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#### **Cardiac tumors**

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#### 1. Learning objectives

To illustrate the CT and MR imaging findings of various types of cardiac tumors diagnosed at our Institution. To outline the advantages and disadvantages of CT and MR for cardiac tumor diagnosis. To discuss the pertinent literature on distinguishing clinical and imaging features of each type of cardiac tumor presented.

#### 2. Background

Cardiac masses are rare. The estimated cumulative prevalence of primary cardiac tumors at autopsy is only 0.002%–0.3%. The prevalence of metastasis to the heart is 1,2%, approximately 40 times higher than that of primary cardiac tumors, being found at autopsy in 10-12 % of patients with known malignancies.

About 75% of primary cardiac tumors are benign, the most common in adults being Myxoma. Malignant cardiac tumors constitute approximately 25% of all primary cardiac tumors, the most common in adults being Angiosarcomas (33%).

Cardiac tumors are frequently asymptomatic. Their clinical presentation may also be similar to that of much more common disorders such as heart failure, stroke and coronary artery disease, including symptoms like dispnea, peripheral edema and thoracic pain, thus frequently delaying the definitive diagnosis.

Innocuous, low cost and widely available transthoracic and transesophageal echocardiography provide high-resolution, real-time images. Transthoracic echocardiography, being non-invasive, is frequently the first imaging modality used when a cardiac tumor is suspected. Both are limited, though, by operator experience, by a restricted field of view and low soft tissue contrast.

CT and MR allow evaluation of the hole mediastinum and possible extracardiac extent of the tumor. These methods are also more flexible in the selection of image planes and allow accurate comparison between examinations. They are very important for prognosis determination and surgery planning. CT offers the possibility of hole thoracic evaluation, including the lungs, and detection of calcifications. MR has better soft tissue contrast resolution and allows better tissue characterization. Disadvantages of these two imaging modalities include the need for electrocardiographic gating, which, in the presence of arrhythmia, may lead to acquisition artifacts.

In this educational exhibit, we review the clinical, morphological and CT and MR imaging findings of several different types of benign and malignant cardiac tumors and tumor-like lesions, including atrial myxomas, lipoma, lipomatous hipertrophy of the interatrial septum, fibroma, rhabdomyoma, bronchogenic cyst, pericardial hamartoma, metastasis, undifferentiated sarcoma and lymphoma, based on selected images of cases diagnosed at our Institution between 1992 and 2008.

#### 3. Imaging findings OR Procedure details

#### **BENIGN ATRIAL TUMORS**

#### MYXOMA

Myxoma is the most common primary cardiac tumor, comprising approximately 50% of all primary

cardiac masses. It is slightly more common in male patients, with a mean age onset of 50 years. 75% of myxomas occur in the left atrium, typically at the level of the fossa ovalis, in the interatrial septum; 18% on the right atrium and only 7% on the ventricles. Although they can be asymptomatic, there is a classic triad of obstructive symptoms related to the chamber where the tumor is based; constitutional symptoms, such as fever, malaise, weight loss and anemia; and embolic events, involving mainly the brain, kidneys and lower limbs with left-sided myxomas, and the pulmonary circulation with right-sided myxomas. These tumors usually present as an endocardial-based pedunculated polypoid mass, with a smooth or villous surface, moving freely inside the atrium, sometimes protuding to the ventricle during diastole. An important differential diagnosis is the presence of an intracavitary thrombus. However, thrombus do not have a pedicle or enhance after IV contrast (**Figure 3**).

The **CT** imaging characteristics consist of a spherical or ovoid mass with a lobulated contour, hipo or isodense compared with the myocardium, usually heterogeneous, sometimes with coarse or punctuate calcifications, and heterogeneous enhancement. There are some possible associated findings such as chamber dilation, signs of cardiac failure or of embolic events (**Figure 1**).





93 yo male patient with left atrial (LA) myxoma. CT Pulmonary Angiogram performed in the Emergency Department for suspected, not confirmed, pulmonary thromboembolism. a) A well defined, soft-tissue density, slightly enhancing mass (arrow), located in the LA wasfound. b) Sagital reformatted view, better depicting the lesion's pedicle.

