COLLECTING CANCER DATA: GIST/SARCOMA

2017-2018 NAACCR WEBINAR SERIES

Q&A

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

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Fabulous Prizes






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AGENDA

- Overview
- Epi Moment
- Quiz 1
- Staging
  - Bone
  - Soft Tissue
  - GIST
- Treatment
- Quiz 2
- Case Scenarios

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SARCOMA

ANATOMY

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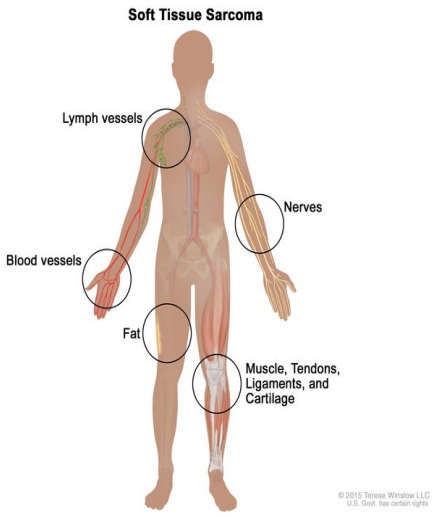
SOFT TISSUE SARCOMA

- Soft tissues include:
  - Muscles
  - Tendons
  - Fat
  - Blood vessels
  - Lymph vessels
  - Nerves
  - Tissues around joints



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SOFT TISSUE SARCOMA

- Over 50 different types
  - Angiosarcoma – blood vessels or lymph vessels; following radiation
  - Undifferentiated pleomorphic sarcoma – most often in arms or legs (previously malignant fibrous histiocytoma MFH)
  - Spindle cell sarcoma – descriptive name based on the appearance of the cells
  - Liposarcoma – fat cells; most frequently seen

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SOFT TISSUE SARCOMA

- **Pleomorphic dermal sarcoma**
  - Arise in sun-damaged skin
    - Head, Neck, Scalp
  - Negative: S100, Multiple CK, Desmin, CD34
  - Behavior likely more aggressive but limited follow up due to patient advanced age at presentation
- **Atypical Fibroxanthoma (AFX)**
  - Arise in sun-damaged skin
    - Ear, Nose, Forehead, Cheek
  - Negative: S100, multiple CK, Desmin, CD34
  - Behavior is almost invariably a benign behavior with only rare local recurrence

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SOFT TISSUE SARCOMA

- Question
  - What is the appropriate histology code for a final diagnosis or undifferentiated pleomorphic sarcoma and/or pleomorphic sarcoma, undifferentiated? Does the Other Sites MP/H Rule H17 apply in this case, which results in coding higher histology 8805/3? Or does the "undifferentiated" statement only refer to grade, which results in coding histology to 8802/3 (pleomorphic sarcoma)?

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ANSWER

- SINQ 20160044
  - Assign 8802/34 to pleomorphic cell sarcoma/undifferentiated pleomorphic sarcoma. Pleomorphic is more important than undifferentiated when choosing the histology code in this case. Undifferentiated can be captured in the grade code.

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SARCOMA

- **2018 Histology – New Terms**
  - 8571/3 Carcinoma with chondroid differentiation (C50._)
    Carcinoma with osseous differentiation (C50._)
    Metaplastic carcinoma with chondroid differentiation (C50._)
    Metaplastic carcinoma with osseous differentiation (C50._)

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SARCOMA

- **2018 Histology – New Terms**
  - 8801/3 Undifferentiated spindle cell sarcoma
  - 8802/3 Undifferentiated pleomorphic sarcoma
  - 8803/3 Undifferentiated round cell sarcoma
  - 8804/3 Undifferentiated epithelioid sarcoma
  - 8805/3 Undifferentiated uterine sarcoma
  - 8830/3 Undifferentiated high-grade pleomorphic sarcoma

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OSTEOSARCOMA

- Most common malignant bone tumor
  - Arise from osteoblasts
- Typically occurs in long bones
- Mutation in TP53 are most common
- Distant metastasis occur in ~20%
  - Lung is most common site

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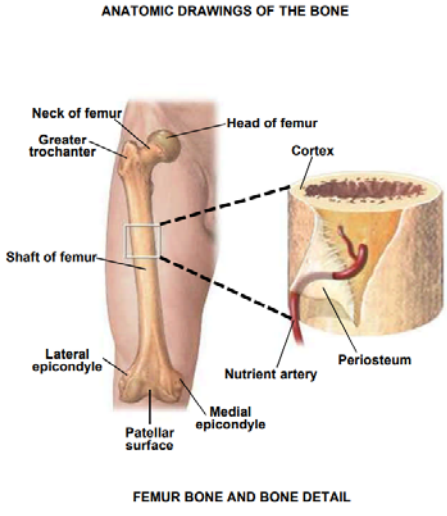
OSTEOSARCOMA

- Periosteum
  - 2 layers
    - Fibrous connective tissue
    - Inner osteogenic
- Medullary cavity
  - Bone marrow



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

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SOFT TISSUE SARCOMA GRADE

- FNCLCC grading system
  - Differentiation
  - Necrosis
  - Mitotic rate
- Histologic Grade:

GX	Grade cannot be assessed
G1	FNCLCC grade score of 2 or 3
G2	FNCLCC grade score of 4 or 5
G3	FNCLCC grade score of 6, 7, or 8

GIST

ANATOMY

WHAT IS GIST?

