MALIGNANT SOFT-TISSUE TUMOURS

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MALIGNANT SOFT TISSUE TUMOURS

- Synovial Sarcoma
- Epithelioid Sarcoma
- Liposarcoma
- Malignant Fibrous Histiocytoma
- Fibrosarcoma
- Rhabdomyosarcoma
- Malignant Neurilemoma
- Soft-tissue Osteosarcoma
Synovial Sarcoma

- Malignant soft-tissue sarcoma arising from mesenchymal cells of synovial differentiation
- 20-40 M>F
- Occurs periarticular tissues of the extremities/ peri-tendinous tissues of the hand and foot
- Rarely occurs intra-articularly
Synovial Sarcoma

- Slow-growing, modestly-symptomatic soft-tissue mass that occurs deep to the deep fascia
- Mostly in the lower extremity; knee joint, foot or thigh
- Clinically the mass has a firm consistency which appears fixed to the deeper structures
- In the hands and feet, commonly mistaken for a ganglion
 lexicographic("Significant incidence of local node involvement; Metastases to lung")
Synovial Sarcoma

- **XRAY:**
  - Lobulated soft-tissue shadow
  - Irregular spotty calcification

- **CT SCAN:**
  - Homogenous soft-tissue mass
  - Significant enhancement with contrast
Synovial Sarcoma

- GROSS FEATURES:
  - Psuedocapsule
  - Rubbery consistency
  - Greasy feel
  - Calcification
  - Evidence of haemorrhage/cysts
Synovial Sarcoma

MICROSCOPIC:

2 types:

1. BIPHASIC (classic)
   - Both a spindle cell and an epithelial component

2. MONOPHASIC
   - Spindle-cell population that lack gland-forming components
Synovial Sarcoma

**TREATMENT:**

- **SURGICAL:** Depending on response to adjuvant therapy:
  - Responsive - wide surgical excision
  - Unresponsive - radical margins

- **CHEMOTHERAPY:**
  - Modest incidence of partial response

- **RADIOTHERAPY:**
  - Most have satisfactory response
Epithelioid Sarcoma

- Fully malignant painless soft-tissue sarcoma
- Young adults; M>F
- Superficial or deep tendon sheaths of hand/wrist/fingers
- ?Synovial origin
Epithelioid Sarcoma

MICROSCOPIC:
- nodular growth
- densely eosinophilic
- epithelial appearance
- Stains +ve for epithelial markers

High tendency to recur/ mets to lung

Treatment - wide excision
Liposarcoma

- Primary malignant tumour arising from fat
- Most common soft-tissue tumour
- 40-60 yrs; M>F
- Thigh and retroperitoneum; Rare in bone
Liposarcoma

Several histogenic subtypes but two main types:
- MYXOID (50%)
  - Specific cytogenetic marker: $t(12;160(q13;p11.2))$
  - Generally low-grade stage I lesion
- PLEOMORPHIC (30%)
  - No specific marker
  - Usually high grade stage II lesion
Liposarcoma

◊ Clinically:
  – Slow-growing
  – Deep-seated
  – Ill-defined mass
  – Seldom tender or painful
  – Often huge at presentation
  – Commonly fixed to deeper structures but not skin
  – Does not transilluminate
Liposarcoma

- XRAY:
  As for other soft-tissue sarcomas:
  - non-specific
  - homogenous mass with same density as adjacent muscle

- ISOTOPE SCAN:
  - ↑ uptake (ionic Ca++)

- CT SCAN:
  - Less radiodense than surrounding muscle
  - very hypervascular
Liposarcoma

GROSS FEATURES:

- Deceptively encapsulated
- Lobulated
- MYXOID:
  - Soft and myxomatous
  - richly vascular
- PLEOMORPHIC:
  - Infiltrative
  - More fatty
  - Cystic/haemorrhagic change
Liposarcoma

HISTOLOGICALLY:

- **MYXOID:**
  - Sheets of lipocytes/blasts
  - Myxomatous matrix
  - Myriads of branching capillaries
  - “Road Map” appearance

- **PLEOMORPHIC:**
  - More cellular
  - Lacks matrix/capillaries
  - Gigantic bizarre lipoblasts

- Oil red O stain and S-100 antigen positive
Liposarcoma

TREATMENT:

- Surgical:
  - Stage I (myxoid) - wide margin or marginal margin after satisfactory pre-op radiation
  - Stage II - radical/wide margin

- Radiotherapy:
  - Used extensively pre-op to facilitate limb salvage

- Chemotherapy:
  - Generic therapy used but little experience

Survival 5 years: Myxoid 90%; Pleomorphic 60%

High incidence of a second lesion (30-50%) in the retroperitoneal region
Malignant Fibrous Histiocytoma

- Primary malignant neoplasm containing both fibrous and histiocytic components
- 40-70; M>F
- Lower extremity (thigh)
- Majority deep to deep fascia
- High-grade lesion
  - mets to lung (90%)
  - Overall survival 50%
Malignant Fibrous Histiocytoma

- Clinically:
  - Soft tissue mass:
    - Aggressively enlarging
    - Deep
    - Mildly symptomatic