The **MR** imaging findings include the presence of a spherical or ovoid mass with lobulated contours, with heterogeneous signal intensity, predominately isointense with respect to the myocardium on T1-weighted images and increased signal intensity on T2-weighted images, with heterogeneous enhancement after gadolinium. There is a better correlation between the point of attachment of the Myxoma described on MR than with CT imaging, when compared with the surgical description (**Figure 2**, [Video 1] video 1).



Same patient as in Video 1. Cardiac MR. a) SSFP image shows the myxoma (\*) has moved to the left ventricle during diastole b) IRGE image shows no areas of delayed enhancement.



23 yo male patient with chronic renal insufficiency and acute rejection to a kidney transplant. Post-contrast abdominal CT scan shows 2 homogeneous, non-enhancing, hypodense atrial images sugestive of thrombus. These findings were confirmed with a transthoracic echocardiogram.

#### LIPOMA

Lipomas are rare, comprising about 14% of all primary benign cardiac masses. They are similar to extracardiac lipomas and are located on the subendocardial region in about 50% of cases. The left atrium and ventricle are the most frequent locations. These tumors can grow to a large size without any clinical maifestations or cause clinical symptoms related to obstruction, when subendocardial, compression on the ventricles or displacement of the lungs, when subepicardial. They are also associated with atrial fibrillation, ventricular tachycardia or atrioventricular block.

The **CT** findings include te presence of an homogeneous, well defined, hipodense mass that does not enhance after contrast.

The **MR** findings include the presence of a well defined mass with increased and homogeneous signal intensity on T1 and T2-weighted images that decreases in fat-saturated sequences, with no soft-tissue component and no enhancement after gadolinium (**Figure 4**).



30 yo male patient. Mass sugestive of lipoma detected on transesophageal echocardiogram. Cardiac MR showed a well-defined, mobile lesion (\*) contacting the right atrium, the ascending aorta and the superior vena cava a) The lesion was hyperintense on T1-weighted images. It was hypointense on T1-Fat supressed and T2-weighted images.

Much more common is the **lipomatous infiltration of the interatrial septum** associated with obesity and advanced age, not representing a true neoplasm, it is said to be present when there is a depositon of fat exceeding 2 cm of thickness on this location (**Figures 5** and **6**).



61 yo female patient with morbid obesity and lipomatous hypertrophy of the interatrial septum (IAS). CT scan performed for staging of folicular lymphoma. a) Post-contrast CT shows non-enhancing, well defined, homogeneous fat density mass confined to the IAS (between arrows). b) Axillary and upper mediastinal lymphadenopathy (\*) are also shown.





Same patient as in Figure 4. Cardiac MR a) T1-weighted image showing a well defined, homogeneous mass confined to the IAS (between arrows). b) T1 - fat supressed - weighted image shows that the lesion now appears hypointense (between arrows). c) Mediastinal lymphadenopathy (\*) are also shown, on a sagital plane.

#### PAPPILLARY FIBROELASTOMA

Papillary fibroelastomas are small (usually less than 1 cm), avascular tumors composed of muscular cells, fibrous and smooth elastic tissue, attached to the endocardium by a short pedicle. They constitute about 10% of all primary cardiac tumors and are located on the valves' surface in about 90% of cases, the aortic and mitral valve being the most common locations (54%). There is no gender preference, the mean age at presentation is 60 years and they are generally asymptomatic, although symptoms may occur when thrombi gathered around the tumors embolize.

Echocardiography is usually diagnostic where they appear as small mobile vibrating masses attached to a valve. **CT** and **MR** are usually not helpful given their small size and mobility.

#### **BENIGN VENTRICULAR TUMORS**

#### RHABDOMYOMA

Rhabdomioma represents 20% of all primary cardiac tumors and is the most common benign congenital tumor, representing 90% of all primary cardiac tumors occurring in chidren. It is associated with tuberous sclerosis in 50 to 86% of cases and also to cardiac disrhythmia and non-imune hydrops. It is as anomalous benign proliferation of tissue displaying typical "spider cells" on histopathology and may appear as single or multiple solid, circumscribed, nonencapsulated tumors, most often located in the left ventricular myocardium, sometimes projecting into a cardiac chamber (**Figure 7**). They typically regress with age (**Figure 8**).





