

RADY 401 Case Presentation

William King, August 2019

Focused patient history and workup

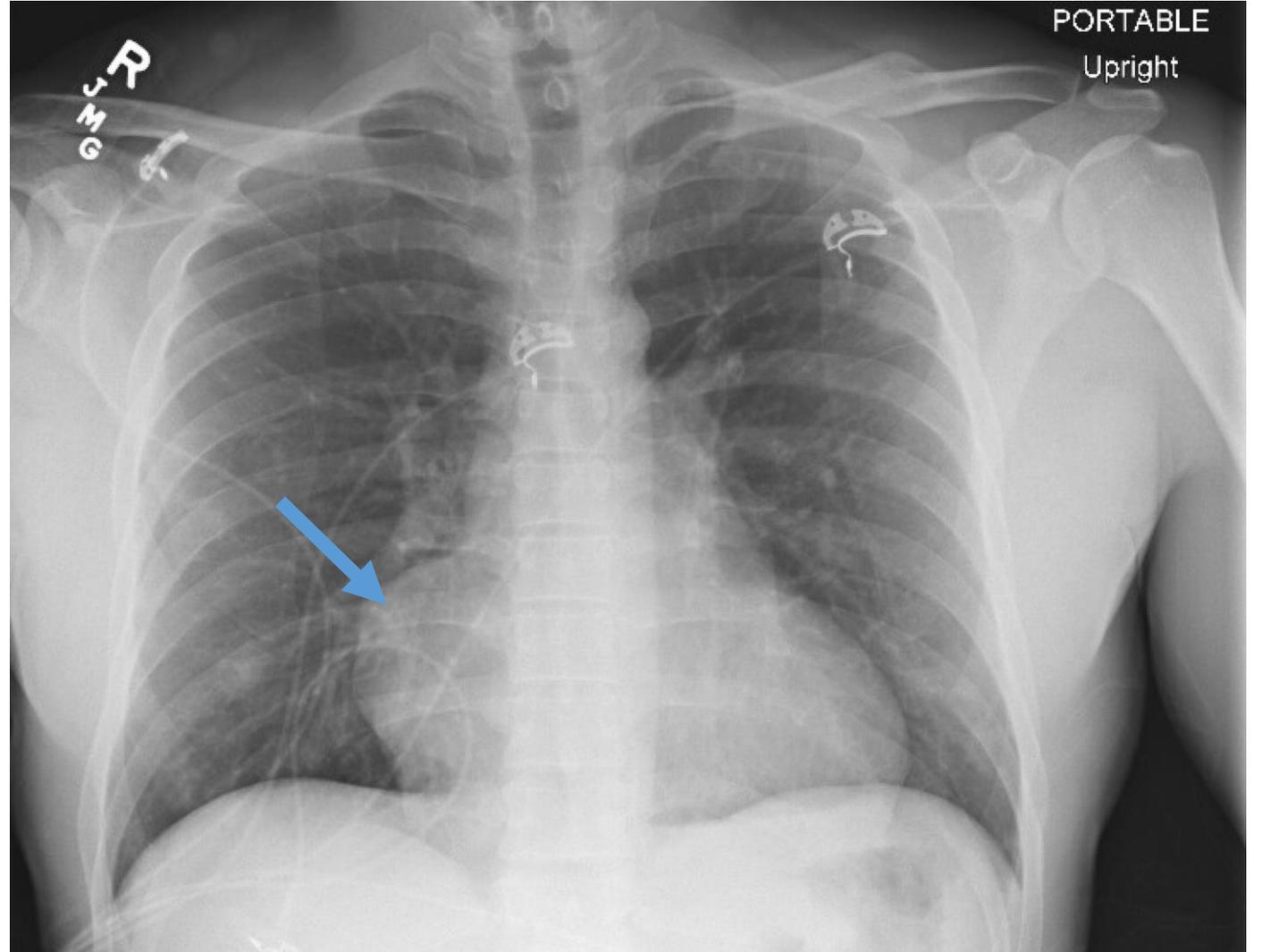
- 32-year-old man with history of pericardial effusion s/p drainage presents to Womack Army Medical Center with worsening dyspnea on exertion and pleuritic chest pain
- Found to have peripheral edema, but a BNP of 28. Kidney and liver function normal
- Troponin 0.4
- Transthoracic echocardiogram 9/21/18: “Large heterogeneous echodensity (4.5cm in diameter) surrounding the RV and RA, with the appearance of invasion into the RA wall. This is concerning for malignancy.”
- Transferred to UNC for “cardiac mass”

