## Case Scenario 1

A 65 year old white female presents with a urinary tract infection and abdominal pain.

**CT Abdomen/Pelvis:**

There is a small sliding hiatal hernia.

There is a solid mass with heterogeneous density and a markedly heterogeneous enhancement pattern seen in left upper quadrant between the spleen and the left kidney, which is displaced caudally. Mass measures 11.6 cm x 11.8 cm x 14.6 cm. No hemorrhage or calcifications are detected within it. Small amount of free fluid is seen along its caudal margin. Left adrenal gland is not separately identified. Aside from displacement, spleen and left kidney are normal.

The liver, gallbladder, bile ducts, pancreas, and the right adrenal gland are normal. The right kidney contains small cysts. It is otherwise negative.

No evidence of small or large bowel obstruction. Apex is identified. No inflammatory changes are seen.

Impression:

1. Large left upper quadrant tumor mass. Differential diagnosis includes pheochromocytoma, adrenal carcinoma or other adrenal mass, lymphoma, exophytic left-sided renal cell carcinoma and metastatic disease.
2. There is a 2.3 cm x 3.7 cm x 4.4 cm subcutaneous cyst along the left costal margin. This could be confirmed with sonography if appropriate.
3. Small sliding hiatal hernia.

**Operative Report:**

1. RESECTION OF A LARGE LEFT-SIDED RETROPERITONEAL MASS.
2. SPLENECTOMY.
3. TAKEDOWN OF SPLENIC FLEXURE.

This was a very large lesion, which was heavy. I am suspecting it weighs 8-10 pounds. It was, however, separate from the kidney and actually was separate from the adrenal gland. We could see the adrenal gland once the mass was removed, and it remained intact. The mass was adherent to the lower pole of the spleen. There was a fibrous capsule which was also resected after the primary tumor had been removed, once the pathologist told us that this was sarcoma. We also removed the spleen due to the tissue diagnosis. There was some inferior pole bleeding, and we opted to remove the spleen based on the sarcomatous nature.

**Pathology:**

* Soft tissue, abdominal mass excision:
	+ High grade pleomorphic liposarcoma (weight = 2,200 gm, measuring 22.0 x 16.0 x 15.0 cm)
* Spleen: no significant abnormalities
* Soft tissue “tumor capsule” excision: involvement by high grade pleomorphic liposarcoma
	+ Addendum report is issued to clarify the status of surgical excision margins.
		- Inked margins are free of tumor; however, pleomorphic liposarcoma is present 0.1 mm to less than 0.1 mm from inked resection margins of the specimen (part A).
		- Staging form is present and completed as pT2NXM0 stage III

**Treatment Summary:**

Patient received 45 Gy of external beam radiation utilizing 18 MV in 25 sessions using to the tumor bed in the upper abdomen utilizing parallel opposed RAO/LPO oblique fields. She received an additional 5.4 Gy in 3 treatments as a boost to the tumor bed, again utilizing 18 MV photons in opposing RAO/LPO oblique fields for a total of 50.4 Gy in 28 sessions to the tumor bed. Treatments proceeded from May 19 to June 26. She tolerated the treatments exceptionally well experiencing no nausea, vomiting, diarrhea, or pain.

**5 month follow up CT abdomen/pelvis:**

1. Interval development of small bilateral pleural effusions with bilateral lower lobe atelectasis.
2. Decrease in size of several left subphrenic fluid collections since which may represent postoperative seromas.
3. Development of multiple peritoneal masses throughout the abdomen especially on the right side, most consistent with peritoneal metastatic disease.
4. Small amount of ascites.
5. Interval enlargement of left adnexal cystic lesion although adjacent previously described coarse calcifications remained stable.
6. Patient treated palliatively with gemcitabine and taxotere.

Patient expired 6 months later.