21 yo female patient with disrythmia. Cardiac MR shows a well defined, rounded mass located in the interventricular septum (between arrows), compatible with a rhabdomyoma. The mass is isointense with the miocardium on T1-weighted images (a), enhancing less than the surrounding myocardium and heterogeneously after gadolinium (b) and hypointense on T2-weighted images (c).









23 yo female patient with tuberous sclerosis and an involuted left ventricle rhabdomyoma with adipose substitution (arrow). Cardiac MR. a) T1-weighted image showing a hyperintense linear lesion on the lateral wall of the left ventricle. b) T1-weighted image after gadolinium shows that the lesion does not enhance significantly. c) On T2-weighted images, the lesion appears hypointense. d) and e) IRGE images, long and short axis, respectively, showing delayed enhancement of the lesion.

#### **FIBROMA**

Fibroma is a rare tumor that primarily affects children, being the 2<sup>nd</sup> most common benign primary cardiac tumors in children after rhabdomioma. It may occur as part of Gorlin or basal cell nevus syndrome. Although one third are incidentally found, these tumors are sometimes associated with arrhythmias, heart failure and sudden death. Fibromas are composed of fibroblasts and large amounts of collagen and are tipically located in the ventricles, most often the ventricular septum and left ventricular free wall.

At **CT**, they appear as a soft-tissue density, homogeneous mass, frequently with dystrophc calcifications, either well circumscribed or infiltrative, with litlle or no enhancement.

At **MR** these tumors are usually homogeneously hypointense on T2-weighted images and isointense when compared to the myocardium n T1-weighted images, showing little or no enhancement after gadolinium (**Figures 9** and **10**).







41 yo male patient with a history of acute myocardial infarction. Cardiac MR showing a fibroma in the ventricular septum a) T1-weighted image shows an oval mass in the interventricular septum that is isodense to the myocarium (arrow) b) T1-weighted image after gadolinium shows minimal enhancement (arrow) c) IRGE shows delayed enhancement of the lesion (arrow) d) Subendocardial delayed enhancement at the inferior wall of the left ventricle related to previous infarct (arrow).







72 yo male with a history of acute myocardial infarction and congestive heart failure. Transthoracic echocardiogram detected an encapsulated ecogenic mass protuding to the interatrial septum. Cardiac MR shows an infero-septal, left ventricle based fibroma (between arrows). a), b) and c) On T1-weighted images (axial, coronal and sagital plane, respectively) the lesion was slightly hypointense. d) Post-gadolinium T1-weighted image shows that the lesion enhances less than the myocardium. e) the lesion was hypointense on T2-weighted images.

#### **BENIGN PERICARDIAL TUMORS**

#### PERICARDIAL CYSTS

Pericardial cysts are usually located in the right cardiophrenic space and may be congenital or iatrogenic. They are usually asymptomatic but may rarely cause symptoms like dyspnea, arrhythmia or retrosternal pain.

At **CT**, they present as a homogeneous, well-defined, fluid attenuation lesion with a smooth wall, although sometimes the attenuation may be greater.

At **MR**, they have a characteristic low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (**Figure 11**).

These lesions do not enhance after IV contrast.





Incidentally found pericardial cyst in a 78 yo woman. a) Axial T2 Haste weighted image and b) Coronal T2 Haste weighted image showing an homogeneous, well-defined, hyperintense image in the left cardiophrenic space (an unusual location).



6 yo child with pericardia hamartoma. a) Thoracic CT shows nodular hipodense image with corse peripheral calcifications. b) coronal reformat image showing the longitudinal extension of the lesion.

#### **MALIGNANT TUMORS**

#### **METASTASIS**

Metastases are 20-40 times more prevalent than primary cardiac tumors, the most common primary sources being bronchogenic carcinoma, lymphoma, leucemia, breast carcinoma, esophagus carcinoma melanoma, sarcoma, hepatoma, adrenal adenocarcinoma and renal cell carcinoma. They may involve the heart by contiguous extension, lymphatic or hematogenous spread. The commonest site of involvement is the pericardium with or without invasion of the underlying myocardium (**Figure 13**). In approximately one-third of patients with **cardiac** involvement, death will be directly attributable to the metastases as a result of pericardial tamponade, congestive **cardiac** failure, or coronary artery invasion Presenting symptoms may include shortness of breath, chest wall pain, and peripheral edema. As with other **cardiac** malignancies, arrhythmias may also be a feature. There are no specific appearances of metastases or direct extension to the heart.







