CT and MR Imaging of Primary Cardiac Malignancies[1]

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LEARNING OBJECTIVES

After reading this article and taking the test, the reader will be able to:

- Discuss the relative merits of echocardiography, CT, and MR imaging in the evaluation of primary cardiac malignancies.
- Identify features of atrial masses that are suspicious for malignancy.
- Identify clinical and radiologic features that may help differentiate various primary cardiac tumors.

Primary cardiac malignancies are rare tumors that are difficult to diagnose clinically. Different primary cardiac malignancies may have different clinical, morphologic, and radiologic features and intracardiac locations. Angiosarcoma is the most common primary cardiac malignancy. It tends to occur in the right atrium and involve the pericardium. Because of its tendency to hemorrhage, angiosarcoma often demonstrates areas of increased signal intensity with T1-weighted sequences. Undifferentiated sarcomas typically occur in the left atrium and have variable epidemiologic and radiologic features. Rhabdomyosarcoma is the most common primary cardiac malignancy in children and is more likely than other primary cardiac sarcomas to involve the valves. Primary cardiac osteogenic sarcoma almost always occurs in the left atrium and frequently demonstrates calcification. Certain features (eg, broad base of attachment, origin at a site other than the atrial septum) help differentiate this tumor from left atrial myxoma. Leiomyosarcoma favors the left atrium and tends to invade the pulmonary veins and mitral valve. Fibrosarcoma also tends to occur in the left atrium and is often necrotic. Liposarcoma is very rare and usually manifests as a large, infiltrating mass. Foci of macroscopic fat are occasionally seen. Primary cardiac lymphoma occurs more commonly in immunocompromised patients, frequently involves the pericardium, and, unlike other primary cardiac malignancies, may respond to chemotherapy. The advent of cross-sectional imaging has allowed earlier detection of primary cardiac malignancies as well as more accurate diagnosis and characterization.

Abbreviation: AFIP = Armed Forces Institute of Pathology

Index terms: Angiosarcoma, 51.324 • Heart, CT, 51.1211 • Heart, MR, 51.1214 • Heart, neoplasms, 51.321, 51.3221, 51.3231, 51.324, 51.329, 51.34 • Leiomyosarcoma, 51.325 • Liposarcoma, 51.371 • Lymphoma, 51.34 • Osteosarcoma, 51.324 • Rhabdomyosarcoma, 51.321

RadioGraphics 1999; 19:1421–1434

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■ INTRODUCTION

Primary cardiac malignancies are rare. Metastases to the heart are 20–40 times more common than primary tumors, only 25% of which are malignant (1). In one autopsy series, the prevalence of primary cardiac malignancies was only 0.001%–0.28% (2). Primary cardiac malignancies present a clinical dilemma: Not only are they unusual, but they are also often asymptomatic until they become large, and even then they produce nonspecific symptoms. Before the advent of cross-sectional imaging, primary cardiac malignancies were rarely diagnosed before death. Today, they are being discovered in living patients, sometimes incidentally, and their proper characterization has become important in guiding management.

To date, echocardiography has been the modality of choice for diagnosis of intracardiac disease. Transthoracic echocardiography is inexpensive, noninvasive, and clearly depicts heart morphology in a variety of imaging planes. It also provides a large amount of functional information. Echocardiography allows real-time imaging and can show tumor mobility and distensibility, features that are typically seen in atrial myxomas and less often in sarcomas (3). Doppler ultrasound can provide velocity measurements that can be used to estimate chamber pressures. The effectiveness of transthoracic echocardiography is mainly limited by the available imaging window, which can vary with patient body habitus and operator experience.

Transesophageal echocardiography is invasive but circumvents the problem of acoustic window often found with transthoracic echocardiography. Some extracardiac structures such as the descending aorta and pulmonary veins can be imaged. Because it is not necessary for reflected sound to penetrate the chest wall with transesophageal echocardiography, higher-frequency transducers may be used and better resolution is achieved. However, soft-tissue characterization remains limited compared with that achieved with computed tomography (CT) and magnetic resonance (MR) imaging, and myocardial disease such as tumor infiltration is not clearly depicted. Most important, both transesophageal and transthoracic echocardiography provide only limited views of the mediastinum and cannot be used to evaluate extracardiac manifestations of disease.

CT can be used to accurately image the heart and surrounding mediastinum. CT provides better soft-tissue contrast than echocardiography, can depict calcification and fat, and may allow tissue diagnosis of some masses such as lipomas (4). Although spatial resolution of CT has improved with the development of faster imaging techniques, particularly electron beam CT, this modality is still far inferior to echocardiography in the depiction of small moving structures such as the cardiac valves. Unlike echocardiography, CT does not allow true real-time imaging, and imaging planes are limited to those allowed by angulation of the gantry.

