Case 1 – Sarcoma Tumor Board

34F who presented in 2011 with right knee pain.

PMHx: none

Exam: Tender to palpation over the patella. Mild edema.

FINAL PATHOLOGIC DIAGNOSIS

Right patellar tumor, biopsy: Giant cell tumor of bone
Case 1 – Sarcoma Tumor Board

Question: What do you recommend next?

A. Chemotherapy
B. Radiation therapy
C. Surgical resection
D. Observation

Right patellar curettage, burring, hydrogen peroxide application, Argon beam coagulation and packing with bone cement.

Patient elects not to follow up post-resection!

Case 1 – Sarcoma Tumor Board

She presents with a left shoulder mass and pleural-based lung nodules in 2018

No abnormality in the right knee

Case 1 – Sarcoma Tumor Board

Question: What do you recommend next?

A. Chemotherapy
B. Biopsy the shoulder mass
C. Biopsy the lung nodules
D. Biopsy both lesions
**Case 1 – Sarcoma Tumor Board**

**Pathology**

*Left shoulder, biopsy:*  
Dermatofibrosarcoma protuberans (DFSP)  
COL1A1-PDGFβ rearrangement

*Pleural-based nodule, right, needle biopsy:*  
Scant histiocytic proliferation with giant cells consistent with metastatic giant cell tumor (H3F3A positive)

**Question: What do you recommend next?**

A. PDGFR inhibitor  
B. Denosumab  
C. Resection of the shoulder DFSP  
D. Resect DFSP f/b Denosumab

**Medical Management of DFSP**

- PDGFR directed therapy: Imatinib, Nilotinib, etc. (Rutkowski, JCO 2010)

**Medical Management of GCT**

- RANKL monoclonal antibody: Denosumab (Thomas D., Lancet Oncol 2010)

**Surgical Resection of the DFSP**

Denosumab treatment initiated for metastatic GCT.
**Case 1 – Sarcoma Tumor Board**

Questions?

---

**Case 2 – Sarcoma Tumor Board**

32M with two years of right hip and low back pressure, presents to the ED with increasing right leg pain and difficulty ambulating.

PMHx: none

Exam: Posterior radicular leg pain with numbness.

---

**Case 2 – Sarcoma Tumor Board**

Gluteal mass, needle core biopsy:
Alveolar soft part sarcoma (TFE-3 positive, Pax-8 negative)

ASPL-TFE3 translocation positive

Typically presents with a slow growing painless mass. Extremities (60%), Trunk/RP (~25%), H+N (12%), Other (10%)
Case 2 – Sarcoma Tumor Board

Question: What do you recommend next?

A. Additional imaging  
B. Surgical resection  
C. Chemotherapy  
D. Radiation therapy

24% 7% 48% 21%

Metastasis frequently occurs in ASPS (~55% with synchronous metastasis)

Metastatic patterns in ASPS (Paoluzzi, JAMA Onc 2018):

- Lungs (90%)
- Bones (26%)
- Brain (11-19%)

Case 2 – Sarcoma Tumor Board

Radiation therapy:

Hypofractionated radiotherapy (52.5 Gy in 15 fractions) for pain palliation
**Case 2 – Sarcoma Tumor Board**

**Question:** What do you recommend next?

A. Chemotherapy  
B. Immunotherapy  
C. Targeted therapy  
D. Observation

![Bar chart showing percentages]

40 months of Sunitinib therapy (Stacchiotti S., Clin Can Res 2009, Ann Oncol 2011)
- Partial response in pelvis and lungs.
- Last 10 months stable disease.

**Evolving treatment paradigms for ASPS**

**TKIs with VEGF inhibition:**
- sunitinib, cediranib, pazopanib, tivantinib  
  (PR 20-40%, SD ~50%)

**Immune checkpoint inhibitors:**
- Phase 2 - Atezolizumab: PR 42%, SD 47% (Coyne, CTOS 2018)
- Anti-PDL1 based therapy (Groisberg, 2017; Conley, 2017)

**Combination therapies:**
- Phase 2 - axitinib + pembrolizumab: PR 44%, SD 33% (Wilky CTOS 2017)

**Case 2 – Sarcoma Tumor Board**

Patient currently enrolled on Atezolizumab trial at UC Davis

End of Case
Case 2 – Sarcoma Tumor Board

Questions?

Case 3 – Sarcoma Tumor Board

57M presents with urinary frequency and suprapubic pressure
Exam: palpable suprapubic mass

Case 3 – Sarcoma Tumor Board

PATHOLOGIC DIAGNOSIS

"Pelvic mass", core biopsy: Morphologically low-grade spindle cell proliferation most consistent with desmoid fibromatosis
**Case 3 – Sarcoma Tumor Board**

Question: What do you recommend next?

A. Radiotherapy  
B. Medical therapy  
C. Surgical resection  
D. Observation

[Bar graph showing 72% success rate for surgery, 25% for medical therapy, and 3% for observation]

--

**Case 3 – Sarcoma Tumor Board**

Aggressive fibromatosis (Desmoid) are locally aggressive but no known metastatic capacity

Local recurrence high after surgical resection

NCCN recommends observation as a primary therapeutic intervention for asymptomatic, non-life threatening, resectable desmoid.

