Cardiac Tumors: Clinical and Imaging Evaluation of Benign and Malignant Lesion

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Disclosures

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- **Stockholder:** Prova, Inc.
- **Off-label use:** Magnetic resonance imaging for cardiac stress testing and Gadolinium contrast for use in cardiac studies.
Case 1

- 46 yo with mass observed on cardiac CT.
This finding is: a) right atrial thrombus, b) right atrial angiosarcoma, c) right atrial myxoma, d) lipomatous hypertrophy of the interatrial septum.
Pericardial Cyst

• Benign congenital fluid filled structures
• Incidental finding
• Low signal intensity on T1
• High signal intensity on T2
• No contrast uptake on 1st Pass Perfusion or LGE

T1 Dark Blood  T2 Dark Blood  T2 Dark Blood Fat Sat
Case 2: Simultaneous Echo and MRI in 4-chamber views. 
You suspect: 
“Pseudo-tumors” 

*Relatively benign conditions: Pericardial cyst, lipomatous hypertrophy, other*

- Most often found in the right atrium
- Remnants of in-utero structures, congenital anomaly, acquired pathology
  - Lipomatous hypertrophy of the interatrial septum
    - ? Conversion to lipoma
  - Crista terminalis
  - Eustacian valve remnant
  - Chiari network - filamentous

*Images from BMJ & AccessAnesthesiology-McGraw/Hill*
75% of primary cardiac tumors are not malignant

*Atrial myxomas represent 75% of these*

Of the 25% of primary cardiac tumors that are malignant, sarcomas represent 75%.

Of note, most common malignant tumors are metastases (e.g., lymphoma, melanoma, breast, thyroid, etc.)
Case 3: 76 yo woman with asthma and hypothyroidism presents with 2 episodes of right arm numbness and slurring of speech of 2 hours duration in past 2 weeks. House officer swears to hear two second heart sounds. Your diagnosis?

1. Right atrial thrombus
2. Left atrial thrombus
3. Right atrial myxoma
4. Left atrial myxoma
5. Right atrial angiosarcoma
6. Left atrial angiosarcoma
### Signs and Symptoms of Cardiac Myxoma

<table>
<thead>
<tr>
<th>Variable</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Symptoms</strong></td>
<td></td>
</tr>
<tr>
<td>Dyspnea</td>
<td>~70</td>
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<tr>
<td>Paroxysmal dyspnea</td>
<td>~25</td>
</tr>
<tr>
<td>Syncope</td>
<td>~20</td>
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<tr>
<td>Palpitations</td>
<td>~20</td>
</tr>
<tr>
<td>Chest pain</td>
<td>~10</td>
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<tr>
<td>Embolic event</td>
<td>~30</td>
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<tr>
<td>Fever</td>
<td>~20</td>
</tr>
<tr>
<td>Weight loss</td>
<td>~15</td>
</tr>
<tr>
<td><strong>Signs</strong></td>
<td></td>
</tr>
<tr>
<td>Mitral systolic murmur</td>
<td>~50</td>
</tr>
<tr>
<td>Mitral diastolic murmur</td>
<td>~40</td>
</tr>
<tr>
<td>Loud S1</td>
<td>~40</td>
</tr>
<tr>
<td>Tumor plop</td>
<td>~15</td>
</tr>
<tr>
<td><strong>Laboratory Data</strong></td>
<td></td>
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<tr>
<td>Elevated ESR</td>
<td>~30</td>
</tr>
<tr>
<td>Anemia</td>
<td>~30</td>
</tr>
<tr>
<td>LA enlargement on CXR</td>
<td>~10</td>
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</tbody>
</table>
CARDIAC MYXOMAS: LOCATION

- 75% Left atrium
- 15-20% Right atrium
- 3-4% in RV and 3-4% in LV- usually from the ventricular free wall not interventricular septum
- Usually originate from the interatrial septum at the border of the fossa ovalis
- Multiple tumors and abnormal locations more common with familial forms
MYXOMAS:

polypoid, smooth, round with mucin globules and hemorrhagic
CARDIAC MYXOMA

• Embolism occurs in 30-40% of patients in LA position
  – Coronary artery embolism- rare but should be considered in young patients without CAD history
• Clinically evident pulmonary emboli generally uncommon with RA myxomas
• Risk of embolism increases if myxoma becomes infected
• Treatment is surgery (1-3% complication rates) with 85% 20 year survival and.
• Recurrence is uncommon, usually within first 5 yrs (about 40 case reports in literature)
  – Incomplete resection, intraoperative displacement, embolization, and multifocal genesis
A 43 year old female with a PMH of HTN and mild rheumatoid arthritis

Sudden onset of chest pain, and pre-syncope at an airport.