List of imaging studies

- Transthoracic echocardiogram
- Portable AP (anterior-posterior) chest radiograph
- Chest CT
- PET-CT of the chest

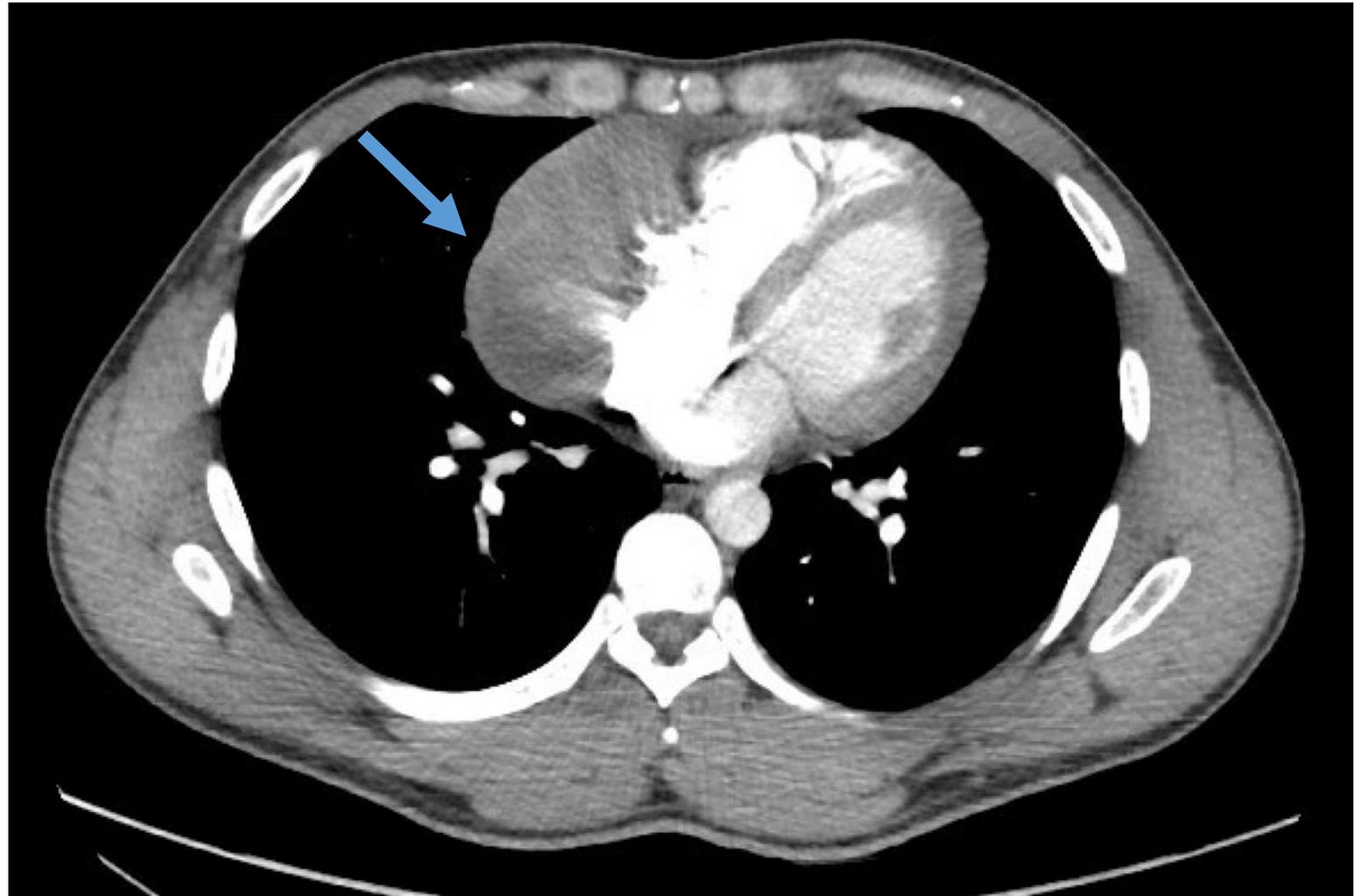
Portable AP chest radiograph UNC

- “Right paracardiac mass and bilateral parenchymal and subpleural nodular opacities, better assessed on the prior day outside the chest.”



