Primary Tumors of the Heart

Mitul B. Kadakia, HMS III
Gillian Lieberman, M.D.
Beth Israel Deaconess Medical Center
Objectives

- Background & Patient Presentation
- Radiological Tests used to identify cardiac tumors
  - Indications, Advantages, & Disadvantages
  - Normal Scan and Anatomy
  - Our patient
  - Effect on differential
Background

- Extremely rare (incidence 0.01%-0.02% in autopsy series)
- Often missed or misdiagnosed
- >75% benign
Clinical Presentation

INTRACAVITARY
- Heart Failure (CHF or low-output)
- Embolic event (PE, stroke)

INTRAMURAL
- Heart Failure (systolic or diastolic dysfunction)
- Arrhythmia
Patient C.H.

- 3 year old male with history of dyspnea and cyanosis.
- EKG notable for periods of supraventricular tachyarrhythmia
Differential Diagnosis

- Valvular Heart Disease (Congenital, AR, AS, MR, MS)
- Hypertrophic cardiomyopathy
- Heart Failure
- Arrhythmia
- Ischemic Heart Disease
- Pulmonary Embolism
- Pulmonary HTN
- Pneumonia
- Cardiac Mass
## Differential of Cardiac Mass in Children

### Benign

<table>
<thead>
<tr>
<th>Tumor</th>
<th>General Appearance</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Rhabdomyoma (42%)</strong></td>
<td>Multiple, small, Intramural masses</td>
</tr>
<tr>
<td><strong>Fibroma (18%)</strong></td>
<td>Intramural, large, Solid mass, ant. LV</td>
</tr>
<tr>
<td><strong>Myxoma (17%)</strong></td>
<td>Mobile, pedunculated</td>
</tr>
<tr>
<td></td>
<td>LA &gt; RA</td>
</tr>
<tr>
<td><strong>Teratoma (12%)</strong></td>
<td>Multiple tissue layers</td>
</tr>
<tr>
<td></td>
<td>In pericardium</td>
</tr>
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### Malignant

- Rhabdomyosarcoma (41%)
- Fibrosarcoma (18%)

### Other

- Lipomatous Hypertrophy
- Endocarditis
- Cyst
- Thrombus
- Metastatic CA

Braunwald, *Heart Disease 7th ed.*; Sparrow, *Radiographics 2005*
Menu of Tests

- Chest Roentgenogram
- Computerized Tomography
- Echocardiography
- Magnetic Resonance Imaging
- Nuclear Medicine
- Angiography
Chest Roentgenogram

- Give first “hints” of tumor
- Can look for cardiac chamber enlargement, pericardial effusion, calcifications
- Associated pulmonary changes may be seen
- Limited information on tissue differentiation, morphology, location, mobility, infiltration
Normal Chest X-Ray

Zaroug et al. Mayo Clinic Proceedings 2004

Braunwald, Heart Disease 7th ed.
CT Scout – Patient C.H.

Heart Diameter > 50% of thorax diameter

Courtesy Dr. Jason Handwerker, BIDMC/Children's
Computerized Tomography

- Can be used to assess size, location, myocardial infiltration, intraluminal invasion (with contrast), and effect on surrounding structures.
- Shows calcifications
- Views include mediastinum and surrounding structure
- Faster, Increased availability
- High Resolution
CT - Limitations

- Recent advances have improved CT soft tissue discrimination, but still not as good as MR.
- Limited planes
- +Radiation
Computerized Tomography - Normals

Braunwald, *Heart Disease 7th ed.*
Non-Contrast, Axial CT – Patient C.H.

Abnormal Calcification

Homogeneous, soft tissue attenuation, enlargement of heart

Courtesy Dr. Jason Handwerker, BIDMC/Children's
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## Which Mass is it?