74 yo male patient with a history of progressive dispnea and peripheral edema for the past 3 months. CT scan showed right central hilar mass with associated right lower lobe colapse and bilateral pleural effusion. The mass invaded the mediastinum. Thoracic MR requested to better depict mediastinal invasion. a), b) and c) are T2 Haste weighted images, a and b in an axial planes (B at a higher level than A) and c) in a sagital plane. We can see an intermediate signal mass involving and invading the right atrium and the emergence of the aorta and pulmonary arteries (\*). Upper mediastinal lymphadenopathies (°) and bilateral pleural effusion (+) are also shown.

#### ANGIOSARCOMA

Angiosarcoma is the most common primary cardiac malignancy in adults, representing 37% of all primary cardiac malignancies, is two times more common in men than in women. It is composed of ill-defined anastomotic vascular spaces lined by atypical endothelial cells. It's located in the right atrium in 80% of cases, growing either as a well-defined mass protruding into the right atrium or as an infiltrating mass extending into the pericardium, the tricuspid valve or the superior or inferior vena cava. They are frequently advanced and metastatic at the time of presentation, the lung being the most common site for metastatic disease. Angiosarcomas cause symptoms by obstructing right cardiac filling and causing pericardial tamponade.

At **CT**, they present as a low attenuation right atrial mass, either ill-defined or nodular, with heterogeneous enhancement after contrast. Pericardial thickening and effusion may be seen.

At **MR**, angiosarcoma presents as a large heterogeneous mass in the right atrium, sometimes involving the pericardium causing pericardial thickening or nodularity and hemorrhagic pericardial effusion. It is heterogeneous on T1-weighted images, with areas of intermediate, low and high signal intensity due to the presence of tumor tissue, necrosis and methemoglobin, respectively. They are heterogeneous, predominantly hyperintense on T2-weighted images. The enhancement after gadolinium is marked at the surface – sunray appearance – and heterogeneous.

#### **UNDIFFERENTIATED SARCOMA**

Undifferentiated sarcoma is a malignant tumor with no specific histologic features, representing 24-38% of all primary cardiac malignancies, with a mean age of presentation of 45 years. About 80% occur in the left atrium and some involve the cardiac valves. The most common clinical manifestations are dyspnea, chest pain and weight loss.

At **CT** they may appear as irregular, hipodense, intracavitary myocardial masses (**Figures 14** and **15**).





54 yo male patient with a history of dispnea, fatigue and malaise. a) CT scan performed before intravenous contrast (IVC) shows hypodense, heterogeneous, rouded mass occupying the right atrium (\*). b) CT scan after IVC shows that the mass (\*) enhances heterogeneously and infiltrates the myocardium at the level of the atrioventricular septum. Pathological analysis characterized the mass as an undifferentiated sarcoma.





63 yo male with disrythmia. a) Thoracic CT scan performed before IV contrast shows a hypodense mass on the left atrium (arrow). b) CT scan after IV contrast shows that the mass is lobulated and enhances heterogeneously (arrow). Pathological analysis revealed an undifferentiated sarcoma.

At **MR**, they may show as polypoid, isointense, infiltrative masses on T1-weighted images (**Figure 16**). The tumor may also manifest as a hemorrhagic mass replacing the pericardium, similar to angiosarcoma.



55 yo male patient with biopsy proven undifferentiated sarcoma of the left ventricle. a) T1-weighted image showing an heterogeneous mass with some hyperintense areas infiltrating the wall of the left ventricle.

Other, rarer primary sarcomas of the heart are **malignant fibrous histiocytoma** (11% - 24%), **leiomyosarcoma** (8% - 9%), **rhabdomyosarcoma** (4% - 7%) and **osteosarcoma** (3% - 9%).

