Mesenchymal Tumors

Focus on Vascular Tumors
- Benign and the “Probably Benign”
  - Newly-described and variant lesions
- Malignant
  - Distinction of benign/low grade lesions from Angiosarcoma
  - What is NOT Angiosarcoma

Focus on Angiomyolipoma: Problem variants that still lead to diagnostic errors
- Epithelioid, inflammatory, trabecular

Vascular Tumors

The Benign and Probably Benign

Hemangioma Variants
Vascular Malformations

Cavernous Hemangioma (CH)

Not true arterial or venous architecture
- No organized muscle bundles
- No elastic laminas
- Not capillary-like
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Cavernous Hemangioma:
What is often “not seen”….

- Hemangioma-like vessels (HLV) in adjacent liver commonly seen with giant CH
  - Low mitotic/proliferative rate <5%
  - Present in almost 80% (16/19) of CH >5 cm
  - Retain composition of vascular walls in CH

Sclerosis within Cavernous Hemangioma

- Sclerosis of thrombosed, ischemic zones with scar formation.
  - “Neo-vessels”
  - Recanalized channels

Cavernous Hemangioma-like vessels in adjacent liver
Giant Cavernous Hemangioma

38 yr old woman, in liver failure.
- Explant, right lobe with giant hemangioma
- Left Lobe: Smaller, irregularly shaped CHs and transitional areas with HLVs admixed with liver, as well as smaller hemangiomas

“Metatastatic” and “Invasive” Cavernous Hemangioma

Lesion extending into hilum around arteries, nerves and ducts

Omental Lesion

Cavernous Hemangioma Variant

Diagnoses: Giant Cavernous Hemangioma and Cavernous Hemangiomatosis
- CH-like vessels throughout liver
- Lung, spleen, omentum involved with CH-like lesions

Vascular Malformations

- Hereditary Hemorrhagic Telangiectasia (HHT) arterial-venous malformations
  - also known as Osler-Weber-Rendu

- Other Arterial and Venous Malformations with similar features
  - (may or may not be HHT)

Vascular Malformations

Spectrum:
Early, mild
To
Late, severe

Early or mild lesions can look much different than advanced or severe lesions probably primarily due to thrombosis and ischemic effects.

Vascular Malformations

Contributors and co-authors of 2 abstracts:


Vascular Malformations:
Early Lesions or Mild Involvement

Periportal fibrosis, Elastochrome stain
Periductal fibrosis (as early ischemic lesion)
Vascular Malformations: More Severe or Advanced Lesions

- Extension of lesions into sinusoids
- Thrombosis within vessels and sinusoids

Vascular Malformations: Severe sinusoidal changes

- Hemangioma-like changes, extensive sinusoidal dilation
- Cavernous hemangioma-like transformation

Small Vessel Hemangioma

- Rare
- Newly described
- Small vascular channels with thin walls
- Bland endothelial cells with low proliferative rate (<10% CH <5%)
- Intermediate tumor cell density
- Irregular “infiltrative” growth pattern at border
- Abnormal liver architecture mimics HCC
- Scaffolding effect mimics angiosarcoma
Small Vessel Hemangioma

- Small channels, thin walls, bland nuclei
- Only focal fibrotic areas
- Center of lesion, bland endothelial cells
- Edge of lesion, with altered cell plate width

Small Vessel Hemangioma

- Small channels with thin walls, no organized muscle
- Low Mib1 (Ki-67) rate
- Edge of lesion, trichrome
- Edge of lesion, reticulin
**Small Vessel Hemangioma**

- Small vessel hepatic hemangioma (SVH): Exact outcome not definitive, so now recommending excision and followup.
- **Differentiation from angiosarcoma**: AS has higher proliferative rate (>15%) and subset + for P53 and GLUT1, but negative in small vessel hemangioma.

**References**

**Epithelioid Hemangioendothelioma**

- **Elastochrome stain: Central vein involvement by tumor**
- **Elastochrome: trichrome plus EVG stain**
  - highlights vein wall elastic fibers

- **Epithelioid Hemangioendothelioma**
  - Angiosarcoma-like pattern of scaffolding growth
Angiosarcoma

- Most aggressive form of vascular malignancy
- Highest proliferative rate
- Epithelioid or spindle cell forms
- Cystic and/or solid
- Known for the typical feature of "scaffolding" growth pattern

Angiosarcoma

- Epithelioid pattern
- High MiB1 (Ki-67) rate

Angiosarcoma

- Scaffolding growth pattern along sinusoids
- CD34 and expanded sinusoidal growth

Angiosarcoma

- Cystic change (upper right)
- Congestion
- Necrosis
- Sinusoidal growth
Angiosarcoma

Scaffolding pattern of growth surrounds hepatocytes

Sinusoidal growth results in anastomosing channels and pseudopapillary pattern

Angiosarcoma: Highlights

- High proliferative rate and cytologic atypia
- Early pattern of growth typically along sinusoids (scaffold-like); atypical endothelial cells, dilated sinusoids
- Later pattern of growth can be pseudopapillary to solid; irregularly-shaped blood filled spaces
- Lacks the stromal prominence of epithelioid hemangioendothelioma, but overlapping cases may be seen
Undifferentiated (Embryonal) Sarcoma of the Liver

What else is NOT angiosarcoma

Typically younger patients; tumor of uncertain etiology
Can be cystic due to necrosis/degneration with irregular edges! (Pattern similar to angiosarcoma scaffolding)

Immunohistochemistry
- Reactive with alpha-1-antitrypsin, alpha-1-antichymotrypsin, vimentin
- Occasional cytokeratin positivity
- Some CD10 and p53 positivity
- Negative hepatocyte-Ab, muscle, S-100 and CD34

- Glypican-3 can be positive in giant cells (personal observation)

Cystic areas common
Related to extensive necrosis
(right)
**Undifferentiated sarcoma, tumor edge with growth along sinusoids**

- PASD + globules
- Also Alpha-1-antitrypsin +

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**Undifferentiated Embryonal Sarcoma**

**Problem with Literature Search**


- **THIS IS NOT THE CORRECT DIAGNOSIS** as per three expert consultants

- Authors got confused about peripheral growth

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**Angiomyolipoma**

**Problem variants**

- Epithelioid, Trabecular, and Inflammatory

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**Problem Case**

- 37-year-old woman
- 11 cm pedunculated mass
- No cirrhosis or other risk factors for HCC
- Mass noted during routine gynecologic exam, no symptoms
HCA, HCC?

Reticulin Stain: too much loss for HCA

Keratin and HMB-45

Angiomyolipoma, epithelioid variant

Angiomyolipoma

Classic features:
- Fat
- Epithelioid cells
- Spindle cells

Angiomyolipoma

- Epithelioid Cells
- Spindle Cells

Angiomyolipoma

- HMB-45: stains stronger on epithelioid cells
- SMA: usually stains spindle cells

Problem Case: Trabecular Angiomyolipoma
Problem Case: Trabecular Angiomyolipoma

Problem Case: Inflammatory Angiomyolipoma

Focal dense to scattered diffuse T-cell infiltrate

Problem Case: Angiomyolipoma, Inflammatory and Trabecular

Case with both inflammatory and "trabecular" background

Problem Case: Angiomyolipoma, Inflammatory and Trabecular

HMB-45

SMA
Angiomyolipoma, Mixed variant

Fatty areas

Trabecular areas

Angiomyolipoma, Mixed variant

Inflammatory areas, 10x

THANKS FOR THE INVITATION TO PRESENT THIS TOPIC

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HMB-45
Inflammatory foci with absent staining
(SMA only rare + cell, not shown)