Paramedics were alerted and she was found to have inferior ST-segment elevations on an ECG. Cardiac catheterization laboratory and was found to have no significant coronary artery lesions. She was diagnosed with coronary vasospasm and treated with diltiazem, carvedilol, aspirin, clopidogrel, nitroglycerin, simvastatin, and lisinopril.
• She did not have recurrent chest pain until seven months after the initial discharge when she had another episode of chest pain with pre-syncope. She developed significant anxiety about her condition so presented for a second opinion.
You suspect:
A) Papillary fibroelastoma,  B) Aortic valve vegetation,  C) Aortic leaflet thrombus,
D) Marantic aortic valve endocarditis,  E) Partially flail right coronary cusp of the aortic valve
PAPILLARY FIBROELASTOMA

- Most common tumor of the cardiac valves
  - Usually aortic or mitral
- Most patients have concomitant valvular disease
- Potential to embolize in up to 25% of patients
  - Rarely aortic valve tumors can partially obstruct a coronary arterial orifice leading to MI
- Complete resection recommended
  - Anticoagulation recommended if surgery not feasible
MALIGNANT CARDIAC TUMORS

- ~25% of cardiac tumors

- ~75-95% of primary malignant cardiac tumors are sarcomas
  - As a result these are second to myxomas in overall frequency of primary cardiac tumors

- Metastatic disease is the most common malignant tumor involving the heart with lymphomas predominating
ANGIOSARCOMA

- Most common right heart malignancy
- High predilection for the atrioventricular groove
- Invades the pericardium causing hemorrhagic effusion
- Very high rate of rapid proliferation and metastatic spread
- Nodular, heterogeneous, hyperintense tumor that can completely incase the heart
Case

• 39 yo woman with past medical history of treatment for breast CA presents with 3 episodes of dizziness, fatigue, and left sided weakness. Recently, she was introduced to several anxiolytic medications. During treatment for breast cancer, she received Herceptin and during a 12 month surveillance echocardiogram, the lab notified you of a concern for a left ventricular mass.

• A cardiac MRI study was ordered.
You suspect:

1) Multiple cardiac thrombi
2) A primary cardiac malignancy
3) Breast cancer that metastasized to the heart and pericardium
4) Evolution of a second malignancy related to prior Herceptin use
History

- This 67 yo man with hypertension, diabetes, and 40 pk yr history of smoking received 340 mg/m2 of doxorubicin for treatment of non-Hodgkin lymphoma. After anthracycline therapy, he received mediastinal radiation. On several occasions he developed some chest pain and dyspnea, but felt much of this was related to his cancer treatment. Two months after completing his radiation treatment he continued with dyspnea on exertion at 2 blocks, and 2 pillow orthopnea. BP: 138/84, HR 94 bpm. He underwent a cardiac imaging study and you are asked to interpret the findings.
After image review, you feel the **most likely** diagnosis is:

1) Anthracycline induced cardiomyopathy
2) Tumor metastasis of his lymphoma
3) Evidence of an anterior myocardial infarction complicated by LV apical thrombus
4) An anthracycline induced second primary malignancy
5) Evidence of a check-point inhibitor related autoimmune myocarditis.
## Imaging Features of Cardiac Tumors

