CARDIAC MASSES

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An Intensive Review of Echocardiography

TUMORS OF THE HEART

• Primary
  – Rare (<5% of all cardiac tumors)
  – Most common: myxoma (more often in left atrium)

• Metastatic
  – More common (>95%)

Cardiac Tumors

• Primary tumors in adults arise most frequently from the endocardium, followed by myocardium and least often from pericardium
• Metastatic lesions arise commonly from the pericardium, followed by myocardium and endocardial lesions occur only through venous extension

Common Types of Primary Tumors of the Heart

• Benign (75%)
  – Myxoma
  – Rhabdomyoma
  – Fibroma
  – Lipoma and lipomatous hypertrophy of the atrial septum
  – AV node tumor
  – Papillary fibroelastoma
  – Hemangioma

• Malignant (25%)
  – Angiosarcoma
  – Rhabdomyosarcoma
  – Fibrosarcoma

Clinical Features of Cardiac Tumors

• Constitutional:
  – Due to embolic phenomenon
  – Due to secretion of Interleukin-6
  – Fever, weight loss

• Embolic:
  – Due to tumor fragmentation
  – Due to thromboembolism from tumor surface
  – Recurrent pulmonary emboli in right sided tumors

• Obstructive:
  – Dyspnea, PND
  – Hemoptysis
  – Sudden death

Differentiation of Left Atrial Myxoma from Mitral Valve Disease

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Myxoma</th>
<th>Mitral Valve Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>History</td>
<td>Short duration, Constitutional symptoms, Syncope</td>
<td>Chronic, no constitutional symptoms, syncope rare</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Episodic</td>
<td>Progressive</td>
</tr>
<tr>
<td>Physical Exam</td>
<td>Tumor plop, variable murmurs with position, other valve disease unusual</td>
<td>Constant murmur, associated valve disease common</td>
</tr>
<tr>
<td>EKG</td>
<td>Sinus rhythm</td>
<td>Atrial fibrillation</td>
</tr>
<tr>
<td>Chest X ray</td>
<td>Tumor Calcification</td>
<td>Valve calcification</td>
</tr>
<tr>
<td></td>
<td>Small left atrium</td>
<td>Dilated left atrium</td>
</tr>
<tr>
<td>Echocardiogram</td>
<td>Characteristic findings</td>
<td>Characteristic findings</td>
</tr>
</tbody>
</table>
Location of Cardiac Myxomas

- Left atrium 75%
- Right atrium 15%
- Right ventricle 5%
- Left ventricle 5%

Familial Cardiac Tumors

- Approximately 10% of myxomas
- Autosomal dominant
- Carney Syndrome:
  - Myxomas in non-cardiac locations (breast or skin) - 67%
  - Pigmentation of skin (lentigines or pigmented nevi) - 67%
  - Endocrine tumors (pituitary adenomas, adrenocortical disease or testicular tumors) - 33%
  - Patients usually younger, less female preponderance, multiple masses, recurrence after surgery, ventricular site

Familial Cardiac Tumors (contd)

- NAME syndrome:
  - Nevi, Atrial myxoma, Myxoid neurofibroma, Epithelides
- LAMB syndrome:
  - Lentigines, Atrial Myxoma, Blue nevi
- Cardiac Rhabdomyomas:
  - Associated with tuberous sclerosis syndrome (hamartomas in multiple organs, epilepsy, mental deficiency, adenoma sebaceum)

Lipomatous Hypertrophy of the Atrial Septum

- Accumulation of non-encapsulated adipose tissue (fetal & adult type) in atrial septum
- Atrial hypertrophy may protrude into RA
- More common in elderly obese women
- Associated with supraventricular arrhythmias
- Spares the fossa ovalis region

Mesotheliomas of the AV Node

- Cystic tumor
- Rare cause of sudden death or complete heart block, VF or cardiac tamponade
- Diagnosed at autopsy

Papillary Fibroelastoma

- Benign tumors that arise from cardiac valves
- Has frond-like projections and 'shimmering' mobility of its fronds on real-time examination
- Can cause valvular incompetence, thromboembolic complications and coronary obstruction when it occurs on aortic valve
Metastatic Cardiac Tumors

- Most common cardiac tumors (20:1)
- Origin:
  - Carcinoma of breast
  - Carcinoma of lung
  - Malignant melanoma
  - Leukemia
  - Lymphoma
- Pericardial metastases cause pericarditis, pericardial effusion or pericardial tamponade

Neoplastic Pericarditis

- 10% patient dying of metastatic cancer have pericardial metastasis
- 5% have myocardial metastases
- Tumors that cause pericarditis:
  - Lung carcinoma 45%
  - Breast carcinoma 20%
  - Hodgkin disease, leukemia, lymphoma 15%
  - Other carcinomas 10%
  - Melanoma 5%
  - Sarcoma 5%
  - Others 5%
- Nephroblastoma (Wilms tumor) causes pericarditis in children

Neoplastic Pericarditis (cont.)