Like CT, MR imaging can depict the extracardiac extent of disease. Although MR imaging cannot show calcification, it allows better overall soft-tissue characterization than CT and is the modality of choice for evaluating abnormalities intrinsic to the myocardium. In addition, MR imaging is more versatile than CT in that it allows imaging in multiple planes and can provide some functional information such as flow direction and flow velocity in large vessels. The main disadvantage of MR imaging is its susceptibility to motion artifact. High-quality cardiac MR imaging is highly dependent on regular electrocardiographic rhythms and cardiac gating.

Although CT and MR imaging can provide important information about cardiac malignancies, these tumors are rare and the cardiac applications of CT and MR imaging are relatively new. We reviewed pathology records at our institution and selected cases of primary cardiac malignancy for which CT or MR imaging was available. In this article, we present CT and MR imaging findings in eight types of primary cardiac malignancies: angiosarcoma, undifferentiated sarcoma, rhabdomyosarcoma, osteogenic sarcoma, leiomyosarcoma, fibrosarcoma, liposarcoma, and lymphoma. We also discuss the pertinent literature with emphasis on distinguishing clinical features, tumor location, gross morphology, and imaging characteristics (Table).

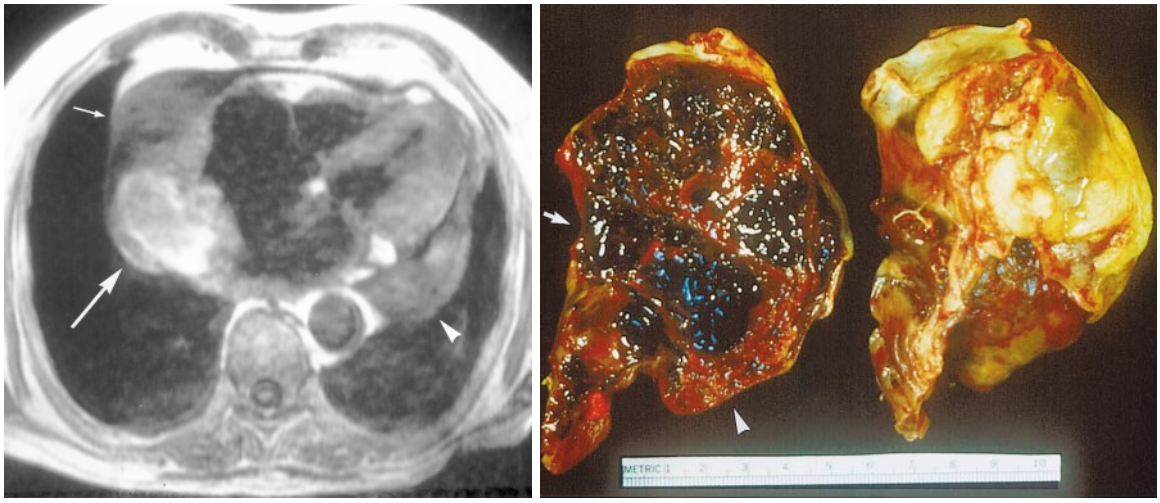
■ ANGIOSARCOMA

Angiosarcoma is a tumor of endothelial cells. It is so named because the endothelial cells line ill-defined anastomotic vascular spaces, although there may also be large avascular areas of spindle cells (1). Angiosarcoma is the most common cardiac sarcoma in surgical studies (37% of cases) (1) and occurs slightly more often in males (1, 5,6). Glancy et al (5) and Jannigan et al (6) described the tumor as typically occurring in middle-aged men, but a wide patient age range has been reported (1,7). Because the tumor tends to occur in the right atrium and involve the pericardium, patients usually present with right-sided heart failure or tamponade (1,5,6), often with superimposed systemic signs such