- Rare type of soft tissue sarcoma
  - Develop in muscle layer of gut rather than mucosa
  - Grow outward (exophytic)
- Described as a distinct entity in 1998
  - Umbrella term for most mesenchymal tumors of stomach and intestine
  - Most tumors historically called leiomyosarcoma are now classified as GISTs

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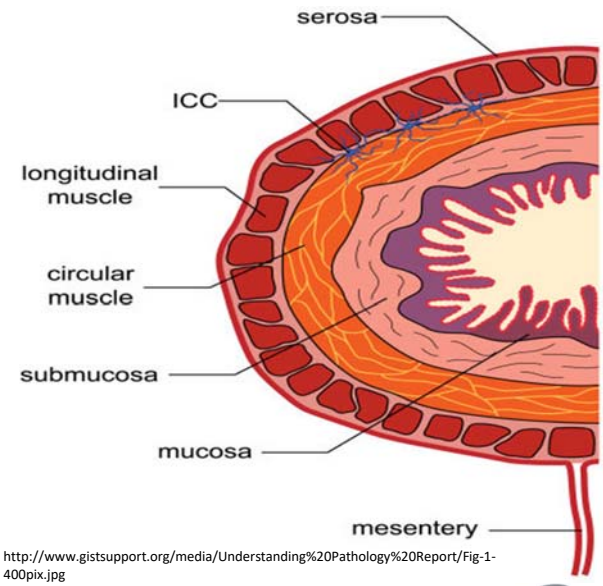
GIST

- Interstitial cells of Cajal
  - "Pacemaker cells"
  - Sends signals to move food and liquid through system (peristalsis)



http://www.gistsupport.org/media/Understanding%20Pathology%20Report/Fig-1-400pix.jpg

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ONCOGENIC MUTATIONS

- ~85% of GIST contain oncogenic mutations in one of two receptor tyrosine kinases
  - KIT-Mutant GIST
  - PDGFRA (Platelet-derived Growth Factor Receptor Alpha)
- Wild Type GIST
  - ~12-15% GIST contain no genetic mutation of KIT or PDGFRA

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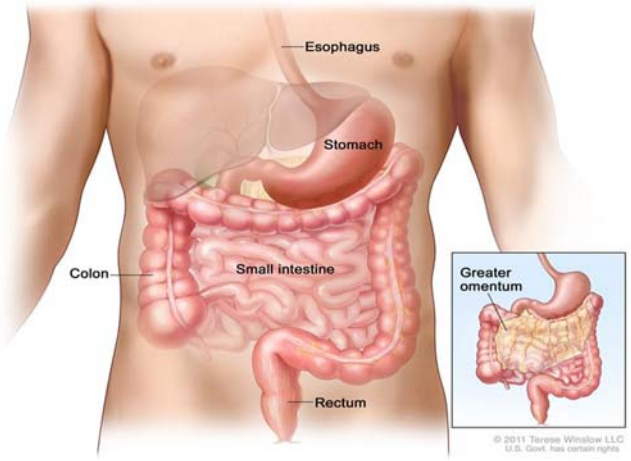
TUMOR LOCATION

Stomach	60%
Small Intestine	30%
Rectum	3%
Colon	1-2%
Esophagus	<1%
Omentum/Mesentery	Rare



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GIST

- How do you determine if a GIST is malignant i.e. reportable?
  - GIST, NOS is a borderline tumor (/1)
    - If your state or facility requires collection of these GISTs, you should follow their requirements
- Don't determine reportability based on staging
  - AJCC staging forms are used on all GISTs

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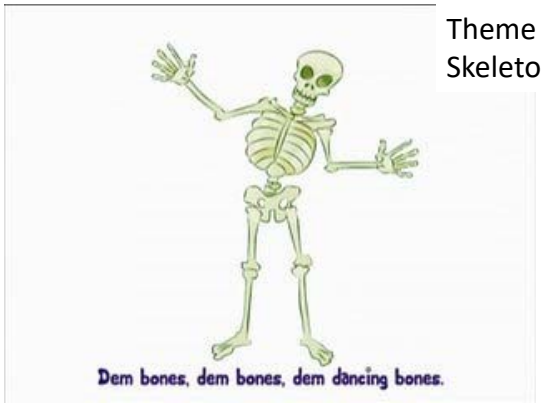

Theme Song: The Skeleton Dance

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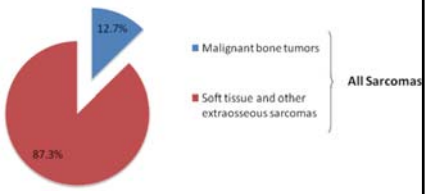
COLLECTING CANCER DATA: SARCOMA

