DERMATOPATHOLOGY?
A BRIEF TOUR OF UNUSUAL CASES ENCOUNTERED IN THE
DERMATOPATHOLOGY DIVISION

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Case 1

- 46 y.o female recently presented with a recurrent mass on her left heel
- Lesion initially biopsied 15 years ago in 2001
Diagnosis?
A. Myxoid neurofibroma  
B. Low grade fibromyxoid sarcoma  
C. Superficial acral fibromyxoma  
D. Dermatofibrosarcoma protuberans

Have you encountered this lesion in your practice?
A. YES  
B. NO

Superficial Acral Fibromyxoma  
(Digital Fibromyxoma)

- First described in 2001 (Fetsch et al.)
- Benign myxoid mesenchymal tumor
- Wide age range (mean = fourth decade)
- Male predilection (2:1)

- Subungual or periungual regions of fingers and toes
  - Also other acral sites (palms / soles)
- Usually less than 2 cm (range 0.5 to 5 cm)
- Can be painful
Superficial Acral Fibromyxoma
(Digital Fibromyxoma)

- Benign
- Rarely erode bone
- Can recur if incompletely excised

CD34
Case 2

- 50 y.o. female presented with an abdominal wall mass in the subcutaneous adipose tissue 8 years after having a lesion removed from her right hand.
Which diagnosis can you exclude?

A. Metastatic malignant melanoma
B. Metastatic poorly differentiated squamous cell carcinoma
C. Metastatic clear cell sarcoma

A. Metastatic malignant melanoma
C. Metastatic clear cell sarcoma

What would you do next?

A. Nothing – the answer is already clear
B. Additional immunohistochemical stains
C. Fluorescence in situ hybridization (FISH) analysis
D. Flow cytometry

FISH

“Positive for EWSR1 gene rearrangement in 199/200 nuclei”
Clear Cell Sarcoma

- Melanin-producing soft tissue sarcoma
- Sometimes referred to as “malignant melanoma of soft parts” – not accurate as lesions are separate entities

Clear Cell Sarcoma

- Young adults (median 30 yrs)
- Deep soft tissue of extremities typically in association with tendons or aponeuroses
  - Foot and ankle (40%)
  - Knee, thigh and hand (30%)
- Skin usually not involved
Clear Cell Sarcoma

- Immunohistochemistry
  - S-100 protein positive
  - Positive for markers associated with melanin production (Melan-A, HMB-45, MiTF, etc)

- Electron microscopy
  - Melanosomes

Clear Cell Sarcoma FISH

- t(12;22)(q13;q12)
- Results in EWSR1-ATF1 fusion protein
  - Activates promoter for MiTF
    - Melanin production
    - Cell proliferation
- Found in most clear cell sarcomas, but not in melanoma

Clear Cell Sarcoma

- Wide excision
- Poor prognosis (survival rate 10% at 20 yrs)
  - Prolonged course
  - Metastases to lymph nodes, lung, bone
    - Late metastases possible

Case 3

- 65 y.o. male presented with a 6 mm left elbow mass
Diagnosis?
A. Glomangioma
B. Myopericytoma
C. Leiomyosarcoma
D. Neurothekeoma

Myopericytoma
**Myopericytoma**

- Benign perivascular myoid tumor
- Wide age range
- Painless, slow growing mass in subcutaneous tissue or dermis
- Most common in distal extremities
  - Also proximal extremities, neck, trunk, etc.
  - Rarely visceral or intracranial

**Myopericytoma**

**Immunohistochemistry**

- Smooth muscle actin positive
- h-caldesmon positive
- Desmin negative or only focally positive
References


• Dim DC, Cooley LD, Miranda RN. Clear Cell Sarcoma of Tendons and Aponeuroses. Arch Pathol Lab Med. 2007;131:152-156.