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<td>Teratoma</td>
<td>Multiple tissue layers In pericardium</td>
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### Malignant

- Rhabdomyosarcoma
- Fibrosarcoma

### Other

- Lipomatous Hypertrophy
- Endocarditis
- Cyst
- Thrombus
- Metastatic CA
Echocardiography

- Screening test of choice, portable, quick
- Can assess mobility, size, location, and attachments.
- Real-time images
- Through color doppler, can assess impact on myocardial blood flow.
- Transthoracic vs. Transesophageal
Echocardiography - Limitations

- Limited by operator experience, body habitus
- Limited views
- Limited soft tissue characterization
- Invasive, with associated risks (TEE)
TTE - Normal

Braunwald, Heart Disease 7th ed.
TTE, 4 chamber view – Companion
Patient #1

Hyperechogenic mass filling almost entire LV
Small central calcifications, with acoustic shadowing

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### Malignant

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- Lipomatous Hypertrophy
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Magnetic Resonance Imaging

- Superior soft tissue characterization, multiple planes, no ionizing radiation
- Specialized sequences allow assessment of detailed morphological information, suppression of tissue (i.e. fat), demonstration of fluid, vascularity
- Views include mediastinum and surrounding structure
- Helpful in surgical planning
- 3-D imaging allows for evaluation of cardiac function, blood flow, and mobility of the mass
MRI - Limitations

- Calcifications not seen
- Time consuming, Expensive
- Claustrophobia
- Requires EKG gating
Cardiac MRI

- Gated to “R” wave
- “Black-Blood” and “Bright-Blood” images
- “Cine” images

http://dcmrc.mc.duke.edu/resources_physicians/indications/tumor_mass/tumor.html
Large, homogenous, isointense mass arising from apex of left ventricle. Well-defined, sharp borders with no pericardial infiltration.

Endocardium
Gradient Echo (“Bright Blood”) – Patient C.H.

Large, homogenous, isointense mass arising from apex of left ventricle. Well-defined, sharp borders with no pericardial infiltration.

Endocardium
Well-defined, homogenous, isointense mass arising from anterior left ventricular wall

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Braunwald, Heart Disease 7th ed.; Sparrow, Radiographics 2005
Cardiac Fibroma

- Benign connective tissue tumor, arises from fibroblasts
- Children <10 years of age
- 3-10 cm. Most commonly in anterior free wall of LV. Can be in posterior LV or RV.
- Increased incidence in Gorlin Syndrome (nevoid basal cell CA, medulloblastomas, fibrous histocytomas, skin and skeletal abnormalities)
- Treatment – Surgical excision

Braunwald, *Heart Disease 7th ed*
# Summary of Radiological Tests

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<thead>
<tr>
<th>Test</th>
<th>Value</th>
<th>Limitations</th>
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<td>CXR</td>
<td>Heart enlargement</td>
<td>Location, Mobility, Infiltration, Morphology, Tissue Diff.</td>
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<td>Lung findings</td>
<td></td>
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<tr>
<td>CT</td>
<td>Calcifications, size, location, infiltration, faster, ↑resolution</td>
<td>Soft tissue differentiation, planes Radiation</td>
</tr>
<tr>
<td>Echo</td>
<td>Real-time, good screen, mobility, attachments, size, motion, location</td>
<td>Invasive (TEE), operator-based, Soft tissue differentiation, views</td>
</tr>
<tr>
<td>MRI</td>
<td>Soft tissue differentiation, morphology, location, size, infiltration, surgical planning, diff. planes, tissue suppression</td>
<td>Time-consuming, calcifications not seen, EKG gating</td>
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Summary

- Cardiac tumors are rare, but important not to miss
- Be aware of other, more common cardiac masses that may mimic tumors
- Radiological tools can allow characterization of cardiac masses and tumors with high degree of certainty
- Each test is complementary and tells a part of the story
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- Dr. Gillian Lieberman
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- Dr. Kamlesh Dave
- Fellow students
References

- Shapiro, LM. Cardiac tumours: diagnosis and management. Heart 2001; 85:218-222.