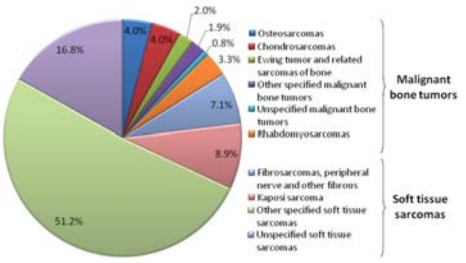
EPI MOMENT: RECINDA SHERMAN

JANUARY 11, 2018

EPIDEMIOLOGY OF SARCOMAS

- Large grouping of distinct cancers
  - 50+ distinct histologies; putative mesenchymal origin
  - Combined & studied as group
- Rare in adults (<2%); Top 5 for Pedi (21%)
- Majority soft tissue (87%); malignant bone (13%)
  - Soft: muscles, joints, fat, nerves, deep skin, blood vessels
  - Bone: commonly in cartilage
- Prognosis generally poor; esp soft
  - Delayed diagnosis: arise anywhere, lack of specific symptoms
  - No population based screening
  - Poor survival adults; better for pedi





Figures from: Burningham, Zachary, Mia Hashibe, Logan Spector, and Joshua D. Schiffman. "The Epidemiology of Sarcoma." Clinical Sarcoma Research (2012). BioMed Central Ltd. Web. 10 Feb. 2016.

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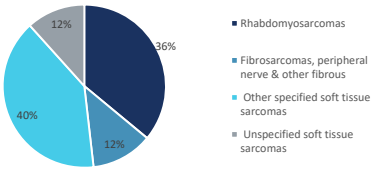
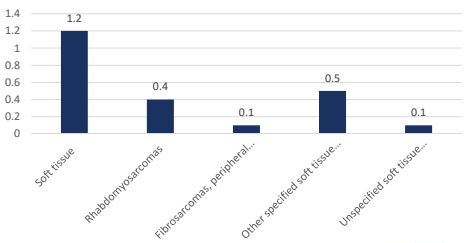
PEDIATRIC SARCOMAS (SOFT)

- Rhabdomyosarcoma – most common soft tissue for peds
  - Skeletal muscle
  - 50% occur <10; slightly more common in males
    - Often presents as painless mass; risk factor Li-Fraumeni syndrome
  - 5 year survival 70%; dependent upon location, stage, and histology—often lymph node involvement
    - Embryonal better prognosis than alveolar subtype; 20% present metastatic with 5 year survival 30-40% vs 80% for local
- Other
  - Fibrosarcoma – historically 2/3rds of sarcomas
    - now 12% due to better classification (proportion changed but not risk)
  - Liposarcoma – often large tumors; common among adults but <5% of ped sarcomas
  - Synovial sarcoma –4th most common; 2x more common in males
    - origin mesenchymal not synovium; largely genetic
  - Malignant peripheral nerve sheath tumors – grouped into Brain CNS category for Epi
  - Alveolar Soft Part Sarcoma (ASPS) – rare, slow growing; generally mets at dx
  - Mesenchymoma – rare but highly aggressive

Sarcoma (Soft), 0-19, CiNA 2014



Sarcomas (Soft), Rate per 1,000,000, CiNA 2014



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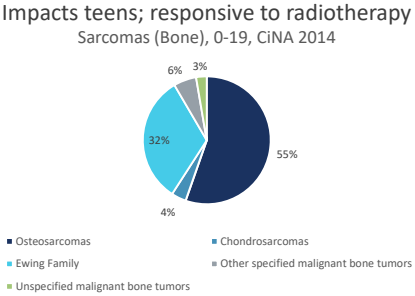
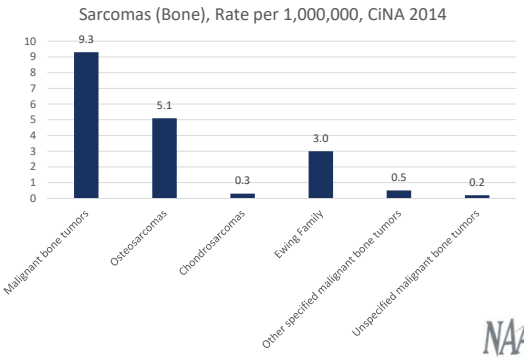
PEDIATRIC SARCOMAS (BONE)

- Osteosarcoma – most common
  - Generally on edges of "long" bones; 2nd most common location upper arm near shoulder
  - Surgery & Chemo
- Chondrosarcomas
- Ewing Family – 10-15% of bone sarcomas in peds
  - Ewing sarcoma, extraosseous Ewing, PNET, Askin)
  - Impacts teens; responsive to radiotherapy





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RISK FACTORS

- Varied causes; distinct disease; limited studies & understanding
  - Environmental, genetic (synergistic)
- Genetic
  - Age: Soft: Rate high <5 for soft, lower but steady increase 6-49; 50+ high; Bone: Rate stable across ages; Rate high in YA (osteosarcoma, Ewing)
  - Race: Ewing (9x more common white vs black); but soft tissue higher for blacks
  - Hx of hernia - Ewing sarcoma (children)
  - Growth "spurts" – osteosarcoma (children)
  - Genetic syndromes (Li-Fraumeni; neurofibromatosis/von Recklinghausen dx, retinoblastoma)
    - Non currently described for Ewing but likely genetic
- Environmental
  - HIV+ for KS (but HHV8 is causal, HIV & EBV))
  - Radiation exposure - Bone cancer; Tx and atomic (Japanese)
  - Occupational exposures --herbicides (soft tissue)