*Patient expresses desire to keep his ileal pouch

**Case 3 – Sarcoma Tumor Board**

Liposomal doxorubicin x 6 cycles

Now asymptomatic

Observation for 9 months. No progression
**Case 3 – Sarcoma Tumor Board**

**Medical management of Desmoid**

**Stability:**
- NSAIDs (Sulindac, Celecoxib, etc),
- anti-hormonal (tamoxifen, raloxifene),
- Imatinib

**Objective responses:**
- liposomal doxorubicin
- Sorafenib (Gounder M., NEJM 2018)
- Gamma secretase inhibitor (investigational)

---

**Case 4 – Sarcoma Tumor Board**

47F who presented in May 2015 with abdominal pain and cramping

PMedHx: Stage IB cervical (SCC) cancer in 2007 s/p hysterectomy and pelvic radiation therapy (5040 cGy to paraaortic and right hemipelvis and 4500cGy to entire pelvis using 180cGy fractions)

PSurgHx: radical hysterectomy with pelvic and RP lymphadenectomy

May 2015: Pelvic malignancy with potential invasion into bowel (SBO) and bladder.
Case 4 – Sarcoma Tumor Board

Surgical resection (May 2015):
Surgical excision of the tumor with superficial excision of bladder dome and small bowel resection, primary anastomosis, and appendectomy.

Post-op course c/b poor wound healing, poor nutritional status and diarrhea.

Pathology:
11.7 cm undifferentiated spindle cell sarcoma involving the small intestine wall and mesenteric fat. Resection margins were negative (0.3 cm from bladder margin).

Radiation-associated soft-tissue sarcomas (RAS) (Gladdy RA., JCO 2010)

STSs are one of the most common Radiation-associated tumors.
83% are high grade, deep (87%), and truncal (61%).

Median interval between radiation and RAS development is 10yrs (median dose 54Gy)

RAS associated with worse DSS than sporadic STSs (HR 1.7)

Worse 5-yr DSS for pleomorphic RAS vs sporadic pleomorphic sarcoma (44% vs 66).

October 2015

Recurrent/residual pelvic mass

Question: What do you recommend next?
A. Radiotherapy
B. Chemotherapy
C. Surgical resection
D. Chemoradiation
**Case 4 – Sarcoma Tumor Board**  
**December 2015**

Completes 3 cycles of gemcitabine and docetaxel

Disease progression

---

**Case 4 – Sarcoma Tumor Board**  
**October 2015**

Question: What do you recommend next?

A. Radiotherapy  
B. Chemotherapy  
C. Surgical resection  
D. Chemoradiation

---

**Case 4 – Sarcoma Tumor Board**  
**May 2016**

Completes 4 cycles of Doxorubicin + Ifosfamide

Cycle 4 c/b GNR septic shock

---

**Case 4 – Sarcoma Tumor Board**  
**July 2016**

**Surgical resection:**
Resection of the mass with ileocectomy with primary anastomosis of the ileum to the ascending colon.

“the main mass was anterior to the rectum and mobile but stuck to the anterior rectum.”

**Intraoperative radiation therapy (IORT)**
15 Gy to the anterior rectum (site of adherence).
**Case 4 – Sarcoma Tumor Board**

July 2016

**Pathology:**
Undifferentiated spindle cell sarcoma, grade 3, viable pleomorphic cells noted, negative margins

**Follow up:**
Elects to forgo adjuvant therapy. NED for 30 months post-op

---

**Case 5 – Sarcoma Tumor Board**

65F with a 6 month history of “swelling/bruise-like” lesions along the right upper lateral and posterior chest wall, below the breast.

**PMHx:** 1993 – Right sided breast cancer s/p right lumpectomy, adjuvant radiation and chemotherapy (AC).

**Exam:**
Right upper chest (below the lumpectomy scar line) there was a 4 x 2 cm purpuric firm plaque.
Right upper back there was a 4 x 5 cm violaceous cutaneous mass.
Case 5 – Sarcoma Tumor Board

Question: What do you recommend next?

A. Incisional biopsy
B. Image-guided biopsy
C. Fine needle aspiration
D. Excision of the masses

Pathology:
Clusters of spindle cells with vasoformative growth (CD34, CD31, and FLI1+)
FNA of the right back – cutaneous angiosarcoma with high mitotic rate.
FNA of the lateral chest wall – cutaneous angiosarcoma with high mitotic rate.

Radiation-associated angiosarcoma (RAAS)
– 15% of radiation-associated sarcomas (Brady MS., Arch Surg 1992)
  - Median latency period (7 years)
  - Cutaneous RAAS are associated with MYC amplification (patients tumor harbors a MYC amp) compared to sporadic cases and other atypical vascular lesions after radiation (Mentzel T., Mod Pathol 2012; Manner J., Am J Pathol., 2010)

Case 5 – Sarcoma Tumor Board

Question: What do you recommend next?

A. Wide surgical resection
B. Neoadjuvant chemotherapy
C. Neoadjuvant radiotherapy
D. Concurrent chemoradiation
Case 5 – Sarcoma Tumor Board

Neoadjuvant chemotherapy with weekly paclitaxel 80mg/m² x 3 cycles (ANGIOTAX study, JCO 2008).

Case 5 – Sarcoma Tumor Board

Question: What do you recommend next?

A. Surgical resection
B. Additional chemotherapy
C. Neoadjuvant radiotherapy
D. Concurrent chemoradiation

Surgical Resection

Skin and soft tissue, right posterior back, excision:
1. Residual/recurrent angiosarcoma, scattered microscopic foci largest individual focus 0.2 cm; negative margins, necrosis present.

Skin and soft tissue, right inferior breast, excision:
1. Residual/recurrent angiosarcoma, scattered microscopic foci, FNCLCC Grade 3; negative margins, necrosis present.
Case 5 – Sarcoma Tumor Board

Question: What do you recommend next?

A. Adjuvant chemotherapy
B. Adjuvant radiotherapy
C. Observation
D. Concurrent chemoradiation

Adjuvant chemotherapy reduces risk of local recurrence (HR 0.35, p=0.012) (Torres KE., Ann Surg Oncol 2013)

Receives Paclitaxel for 3 additional cycles.
NED 1 year post resection

End of Case 5