Characteristic Features of Primary Cardiac Malignancies

Type of Malignancy	Percentage of Primary Cardiac Sarcomas	Epidemiologic Features	Most Common Clinical Manifestation	Most Common Chamber of Origin	Hemorrhage	Calcification	Pericardial Involvement	Valvular Involvement	Other Distinguishing CT or MR Imaging Characteristics
Angiosarcoma	37	Variable, but classically affects middle-aged men	Right-sided heart failure or pericardial tamponade	Right atrium	Typical	No	Frequent, often diffusely infiltrated, may arise primarily in pericardium	Occasional	Areas of increased T1 signal intensity may be focal ("cauliflower" appearance) or linear along pericardium ("sunray" appearance)
Undifferentiated sarcoma	≤24*	Variable	Pulmonary congestion	Left atrium	Occasional	No	May arise primarily in pericardium	Occasional	Variable morphology (may be infiltrative or masslike)
Rhabdomyosarcoma	4–7	Most common primary cardiac malignancy in infants and children, slight male predilection	Variable	Equal prevalence in all chambers	Occasional	No	Occasional, but a portion of the tumor should involve myocardium	More likely to involve valves than are other sarcomas	Variable morphology (may be cystlike or demonstrate large areas of central necrosis
Osteosarcoma	3–9	Variable	Pulmonary congestion	Left atrium	Unusual	Typical (degree of calcification varies)	Occasional	Occasional	None
Leiomyosarcoma	8–9	Variable	Pulmonary congestion	Left atrium	Unusual	Rare (n = 1)	Likely to involve mitral valve	Likely to extend into pulmonary veins	None
Fibrosarcoma	5	Variable	Pulmonary congestion	Left atrium	Occasional	No	May arise primarily in pericardium	Occasional	Often lobulated, may demonstrate large areas of necrosis
Liposarcoma	<1	Variable	Pulmonary congestion, arrhythmias	Left or right atrium	Common	No	May arise primarily in pericardium	Occasional	Foci of macroscopic fat sometimes seen
Lymphoma	Not applicable	More common in immunocompromised patients	Pulmonary congestion	Right atrium	Rare	No	Frequent, may manifest as pericardial effusion	Unusual	Less likely to demonstrate necrosis and extend into cardiac chamber than sarcomas

*More common in older literature.



a. **b.**

Figure 1. Angiosarcoma in a 79-year-old man. **(a)** T1-weighted MR image shows a heterogeneous mass (large arrow) involving the right atrial free wall and interatrial septum with extension into the left atrium. There is a loculated pericardial effusion adjacent to the right atrial free wall (small arrow) and pericardial thickening posteriorly (arrowhead). **(b)** Photograph of a surgical specimen shows intraluminal, polypoid, hemorrhagic tumor nodules (arrow). Atrial wall trabeculae are seen in the lower right portion of the specimen (arrowhead). Scale is in centimeters.

as fever and weight loss (8). Pericardiocentesis yields bloody fluid that often does not contain malignant cells, even when the tumor has invaded the pericardium (9–14).

Prognosis is poor, which may be due in part to the delay in diagnosis (15). In two studies, patients had metastatic involvement at presentation in 66% and 89% of cases (5,6). There have been recent reports of survival from 12 to 30 months with some combination of chemotherapy, surgery, radiation therapy, and transplantation (10,16,17). Others studies have shown poor results with chemotherapy (18), and the overall experience with heart transplantation is poor (19,20).

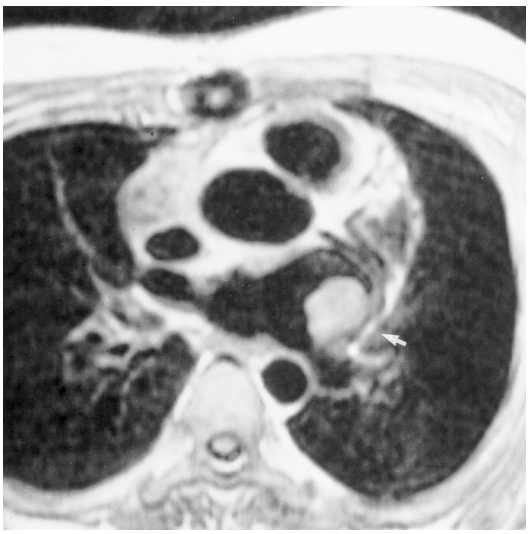
Two main morphologic types have been described in angiosarcoma (6). The first is a well-defined mass protruding into a cardiac chamber, usually the right atrium, often sparing the atrial septum (1). At gross examination, the tumors are hemorrhagic, necrotic, and often adherent to the pericardium (Fig 1) (1). CT often shows a low-attenuation right atrial mass, which may be irregular or nodular (21) and usually arises from the right atrial free wall (13,14,22). Areas of central necrosis in communication with a cardiac chamber have been seen at both CT and MR imaging (23,24). CT has shown tumor infiltration of the myocardium, compression of cardiac chambers (25), direct extension into the pericardium (26), and involvement of the great vessels (9,13). Contrast material enhancement is heterogeneous with both modalities (21,23).

The second morphologic type is a diffusely infiltrative mass extending along the pericardium (6). The pericardial space may be obliterated with hemorrhagic, necrotic tumor debris (27,28), which may appear at CT as pericardial effusion or thickening (8).