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| * **What is the primary site?**

Soft tissue of abdomen – C49.4* **What is the histology?**

Pleomorphic liposarcoma – 8854/3 | * **What is the grade/differentiation?**

High grade – 4 (9 if following CoC rules)* **What is grade path system/grade path value?**

Blank |
| **Stage/ Prognostic Factors** |
| CS Tumor Size | 220 | CS SSF 9 | 988 |
| CS Extension | 120 | CS SSF 10 | 988 |
| CS Tumor Size/Ext Eval | 3 | CS SSF 11 | 988 |
| CS Lymph Nodes  | 000 | CS SSF 12 | 988 |
| CS Lymph Nodes Eval | 0 | CS SSF 13 | 988 |
| Regional Nodes Positive | 98 | CS SSF 14 | 988 |
| Regional Nodes Examined | 00 | CS SSF 15 | 988 |
| CS Mets at Dx | 00 | CS SSF 16 | 988 |
| CS Mets Eval | 0 | CS SSF 17 | 988 |
| CS SSF 1 | 200 | CS SSF 18 | 988 |
| CS SSF 2 | 000 | CS SSF 19 | 988 |
| CS SSF 3 | 000 | CS SSF 20 | 988 |
| CS SSF 4 | 000 | CS SSF 21 | 988 |
| CS SSF 5 | 988 | CS SSF 22 | 988 |
| CS SSF 6 | 988 | CS SSF 23 | 988 |
| CS SSF 7 | 988 | CS SSF 24 | 988 |
| CS SSF 8 | 988 | CS SSF 25 |  |
| **Treatment** |
| Diagnostic Staging Procedure | 00 |  |  |
| **Surgery Codes** |  | **Radiation Codes** |  |
| Surgical Procedure of Primary Site | 25 | Radiation Treatment Volume | 17 |
| Scope of Regional Lymph Node Surgery | 0 | Regional Treatment Modality | 25 |
| Surgical Procedure/ Other Site | 2 | Regional Dose | 04500 |
| **Systemic Therapy Codes** |  | Boost Treatment Modality | 25 |
| Chemotherapy | 00 | Boost Dose | 00540 |
| Hormone Therapy | 00 | Number of Treatments to Volume | 28 |
| Immunotherapy | 00 | Reason No Radiation | 0 |
| Hematologic Transplant/Endocrine Procedure | 00 | Radiation/Surgery Sequence | 3 |
| Systemic/Surgery Sequence | 0 |  |  |

## Case 2

MRI Left Femur:

There is a sharply demarcated lesion within the femoral epiphysis that extends through the metaphysis into the diaphysis. This lesion is slightly hyperintense to muscle on T1 contains very mixed predominantly bright and somewhat macronodular T2 signal and has associated extensive reactive joint effusion with intramuscular edema throughout the vastus lateralis extending into the proximal aspect of the thigh along the IT band. The proximal extent from the intercondylar femur extends for 8.5 cm. There is no mid-diaphyseal or proximal femoral lesion. IV contrast not administered.

 The lesion does have a cortical breach laterally at the level of the metaphysis with soft tissue component extending approximately 14 mm beyond the peripheral cortical margin. Extensive cortical destruction is noted. There is preserved signal intensity within the tibia and fibula.

Intramuscular edema also involves both portions of the vastus medialis at the junction middle and distal third of the thigh as well as lateral hamstring short head of the biceps femoris. The mass is separate from the neurovascular bundle. There are a few small popliteal fossa lymph nodes noted.

IMPRESSION:

Diffuse signal abnormality, primary bone neoplasm of the femoral epiphysis and diametaphysis with associated lateral extension into the quadriceps. Note this lesion has significantly different signal characteristics than the T9 vertebral body lesion and has somewhat of a chondroid pattern on T2 rather than dense osteoid.

Bone scan:

1. Abnormally increased tracer accumulation in the distal left femur corresponding to patient's known mass lesion.
2. Three abnormal but somewhat nonspecific sites of tracer accumulation -- one involving the body of T9, another in the left superior orbital rim, and the third in the left lateral side of the mandible. Differential considerations include fibrous dysplasia, Paget's disease, and metastatic disease.

CT Facial Bones:

IMPRESSION: Diffuse very subtle increased medullary sclerosis of the non-aerated left frontal bone in the region of the frontal sinus. Perhaps fibrous dysplasia and a polyostotic process including the T9 vertebral body is a precursor to the known differentiated femoral malignancy. This CT appearance is not consistent with disseminated metastatic disease

MRI Thoracic Spine:

IMPRESSION: Diffuse marrow replacing process at T9. In light of the very subtle CT findings in the left frontal sinus, more aggressive findings and cortical breach of the distal femur, some type of fibrous or other osseous osteoid dysplasia as an underlying polyostotic cause could be involved in this young woman. Core biopsy could be considered.