- Radiographic:
  - No distinct features
  - Rarely calcifies
  - CT shows isointense mass
Malignant Fibrous Histiocytoma

- **HISTOLOGICALLY:**
  - Highly variable
  - Classically storiform pleomorphic pattern
  - Plump spindle cells
  - Large bizarre histiocytic cells
  +ve stain for Vimentin and A1-AT antibodies
Malignant Fibrous Histiocytoma

TREATMENT:

♦ Radiation:
  – Very useful adjuvent - limb salvage

♦ Chemotherapy:
  – Investigative stages only - limited response

♦ Surgical:
  – Stage I - Wide margin without pre-op adjuvent
  – Stage II - Wide margin with satisfactory pre-op adjuvent
  – Stage III - Radical margin with unsatisfactory response to pre-op adjuvent
Fibrosarcoma

- Primary malignant neoplasm arising from *fibroblasts*
- 30-50; M>F
- Thigh, leg, arm, trunk
- Wide gamut of behaviour clinically
- Deep slow-growing fixed mass
- Often painless
Fibrosarcoma

- Radiographically:
  - Deep mass with same density as adjacent tissues
  - 10% calcification
- Isotope Scan:
  - May show a modest increase in uptake
- CT Scan:
  - Isointense homogenous mass
Fibrosarcoma

**GROSS FEATURES:**
- Greyish-white
- Rubbery in consistency
- Smaller lesions appear well encapsulated
- Larger lesions - infiltrative
**Fibrosarcoma**

**MICROSCOPIC FEATURES:**
- Vary from well to poorly differentiated
- Masses of fibroblastic tissue
- Scattered atypical and mitotic cells
- Masson stain identifies collagen as the matrix being produced (green)
Fibrosarcoma

TREATMENT:

- Radiation:
  - Given pre-op; Often limb-salvaging
  - 5400 rads over 21 days to entire compartment
  - Intent is to stimulate encapsulation
- Chemotherapy: no distinct role
- Surgery: As for MFH
Rhabdomyosarcoma

- Primary malignant tumour derived from striated muscle
- Two forms:
  1. Pleomorphic: (30%)
     - 50-70yrs; M>F
     - Proximal extremities
  2. Embryonal: (70%)
     - 0-15yrs; M>F
     - Head/neck/GU/retroperitoneal; occas. extremities
Rhabdomyosarcoma

- Associated chromosomal abnormality - translocation chromosomes 2 and 13
- High grade with rapid growth
- Local recurrence, distant mets (lung) common - uniformly fatal
- Majority present stage II-B as an enlarging intramuscular mass
Rhabdomyosarcoma

- Radiographically:
  - Obscure intramuscular mass
  - Calcification/ossification not seen
- MRI better than CT scan
  - Moderate int. T-1
  - Much brighter T2
Rhabdomyosarcoma

MICROSCOPICALLY:

1. Embryonal
   - Small blue round cells
   - Clusters; no real pattern
   - Well diff. - cross striations
   - Poorly diff. - EM or +ve reactions for desmin/myoglobin

2. Pleomorphic
   - Spindle cells with marked variation in size
   - Bizarre giant cells that resemble a wrist watch, tadpole or belt buckle
Rhabdomyosarcoma

TREATMENT:

- **Radiation:**
  - Excellent palliation but rarely curative
  - Permits limb-salvage and ↓ risk of local recurrence when given pre-operatively

- **Chemotherapy:**
  - Almost always satisfactory response pre-op
  - When wide margin not obtained, given post-op for 2 years (VAC - Vincristine, Cytoxan, Adriamycin, Actinomycin); has ↑ 5-year survival from 10 to 80% for stage I and II lesions

- **Surgery:**
  - Limb salvage practical (after chemo, radiation) in the majority of cases
Malignant Neurilemoma

- Spindle-cell sarcoma
- Arises from a nerve or neurofibroma
- Accounts for 10% of all soft-tissue sarcomas
- 50% occur in association with neurofibromatosis
- 25-40yrs; presents as a painful mass
- Mostly seen in relationship to major nerves of brachial or sciatic plexus

Therefore, proximal upper or lower extremity
Malignant Neurilemoma

MICROSCOPICALLY:

♦ Most resemble fibrosarcomas
  – but nuclei in MN have a wavy outline (fibrosarcoma - symmetrical)
♦ Mature islands of bone or cartilage
♦ Nodular or plexiform arrangement
♦ Most useful stain is S-100 protein for differentiation:
  ◆ Fibrosarcoma
  ◆ Synovioma
  ◆ Leiomyosarcoma
Extraskeletal Osteosarcoma

- Rare; 40-60 yrs
- Usually intramuscular
- Small, round to ovoid lesions
- Finely trabeculated bone formation
- Prognosis poor with early metastases
Extraskeletal Osteosarcoma

- **Grossly:**
  - Soft to firm
  - Gritty
  - Haemorrhage/necrosis
  - May be infiltrative

- **Microscopically:**
  - Either fibroblastic or extremely cellular
  - Fine lace-like osteoid and mineralized bone