Because of their propensity toward hemorrhage and necrosis, angiosarcomas typically have heterogeneous signal intensity on MR images (24). Areas of increased signal intensity on T1-weighted images may be focal (21) or peripheral (10) and are thought to represent blood products (21). Kim et al (29) described local nodular areas of increased signal intensity interspersed within areas of intermediate signal intensity on T1- and T2-weighted images as having a "cauliflower" appearance. In cases with diffuse pericardial infiltration, linear contrast material enhancement along vascular lakes gives what some authors have described as a "sunray" appearance (12).

■ UNDIFFERENTIATED SARCOMA

Undifferentiated sarcomas are malignant neoplasms without specific histologic features. Results of immunohistochemical staining for multiple markers are generally negative (1). In earlier literature, these tumors are often referred to by histologically descriptive names such as pleomorphic sarcoma, round cell sarcoma, or spindle cell sarcoma. Before the era of immunohistochemical staining and electron microscopy,



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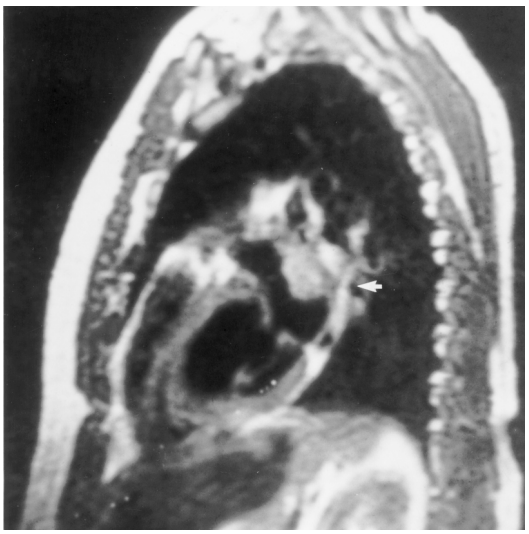

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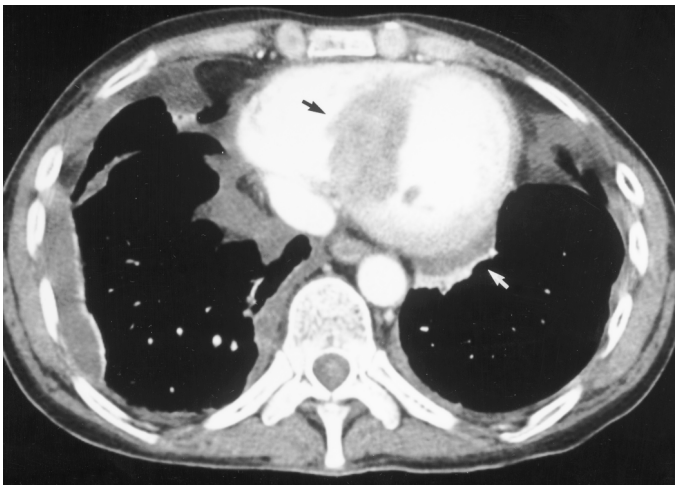

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Figures 2, 3. **(2)** Undifferentiated sarcoma in a 36-year-old man. Axial **(a)** and sagittal **(b)** T1-weighted MR images show a broad-based, lobulated mass arising from the left lateral wall of the left atrium (arrow). The mass is isointense relative to myocardium. **(3)** Undifferentiated sarcoma in a 42-year-old man. Electron beam CT scan shows irregular thickening of the interventricular septum (black arrow) and pericardial effusion (white arrow).

