soft tissue 4.0.0.0, June 2017)

Checklist for standard reporting of soft tissue resection specimen defined by CAP cancer protocol (2)

Pathologic stage classification

TNM Descriptors (required only if applicable) (select all that apply)

___ m (multiple)
___ r (recurrent)
___ y (posttreatment)

Primary Tumor (pT)

Head and Neck

___ pTX: Primary tumor cannot be assessed
___ pT1: Tumor ≤2 cm
___ pT2: Tumor >2 to ≤4 cm
___ pT3: Tumor >4 cm
___ pT4: Tumor with invasion of adjoining structures
___ pT4a: Tumor with orbital invasion, skull base/dural invasion, invasion of central compartment viscera, involvement of facial skeleton, or invasion of pterygoid muscles
___ pT4b: Tumor with brain parenchymal invasion, carotid artery encasement, prevertebral muscle invasion, or central nervous system involvement via perineural spread

Trunk and Extremities

___ pTX: Primary tumor cannot be assessed
___ pT0: No evidence of primary tumor
___ pT1: Tumor 5 cm or less in greatest dimension
___ pT2: Tumor more than 5 cm and less than or equal to 10 cm in greatest dimension
___ pT3: Tumor more than 10 cm and less than or equal to 15 cm in greatest dimension
___ pT4: Tumor more than 15 cm in greatest dimension

Abdomen and Thoracic Visceral Organs

___ pTX: Primary tumor cannot be assessed
___ pT1: Organ confined
___ pT2: Tumor extension into tissue beyond organ
___ pT2a: Invades serosa or visceral peritoneum
___ pT2b: Extension beyond serosa (mesentery)
___ pT3: Invades another organ
___ pT4: Multifocal involvement
___ pT4a: Multifocal (2 sites)
___ pT4b: Multifocal (3-5 sites)
___ pT4c: Multifocal (>5 sites)

Retroperitoneum

___ pTX: Primary tumor cannot be assessed
___ pT0: No evidence of primary tumor
___ pT1: Tumor 5 cm or less in greatest dimension
___ pT2: Tumor more than 5 cm and less than or equal to 10 cm in greatest dimension
___ pT3: Tumor more than 10 cm and less than or equal to 15 cm in greatest dimension
___ pT4: Tumor more than 15 cm in greatest dimension

Orbit

___ pTX: Primary tumor cannot be assessed
___ pT0: No evidence of primary tumor
___ pT1: Tumor ≤2 cm in greatest dimension
___ pT2: Tumor >2 cm in greatest dimension without invasion of bony walls or globe
___ pT3: Tumor of any size with invasion of bony walls
___ pT4: Tumor of any size with invasion of globe or periorbital structures, including eyelid, conjunctiva, temporal fossa, nasal cavity, paranasal sinuses, and/or central nervous system

Regional Lymph Nodes (pN)

___ pN0: No regional lymph node metastasis or unknown lymph node status
___ pN1: Regional lymph node metastasis

Distant Metastasis (pM) (required only if confirmed pathologically in this case)

___ pM1: Distant metastasis
Specify site(s), if known: _____

Additional Pathologic Findings

Specify: _____

Ancillary Studies (required only if applicable)

Immunohistochemistry (specify): _____
___ Not performed
Cytogenetics (specify): _____
___ Not performed
Molecular Pathology (specify): _____
___ Not performed

Preresection Treatment (select all that apply)

___ No known preresection therapy
___ Chemotherapy performed
___ Radiation therapy performed
___ Therapy performed, type not specified
___ Not specified

Treatment Effect

___ No known presurgical therapy
___ Not identified
___ Present
Specify percentage of viable tumor (compared with pretreatment biopsy, if available): ____%
___ Cannot be determined

Comment(s)

Contents

- Introduction
- Bone
- Soft tissue sarcoma
- **Summary**
  - **Limitation in 8th AJCC cancer staging manual**
  - **Pathologist's role**
  - **Summary**

Limitation in 8th AJCC cancer staging manual

- An important **limitation** of 8th AJCC cancer staging manual is **lack of level I evidence** to support the staging proposals.

- **Further improvements** and **refinements** in the T- and overall staging for bone and soft tissue sarcoma will be necessary.

- The future 9th edition will be strengthened by a coordinated international effort to rigorously **analyze, improve,** and **validate the staging system.**

Pathologist's role

- Pathologists play a central role in the **diagnosis** and **staging** of tumors. Accurate microscopic diagnosis is essential to the evaluation and treatment of cancer.

- Pathologists must accurately report several **anatomic**, **histologic**, and **morphologic** characteristics of tumors, as well as **key histologic** features.

- **Pathological reporting** is best accomplished by using standardized nomenclature in a structured report, such as the synoptic reports or **cancer protocols** defined by CAP.

(AJCC 8th ed., 2017)

Summary

- In 8th edition AJCC cancer staging of bone and soft tissue sarcomas, **new separating staging system** based on anatomic sites and **new chapters/staging system** were reviewed.

- **Accurate staging** for cancer is the basis for clinical practice, treatment strategy, and prognosis determination.

- Cancer staging requires the **collaborative effort** of many professionals, including the physician, pathologist, radiologist, cancer registrar, and others.

Thank you for your attention