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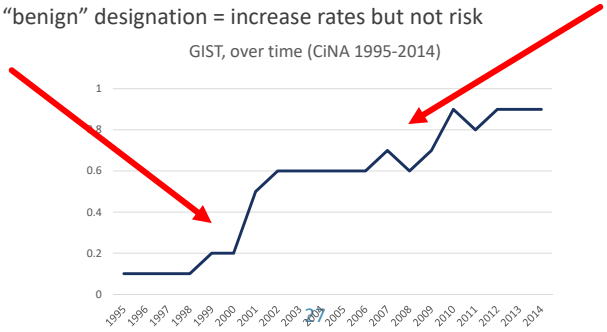
THE GIST ON G.I.S.T.[h/t Brad Wohler, FCDS]

- 2001 ICD-O-3 specific code
- Rare, digestive tract soft tissue sarcoma
  - Adult cancer; 50+
  - Used to believe origin was nerve or muscle cells
    - Now understood to arise from interstitial cells of Cajal (ICC) or precursor—the "pacemakers" of digestion; occur from esoph to anus but over ½ in stomach
  - Moving away from "benign" designation = increase rates but not risk

APC 7.8%*



QUESTIONS?

QUIZ 1

STAGING

SUMMARY STAGE

AJCC

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SUMMARY STAGE

2000 & 2018

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SUMMARY STAGE 2000-MUSCULOSKELETAL SYSTEM

- Bones, joints, and articular cartilage
  - C40.0-C40.3, C40.8-C40.9, C41.0-C41.4, C41.8-C41.9
- Peripheral nerves and autonomic nervous system; connective, subcutaneous, and other soft tissues
  - C47.0-C47.6, C47.8-C47.9, C49.0-C49.6, C49.8-C49.9
- Retroperitoneum and peritoneum
  - C48.0-C48.2, C48.8

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

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SUMMARY STAGE 2000-GIST

- Use location of the tumor to determine to summary stage chapter.
  - GIST of the stomach
    - Use summary stage chapter Stomach
  - GIST of the Ileum
    - Use Summary Stage chapter Small Intestine

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SUMMARY STAGE 2018

- GIST will have it's own chapter
- Sarcoma chapters have not yet been defined

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AJCC STAGE

REVIEW OF 7TH & 8TH EDITION

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BONE

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7TH EDITION CHAPTERS

Chapter Title	Chapter
Gastrointestinal Stromal Tumor	16
Bone	27
Soft Tissue Sarcoma	28

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8TH EDITION CHAPTERS

Chapter Title	Chapter Number	AJCC ID
Bone	38	38.1, 38.2, 38.3
Introduction (Information only)	39	NA
Head and Neck	40	40
Trunk and Extremities	41	41
Abdomen and Thoracic Visceral Organs	42	42
GIST	43	43
Retroperitoneum	44	44
Unusual Histologies and Sites (Information only)	45	45

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BONE-RULES FOR CLASSIFICATION

- Clinical
  - Imaging (MRI) followed by biopsy
- Pathologic
  - Resection of the primary tumor
  - cN may be used to assign the pathologic stage

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7TH EDITION-SIZE OF THE PRIMARY TUMOR

- Is the tumor 8cm or less?
- Are there discontinuous tumors present in the bone?

https://basicmedicalkey.com/osteosarcoma-4/

See page 286

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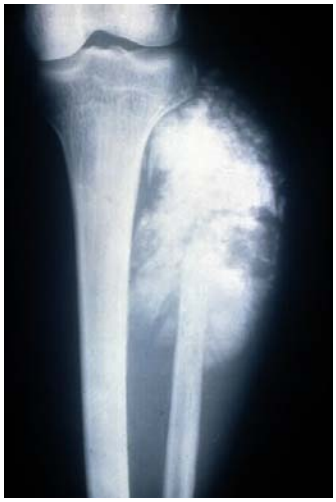
8TH EDITION- LOCATION, LOCATION, LOCATION

- Where is the tumor?
  - Appendicular skeleton, trunk, skull, facial bones
  - Spine
  - Pelvis



http://www.connectedkansaskids.com/diagnoses/osteosarcoma.html

See page 476

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APPENDICULAR SKELETON, TRUNK, SKULL, FACIAL BONES

- Is the tumor 8cm or less?
- Are there discontinuous tumors present in the bone?

See page 476

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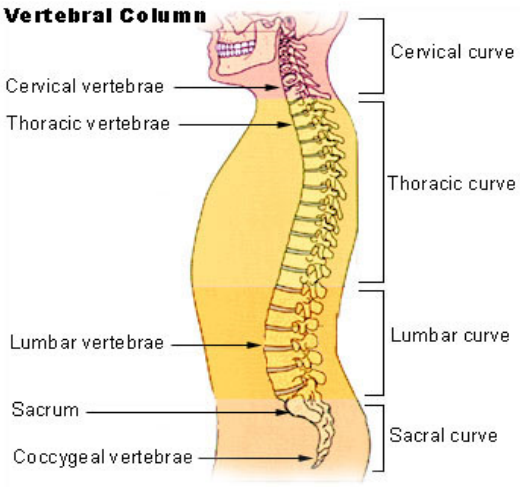
SPINE

- How many vertebral segments are involved?
- Is there spinal canal involvement?
- Is there involvement of the great vessels?