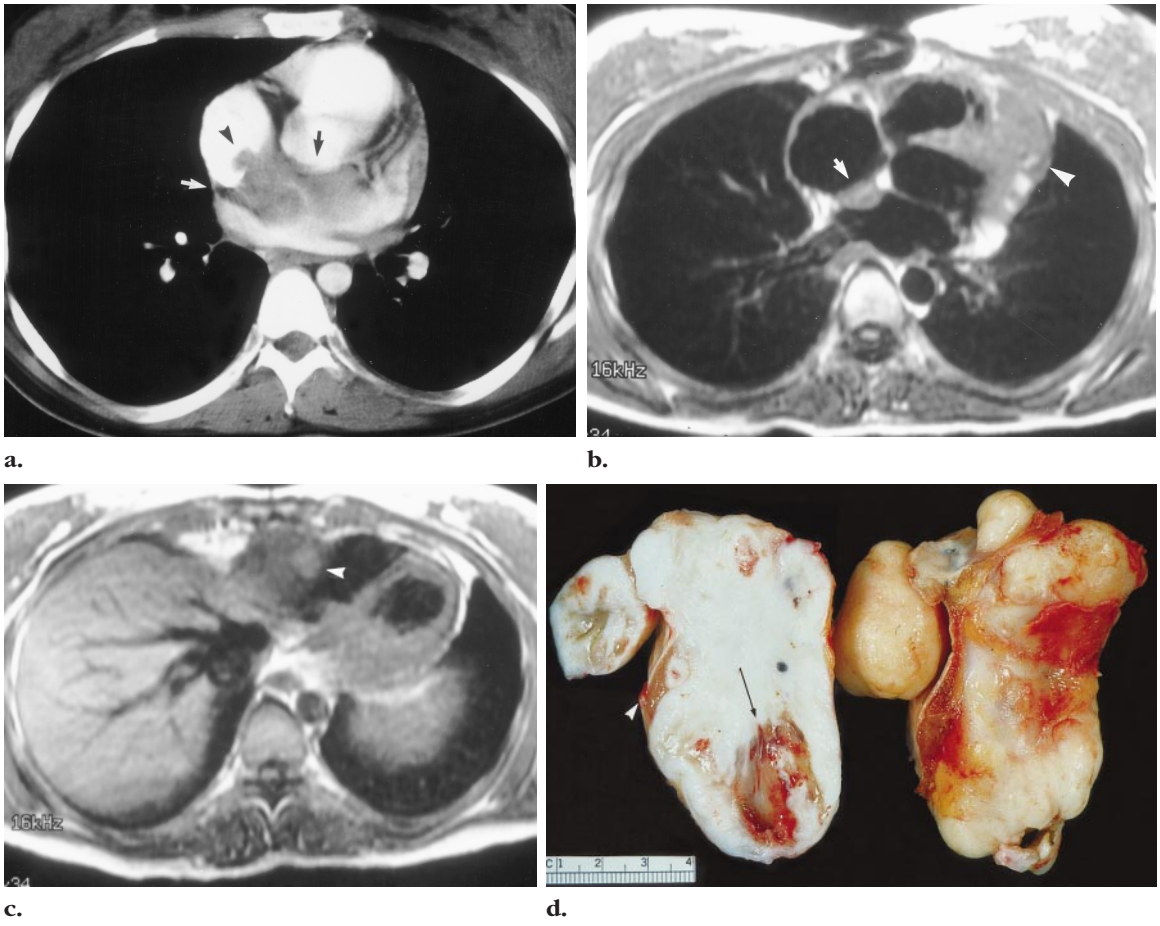
the prevalence of unclassifiable primary cardiac sarcoma was as high as 50%; in recent series, however, its reported prevalence varies from 0% to 24% (1). The highest prevalence was reported in a study from the Armed Forces Institute of Pathology (AFIP) (1); the authors state that this finding may be due to biases inherent in AFIP practice.

The tumors have diverse clinical manifestations, although the most common is pulmonary congestion (1). In the AFIP series (1), the mean age at presentation was 45 years, but an age spectrum ranging from neonates to the elderly has been reported (1,30). In a study by Tazelaar et al (31), clinical outcome was not significantly different between patients with undifferentiated and differentiated primary cardiac malignancies, although outcome was poor for all patients.

In the most recent AFIP series (1), 81% of unclassified sarcomas arose in the left atrium. The tumors may take the form of discrete myocardial masses (32), which have appeared as large, irregular, low-attenuation intracavitary lesions at CT (33,34) or as polypoid masses that are isointense relative to myocardium at MR imaging (Fig 2). Tumor infiltration of myocardium may appear as thickening and irregularity (Fig 3). The tumor may also manifest as a hemorrhagic mass replacing the pericardium, similar to angiosarcoma (30). Several reports have described a tendency to involve the valves (35–37).

■ RHABDOMYOSARCOMA

Rhabdomyosarcoma is a malignant tumor of striated muscle. Two main forms are seen at histologic analysis: embryonal neoplasms, which occur in infants, children, and young adults, and the adult types of rhabdomyosarcoma, which are more pleomorphic and are much less common in the heart (1).