<table>
<thead>
<tr>
<th>Cardiac Tumor</th>
<th>Echocardiography</th>
<th>CT</th>
<th>MRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myxoma</td>
<td>Mobile tumor</td>
<td>Narrow base of attachment</td>
<td>Heterogeneous</td>
</tr>
<tr>
<td></td>
<td>Narrow stalk connected to fossa ovalis</td>
<td>Heterogeneous, low attenuation</td>
<td>Primarily isointense on T1, with areas</td>
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<tr>
<td></td>
<td>Heterogeneous with hyperechoic and</td>
<td>Occasionally with calcification</td>
<td>of hypointensity and hyperintensity</td>
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<tr>
<td></td>
<td>hyperechoic foci</td>
<td></td>
<td>Hyperintense on T2</td>
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<tr>
<td>Papillary</td>
<td>Mobile mass</td>
<td>Difficult to see</td>
<td>Heterogeneous</td>
</tr>
<tr>
<td>fibroelastoma</td>
<td>Short pedicle</td>
<td></td>
<td>enhancement</td>
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<tr>
<td></td>
<td>“Shimmering” edges</td>
<td></td>
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<tr>
<td>Lipomas</td>
<td>Intramural hyperechoic mass</td>
<td>Homogeneous</td>
<td>Hyperintense on T1</td>
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<tr>
<td></td>
<td></td>
<td>Low (fat) attenuation</td>
<td>+ Signal with fat suppression</td>
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<td></td>
<td></td>
<td></td>
<td>No enhancement</td>
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<tr>
<td>Rhabdomyomas</td>
<td>Multiple small, lobulated hyperechoic</td>
<td>Homogeneous</td>
<td>Isointense on T1</td>
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<tr>
<td></td>
<td>intramural masses</td>
<td></td>
<td>Hyperintense on T2</td>
</tr>
<tr>
<td>Fibromas</td>
<td>Intramural large, solid mass</td>
<td>Homogeneous, low attenuation</td>
<td>Isointense on T1</td>
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<tr>
<td></td>
<td>Central hyperechoic foci</td>
<td>Calcification</td>
<td>Hyperintense on T2</td>
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<tr>
<td>Toratomas</td>
<td>Very heterogeneous</td>
<td>Very heterogeneous</td>
<td>Minimal enhancement</td>
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<tr>
<td></td>
<td>Pericardial effusion</td>
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<tr>
<td>Hemangiomomas</td>
<td>Hyperechoic</td>
<td>Heterogeneous</td>
<td>Isointense on T1</td>
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<tr>
<td></td>
<td></td>
<td>Calcification</td>
<td>Hyperintense on T2</td>
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<td></td>
<td></td>
<td>Marked enhancement</td>
<td>Marked enhancement</td>
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<tr>
<td>Angiosarcoma</td>
<td>Mass protruding into right atrium</td>
<td>Low attenuation</td>
<td>Infiltrative</td>
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<tr>
<td></td>
<td>Pericardial effusion</td>
<td></td>
<td>Heterogeneous</td>
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<td></td>
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<td></td>
<td>Nodular areas of hyperintensity on T1</td>
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<td></td>
<td></td>
<td>Linear areas of enhancement</td>
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<tr>
<td>Other sarcomas</td>
<td>Left atrial mass</td>
<td>Low attenuation</td>
<td>Infiltrative and heterogeneous</td>
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<tr>
<td></td>
<td>Broad base of attachment to posterior</td>
<td>+ Calcification</td>
<td>Variable intensity on T1</td>
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<tr>
<td></td>
<td>atrial wall</td>
<td></td>
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<tr>
<td>Lymphoma</td>
<td>Hyperechoic masses</td>
<td>Low attenuation</td>
<td>Infiltrative</td>
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<tr>
<td></td>
<td>Pericardial effusion</td>
<td></td>
<td>Isointense to hypointense on T1</td>
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<td></td>
<td></td>
<td></td>
<td>Heterogenous enhancement</td>
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Summary for evaluation of masses (1)

• Some of the more frequently seen non-cancerous masses involving the heart and pericardium include:
  – Pseudo-tumors of the right atrium
  – Pericardial cysts
  – Cavitary thrombi
  – Lipomatous hypertrophy of the interatrial septum
  – Valvular and device related vegetations

• Some of the more frequently seen non-malignant primary cancers of the heart in adults include:
  – Myxomas (most common in the atria)
  – Papillary fibroelastomas (valve leaflet tumors)
  – Lipomas (round fat tumors – SVC compression over time)
Some of the more frequently seen malignant tumors (invade across tissue planes and irregular borders) involving the heart and pericardium include:

- **Metastatic disease**
  - Lymphoma, melanoma, breast, thyroid

- **Sarcomas**
  - Angiosarcoma of right heart
  - Leiomyosarcoma (invasive fat tumor)
  - Osteosarcoma
Questions?
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