Una Patología para el siglo XXI
Patología molecular, imagen digital y gestión
Cádiz
22-24 mayo 2013

Andrew E. Rosenberg, M.D.
Myoepithelioma of Bone and Soft Tissue

E. Rosenberg, M.D.
Andrés
Myoepithelioma

**Definition:** A neoplasm that demonstrates a myoepithelial phenotype

**Myoepithelial cell:** A normal cell located within an ductal acinar structures located between the lining epithelial cell and the underlying basement membrane. Demonstrate the ultrastructural and immunohistochemical features of an epithelial cells with some characteristics of smooth muscle.

**Neoplasms with Myoepithelial Cells:**
- Mixed tumors (salivary gland, skin)
- Myoepithelioma (salivary gland, soft tissue, bone, breast)
Myoepithelioma

**Definition:**
A neoplasm that demonstrates a myoepithelial phenotype.

**Myoepithelial cell:**
A normal cell located within ductal acinar structures located between the lining epithelial cells and the underlying basement membrane. Demonstrate the ultrastructural and immunohistochemical features of an epithelial cell with some characteristics of smooth muscle.

**Neoplasms with Myoepithelial Cells:**
- Mixed tumors (salivary gland, skin)
- Myoepithelioma (salivary gland, soft tissue, bone, breast)

**Immunohistochemistry:**
- Use markers such as p63 to identify myoepithelial cells.
Myoepithelial Neoplasms

**Morphology**
- Myoepithelioma
  - Spindle, plasmacytoid, epithelioid, clear cells, myxoid or hyalinized stroma
- Mixed Tumor
  - Presence of ductal structures

**Immunophenotype**
- Myoepithelioma
  - Keratin, EMA, S-100, GFAP, P63, calponin, SMA, desmin
- Mixed Tumor
  - Keratin, EMA, S-100, GFAP, P63, calponin, SMA, desmin

**Genetics**
- Myoepithelioma
  - Rearrangement of *EWSR1* - soft tissue
- Mixed Tumor
  - Rearrangement of *PLAG1* or *HMGA2* - Salivary gland
Myoepithelioma - Clinical Features

- Equal gender distribution
- Wide age range
- Peak incidence in 4th decade
- Usually painless mass - soft tissue; pain - bone
## Myoepithelioma - Anatomic Distribution

<table>
<thead>
<tr>
<th>Soft Tissue - Subcutaneous &gt;&gt; deep</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Limbs/limb girdles</td>
<td>75%</td>
</tr>
<tr>
<td>Head and neck</td>
<td>15%</td>
</tr>
<tr>
<td>Trunk</td>
<td>10%</td>
</tr>
</tbody>
</table>

## Bone

| Axial skeleton                    | 60%|
| Long bones – tibia                | 30%|
| Jaw bones                         | 10%|
Myoepithelioma – Macroscopic Features

Most tumors 2-5 cm
Occasionally >10 cm
Grossly well-circumscribed
Glistening/gelatinous
Myoepithelioma

16 year old female

31 year old female
Myoepithelioma

49 year old male
Myoepithelioma –
Histologic Features

Heterogeneous – similar to salivary gland lesions
Typically lobulated/multinodular
Often reticular
Myxoid stroma common
Occasionally solid
Often mixed patterns
Myoepithelioma – Histologic Features

Epithelioid, spindled, plasmacytoid (hyaline), or clear cells
Often mixed cell types
Metaplastic cartilage/bone in 15%
Myoepithelioma
<table>
<thead>
<tr>
<th>Protein</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Keratins</td>
<td>60%</td>
</tr>
<tr>
<td>S-100 protein</td>
<td>85%</td>
</tr>
<tr>
<td>EMA</td>
<td>60%</td>
</tr>
<tr>
<td>GFAP</td>
<td>50%</td>
</tr>
<tr>
<td>Calponin</td>
<td>50%</td>
</tr>
<tr>
<td>SMA</td>
<td>40%</td>
</tr>
<tr>
<td>p63</td>
<td>20%</td>
</tr>
</tbody>
</table>
Myoepithelioma – Genetics

45% have rearrangement of EWSR1

EWSR1 – POU5F1 (8%) t(6;22) – clear cells
EWSR1 – PBX1 (8%) t(1;22) – clear cells, epithelioid
EWSR1 – ZNF444 (1.5%) t(19;22) – epithelioid/clear/spindle

EWSR1 negative tumors are more often benign
45 year old woman
Myoepithelioma: - Criteria for Malignancy

Nuclear atypia
Mitotic activity
Necrosis
Malignant Myoepithelial Tumors

Soft tissue – children; bone - adults
Similar morphologic and immunophenotypic features
May contain an undifferentiated round cell component
Malignant Myoepithelioma
Malignant Myoepithelioma
# Myoepithelial Tumors – Clinical Behavior

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Recurrence</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myoepithelioma</td>
<td>20%</td>
<td>Rare</td>
</tr>
<tr>
<td>Myoepithelial carcinoma</td>
<td>40%</td>
<td>40-50%</td>
</tr>
</tbody>
</table>

Sites of metastasis: lung, lymph nodes, bone, brain, soft tissue
Myoepithelial Tumors - Differential Diagnosis

- Ossifying fibromyxoid tumor
- Epithelioid schwannoma
- Chondromyxoid fibroma
- Extraskeletal myxoid chondrosarcoma
- Epithelioid MPNST
- Metastatic carcinoma
- Proximal-type epithelioid sarcoma
- Conventional chondrosarcoma
- Ewing sarcoma
Extraskelatal Myxoid Chondrosarcoma

Distinctive rare myxoid sarcoma
Usually arise in deep soft tissues
Proximal extremities (thigh), trunk
High rate of lung metastasis (late)
NOT cartilaginous
Extraskeletal Myxoid Chondrosarcoma
Chondromyxoid Fibroma

<1% of primary bone tumors
75% younger than 30 years
pain / swelling
75% involve lower extremity
95% centered in metaphysis
Ilium / metatarsal
Myoepithelioma
Myoepithelial Tumors of Bone and Soft Tissue

Uncommon tumor
Benign > malignant
Morphologic heterogeneity
Express keratin, EMA, S100, SMA
50% have rearrangement of EWSR1
Treatment is complete excision