## Management of Extremity Sarcomas

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### **Declarations**

none



#### Case 1



- 23 year old female
- Right hand dominant
- 1 year history of soft tissue mass in right antecubital fossa
- Swelling in right hand
- Neurovascularly intact
- Mass is mobile over bone



#### Case 2



- 16 year old male
- 4 week history of pain and swelling in left wrist
- Night pain
- Large, firm soft tissue mass fixed to bone
- No constitutional symptoms



## Presentation of extremity sarcomas

- Pain
- Soft tissue mass
- Pathologic fracture
- Incidental finding







## **Staging**

#### **Bone Sarcomas**

- Plain radiographs
- MRI of <u>entire</u> bone
- CT thorax
- Total body bone scan
- ALP, LDH in osteosarcoma/Ewing
- ? PET

#### **Soft Tissue Sarcomas**

- MRI of mass
- CT thorax
- Total body MRI in myxoid liposarcoma
- Imaging of lymph nodes in clear cell sarcoma, angiosarcoma, rhabdomyosarcoma, epithelioid sarcoma, extraskeletal myxoid chondrosarcoma
- ? PET



## **Biopsy – bone sarcoma**

- Confer with treating surgeon
- image guided core needle biopsy
- Ultrasound guided if soft tissue mass
- CT guided if intraosseus
- Must be reviewed at host sarcoma site by sarcoma pathologist

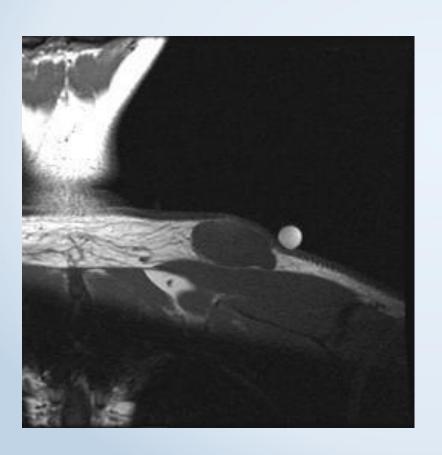


## **Biopsy – soft tissue sarcoma**

- Confer with treating surgeon
- > 5 cm core needle biopsy
- Deep to fascia core needle biopsy
- Adjacent to critical structures (vessels, nerve, bone) – core needle biopsy
- Otherwise can do excisional biopsy
- Must be reviewed at host sarcoma site by sarcoma pathologist

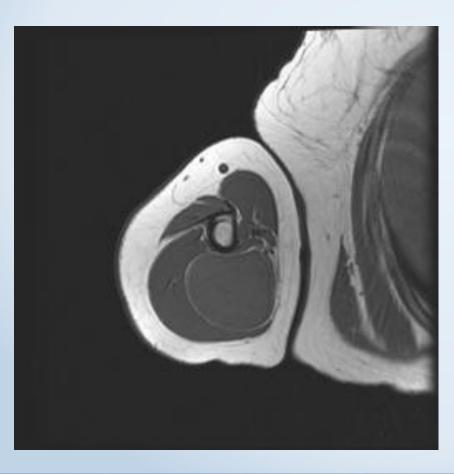


### **Management of Soft Tissue Sarcoma**



- Wide surgical resection
- If we look at imaging and feel that we can resect tumor
  - with adequate wide margins (2 cm)
  - without sacrificing critical structures or
  - significantly affecting function
- surgery alone is usually all that is necessary -usually only reserved for small, superficial tumors, irrespective of tumor grade

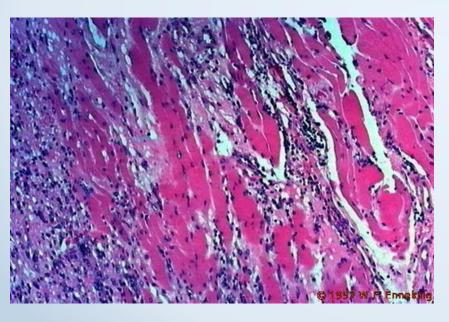
## **Management of Soft Tissue Sarcoma**



- If wide resection means
  - sacrifice of critical structures or
  - severe functional consequences
- Radiation will be used almost always in deep lesions
- Therefore radiation is necessary if marginal margins are anticipated



#### **Radiation for Soft Tissue Sarcoma**



- Rationale allows for a less extensive operation, preserving critical structures
- Treats microscopic disease that is often found in surrounding edema
- Tends NOT to shrink tumour (exception – myxoid liposarcoma)

#### **Radiation for Soft Tissue Sarcomas**

- Like surgery in that it achieves local control of the tumour only
- It does not affect the development of metastases
- Local control with marginal excision + radiation is equivalent to wide excision, but function is better
- Can be administered preoperatively or postoperatively – no recurrence advantage
- We prefer preoperative irradiation

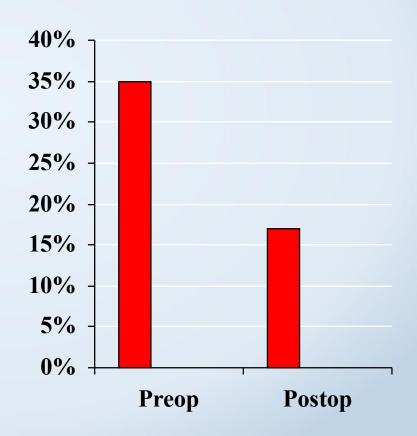


## NCIC SR-2 Randomized Clinical Trial-O' Sullivan et al

- STS patients seen by surgeon and rad onc with decision for combined Rx
- Randomized to receive preop or postop
- End points- acute wound complications, function
- Complications defined as repeat surgery or deep wound packing beyond 6 weeks