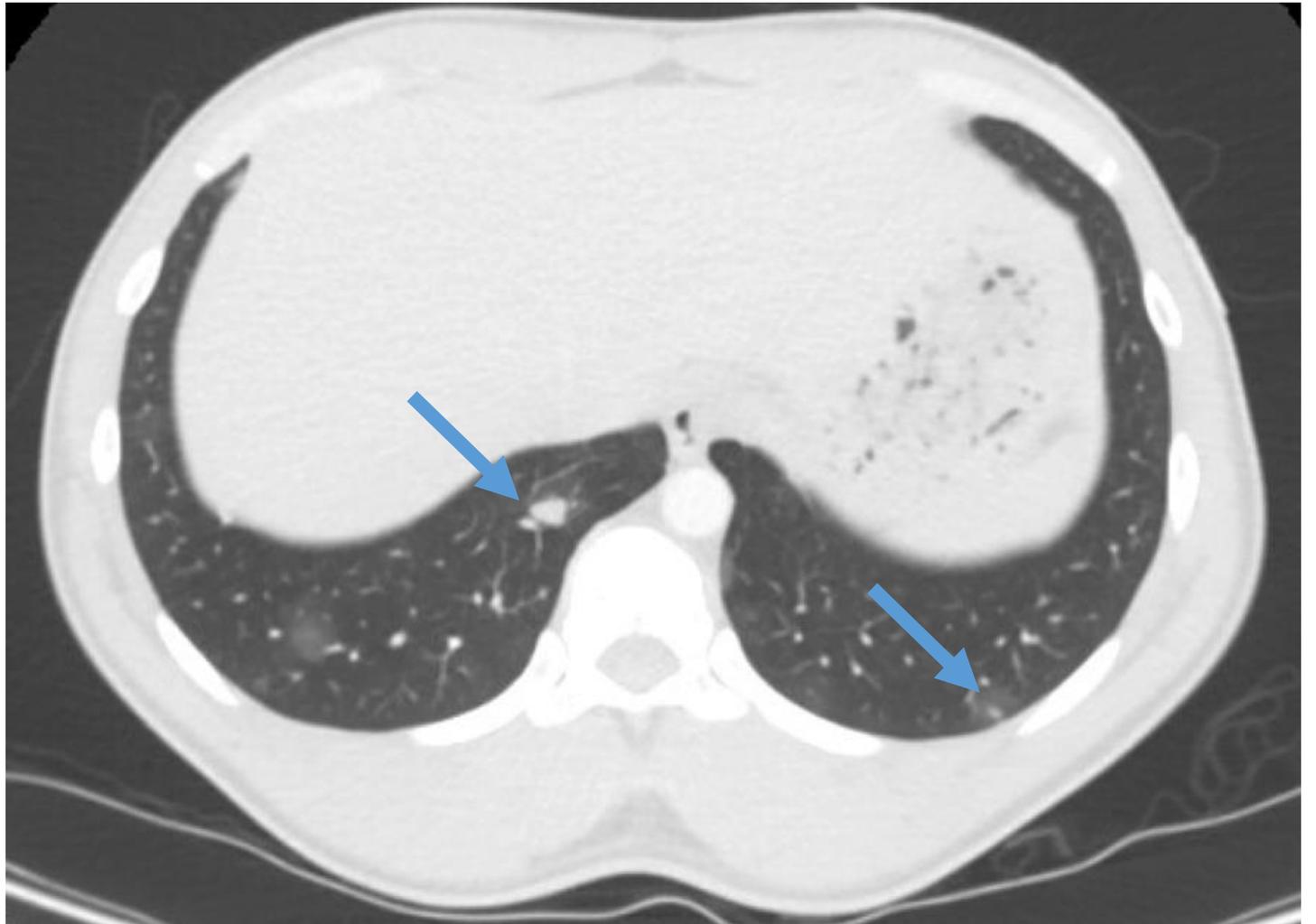
Chest CT from outside hospital

- "9.3 x 4.7 cm soft tissue lesion along the right heart wall, with mild mass effect on the lumen of the right atrium and right ventricle. No definite evidence of intracardiac thrombus."
- "Differential includes lymphoma and angiosarcoma and metastasis."



