Greatest Clinicopathologic Cases

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DISCLOSURES

I do not have any relevant relationships with industry.
Case #1

• 69 year old female
• “Draining lumps” on left leg x months, painful
• Bedbound with decubitus ulcers but feels these look different
• Denies systemic symptoms
• PMH: CAD, PVD, HTN, HLD, DM, Afib, morbid obesity
• Significant Medications: warfarin, metoprolol, furosemide, omeprazole, baclofen, meloxicam, loperamide, flecainide, tramadol
Poll: You decide to proceed with a 4mm punch biopsy of the area. What is your leading diagnosis?

A. Calciphylaxis
B. Diffuse dermal angiomatosis
C. Deep fungal infection
D. Medium vessel vasculitis
E. Warfarin-induced skin necrosis
Poll: Given the pathologic findings what is your leading diagnosis?

A. Calciphylaxis
B. Diffuse dermal angiomatosis
C. Deep fungal infection
D. Medium vessel vasculitis
E. Coumadin necrosis
Given the pathologic findings what is your leading diagnosis?

A. Calciphylaxis  
B. **Diffuse dermal angiomatosis**  
C. Deep fungal infection  
D. Medium vessel vasculitis  
E. Coumadin necrosis
Diffuse Dermal Angiomatosis

• Cutaneous benign reactive vascular proliferation
• Considered to be a variant of cutaneous reactive angioendotheliomatosis
• Possibly due to local tissue hypoxia → reactive angiogenesis

• Classically associated with:
  • Severe (peripheral) atherosclerotic disease
  • Smoking history
  • Arteriovenous fistula
- Painful, violaceous plaques
- Often ulcerated
- Lower extremities
• Diffuse dermal proliferation of endothelial cells
  • Often spindled with vacuolated cytoplasm
• Interstitially arranged between collagen bundles
• Cells + for endothelial markers: CD34, CD31, Erg, *Ulex europaeus*

Diffuse Dermal Angiomatosis of the Breast

• Associated with large, pendulous breasts
• Lack a history of a vaso-occlusive disease process
• Possibly traumatic ulcerations of these fat-rich areas → subsequent angiogenesis

DDA: Treatment

• **Reversal of underlying hypoxemia-inducing condition**
• Oral corticosteroids
• Isotretinoin: antiangiogenic effect
Case #2

• 65 year old female
• New non-painful lump on the left arm for “many months”
• 5mm soft dermal nodule
• Outside ultrasound: “Indeterminate hypoechoic mass... probably a lymph node”
• Consistent with lipoma → sent for excision
Poll: What is the best diagnosis?

A. Dermatofibrosarcoma protuberans
B. “Low fat” spindle cell lipoma
C. Desmoplastic melanoma
D. Solitary fibrous tumor
E. Angioleiomyoma
What is the best diagnosis?

A. Dermatofibrosarcoma protuberans
B. “Low fat” spindle cell lipoma;
C. Desmoplastic melanoma
D. Solitary fibrous tumor
E. Angioleiomyoma
Poll: Which gene fusion is associated with this condition?

A. NAB2-STAT6
B. COL1A2-PDGFB
C. EWSR1-ETS
D. SYT-SSX
Which gene fusion is associated with this condition?

A. NAB2-STAT6: Solitary fibrous tumor
B. COL1A2-PDGFB: Dermatofibrosarcoma protuberans
C. EWSR1-ETS: Ewing's Sarcoma
D. SYT-SSX: Synovial Sarcoma
Solitary Fibrous Tumor

- Spindle cell mesenchymal tumor
- Originally described as a pleural tumor
- Also reported in:
  - Soft tissue (40% of extrapleural cases; head and neck)
  - Skin
  - CNS
  - Liver
  - Kidney
  - Thyroid
Histopathology

• Fascicles of spindle cells
• Alternating hypo and hypercellular areas
• CD34+ → may be confused with DFSP and spindle cell lipoma
• NAB2-STAT6 fusion gene
  • NAB2 is a transcriptional repressor of the early growth response (EGR) transcription factor, and its fusion with STAT6 converts the repressor into a transcriptional activator
Superficial SFT

• Dermal & Subcutaneous

• Feasel et al., American Journal of Surgical Pathology, 2018:
  • Review of 26 cases
  • Female
  • Cephalic predominance
  • Generally low risk/indolent
<table>
<thead>
<tr>
<th>Risk factor</th>
<th>Score</th>
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<tr>
<td>Age</td>
<td></td>
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<tr>
<td>&lt; 55</td>
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<tr>
<td>≥ 55</td>
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<tr>
<td>Tumor size (cm)</td>
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<tr>
<td>Mitotic count (/10 high-power fields)</td>
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<td>≥ 4</td>
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<td>Tumor necrosis</td>
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<td>Intermediate</td>
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<tr>
<td>High</td>
<td>6–7</td>
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- Low risk: No metastases at 10 years
- Intermediate risk: 10% 5 and 10 year metastatic risk
- High risk: 73% 5 year metastatic risk
Treatment

• Low risk of metastasis
• Complete surgical excision is generally curative
• Often incompletely excised: 25-36% can recur