



# AMSER Rad Path Case of the Month:

59 yo male with history of renal cell carcinoma s/p left radical nephrectomy presents for surveillance CT of the abdomen/pelvis

Austin Snyder, MS3

Harvard Medical School

Paige Parrack, MD

Brigham and Women's Hospital, Department of Pathology

Angela Ann Giardino, MD

Brigham and Women's Hospital, Department of Radiology



### **Patient Presentation**

Clinical history:

 59 yo male w/ history of multifocal unclassified renal cell carcinoma (as well as separate papillary adenoma, angiomyolipoma and adrenal adenoma) s/p left radical nephrectomy presents for surveillance abdominal CT.

Family history:

Colon cancer (paternal uncle and paternal grandfather)

Social history:

• Non-smoker, works as a teacher

Physical exam:

• No abdominal masses or tenderness, no CVA tenderness, no lymphadenopathy

#### Laboratory Data

• CBC, BMP, LFTs, and PT-INR unremarkable

# ACR Appropriateness Criteria

- Per NCCN guidelines for unclassified RCC s/p left radical nephrectomy, patient underwent surveillance imaging every 6 months.
- Imaging modality was selected using the American College of Radiology Appropriateness Criteria (see right).

American College of Radiology ACR Appropriateness Criteria <sup>®</sup> Post-Treatment Follow-up and Active Surveillance of Clinically Localized Renal Cell Cancer <u>Variant 1:</u> Follow-up for clinically localized renal cell cancer; post radical or partial nephrectomy.		
CT abdomen with IV contrast	Usually Appropriate	***
MRI abdomen without and with IV contrast	Usually Appropriate	0
CT abdomen without and with IV contrast	Usually Appropriate	***
CT abdomen and pelvis with IV contrast	May Be Appropriate	***
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	***
CT chest with IV contrast	May Be Appropriate	
MRI abdomen without IV contrast	May Be Appropriate	0
Radiography chest	May Be Appropriate	۲
US abdomen with IV contrast	May Be Appropriate	0
CT abdomen without IV contrast	May Be Appropriate	***
CT chest without IV contrast	May Be Appropriate	⋧⋧⋧
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate (Disagreement)	0
MRI abdomen and pelvis without IV contrast	May Be Appropriate	0
CT abdomen and pelvis without IV contrast	May Be Appropriate	
FDG-PET/CT skull base to mid-thigh	May Be Appropriate	****
US kidney retroperitoneal	May Be Appropriate	0

#### CT Abdomen/Pelvis w/ Contrast 18 months s/p Left Nephrectomy





Axial (unlabeled)

Coronal (unlabeled)

#### CT Abdomen/Pelvis w/ Contrast 18 months s/p Left Nephrectomy



- 13 mm hyperattenuating soft tissue nodule (arrow) adjacent to the right diaphragmatic crus at the level of the upper pole of the right kidney.
- Of note, there was no evidence of tumor recurrence in the left nephrectomy bed.

#### 1<sup>st</sup> Surveillance CT Abdomen/Pelvis w/ Contrast 6 months s/p Left Nephrectomy





Axial (unlabeled)

Coronal (unlabeled)

#### 1<sup>st</sup> Surveillance CT Abdomen/Pelvis w/ Contrast 6 months s/p Left Nephrectomy



- 5 mm hyperattenuating soft tissue nodule (arrow) adjacent to the right diaphragmatic crus at the level of the upper pole of the right kidney.
- Of note, there was no evidence of tumor recurrence in the left nephrectomy bed.

#### Imaging DDx for Retroperitoneal Soft-Tissue Nodule

- Metastatic disease
- Soft tissue sarcoma (angiosarcoma, liposarcoma, leiomyosarcoma, rhabdomyosarcoma, etc.)
- Angiomyolipoma
- Hemangioma
- Lymphoma
- Fibroma
- Germ cell tumor
- Other rare etiologies

### **CT-Guided Core Biopsy**

• Given the progressive increase in size of the nodule, a CT-guided core biopsy was performed to rule out metastatic disease.





## Micro Path



Core biopsy showing irregular, anastomosing sinusoidal vascular channels (arrowheads).

The vascular spaces are lined by bland, endothelial cells with focal hobnail morphology. The lesion is also characterized by intracytoplasmic hyaline globules and fibrin thrombi (below).



# Immunohistochemistry



ERG highlights the endothelial cells lining the vascular channels.

SMA (smooth muscle actin) shows diffuse staining of pericytes adjacent to the endothelium.

