Let’s mention 2 malignant cutaneous sarcomas that may be deceptively bland
Epithelioid sarcoma

- Commonest sarcoma of hand/forearm
- Young 10-40 years
- Nodular/ Necrosis/ Cystic ±
- Low and high molecular weight CK
- SMARCB1 gene (INI1) loss
- High recurrence and metastatic rate incl nodes
Differential Diagnosis

- Rheumatoid nodule
- Necrobiosis lipoidica
- Granuloma annulare
- Fibrous histiocytoma
Clear Cell Sarcoma

- Age 20-40
- Deep soft tissue of distal extremities
- Clinically often benign
- ± melanin 50%
- Low mitotic count – 4 mitoses /10HPF
- Melanoma markers positive
- Clinically, genetically and biologically distinct from malignant melanoma
- T(12;22)
- Neg for BRAF V600
Pitfalls

• Spindle cell sarcoma mimics
  – Spindle cell melanoma
  – Spindle cell carcinoma

• Pseudo sarcomas: Benign tumours mimicking sarcomas

• Sarcomas mimicking benign tumours eg Epithelioid sarcoma
Pseudosarcomas
Entities we may mistakenly call malignant

- Nodular fasciitis
- Proliferative myositis
- Proliferative fasciitis
- Giant cell tumour of tendon sheath
- Pleomorphic lipoma
- Fibrous histiacytoma with monstrous cells
- Cellular schwannomas
**Myxofibrosarcoma**
- Subcutis based
- May extend into skeletal muscle or up into dermis
  - Larger
  - Slow progressive growth

**Nodular fasciitis**
- Attached to fascia
- (into overlying subcut, into muscle from fascia, fascial along fascia)
  - Small <5 cms
  - Rapid growth (weeks)
  - Spindle cells in myxoid stroma
  - Tissue culture like
  - Mitoses ++
  - Extravasated rbc’s and inflammatory cells
  - May be marked collagen
Spindle cell/ Pleomorphic lipoma

- Age 45 – 60
- Site: 80% posterior neck, shoulder, back
- 20% elsewhere
- Gender (90% male)

Micro:
- Spindle/ovoid cells
- Fat content variable
- Multinucleated floret cells
- May be bizarre nuclei
- Mitoses very scanty

Ropey collagen CD34 pos
Mast cells
S100P neg outside of fat
BCL2 pos (non specific)
Loss of 16q material
Proliferative Myositis
Ganglion like cells
Sarcomas mimicking benign tumours

- Leiomyosarcoma
- Desmoplastic melanoma
- Synovial sarcoma
- Epithelioid sarcoma
- Clear cell sarcoma
Summary

• Be aware of the specific entities
• A growing tumour of more than a six months duration is usually malignant
• Do not diagnose a lesion as benign or malignant on the basis of cellularity or mitotic activity
• Use immunohistochemistry as part of a panel
• Get a good history and discuss the case with clinician and Pathology Colleagues
• Be humble! And be careful
4 Difficult myxoid entities
Myxoinflammatory Fibroblastic Sarcoma

- Recently described entity (1998)
- Synonyms: Acral Myxoinflammatory Fibroblastic Sarcoma, Inflammatory Myxohyaline Tumour of Distal Extremities with virocyte or RS like cells, Inflammatory Myxoid tumour of soft parts with bizarre giant cells

- Looks inflammatory with “virocyte” like, ganglion like or RS like cells
- Is a true neoplasm, may recur 5%, may metastasise
Low Grade fibromyxoid sarcoma
Myxofibrosarcoma
<table>
<thead>
<tr>
<th></th>
<th>Myxofibrosarcoma</th>
<th>Myxoma</th>
<th>Fibromyxoid sarcoma</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Site</strong></td>
<td>limbs</td>
<td>Muscle</td>
<td>H&amp;N Limbs</td>
</tr>
<tr>
<td><strong>Site</strong></td>
<td>Dermis 60%</td>
<td>Muscle</td>
<td>Subfascial</td>
</tr>
<tr>
<td><strong>Cellularity</strong></td>
<td>variable</td>
<td>minimal</td>
<td>variable</td>
</tr>
<tr>
<td><strong>Morphology</strong></td>
<td>Less collagen</td>
<td>No collagen</td>
<td>Good collagen</td>
</tr>
<tr>
<td><strong>Vessels</strong></td>
<td>Curvilinear</td>
<td>Almost none</td>
<td>Thick</td>
</tr>
<tr>
<td><strong>Mitoses</strong></td>
<td>Variable</td>
<td>None</td>
<td>Variable</td>
</tr>
<tr>
<td><strong>Collagen Rosettes</strong></td>
<td>None</td>
<td>None</td>
<td>40%</td>
</tr>
<tr>
<td><strong>MUC4 x</strong></td>
<td>Absent</td>
<td>Absent</td>
<td>Positive</td>
</tr>
<tr>
<td><strong>T(7;16) y FUS and CREB3L2.</strong></td>
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<td></td>
<td>T(7;16) y FUS and CREB3L2.</td>
</tr>
<tr>
<td><strong>Prognosis</strong></td>
<td>Variable</td>
<td>Excellent</td>
<td>40% die, 60% recur</td>
</tr>
</tbody>
</table>
Final message

- I don’t call a skin tumour malignant before considering if it is a pseudosarcoma ie a benign lesion mimicking a sarcoma

- I don’t call a spindle skin tumour benign before identifying the nature of the spindle cells
Thank you for listening