CT Chest/Abdomen:

1. Negative for metastatic disease.
2. Patchy linear areas of scarring involving the lingula and left lower lobe.

PET/CT: IMPRESSION:

1. Markedly increased FDG accumulation within the patient's known malignant lesion in the distal left femur.
2. Markedly increased FDG accumulation within the body of T9 corresponding to the abnormalities visualized on MRI and bone scintigraphy. This is highly suspicious for a metastatic lesion.
3. Focus of abnormally increased FDG accumulation in the posterior superior aspect of the left iliac bone which corresponds to a site of abnormal tracer accumulation on yesterday's bone scan. This is highly suspicious for malignancy.
4. No abnormal FDG accumulation to correspond to the sites of increased bone tracer accumulation in the left supraorbital region and lateral the left mandible. This favors non-malignant etiologies for the increased uptake observed in both sites on yesterday's bone scan.
5. Mild FDG accumulation within a small ill-defined region of opacification in the inferior aspect of the left lower lobe, and within a small curvilinear density in the lingula. These are felt to most likely represent focal regions of infiltrate/inflammation however, is not entirely excluded. A followup chest CT in 3 to 6 months may be helpful.

Pathology Report:

* Bone, T9 vertebrae FNA/Bx: involvement by high grade osteosarcoma
* Bone left iliac crest FNA/Bx: involvement by high grade osteosarcoma

Treatment Summary:

* On protocol COG AOST0331-MAP:
	+ Doxorubicin, cisplatin, mannitol followed 3 weeks later by methotrexate, leucovorin rescue

Post chemotherapy MRI femur:

1. Re-demonstration of a distal femur lesion compatible with given history of osteosarcoma. Overall, similar appearance of lesion when compared to prior MRI exam.
2. Mild interval increase in size of 3 popliteal lymph nodes, measuring up to 1 cm.
3. Interval increase of T2 signal in the proximal soleus and lateral head of the gastrocnemius.
4. Interval decrease in size of the enhancing adjacent knee joint effusion.

Patient had widespread metastasis and went on to have more chemotherapy, radiation, and surgeries.

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| * **What is the primary site?**

Femur – C40.2* **What is the histology?**

Osteosarcoma – 9180/3 | * **What is the grade/differentiation?**

9 (high grade from metastatic site)* **What is grade path system/grade path value?**

Blank |
| **Stage/ Prognostic Factors** |
| CS Tumor Size | 085 | CS SSF 9 | 988 |
| CS Extension | 400 | CS SSF 10 | 988 |
| CS Tumor Size/Ext Eval | 0 | CS SSF 11 | 988 |
| CS Lymph Nodes  | 000 | CS SSF 12 | 988 |
| CS Lymph Nodes Eval | 0 | CS SSF 13 | 988 |
| Regional Nodes Positive | 98 | CS SSF 14 | 988 |
| Regional Nodes Examined | 00 | CS SSF 15 | 988 |
| CS Mets at Dx | 40 | CS SSF 16 | 988 |
| CS Mets Eval | 3 | CS SSF 17 | 988 |
| CS SSF 1 | 999 | CS SSF 18 | 988 |
| CS SSF 2 | 999 | CS SSF 19 | 988 |
| CS SSF 3 | 998 | CS SSF 20 | 988 |
| CS SSF 4 | 000 | CS SSF 21 | 988 |
| CS SSF 5 | 988 | CS SSF 22 | 988 |
| CS SSF 6 | 988 | CS SSF 23 | 988 |
| CS SSF 7 | 988 | CS SSF 24 | 988 |
| CS SSF 8 | 988 | CS SSF 25 | 988 |
| **Treatment** |
| Diagnostic Staging Procedure | 01 |  |  |
| **Surgery Codes** |  | **Radiation Codes** |  |
| Surgical Procedure of Primary Site | 00 | Radiation Treatment Volume | 00 |
| Scope of Regional Lymph Node Surgery | 0 | Regional Treatment Modality | 00 |
| Surgical Procedure/ Other Site | 0 | Regional Dose | 00000 |
| **Systemic Therapy Codes** |  | Boost Treatment Modality | 00 |
| Chemotherapy | 03 | Boost Dose | 00 |
| Hormone Therapy | 00 | Number of Treatments to Volume | 000 |
| Immunotherapy | 00 | Reason No Radiation | 1 |
| Hematologic Transplant/Endocrine Procedure | 00 | Radiation/Surgery Sequence | 0 |
| Systemic/Surgery Sequence | 0 |  |  |