Negative: PAX8, HMB-45, MELAN-A

#### Final Dx:

#### Anastomosing Hemangioma

#### Case Discussion

- An anastomosing hemangioma (AH) is a benign neoplastic vascular tumor composed of irregularly anastomosing sinusoidal-like spaces lined by endothelial cells that classically mimics the appearance of an angiosarcoma.
- AHs most commonly arise in the kidneys and retroperitoneal fat, but can also be seen in the adrenals, liver, colon, testes, ovaries, and bones.
- The most common comorbidity associated with AHs is end-stage renal disease.
- AHs are most commonly asymptomatic and found incidentally on imaging as a circumscribed, hyperdense, heterogeneous mass.

## Case Discussion (cont.)

- On histology, AHs appear as irregular capillary-sized vascular spaces with anastomoses lined by a single layer of endothelial cells with hobnail morphology, and intravascular fibrin thrombi.
- Immunohistochemical analysis is typically positive for endothelial markers, including smooth muscle actin and ERG, and negative for HHV-8 (Kaposi sarcoma), HMB-45 and MELAN-A (angiomyolipoma), and PAX-8 (renal cell carcinoma).
- Absence of high-grade features (nuclear atypia, pleomorphism, active mitoses) and plasma cells helps differentiate AH from angiosarcoma.

## Case Discussion (cont.)

- Patients with AHs tend to have great prognoses, given the benign nature of the tumor and absence of metastatic behavior.
- Patients with renal AHs are commonly treated with total nephrectomy given their worrisome appearance on histology, although this is not necessarily the best treatment given that AHs are benign neoplasms and do not metastasize.
- Nonrenal lesions are also typically treated with simple excision, but there does not appear to be recurrence in patients who do not undergo surgical excision, which suggests observation may be better than surgical treatment.
- Efforts to avoid misdiagnosis of AHs as malignant neoplasms is crucial, as AHs can be safely observed, while malignant neoplasms such as angiosarcomas require prompt surgical treatment.

#### Back to Our Patient

 Given that the patient's radiologic and pathologic findings were most consistent with an anastomosing hemangioma rather than a malignant process such as angiosarcoma, the patient was followed with routine CT scans of the abdomen/pelvis, and surgical excision was not performed.

### References

- 1. Cheon, P. M., Rebello, R., Naqvi, A., Popovic, S., Bonert, M., & Kapoor, A. (2018). Anastomosing hemangioma of the kidney: radiologic and pathologic distinctions of a kidney cancer mimic. *Current Oncology*, *25*(3), e220.
- 2. John, I., & Folpe, A. L. (2016). Anastomosing hemangiomas arising in unusual locations. *The American journal of surgical pathology*, *40*(8), 1084-1089.
- 3. Lappa, E., & Drakos, E. (2020). Anastomosing hemangioma: short review of a benign mimicker of angiosarcoma. *Archives of pathology & laboratory medicine*, 144(2), 240-244.
- 4. Lin, J., Bigge, J., Ulbright, T. M., & Montgomery, E. (2013). Anastomosing hemangioma of the liver and gastrointestinal tract: an unusual variant histologically mimicking angiosarcoma. *The American journal of surgical pathology*, *37*(11), 1761-1765.
- Messiou, C., B Med Sci, BMBS, MRCP, FRCR, MD, Moskovic, E, Vanel, D, Morosi, C, Benchimol, R, Strauss, D, . . . Bonvalot, S. (2016). Primary Retroperitoneal Soft Tissue Sarcoma: Imaging appearances, pitfalls and diagnostic algorithm. European Journal of Surgical Oncology, 43(7), 1191-1198.
- 6. O'Neill, Ailbhe C, Craig, Jeffrey W, Silverman, Stuart G, & Alencar, Raquel O. (2016). Anastomosing hemangiomas: Locations of occurrence, imaging features, and diagnosis with percutaneous biopsy. *Abdominal Radiology (New York), 41*(7), 1325-1332.
- 7. Perdiki, M., Datseri, G., Liapis, G., Chondros, N., Anastasiou, I., Tzardi, M., ... & Drakos, E. (2017). Anastomosing hemangioma: report of two renal cases and analysis of the literature. *Diagnostic pathology*, *12*(1), 1-9.
- 8. Rajiah, P., Sinha, R., Cuevas, C., Dubinsky, T. J., Bush Jr, W. H., & Kolokythas, O. (2011). Imaging of uncommon retroperitoneal masses. *Radiographics*, *31*(4), 949-976.
- 9. Zhao, M., Li, C., Zheng, J., & Sun, K. (2013). Anastomosing hemangioma of the kidney: a case report of a rare subtype of hemangioma mimicking angiosarcoma and review of the literature. *International journal of clinical and experimental pathology*, *6*(4), 757.