Chest CT from outside hospital – re planned biopsy

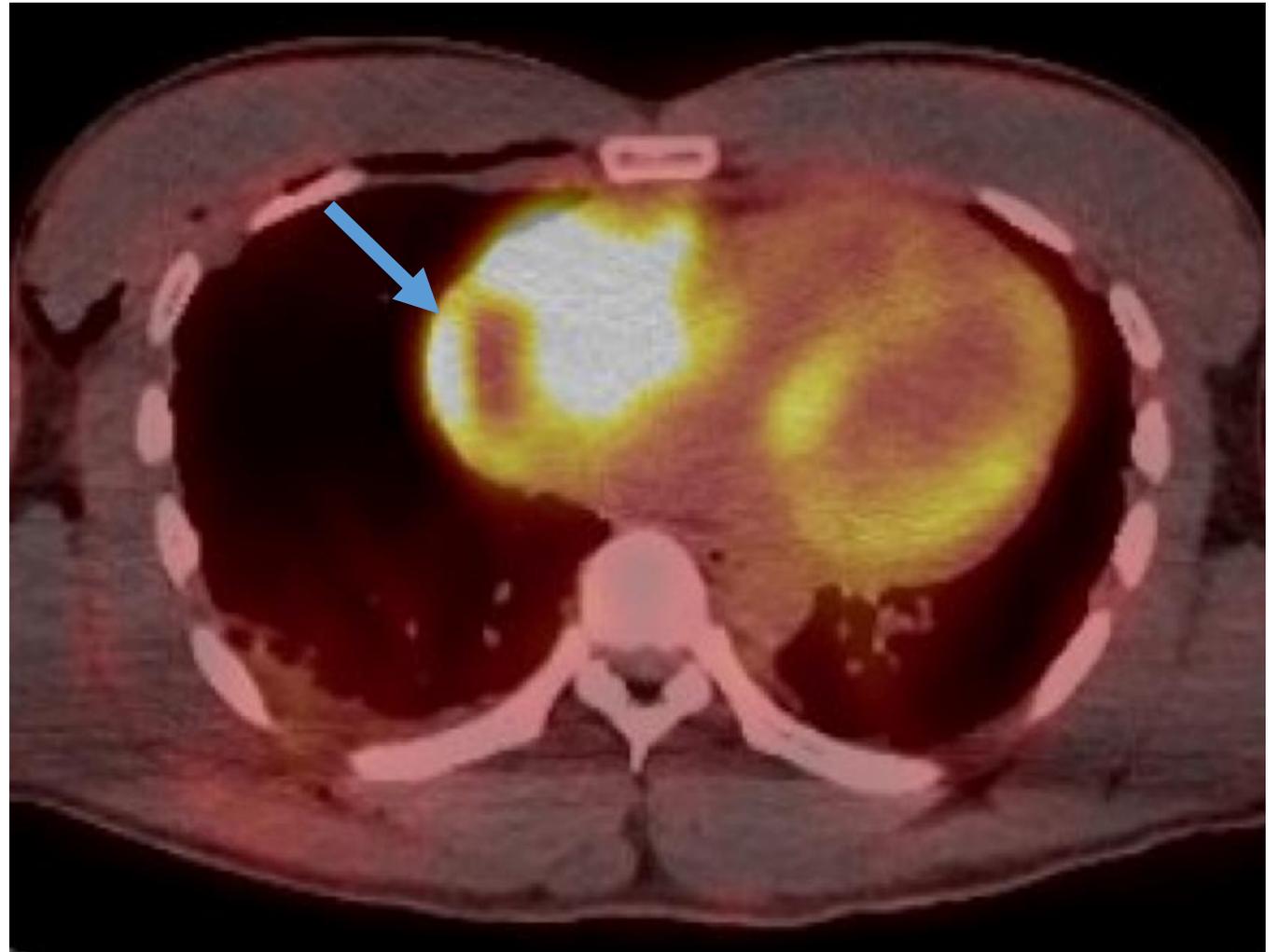
- "Numerous peripheral and subpleural pulmonary nodules with surrounding groundglass, measuring up to 1 cm."
- "Planning prescan CT images, reviewed with Dr. Yu at time of acquisition, did not demonstrate a window to perform the procedure safely without risk of complication."