https://en.wikipedia.org/wiki/Vertebral_column

See page 476

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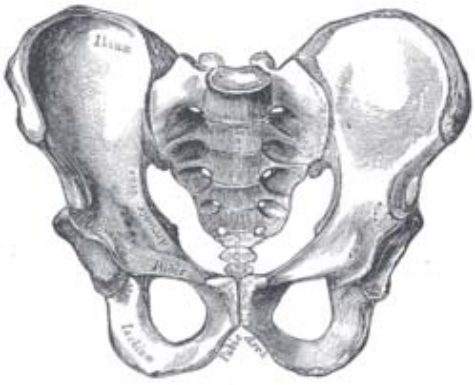
PELVIS

- How many segments of the pelvis are involved?
  - See fig 38.2 on page 474
- Is the tumor 8cm or less?
- Does the tumor cross the sacroiliac joint?
- Does tumor encase the external iliac vessel or cause tumor thrombus?



https://en.wikipedia.org/wiki/Pelvis

See page 476

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METASTASIS

- Lymph node metastasis is rare
  - cN values may be used in the pN data item
- Distant metastasis
  - Lung
    - Solitary tumor
    - Multiple tumors
  - Secondary bone
  - Other

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GRADE

- Cases diagnosed ≤ 2017 use the instructions for Coding Grade for 2014+
  - Two grade
    - Low-2
    - High-4
  - Four grade

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GRADE

- Cases diagnosed ≥ 2018 code Clinical, Pathologic, Post-Therapy Grade

Code	Grade Description
1	G1: Well differentiated, low grade
2	G2: Moderately differentiated, high grade
3	G3: Poorly differentiated, high grade
H	Stated as “high grade” only
9	Grade cannot be assessed (GX); Unknown; Not applicable

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STAGE GROUPING

- Appendicular Skeleton, Trunk, Skull, and Facial Bones
  - Grade is part of stage grouping
- Spine and Pelvis do not have stage group tables.
  - Stage group is 88

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SSF 3/SSDI-PERCENT NECROSIS POST NEOADJUVANT

- Record percentage value of the tumor necrosis post neoadjuvant chemotherapy as recorded in the pathology report from resection of the primary tumor.

Code	Description
0.0	Tumor necrosis not identified/not present
0.1-100.0	0.1 – 100.0 percent tumor necrosis (Percentage of tumor necrosis to nearest tenth of a percent)
XXX.2	Tumor necrosis present, percent not stated
XXX.8	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code XXX.8 will result in an edit error.)
XXX.9	Not documented in medical record No histologic examined of primary site No neoadjuvant therapy No surgical resection of primary site is performed

POP QUIZ 1

- A 73 year old male presented with a mass on his left femur.
  - An MRI showed a single 9cm mass confined to the femur.
  - A biopsy confirmed high grade chondrosarcoma.
  - The patient received neoadjuvant chemotherapy followed by surgical resection of the tumor.
  - Imaging showed post-therapy tumor size of 7cm.
  - The pathology report from the resected specimen showed a 7cm chondrosarcoma grade I. The extent of tumor necrosis was 95%.

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Data Item	7th ed	8th ed
Clinical T	cT2	cT2
Clinical N	cN0	cN0
Clinical M	cM0	cM0
Grade/ Clinical Grade	4	H
Stage	2B	2B
Path T	ypT1	
Path N	Blank or cN0	
Path M	cM0	
Path Grade		9
Stage	2B	

POP QUIZ 1

- A 73 year old male presented with a mass on his left femur.
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Data Item	7th ed	8th ed
Post Therapy T		ypT1
Post Therapy N		cN0
Post Therapy M		cM0
Post Therapy Grade		1
Post Therapy Stage		1A
Percent Necrosis Post Neoadjuvant/ SSF3	095	95.0

SOFT TISSUE

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7TH EDITION CHAPTER 28-SOFT TISSUE SARCOMA

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

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SOFT TISSUE SARCOMA-RULES FOR CLASSIFICATION

- Clinical Staging
  - Based on imaging and clinical evaluation prior to any treatment.
    - Tumor size can be measured clinically or radiographically (MRI or CT)
    - Evaluation for metastasis should be based on imaging. Most likely spot for distant metastasis is lungs.

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SOFT TISSUE SARCOMA-RULES FOR CLASSIFICATION

- Pathologic Staging
  - Based on resection of the primary tumor and clinical/radiologic evaluation for regional and distant metastasis.
    - Tumor size can be based on imaging if an accurate tumor size cannot be obtained from the resected specimen
    - If no lymph nodes removed, cN value may be used in the pN data item

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7TH EDITION CHAPTER 28-SOFT TISSUE SARCOMA

- Is the primary tumor ≤ 5cm?
- Is the tumor superficial or deep?
- No T3 or T4

Pg 297

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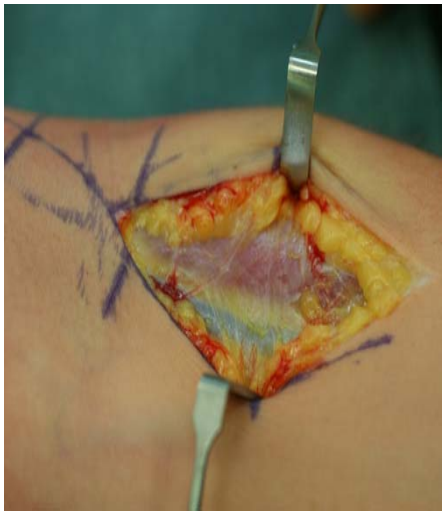
SUPERFICIAL VS DEEP

