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Clinical History

- 55 year old male with large, deep, non-tender left thigh mass.
- Seen at LAC+USC Med Ctr FNA clinic
- No h/o trauma or radiation
Vimentin +
S-100, PanK, CD31 –
Dx: Malignant cells present, favor high-grade sarcoma, recommend biopsy for definitive diagnosis
Work-up and Clinical Course

- **MRI**
  - 8x5cm thigh mass

- **AIM chemotherapy x1 (pre-operative)**
  - Adriamycin (Doxorubicin) + Ifex (Ifosfamide) + Mesna

- **Post chemotherapy MRI 2 weeks later**
  - Increased size to 9.4cm
  - = Failed chemotherapy
Surgery

Wide excision limb-salvage surgery

Specimen

- Left thigh resection specimen
- Grossly well-circumscribed (10cm)
- Soft, variegated with hemorrhage and necrosis.
- Surrounded by skeletal muscle
DIAGNOSIS??
DIAGNOSIS

- Extraskeletal Osteosarcoma, high-grade
  - All surgical margins negative
  - T2bNXMO, Stage III
Extraskeletal Osteosarcoma

- A malignant neoplasm
- Cells have osteoblast phenotype
- Synthesis of osteoid/bone
- Located in the soft tissues
- No attachment to the skeleton including periosteum.
Extraskeletal Osteosarcoma

- 1-2% of all soft tissue sarcomas
- First described in 1941 by Smith
- Patients generally >40 years old
- Site: Thigh, pelvic and shoulder girdles
- Presents as an enlarging soft tissue mass
  - Painful in about 1/3rd of cases
  - Usually present 6-8MO from onset of symptoms
  - Size at presentation: 5-10 cm
  - M:F = 1.9:1

Extraskeletal Osteosarcoma

Microscopic Findings

- Neoplastic osteoid and/or bone
  - Wide variation in amount
  - Foci of neoplastic cartilage common

- Different patterns
  - Fibroblastic (most common)
    - Resembles undifferentiated pleomorphic sarcoma (UPS) or “MFH-like” except with the presence of osteoid
  - Chondroblastic may have better prognosis
Extraskeletal Osteosarcoma Subtypes

- **Osteoblastic**
  - Very cellular, highly pleomorphic tumor cells, numerous mitoses, and lots of osteoid production.

- **Chondroblastic**
  - Giant cell rich (osteoclast-like giant cells)
  - Telangiectatic (markedly dilated blood filled spaces lined by malignant tumor cells)
  - Small cell pattern.

Extraskeletal Osteosarcoma

**Immunohistochemistry**

- Vimentin +
- SMA, EMA, Desmin, S-100 –
  - Main value is to exclude other malignant tumors with metaplastic osteoid/bone
    - Synovial Sarcoma, Epithelioid Sarcoma, Melanoma, MPNST
Extraskletal Osteosarcoma

Differential Diagnosis

- Myositis Ossificans (Early stage)
  - A benign reactive ossifying process in muscle
  - Distinct Zonation Pattern - most important feature to distinguish it from EOS
  - Central immature zone is highly cellular (often confused with EOS)
Myositis Ossificans: Innermost Zone
Myositis Ossificans: Intermediate zone
Myositis Ossificans: Peripheral Zone
Extraskeletal Osteosarcoma

Differential Diagnosis

- Myositis Ossificans
- Malignant tumors with metaplastic bone
- Parosteal Osteosarcoma
- Periosteal Osteosarcoma
- High-grade surface osteosarcoma
Extraskeletal Osteosarcoma

Prognosis

- Poor
- Most patients die from metastatic disease

Treatment

- Surgery
- Preoperative or postoperative multiagent chemotherapy
- Radiation

Prognostic variables

- <5cm, chondroblastic subtype = better prognosis)
Continued Clinical History

MRI 1/08

- Interval development of irregular enhancing mass
- 3/08 re-resection showed recurrent disease

Patient treated with radiation therapy
Parakrama Chandrasoma, M.D.
Nancy Klipfel, M.D.
Yanling Ma, M.D.
Wesley Naritoku, M.D., Ph.D.
Sarka Cernocek, M.D.