## Case Scenario 3

Operative Report:

15 year male presents for biopsy of a mass located near the left knee.

Left distal femur biopsy:

High grade osteoblastic osteosarcoma grade 3(of 3)

MRI Femur:

Again noted is an approximately 4.7 x 3.4 x 10.6 cm enhancing interosseous mass within the distal medial metaphysis and epiphysis of the left femur with an associated medial cortical destruction and a contiguous enhancing mass extending into the medial extraosseous soft tissues which measures approximately 2.2 x 4.8 x 8.0 cm. There also subtle areas of heterogeneous T2 signal hyperintensity within the adjacent bone marrow the distal left femur encompassing approximately the distal third of the length of the shaft. There is a nonenhancing rind of fluid and edema surrounding this interosseus/extraosseous mass. The remainder of the visualized bone marrow and cortex of the proximal two-thirds of the left femur is within normal limits and MR signal characteristics. A moderate left knee joint effusion is once again noted.

There has been interval development of edema interdigitating within the superficial muscle fascicles, as well as fluid tracking superficial to the vastus medialis extending approximately to the level of the mid-thigh without enhancement. There has been interval amount of edema and enhancement within the adjacent medial subcutaneous fat. There has also been an interval development of non-enhancing fluid tracking adjacent to the medial cortex of the left femur extending superiorly to the level of the mid femur. These new findings may be secondary to interval tissue sampling procedure.

 IMPRESSION:

Re-identified mass within the medial aspect of the metaphysis and epiphysis of the left distal femur with associated cortical destruction and mass extending into the adjacent soft tissues.

 No distinct mass or signal abnormalities within the proximal two-thirds of the

 left femur to suggest additional or satellite lesions.

Interval development of areas of fluid and edema within the soft tissues of the mid to distal thigh, which likely represents interval post procedure changes.

Bone scan:

* Markedly increased tracer accumulation corresponding to patient's known mass in the distal medial aspect of the left femur. This is consistent with the patient's biopsy-proven osteosarcoma.
* No additional suspicious sites of abnormal tracer accumulation are visualized elsewhere.

Treatment Summary:

Patient on protocol COG AOST-331-MAP: Doxorubicin, mannitol, cisplatin followed 3 weeks later by methotrexate, leucovorin rescue

Post chemo MRI Left Femur:

There is normal marrow signal within the bony pelvis and proximal femurs. The mid femoral diaphysis also has normal signal intensity.

Abnormal signal extends longitudinally throughout the diametaphysis for a distance of 8.5 cm above the physeal line within the medial thigh.

Architecturally, the tumor is much more heterogeneous in T1 signal intensity and has lesser degree of periosteal extension. There is longitudinal breech through the physeal line into the medial epiphysis below the level of PCL origin. Tumor dimensions transverse 6.3 cm, craniocaudad 10.5 cm and is AP maximum 5.8 cm.

The tumor spares the popliteal neurovascular bundle. Approximately at the level of the quadriceps tendon, there is extension to involve the neurovascular branches of the vastus medialis of the descending genicular group. Post-contrast the lesion demonstrates vigorous enhancement shown coronally. There is no enhancing knee joint effusion. Diffuse subcutis edema which had been shown previously surrounding the vastus medialis has resolved.

 Tumor abuts the origin fibers medial gastrocnemius.

 IMPRESSION:

 Overall little change in volume of diffusely enhancing metaphyseal osteosarcoma.

Tumor margins as described. Inferior extension below the posterior cruciate ligament origin, anteromedial cortical breach and enhancing tumor in immediate proximity to the origin of the medial gastrocnemius.