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



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

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8TH EDITION SOFT TISSUE SARCOMA

- Chapter 39-Introduction
- Chapter 40-Head and Neck
- Chapter 41-Trunk and extremities
- Chapter 42-Abdomen and Thoracic Visceral Organs
- Chapter 44-Retroperitoneum
- Chapter 45-Unusual Histologies and Sites

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SOFT TISSUE SARCOMA-RULES FOR CLASSIFICATION

- Clinical Staging
  - Based on imaging and clinical evaluation prior to any treatment.
    - Tumor size can be measured clinically or radiographically (MRI or CT)
    - Evaluation for metastasis should be based on imaging.

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SOFT TISSUE SARCOMA-RULES FOR CLASSIFICATION

- Pathologic Staging
  - Based on resection of the primary tumor and clinical/radiologic evaluation for regional and distant metastasis.
    - Tumor size can be based on imaging if an accurate tumor size cannot be obtained from the resected specimen
    - If no lymph nodes removed, cN value may be used in the pN data item

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SOFT TISSUE SARCOMA

Head and Neck

- C47.0 Peripheral nerves of head and neck
- C49.0 Connective, subcutaneous, and other soft tissues of head and neck
- C32.9 Larynx
- C02.3 Anterior 2/3 of tongue
- …

Pg 500

Trunk and Extremities

- C47.1 Peripheral nerves of the upper limb and shoulder
- C49.1 Connective, subcutaneous, and other soft tissues of the upper limb and shoulder
- C50.9 Breast

Pg 500

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SOFT TISSUE SARCOMA

Head and Neck

- Is the tumor ≤ 2cm?
- Is the tumor ≤ 4cm?
- Is the tumor invading adjacent structures?
- Stage group 88

Pg 503

Trunk and Extremities

- Is the tumor ≤ 5cm?
- Is the tumor ≤ 10cm?
- Is the tumor ≤ 15cm?
- Is the tumor more than 15cm?

Pg 511

Superficial vs Deep not a factor

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SOFT TISSUE SARCOMA

Abdomen and Thoracic Visceral Organs

- C47.0 Peripheral nerves thorax
- C49.0 Connective, subcutaneous, and other soft tissues of thorax
- C15-C26 Digestive organs
- C34-C37 Intrathoracic organs

Pg 518

Retroperitoneum

- C48.0-C48.8 Retroperitoneum

Pg 532

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SOFT TISSUE SARCOMA

Abdomen and Thoracic Visceral Organs

- Is the primary tumor confined to the organ of origin?
- Does the tumor invade into or through the serosa or visceral peritoneum?
- Does tumor invade another organ?
- Are there multiple tumors?
- Stage group 88

Pg 518

Retroperitoneum

- T, N, M similar to Trunk and extremities, but the stage group slightly different

Pg 532

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GRADE-CLIN, PATH, POST-THERAPY

- Codes 1-3 take priority over A-D.
- Codes A-D are equivalent to a GX when assigning AJCC Stage Group.

Code	Grade Description
1	G1: Sum of differentiation score, mitotic count score and necrosis score equals 2 or 3
2	G2: Sum of differentiation score, mitotic count score and necrosis score of 4 or 5
3	G3: Sum of differentiation score, mitotic count score and necrosis score of 6, 7, or 8
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated, anaplastic
9	Grade cannot be assessed (GX); Unknown; Not applicable

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CS SSF 3/SSDI-BONE INVASION

- Direct tumor extension from the primary sarcoma into adjacent bone.
- This field does not include distant or discontinuous metastases to the skeletal system.
- Information in this field is based on radiology and other imaging techniques.

Code	Description
0	Bone invasion not present/not identified on imaging
1	Bone invasion present/identified on imaging
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error.)
9	Not documented in medical record Bone invasion not assessed or unknown if assessed

POP QUIZ 2

- A patient presents with a 6cm mass in her left breast.
  - MRI: 5.7cm mass in the left breast. No additional masses identified.
  - A core biopsy confirmed fibrosarcoma. Grade could not be assessed due to inadequate specimen.
  - Modified radical mastectomy:
    - 5.7cm fibrosarcoma confined to the breast.
      - Mitotic Count Score: 11 per 10 HPF
      - Tumor Necrosis: 75% tumor necrosis
      - Differentiation Score: 2
      - FNCLCC grade 3
    - No bone involvement

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Data Item	7th ed	8th ed
Clinical T	cT2	cT2
Clinical N	cN0	cN0
Clinical M	cM0	cM0
Grade/ Clinical Grade	4	9
Stage	1B	1B
Path T	pT2	pT2
Path N	Blank or cN0	cN0
Path M	cM0	cM0
Path Grade		3
Stage	3	3A
SSF 3 Bone Invasion	000	0

GIST

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7TH EDITION CHAPTER 28-SOFT TISSUE SARCOMA
8TH EDITION CHAPTER 43-GASTROINTESTINAL STROMAL TUMOR

- No changes in 8th edition
- Follow Rules for Classification for Soft Tissue Tumors

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7TH EDITION CHAPTER 28-SOFT TISSUE SARCOMA
8TH EDITION CHAPTER 43-GASTROINTESTINAL STROMAL TUMOR

