Case 1

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Disclosure of Relevant Financial Relationships

I have no conflicts of interest to disclose.

Clinical History

• A 51 year old man presented with a painless mass in the left calf, which he had had for about one year.

Imaging findings

Clinical History, continued

• Tumor was biopsied.
• Diagnosis of “low grade spindle cell neoplasm” was rendered on scant biopsy material.
• A 5.7 cm tumor was resected with negative margins.
Differential Diagnosis

- Solitary Fibrous Tumor
- Low Grade Fibromyxoid Sarcoma
- Soft Tissue Perineurioma
- DFSP

Diagnosis – Why does it matter?

- Solitary Fibrous Tumor
  - Intermediate malignant potential
  - Metastasize in 5-20% of cases
- Low Grade Fibromyxoid Sarcoma
  - Low grade malignancy
  - High rate of LR if not treated appropriately, infrequently metastasize
- Soft Tissue Perineurioma
  - Benign
  - Low risk of LR, do not metastasize
- DFSP
  - Locally aggressive
  - High risk LR, only metastasize if fibrosarcomatous transformation
Solitary Fibrous Tumor

- Can arise in any anatomic site
  - Deep > superficial
- Any age, M=F
- May be present for long duration prior to presentation

**IHC:** positive nuclear STAT6 (c-tetra), BCL2, CD34, CD99
**Pitfalls:** very rarely express EMA, keratin, actin

Low Grade Fibromyxoid Sarcoma

- Proximal extremities and trunk
  - Deep > superficial
- Mostly young adults but may arise at any age
- May be present for long duration prior to presentation
Dermatofibrosarcoma Protuberans

- Superficial trunk and proximal extremities
- Young to middle age patients
  - Slight male predominance
  - Nodular/multinodular cutaneous mass, slow growing

Soft Tissue Perineurioma

- Most common in extremities and trunk
- Superficial > deep
- Slight female predominance
- Arise at any age, more common in adults.
Perineurioma

- Sclerosing (cutaneous)
- Reticular/microcystic
- Intraneural
- Pseudolipoblastic
- Epithelioid perineurioma
- Hybrid nerve sheath tumors with perineural differentiation
- Etc.

IHC: Positive for EMA, GLUT-1, claudin, +/- CD34
Pitfalls: expression may be focal, may have focal S100

Variants:
- Sclerosing (cutaneous)
- Intraneural
- Pseudolipoblastic
- Epithelioid perineurioma
- Hybrid nerve sheath tumors with perineural differentiation
- Etc.
**Diagnostic pitfalls in bone and soft tissue pathology**

### Morphology

<table>
<thead>
<tr>
<th></th>
<th>Collagen</th>
<th>Blood vessels</th>
<th>Infiltration</th>
<th>Cells</th>
</tr>
</thead>
<tbody>
<tr>
<td>SFT</td>
<td>Often rosy, thick, glassy</td>
<td>Thin, dilated, branching &quot;staghorn&quot;</td>
<td>Usually encapsulated</td>
<td>Variable</td>
</tr>
<tr>
<td>LGFMS</td>
<td>Fine, Giant collagen rosettes</td>
<td>Curvilinear to branching</td>
<td>Usually encapsulated</td>
<td>Very bland with scant cytoplasm, to epithelioid</td>
</tr>
<tr>
<td>Perineurioma</td>
<td>Variable</td>
<td>Usually not prominent</td>
<td>Usually encapsulated</td>
<td>Long delicate bipolar processes at least focally</td>
</tr>
<tr>
<td>DFSP</td>
<td>Inconspicuous</td>
<td>Rich vascular network</td>
<td>Highly infiltrative, poorly circumscribed</td>
<td>Spindled, moderate cytoplasm</td>
</tr>
</tbody>
</table>

### Immunohistochemistry

<table>
<thead>
<tr>
<th></th>
<th>CD34</th>
<th>SMA</th>
<th>SMA</th>
<th>GLUT1</th>
<th>CLAUDIN 1</th>
<th>MUC4</th>
<th>STAT6</th>
</tr>
</thead>
<tbody>
<tr>
<td>SFT</td>
<td>+</td>
<td>+/-</td>
<td>rare</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>*</td>
</tr>
<tr>
<td>LGFMS</td>
<td>-/+</td>
<td>+/-</td>
<td>rare</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Perineurioma</td>
<td>+/-</td>
<td>+/-</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>DFSP</td>
<td>+/-</td>
<td>rare</td>
<td>-</td>
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</tr>
</tbody>
</table>

### Molecular Diagnostics

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>Molecular alteration</th>
<th>Gene fusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>SFT</td>
<td>t(12;16)(p13;q11), t(16;16)</td>
<td>NAB2-STAT6</td>
</tr>
<tr>
<td>LGFMS</td>
<td>t(7;16)(q22;p11), t(11;16)</td>
<td>FUS-CREB3L2, FUS-CREB3L1</td>
</tr>
<tr>
<td>Perineurioma</td>
<td>22q deletions, mutations in NF2</td>
<td></td>
</tr>
<tr>
<td>DFSP</td>
<td>Ring chromosomes, 17q-22q</td>
<td>CD114-PDGFB</td>
</tr>
</tbody>
</table>

### Back to the Case!

**GLUT-1**

IHC Positive: GLUT-1, CD34, Claudin-1

IHC Negative: EMA, S100, STAT6

FISH negative for FUS translocation

**CD34**

**Diagnosis:** Soft Tissue Perineurioma
Follow-up

- Patient is without recurrence after 4 months follow-up.

THANK YOU