Cutaneous Sarcomas

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Soft tissue sarcomas (STS)

- Uncommon but diverse mesenchymal malignancies.
  - Bone
  - Muscle
  - Nerves
  - Cartilage
  - Fibrous tissue
  - Fat
  - Deep skin

- Incidence
  - US: 11,000 (4,000 deaths)
  - Worldwide: 200,000
~ 60 subtypes
Each has its own clinical course.

Cutaneous Sarcomas

Low Risk Lesions
- Atypical Fibroxanthoma
- Cutaneous Primary Leiomyosarcoma

High Risk Lesions
- Angiosarcoma
- Superficial Dermal Sarcoma
- Recurrent DFSP
- Myxofibrosarcoma
- Malignant Peripheral Nerve Sheath Tumor
Focus of Talk

- Angiosarcoma
- DFSP
- Atypical Fibroxanthomma
- Pleomorphic Dermal Sarcoma
History

• 79F with left arm lymphedema following mastectomy, lymph node dissection, and radiation therapy 8 years ago.

• Noted a 2.5 cm purple mass of left forearm.
Scouting biopsies negative

Biopsy = angiosarcoma
10 cm diameter circular resection
Angiosarcoma

- Vascular neoplasms 1% of sarcomas
- Commonly arise face or scalp of elderly patients
- Most are sporadic
- Can occur 3 years after radiation.
- Long-standing lymphedema (Stewart-Treves)
Angiosarcoma: prognosis

• Highly aggressive
  – Local recurrence = 26%
  – Distant metastasis = 22%.

• Worse prognosis associated with older age, epithelioid histology, tumor necrosis.

• 5-year disease specific survival = 48%

**Angiosarcoma: treatment**

- Multimodality approach by a sarcoma team.
  - Do not treat alone.

- May consider neoadjuvant radiation therapy or chemotherapy.

- Resection with wide margins.
Dermatofibrosarcoma Protuberans

- Cell of origin remains unknown
- No known etiologic factors
- 18% of all cutaneous sarcomas
- Translocation t(17:22) leads to fusion between COL1A1 and PDGF-β.
DFSP: clinical presentation

• Slow persistent growth over several years.

• Firm, plaque like lesion with surrounding red to blue discoloration.

• Protuberans secondary to neglected lesion that has fully developed.
DFSP: treatment

• Tumor involves skin and subcutis. Rarely involves underlying muscle.

• Surgery cornerstone in treatment.

• Radiation therapy for unresectable tumors or close margins.

• Imatinib for metastatic disease.
DFSP: local recurrence

- Local recurrence rate is 20-50%.

- Develops within 3 years of initial surgery, although one-third will develop recurrence after 5 years.

- Wide local excision
  - >3 cm, recurrence is 20%
  - <2 cm, recurrence is 41%

- Mohs
  - Recurrence is < 10%

DFSP: case presentation

• 63M presented with recurrent DFPS of the scalp in 2013.

• > 12 resections since 1982.

• CT negative for distant disease.
DFSP: metastasis

• Metastasis is rare (3.4%)
• May be more common if fibrosarcomatous DFSP.
• Usually always recurrent lesions.
• 75% metastasis are to the lung and 25% are to lymph nodes.
• If oligometastasis, metastasectomy.
• May respond to imatinib.
Atypical Fibroxanthoma

- Expansile dermal nodule
- Solitary nodules, < 2 cm
- Predisposing factors
  - Solar: Sun-damaged skin
  - Radiation: w/i 10 years
- Head and Neck

- Does not involve subcutis
- Does not invade fascia or muscle
- Histology resembles UPS
AFX: prognosis and treatment

• Excellent prognosis

• Local recurrence rate = 3%

• Distant metastasis rate = 0%

• Conservative resection; although Mohs has also shown to be effective.
Superficial Undifferentiated Pleomorphic Sarcoma

• Similar to AFX, but...

• Tumor is larger; > 2 cm

• Involves subcutis, penetrates fascia and muscle, or exhibits necrosis or vascular/perineural invasion.
Superficial UPS: semantics

• Important differentiate AFX from Superficial UPS (“metastasizing AFX”).

• Also called Pleomorphic Dermal Sarcoma

• Malignant fibrous histiocytoma (MFH) concept has been questioned. Agreed that these often represent liposarcomas, leiomyosarcomas, etc.

• Only a small group that remains undifferentiated is now referred to as Undifferentiated Pleomorphic Sarcoma. “MFH” term rarely used.
Superficial UPS: prognosis

- Local recurrence rate = 30%
- Distant metastasis rate = 10-20%
  - Skin, lung
- Underscores need to differentiate from AFX.

Superficial UPS: treatment

• Wide local excision with negative margins.

• Radiation therapy should be considered for those tumors > 5 cm.

• Chemotherapy for metastasis.

Summary

• Confirm diagnosis.

• Confirm stage for high risk lesions (angiosarcoma, superficial UPS, multiple recurrent DFSP) with imaging.

• Surgical excision with wide margins offers best chance for cure.

• High risk lesions best treated with multimodality approach.
Confirm Diagnosis

- **Low Risk**
  - Margin negative resection

- **High Risk**
  - Multi-D Team