"The receiver-operating-characteristic analysis revealed an optimal cutoff of 3 morphologic criteria, with a high specificity of 100% and a sensitivity of 70%. Using a threshold of malignancy of 4 or more morphologic criteria increased the positive predictive value to 100% at the cost of a lower sensitivity of 71%." –Rhabhar *et al.*

PET CT UNC

- "Soft tissue mass abutting the right cardiac wall is again visualized and demonstrates heterogeneous and intense uptake, worrisome for malignancy with necrotic areas."



"An SUV_{max} of 3.5 reveals a sensitivity of 100% and specificity of 86%, with a positive predictive value of 94% and a negative predictive value of 100%. With an SUV_{max} of 4.6, the sensitivity drops to 94% and the specificity rises to 100%, with a positive predictive value of 100%." –Rahbar *et al.*

Diagnosis and Treatment

- Underwent biopsy via video-assisted thoracoscopic surgical biopsy of lung nodules
- Pathology showed spindle cell proliferation and extensive hemangiovascular invasion consistent with low-grade metastatic angiosarcoma
- Not a candidate for resection due to extensive involvement of right atrial free wall and numerous pulmonary metastases

Outcome

- Underwent palliative chemotherapy and radiation with gemcitabine + docetaxel + external beam radiation to primary tumor
- Now has innumerable metastases to lungs, liver, as well as concerning lesions in right ischium and spleen
- Dismal prognosis

Imaging discussion

- Progressed through appropriate imaging studies
- Classic presentation of cardiac angiosarcoma:
 - Right heart failure and/or tamponade
 - CT shows low-attenuation right atrial mass arising from right atrial free wall
- Initial read contained cardiac angiosarcoma in differential

Test	Cost	Radiation dose (mSv)
TTE	\$2,467	0
2-view CXR	\$512	0.02
Chest CT	\$4,374	8
PET CT	\$5307	32