Architecturally, the mass is now centrally more heterogeneous in T1 signal with persistent vigorous post-contrast enhancement.

**OPERATION**

RADICAL RESECTION LEFT DISTAL FEMUR BONE TUMOR WITH TOTAL KNEE RECONSTRUCTION USING OSS DISTAL FEMORAL REPLACEMENT ARTHROPLASTY.

**Pathology: Final Diagnosis:**

A) BONE, LEFT DISTAL FEMUR, RESECTION:

 - OSTEOSARCOMA, HIGH-GRADE (GRADE 3 OF 3), OSTEOBLASTIC TYPE,

WITH POST-CHEMOTHERAPY CHANGES.

 - TUMOR MEASURES 10.5 X 5.5 X 5 CM IN MAXIMUM DIMNESIONS.

 - TUMOR SHOWS APPROXIMATELY 80% TUMOR CELL NECROSIS.

 - EVIDENCE OF PRIOR FOCAL TUMOR EXTENSION INTO PERIOSTEAL CONNECTIVE TISSUE IS IDENTIFIED.

 - LYMPH-VASCULAR INVASION IS NOT IDENTIFIED.

 - RESECTION MARGINS FREE FROM MALIGNANCY.

B) BONE, PROXIMAL FEMORAL MARGIN, BIOPSY:

 - NEGATIVE FOR MALIGNANCY.

COMMENT: The sections of the post-chemotherapy left distal femur resection specimen show high grade, osteosarcoma, osteoblastic type, with chemotherapy related changes. Approximately 80% of the tumor shows tumor cell necrosis. There is evidence of prior focal tumor extension beyond the periosteum into the periosteal connective tissue. Lymph-vascular invasion is not seen. All specimen margins are negative. Chemo continued post-op.

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| * **What is the primary site?**

Femur – C40.2* **What is the histology?**

Osteoblastic osteosarcoma – 9180/3 | * **What is the grade/differentiation?**

High grade; grade 3 of 3 – 4* **What is grade path system/grade path value?**

Grade 3 of 3 – 3; 3 |
| **Stage/ Prognostic Factors** |
| CS Tumor Size | 106 | CS SSF 9 | 988 |
| CS Extension | 400 | CS SSF 10 | 988 |
| CS Tumor Size/Ext Eval | 5 | CS SSF 11 | 988 |
| CS Lymph Nodes  | 000 | CS SSF 12 | 988 |
| CS Lymph Nodes Eval | 0 | CS SSF 13 | 988 |
| Regional Nodes Positive | 98 | CS SSF 14 | 988 |
| Regional Nodes Examined | 00 | CS SSF 15 | 988 |
| CS Mets at Dx | 00 | CS SSF 16 | 988 |
| CS Mets Eval | 0 | CS SSF 17 | 988 |
| CS SSF 1 | 047 | CS SSF 18 | 988 |
| CS SSF 2 | 034 | CS SSF 19 | 988 |
| CS SSF 3 | 080 | CS SSF 20 | 988 |
| CS SSF 4 | 000 | CS SSF 21 | 988 |
| CS SSF 5 | 988 | CS SSF 22 | 988 |
| CS SSF 6 | 988 | CS SSF 23 | 988 |
| CS SSF 7 | 988 | CS SSF 24 | 988 |
| CS SSF 8 | 988 | CS SSF 25 | 988 |
| **Treatment** |
| Diagnostic Staging Procedure | 02 |  |  |
| **Surgery Codes** |  | **Radiation Codes** |  |
| Surgical Procedure of Primary Site | 30 | Radiation Treatment Volume | 00 |
| Scope of Regional Lymph Node Surgery | 0 | Regional Treatment Modality | 00 |
| Surgical Procedure/ Other Site | 0 | Regional Dose | 00000 |
| **Systemic Therapy Codes** |  | Boost Treatment Modality | 00 |
| Chemotherapy | 03 | Boost Dose | 00 |
| Hormone Therapy | 00 | Number of Treatments to Volume | 000 |
| Immunotherapy | 00 | Reason No Radiation | 1 |
| Hematologic Transplant/Endocrine Procedure | 00 | Radiation/Surgery Sequence | 0 |
| Systemic/Surgery Sequence | 4 |  |  |