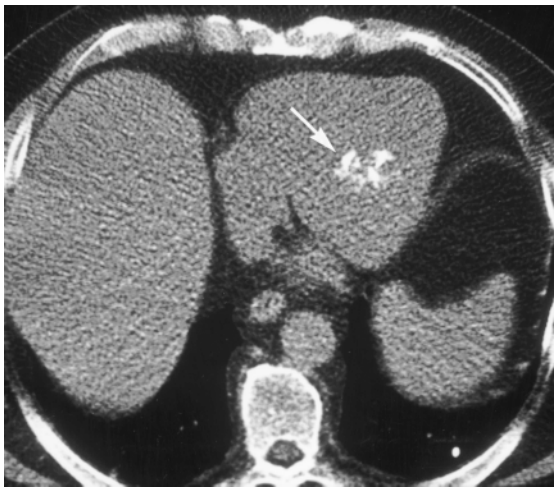
a. b. c. d.

Figure 4. Rhabdomyosarcoma in a 22-year-old woman. **(a)** Contrast material–enhanced CT scan of the chest shows a large, homogeneous soft-tissue mass in the left atrium (black arrow) with extension through the interatrial septum (white arrow) into the right atrium (arrowhead). **(b)** Axial T1-weighted spin-echo MR image obtained 6 months later after tumor debulking shows recurrent tumor along the interatrial septum (arrow) and in the pericardial space (arrowhead). **(c)** Axial T1-weighted spin-echo MR image obtained inferior to **b** shows a new site of intrapericardial tumor implantation along the anterolateral right ventricular free wall (arrowhead). **(d)** Photograph of a specimen obtained at initial tumor debulking surgery shows a fleshy tumor with cystic degeneration (arrow). Remnants of myocardium can be seen at the periphery of the mass (arrowhead). Scale is in centimeters.

Rhabdomyosarcomas account for only 4%–7% of cardiac sarcomas overall (31) but are the most common cardiac malignancy in infants and children (38). They have a slight male predilection, but because there is no predilection for any one chamber, clinical signs and symptoms vary (1).

Rhabdomyosarcomas may arise anywhere in the myocardium (1,39,40) and are more likely than other sarcomas to involve or arise from cardiac valves (39–43). They are often multiple (39,44) and may invade the pericardium (45). Unlike angiosarcoma, however, a portion of the tumor should always involve the myocardium (46), and pericardial involvement is usually in the form of nodular masses rather than sheet-like spread (39).

Gross specimens may be gelatinous and friable (1,40,47), firm and fleshy (48,49), or cyst-like (50). Large areas of central necrosis may be seen at gross examination (51). In one study, these necrotic areas appeared at MR imaging as a large defect in communication with the pericardial space (44). CT may show a smooth (51) or irregular (52) low-attenuation mass in a cardiac chamber (Fig 4a). Signal intensity characteristics at MR imaging are variable. Isointensity relative to myocardium (41) as well as heterogeneous signal intensity and contrast material enhancement have been reported (44). Extracardiac extension is clearly depicted at CT and MR imaging. Extension into the pulmonary arteries has been demonstrated at CT (41), and MR imaging has been used to delineate invasion of the pulmonary artery (53), descending aorta (54), and pulmonary valve (41).



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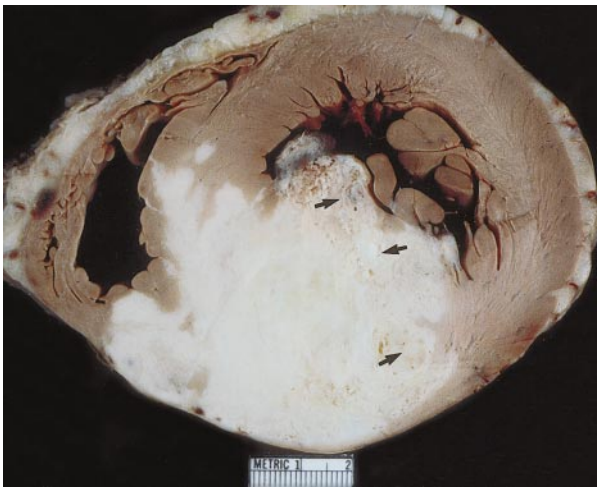

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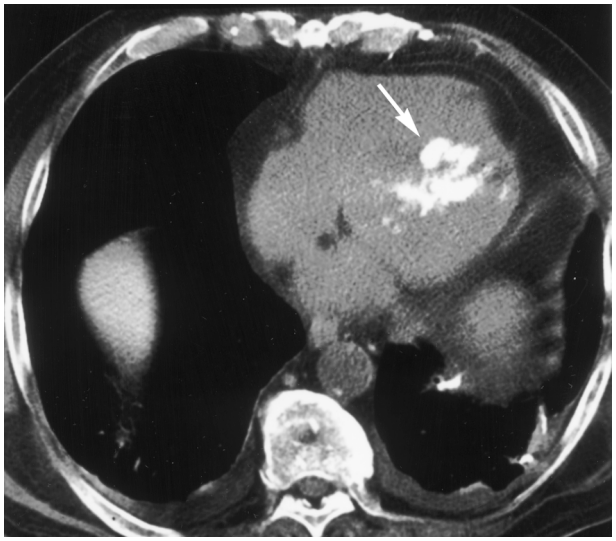