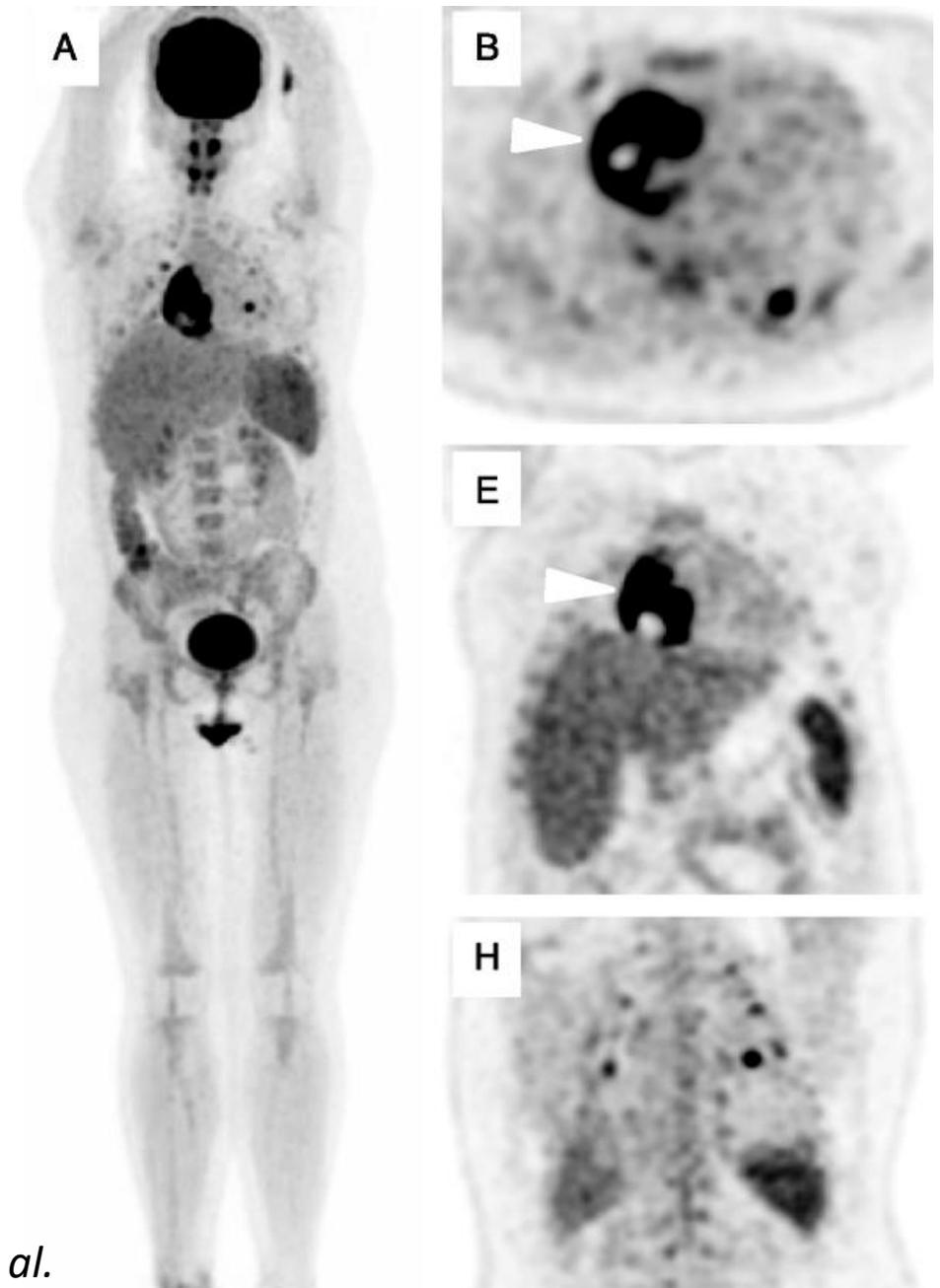
Source: www.fairhealthconsumer.org

Cardiac Angiosarcoma

- Cardiac tumors are extremely rare (<0.1%). Of these, few (~15%) are malignant, but those that are usually (>80%) fatal
- Diagnostic delays are common due to rarity
- Classically presents with right heart failure or cardiac tamponade without heart disease or risk factors for heart failure
- Two morphologic subtypes on imaging:
 1. Low-attenuation mass arising from right atrial free wall
 2. Diffusely infiltrative mass extending along pericardium
- Hematologic metastasis to lungs typically occurs prior to diagnosis

Cardiac Angiosarcoma

- Arises from right atrium
- PET classically shows intense uptake of primary tumor (white arrowheads in figures B and E) and numerous foci of increased metabolic activity (seen in figure H) representing pulmonary metastases



Hod *et al.*

UNC Top Three

- Cardiac tumors are extremely rare (<0.1%). Of these, few (~15%) are malignant, but those that are usually (>80%) fatal. Cardiac angiosarcoma is one such malignant tumor.
- One should suspect cardiac tumor in otherwise healthy patient with single-chamber heart failure.
- Transthoracic echocardiogram is the first-line imaging test.

References

- Muzio DB, Radswiki *et al.* Cardiac angiosarcoma. *Radiopedia*. Accessed 8/7/19. URL: <https://radiopaedia.org/articles/cardiac-angiosarcoma?lang=us>
- Gaasch WH, Vander Salm TJ. Cardiac Tumors. *UpToDate*. Updated May 2019. Accessed 8/7/19.
- Rahbar K *et al.* Differentiation of Malignant and Benign Cardiac Tumors Using ¹⁸F-FDG PET/CT. *J Nuc Med*. 2012 Jun;53(6):856-63.
- Lee CI, Elmore JG. Radiation-related risks of imaging. *UpToDate*. Updated Nov 2017. Accessed 8/7/19.
- Huang B, Law MW, Khong PL. Whole-body PET/CT scanning: estimation of radiation dose and cancer risk. *Radiology*. 2009 Apr;251(1):166-74.
- Janigan DT, Husain A, Robinson N. Cardiac Angiosarcomas: A Review and a Case Report. *Cancer*. 1986 Feb 15;57(4):852-9.
- Chrysohoou C, Lalude O, Stillman A, Lerakis S. A Rare Case of Angiosarcoma of the Left Ventricle Detected by Cardiac Magnetic Resonance Imaging. *Hellenic J Cardiol*. 2015 Sep-Oct;56(5):444-5.
- Hod N, Shalev A, Levin D, Anconina R, Ezroh Kazap D, Lantsberg S. FDG PET/CT of Cardiac Angiosarcoma With Pulmonary Metastases. *Clin Nucl Med*. 2018 Oct;43(10):744-746.