#### LYMPHOMA

Primary cardiac lymphomas are exceedingly rare, are typically of the non-Hodgkin B-cell type, and are confined to the heart or pericardium. They usually occur in immunocompromised patients but are not restricted solely to this group. Presentation is with rapidly worsening heart failure, obstructive symptoms, or arrhythmias. They most commonly involve the right side of the heart, in particular the right atrium, with frequent involvement of more than one chamber and invasion of the pericardium with pericardial effusion. Microscopically, they consist of firm homogeneous nodules, not prone to hemorrhage or necrosis.

The **CT** imaging findings are nonspecific (**Figures 17** and **18**).





63 yo immunocompetent male patient with a history of fatigue and peripheral edema with a month duration. Transthoracic echocardiogram showed pericardial effusion and a lobulated mass located at the tricuspid valve, insinuating to the interatrial septum. a) Thoracic CT before the administration of IVC shows pericardial (\*) and bilateral pleural effusion (+). b) Cardiac CT after IVC shows a hypodense, hypoattenuating, labulated mass located at the level of the tricuspid valve, extending to the interatrial septum. The patient died a few weeks later. Pathological analysis revealed a primary B-Cell lymphoma of the heart.







83 yo woman. CT performed for staging of lymphoma (supraclavicular lymphadenopaties previously biopsed). a) Post-IV contrast CT axial image showing supraclavicular lymphadenopaties (\*). b) Lobulated mass involving the right atrial and ventricular myocardium. c) Coronal reformat image showing the longitudinal extension of the mass.

At **MR** imaging, they are isointense on T1-weighted images and heterogeneously hyperintense on T2weighted images; they demonstrate heterogeneous enhancement after administration of gadolinium contrast material, with areas of low enhancement in the center of the lesion compared to the periphery.

#### **MALIGNANT PERICARDIAL TUMORS**

Most malignant pericardial tumors are **metastasis**. Primary malignancies are pericardial **mesotheliomas**, that represent less than 1% of all mesotheliomas but 50% of all primary pericardial tumors.

#### 4. Conclusion

CT and MR imaging findings help differentiate benign from malignant cardiac tumors, sometimes even further narrowing the differential diagnosis; therefore, influencing their management.

#### **5. Personal Information**

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#### 7. Mediafiles

## Figure 1



93 yo male patient with left atrial (LA) myxoma. CT Pulmonary Angiogram performed in the Emergency Department for suspected, not confirmed, pulmonary thromboembolism. a) A well defined, soft-tissue density, slightly enhancing mass (arrow), located in the LA wasfound. b) Sagital reformatted view, better depicting the lesion's pedicle.



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Same patient as in Figure 4. Cardiac MR a) T1-weighted image showing a well defined, homogeneous mass confined to the IAS (between arrows). b) T1 - fat supressed - weighted image shows that the lesion now appears hypointense (between arrows). c) Mediastinal lymphadenopathy (\*) are also shown, on a sagital plane.







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23 yo female patient with tuberous sclerosis and an involuted left ventricle rhabdomyoma with adipose substitution (arrow). Cardiac MR. a) T1-weighted image showing a hyperintense linear lesion on the lateral wall of the left ventricle. b) T1-weighted image after gadolinium shows that the lesion does not enhance significantly. c) On T2-weighted images, the lesion appears hypointense. d) and e) IRGE images, long and short axis, respectively, showing delayed enhancement of the lesion.



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54 yo male patient with a history of dispnea, fatigue and malaise. a) CT scan performed before intravenous contrast (IVC) shows hypodense, heterogeneous, rouded mass occupying the right atrium (\*). b) CT scan after IVC shows that the mass (\*) enhances heterogeneously and infiltrates the myocardium at the level of the atrioventricular septum. Pathological analysis characterized the mass as an undifferentiated sarcoma.





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55 yo male patient with biopsy proven undifferentiated sarcoma of the left ventricle. a) T1-weighted image showing an heterogeneous mass with some hyperintense areas infiltrating the wall of the left ventricle.





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Video 1



68 yo male patient with fatigue and malaise. Transthoracic echocardiogram detected a mobile, pedunculated mass in the left atrium sugestive of a myxoma. Cardiac MR SSFP video, long axis plane, depicting the myxoma's movement to the left ventricle during diastole.