- Primary pericardial tumors are rare (mesothelioma, pheochromocytoma and sarcoma- fibrosarcoma, angiosarcoma and liposarcoma)
- Hemorrhagic pericardial effusions occur in chronic myeloid leukemia & myelomonocytic leukemic blast crisis due to intrapericardial hemopoeisis
- In breast cancer small, asymptomatic pericardial effusions may be seen due to lymphatic obstruction (50%)

Modes of Spread to Heart

- Via lymphatics (carcinoma of branchus & breast)
- Direct extension
- Via pulmonary veins (carcinoma of bronchus)
- Via venous system (carcinoma of testis & kidney)

Manifestations of Myocardial Metastases

- Asymptomatic
- Cardiac arrhythmias
- Heart block
- Myocardial dysfunction
- EKG changes (ST-T changes)
- Imaging techniques:
  - Echocardiography
  - CT
  - MRI

Incidence of Primary Cardiac Tumors by Mean Age at Presentation

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>%</th>
<th>Age</th>
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<tbody>
<tr>
<td>Rhabdomyoma</td>
<td>2</td>
<td>33 weeks</td>
</tr>
<tr>
<td>Fibroma</td>
<td>2</td>
<td>13 years</td>
</tr>
<tr>
<td>Rhabdomyoma</td>
<td>&lt;1</td>
<td>15 years</td>
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<tr>
<td>Hemangioma</td>
<td>1</td>
<td>31 years</td>
</tr>
<tr>
<td>Paraganglioma</td>
<td>&lt;1</td>
<td>39 years</td>
</tr>
<tr>
<td>Sarcoma (angio)</td>
<td>4</td>
<td>40 years</td>
</tr>
<tr>
<td>Sarcoma (myofibroblastic)</td>
<td>9</td>
<td>41 years</td>
</tr>
<tr>
<td>Myxoma</td>
<td>76</td>
<td>50 years</td>
</tr>
<tr>
<td>Papillary fibroelastoma</td>
<td>5</td>
<td>59 years</td>
</tr>
<tr>
<td>Lipomatous hypertrophy</td>
<td>&lt;1</td>
<td>64 years</td>
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</table>
Heart Tumors that present as Cardiac Masses: Classification

- **Pediatric Heart Tumors**
  - Hamartomas
  - Fibroma
  - Purkinje cell hamartoma/histiocytoid cardiomyopathy
  - Germ cell tumors
- **Benign tumors seen primarily in adults**
  - Non-neoplastic masses
    - Mural thrombi
    - Lipomatous hypertrophy, atrial septum
    - Papillary fibroelastoma
  - Benign neoplasms
    - Myxoma
    - Paraganglioma/phaeochromocytoma

- **Germ cell tumors**
- **Benign tumors seen primarily in adults**
  - Malignant neoplasms
    - Sarcomas
      - Angiosarcoma (mostly right atrium and pericardium)
      - Sarcomas with myofibroblastic differentiation (mostly left atrium)
        - Undifferentiated malignant fibrous histiocytoma
        - Leiomyosarcoma
        - Fibrosarcoma
        - Osteosarcoma
        - Rhabdomyosarcoma
    - Lymphomas
    - Metastasis (usually right sided)
      - Carcinomas
      - Renal cell/hepatocellular, mostly intracavitary
      - Sarcomas
      - Melanomas

Cardiac Tumors by Site

- **Left atrium (cavitary, pedunculated or broad-based attachment)**
  - Myxoma
  - Sarcoma
  - Metastatic (extension of lung primary)
  - Hemangioma
  - Paraganglioma
- **Left atrium (involving wall/pericardium)**
  - Sarcoma
  - Lymphoma
  - Metastasis (renal cell and hepatocellular carcinoma)
  - Hemangioma
  - Paraganglioma

- **Right atrium (cavitary mass)**
  - Myxoma
  - Idiopathic thrombus
  - Lipomatous hypertrophy
  - Metastasis (renal cell and hepatocellular carcinoma)
  - Hemangioma

- **Valve**
  - Papillary fibroelastoma
  - Myxoma
  - Hamartoma
- **Pericardium**
  - Metastasis
  - Mesothelioma
  - Lymphoma
  - Sarcoma

- **Ventricle**
  - Sarcoma
  - Lipoma
  - Hemangioma
  - Myxoma
  - Idiopathic thrombus
  - Metastatic (RV)
  - Lymphoma
  - Rhabdomyosarcoma