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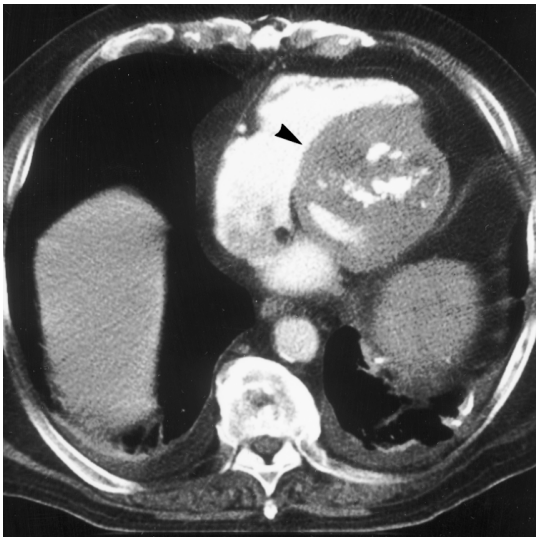


c.

Figure 5. Left ventricular osteosarcoma in a 69-year-old man. **(a)** Unenhanced CT scan of the chest shows dense calcification (arrow), thought to represent chronic calcification of the posterior medial papillary muscle. **(b)** On an unenhanced electron beam CT scan obtained 6 years later, the left ventricular calcifications have increased markedly (arrow). **(c)** Contrast-enhanced CT scan reveals a large, low-attenuation mass occupying the left ventricle (arrowhead). **(d)** Photograph of an autopsy specimen shows pericardial rind and extensive mural involvement from primary osteogenic sarcoma. Foci of calcification are also seen (arrows). Scale is in centimeters.

■ OSTEOSARCOMA

Osteosarcomas in the heart are a heterogeneous group of tumors that contain malignant, bone-producing cells. They may be predominantly osteoblastic or may have chondroblastic or fibroblastic differentiation (1). Cardiac osteosarcomas are uncommon tumors, accounting for only 3%–9% of all cardiac sarcomas (1). Unlike metastatic osteosarcoma, which most often occurs in the right atrium, primary cardiac osteosarcomas arise in the left atrium in the overwhelming majority of cases and are therefore usually accompanied by signs and symptoms of congestive heart failure (55). Primary cardiac osteosarcomas are often aggressive with a very poor prognosis (55,56).

Calcification is often noted in gross surgical specimens. Burke and Virmani (1) described the calcification of cardiac osteosarcoma as "gritty" at pathologic sectioning (1), but some authors have described "stone hard" tumors (57) or the necessity of sectioning tumors with a saw (58). CT may show dense calcifications within a low-attenuation mass (34). However, calcification may also be minimal (55) and in the early stages may be mistaken for benign, dystrophic calcifications (Fig 5).

Mainly because of their left atrial location, primary cardiac osteosarcomas are often confused at radiology with left atrial myxomas,

which are more common and are benign (1). Imaging features that may suggest osteogenic sarcoma include a broad base of attachment (3,58), an aggressive growth pattern such as extension into the pulmonary veins (3,56,58), invasion of the atrial septum (58), or infiltrative growth along the epicardium (57). Localization of the tumor within the left atrium may also suggest the diagnosis. Left atrial myxomas typically arise from the atrial septum near the fossa ovalis (1); Origination of the tumor from another site within the left atrium is atypical and should raise suspicion for a cardiac sarcoma (3). It has been suggested that primary cardiac osteosarcoma should be considered in all atypical left atrial myxomas (55).

■ LEIOMYOSARCOMA

Leiomyosarcoma is a malignant tumor with smooth muscle differentiation. It may arise from smooth muscle bundles lining the subendocardium, but many so-called cardiac leiomyosarcomas likely arise from the smooth muscle of the pulmonary veins and arteries and then spread into the heart (1). Leiomyosarcoma is an uncommon type of cardiac malignancy, constituting only 8%–9% of all cardiac sarcomas (1). Like the other cardiac sarcomas of smooth muscle and fibrous tissue differentiation, leiomyosarcoma has a predilection for the left atrium with cardiac output failure being the most common clinical manifestation (1). Affected patients typically present in the 4th decade of life (1), which is slightly younger than the average age at presentation for primary cardiac sarcoma patients in general. Survival up to 5 years has been reported with a combination of surgery and chemotherapy (59), but overall prognosis is very poor (1).