- Primary tumor is assessed based on size
  - Is the tumor ≤ 2cm?
  - Is the tumor ≤ 5cm?
  - Is the tumor ≤ 10cm?
  - Is the tumor more than 10cm?
- Regional node metastasis is extremely rare
  - cN values may be used in the pN data item
- Distant metastasis is rare, but may occur arise in intraabdominal soft tissue, liver (parenchyma), bone, soft tissues, and skin

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GRADE-CLIN, PATH, P-T

- Codes L and H take priority over A-D.
  - Codes A-D are equivalent to a GX when assigning AJCC Stage Group.
- Record the mitotic rate as Low or High as indicated on the pathology report or CAP protocol. Assume the denominator is 5 square mm if not specified.
  - Low: 5 or fewer mitoses per 5 mm2 (L)
  - High: Over 5 mitoses per 5 mm2 (H)
- SSF for pre-2018 cases

Code	Grade Description
L	Low: 5 or fewer mitoses per 5 mm^2
H	High: Over 5 mitoses per 5 mm^2
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated, anaplastic
9	Grade cannot be assessed; Unknown Not applicable

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GIST STAGE GROUPING

- Mitotic rate strongly influences stage group
- Stage grouping is different for tumors arising in the stomach and tumors arising in the small intestine
  - Primary omental GIST - Gastric Omental table (8th edition)
  - Tumors arising in sites other than stomach/omentum or small intestine should be grouped based on Small Intestine table

7th edition pg 177

8th edition pg 528

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SSF/SSDI-KIT GENE IMMUNOHISTOCHEMISTRY

- KIT immunohistochemistry is a special immunofluorescent stain that turns mutated cells brown and confirms a diagnosis of GIST.
- The presence of the KIT gene also indicates that the patient may respond to Gleevec or Sutent.

Code	Description
0	KIT negative/normal; within normal limits
1	KIT positive
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case
9	Not documented in medical record; Cannot be determined by pathologist; KIT not assessed or unknown if assessed

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SSDI-SCHEMA DISCRIMINATOR

- Since both omental and peritoneal gastrointestinal stromal tumors (GIST) are coded with the same ICD-O-3 topography code (C48.1), this data item must be used to identify the appropriate AJCC stage table.

Code	Description	Stage Table
1	Mesentery; Mesoappendix; Mesocolon; Pelvic peritoneum; Rectouterine pouch; Cul de sac; Pouch of Douglas; Other specified peritoneal site	Small Intestinal, Esophageal, Colorectal, Mesenteric and Peritoneal GIST
2	Omentum	Gastric and Omental GIST
9	Unknown or no information Not documented in patient record	Small Intestinal, Esophageal, Colorectal, Mesenteric and Peritoneal GIST

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POP QUIZ 3

- A patient presents with severe flank pain.
  - CT showed a 12.1 x 5.9cm hypodense mass mesenteric mass suspicious for a solid mass vs large hematoma. No associated lymphadenopathy. Surgery was recommended.
  - Pathology from Surgery
    - 8.2cm, cystic, hemorrhagic malignant spindle cell lesion with coagulative necrosis, most consistent with extra-gastrointestinal stromal tumor
    - CKIT, DOG1 and Vimentin were positive.
    - Mitosis rate 8/50hpf.

Primary Site: C48.1 Mesentery
Histology: 8936/3 GIST

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Data Item	7th ed	8th ed
Schema D 1		1
Clinical T		
Clinical N		
Clinical M		
Grade/ Clinical Grade	**9**	**9**
Stage	**99**	**99**
Path T	**pT3**	**pT3**
Path N	**Blank or cN0**	**cN0**
Path M	**cM0**	**cM0**
SSF 3 Mitotic Count Path Grade	**080**	**H**
Stage	**3B**	**3B**

TREATMENT

SARCOMA/GIST

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TREATMENT - OSTEOSARCOMA

- **Intramedullary**
  - Wide excision
- **Periosteal**
  - Consider neoadjuvant chemotherapy
  - Wide excision
- **High grade Intramedullary**
  - Neoadjuvant chemotherapy
  - If resectable – Wide Excision
    - Adjuvant chemotherapy +/- Radiation
  - If unresectable
    - Radiation
    - Chemotherapy

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TREATMENT OSTEOSARCOMA

- **Metastatic disease at diagnosis**
  - Resectable:
    - Wide excision local tumor
    - Excision of metastasis
    - Chemotherapy
  - Unresectable
    - Chemotherapy
    - Radiation

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SURGERY

C40.0-C41.9; C47.0-C47.9; C49.0-C49.9

- Local excision – 25
  - Excisional biopsy – tumor itself
- Partial resection – 26
  - Wide excision – more healthy tissue removed around tumor
- Radical excision or resection with limb salvage – 30
  - Significant amount of healthy tissue removed

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SURGERY

C40.0-C41.9; C47.0-C47.9; C49.0-C49.9

- Amputation of limb – 40
  - Partial amputation of limb – 41
    - Portion of the arm or leg
  - Total amputation of limb – 42
    - Leg and the hip
    - Arm and the shoulder

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POP QUIZ

- 48yo with large mass on right 3rd toe
- Incisional biopsy right 3rd toe reveals sarcoma
- Decision to amputate the entire right 3rd toe
- Pathology R 3rd toe, amputation: pleomorphic sarcoma, margins negative within 1cm.

