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
• “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."
“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$_{\text{max}}$ 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$_{\text{max}}$ 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

<table>
<thead>
<tr>
<th>Test</th>
<th>Cost</th>
<th>Radiation dose (mSv)</th>
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</thead>
<tbody>
<tr>
<td>TTE</td>
<td>$2,467</td>
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<tr>
<td>2-view CXR</td>
<td>$512</td>
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<td>Chest CT</td>
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<td>8</td>
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<tr>
<td>PET CT</td>
<td>$5307</td>
<td>32</td>
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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