## SR-2 RCT- Results

- Wound healing complications twice as frequent following preop rads
- Other significant variables- tumour size, upper vs. lower extremity

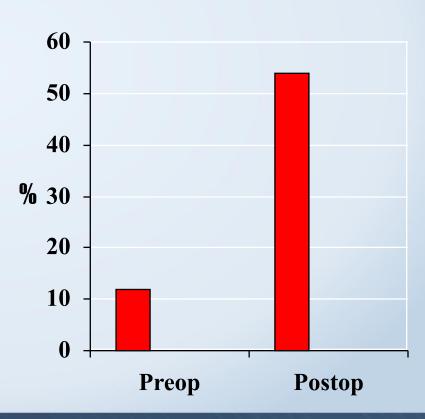




#### Radiation for soft tissue sarcomas

- Late effects of fibrosis measured at 2 years
- Fibrosis correlates with reduced function scores
- This improves long term functional outcome
- Wounds almost always heal

#### RTOG Fibrosis > 2





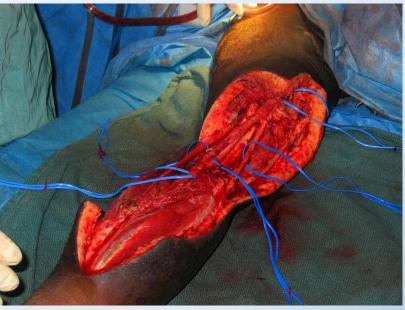
## **Chemotherapy in Soft Tissue Sarcoma**

- Rarely used with resectable localized disease
- Exceptions extraskeletal Ewing sarcoma, embryonal/alveolar rhabdomyosarcoma
- May be used in metastatic disease or advanced/unresectable localized disease
- Targeted therapy
- immunotherapy



### Case 1





## **Management of Osteosarcoma**

- Commonest bone sarcoma
- Historically extremity amputation was treatment of choice
- 95% of patients had amputation
- 90% of patients died from metastatic disease



## **Current Osteosarcoma Treatment Paradigm**

- multiple drug neo-adjuvant chemotherapy
  - Adriamycin
  - Cisplatin
  - high dose methotrexate
- Wide resection
- Completion of adjuvant chemotherapy
- aggressive metastasectomy
- Current cure rate approx. 70%



### Case 2







## Most bone sarcomas occur around the knee







## **Tumour endoprosthesis**

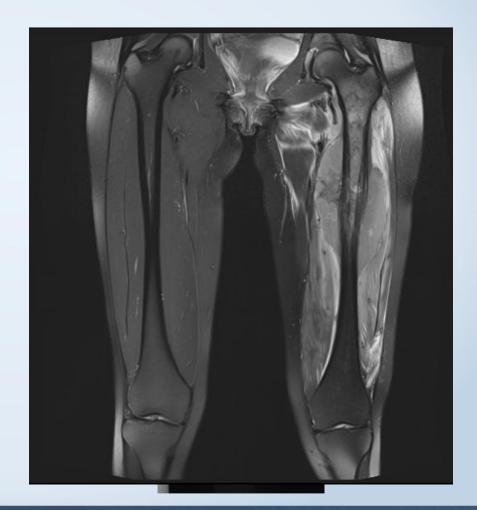






## Management of Ewing sarcoma

- Second most common bone sarcoma in children
- Historically radiation was treatment of choice
- Current treatment involves neoadjuvant chemotherapy and limb salvage surgery when possible
- Radiation reserved for tumours that are unresectable or if limb salvage will have severe functional consequences





# **Chemotherapy and Ewing's Sarcoma**

- Current regimens use:
- VAC (Vincristine, Adriamycin and Cyclophosphamide)
- Addition of Etoposide and Ifosfamide has shown to be of benefit in non-metastatic Ewing's

Grier HE et al, Addition of Ifosfamide and Etoposide to Standard Chemotherapy for Ewing's Sarcoma and Primitive Neuroectodermal Tumor of Bone, NEJM 348(8), 694-701, 2003.

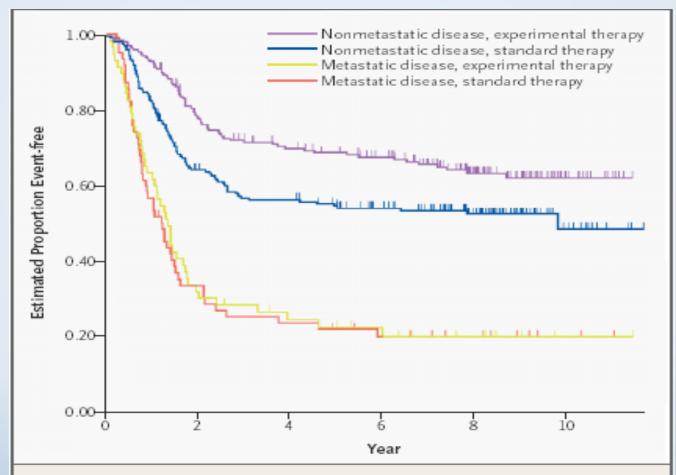


Figure 1. Event-free Survival According to Study Group and the Presence or Absence of Metastatic Disease.

## **Management of Chondrosarcoma**

- Most common bone sarcoma in elderly
- Surgical disease
- No role for adjuvants
- Grade 1 intramedullary curettage
- Grade 1 extramedullary wide resection
- Grade 2/3 wide resection & reconstruction





## Commonest sarcoma in the pelvis





## **Questions?**