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POP QUIZ

- Diagnostic/Staging Procedure:
  - 02
- Surgery Primary Site:
  - 30
- Scope Regional LN Surgery:
  - 0

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RADIATION

- Primary Tumor Volume /Phase I Radiation Primary Treatment Volume

FORDS-2017	STORE-2018	STORE Description
25	80	Skull
24	81	Spine/Vertebral Bodies
37	82	Shoulder
26	83	Ribs
27	84	Hip
28	85	Pelvic Bones
38	88	Extremity bone, NOS
30	90	Skin
31	91	Soft Tissue

RADIATION

- Regional Modality /Phase I Radiation Treatment Modality

FORDS-2017	STORE-2018	Definition
21	01	External beam, photons, low energy
22-27, 31, 41, 42, 43	02	External beam, photons, megavoltage
40	03	External beam, protons
28	04	External beam, electrons
30	05	External beam, neutrons
20	06	External beam, carbon ions
29	09	External beam, NOS

RADIATION

- Regional Modality/Phase I External Beam Radiation Planning Technique

FORDS Modality-2017	STORE-2018	STORE Description
20, 22-27, 29, 30, 40	01	External beam, NOS
21	02	Low energy x-ray/photon
28	03	2-D therapy
32	04	Conformal or 3-D conformal therapy
31	05	Intensity Modulated Therapy
41, 42	06	Stereotactic radiotherapy or radiosurgery, NOS
	07	Stereotactic radiotherapy or radiosurgery, robotic
43	08	Stereotactic radiotherapy or radiosurgery, Gamma Knife
	09	CT guided online adaptive therapy
	10	MR guided online adaptive therapy

POP QUIZ 4

- A 48 year old patient with a recent right 3rd toe amputation for sarcoma presents to discuss radiation treatment.
  - Plan: 6MV photons, IMRT to right foot, surgical bed
- The patient completed radiation on 8/1/2017, right foot surgical bed, 6MV photons, 1800cGy in 6fx

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POP QUIZ

Radiation Data Items	2017	2018
Radiation Primary Tumor Volume	38	88
Radiation Treatment Modality	31	02
External Beam Radiation Planning Technique		05

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SYSTEMIC - CHEMOTHERAPY

- Isolated Limb Infusion
- Ifosfamide and Doxorubicin
  - Mesna – used with Ifosfamide
- MAID – Mesna, Adriamycin, Ifosfamide, Dacarbazine
- Pazopanib (Votrient)
  - Blocks Tyrosine Kinases

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SYSTEMIC – IMMUNOTHERAPY

- Olaratumab (Lartruvo)
  - FDA approved 10/19/16
  - Radiation or surgery not curative
  - In combination with Doxorubicin

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TREATMENT – GIST

Gastric:

- Surgery
  - Small tumors removed laproscopically

Other Sites:

- Surgery
- Targeted Therapy
  - Imatinib (Gleevec) - Chemotherapy

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TREATMENT – GASTRIC SURGERY

- Excisional biopsy (NOS) – 27
  - WITH electrocautery – 22
  - WITH cryosurgery – 23
  - WITH laser ablation – 24
- Laser excision – 25
- Gastrectomy – 30
  - Antrectomy, lower – 31
  - Lower gastrectomy – 32
  - Upper gastrectomy – 33

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TREATMENT – OTHER SITES SURGERY

C15.0-C15.9

- Excisional Biopsy (NOS) – 27
  - WITH electrocautery – 22
  - WITH Cryosurgery – 23
  - WITH Laser ablation – 24
- Partial Esophagectomy - 30

C18.0-C18.9; C19.9; C20.9

- Excisional biopsy – 27
  - WITH electrocautery – 22
  - WITH cryosurgery – 23
  - WITH laser ablation – 24
- Segmental resection - 30

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POP QUIZ 5

- A 52 year old male presents with chronic anemia. He has never had a colonoscopy. Plan: screening colonoscopy
- Colonoscopy – Rectal mass, biopsy performed; no other abnormalities noted
- Rectum mass, biopsy: malignant GIST
- Patient returns for excision of rectal GIST
- Rectum, excision mass: malignant GIST, 3cm, mitoses: 6/50HPF, KIT+

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POP QUIZ 5 CONT.

- Rectal GIST with high mitotic rate, 3cm, and KIT + is here to discuss adjuvant treatment options. Plan is to begin Gleevec
- Patient here for 3mo follow up for rectal GIST, started Gleevec and is doing well with minimal side effects

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POP QUIZ 5 CONT.

Treatment data items	Codes
Surgery Primary Site	27
Scope Regional LN Surgery	0
Chemotherapy	02
Immunotherapy	00
Systemic/Surgery Sequence	3

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QUESTIONS?

QUIZ 2

CASE SCENARIOS

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COMING UP….

- Collecting Cancer Data: Stomach and Esophagus
  - 02/01/2018
- Boot Camp!
  - 3/1/18

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Fabulous Prizes Winners






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CE CERTIFICATE QUIZ/SURVEY

- Phrase

Necrosis

- Link

http://www.surveygizmo.com/s3/4103019/Sarcoma-2018

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