Leiomyosarcomas usually appear as gelatinous, sessile masses at gross examination and may be multiple in 30% of cases (1). At CT, they usually appear as lobulated, irregular, low-attenuation masses in the left atrium (Fig 6) (60, 61); unlike myxomas, however, leiomyosarcomas usually arise from the posterior wall of the left atrium. Leiomyosarcoma tends to invade the pulmonary veins or mitral valve (1). At CT, extension into the veins may appear as low-attenuation filling defects (62,63), and pericardial involvement may appear as pericardial effusion (64). In one reported case, CT showed dystrophic calcifications within the mass (61). Signal intensity characteristics at MR imaging are nonspecific. Intermediate signal intensity on T1-weighted images (62,65) and increased signal intensity on T2-weighted images have been reported (62).

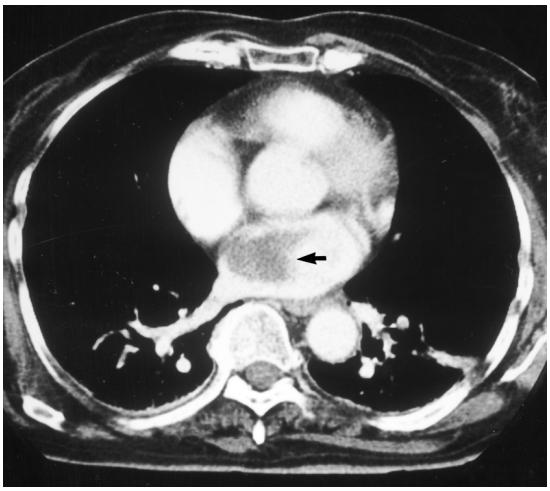


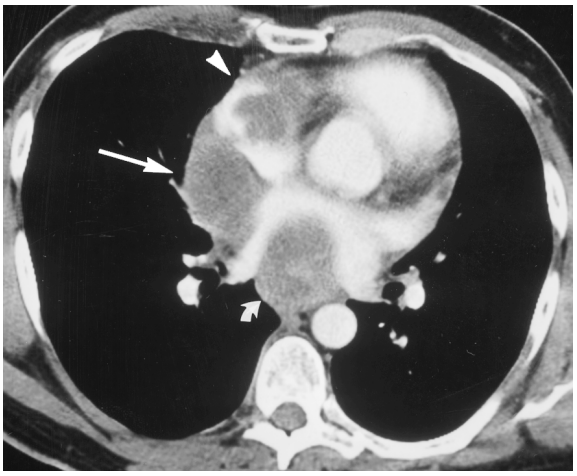
Figure 6. Leiomyosarcoma in a 63-year-old woman. Contrast-enhanced chest CT scan shows a low-attenuation lesion in the left atrium (arrow).

■ FIBROSARCOMA

Fibrosarcoma is a malignant tumor whose cells have morphologic features of fibroblasts. It is a rare cardiac tumor that represents about 5% of cardiac sarcomas in surgical series (1). Like osteosarcoma and leiomyosarcoma, fibrosarcoma tends to occur in the left atrium and usually manifests as congestive heart failure (1). Although one series has shown improved survival and palliation with a combination of surgery and chemotherapy (66), prognosis is generally poor as with all cardiac sarcomas (67). One long-term survivor died of recurrence 12 years after undergoing initial tumor resection (68).

At gross examination, fibrosarcomas have been described as soft, lobulated, gelatinous masses (1,69–71). At CT, low-attenuation tumor may obliterate entire chambers (72,73). Some reports have described necrotic areas in gross pathologic specimens (69), and others have described a similar appearance at CT (74). Fibrosarcoma may infiltrate the pericardium by direct invasion (68) or by the deposition of tumor nodules in the inner pericardium (74). The tumor may also arise primarily in the pericardium and have an appearance similar to that of malignant mesothelioma at gross examination (1). It may be heterogeneous (68) or isointense relative to myocardium on T1-weighted MR images (Fig 7) (75).

Figure 7. Fibrosarcoma in a 43-year-old man. **(a, b)** Contrast-enhanced electron beam CT scans of the chest show a low-attenuation soft-tissue mass filling the right atrium (straight arrow in **a**, large arrow in **b**) and the posterior inferior left atrium (curved arrow in **a**, small arrow in **b**), as well as abnormal soft tissue in the right ventricle (arrowhead in **a**). **(c)** Oblique sagittal CT reconstruction shows a large soft-tissue mass in the left atrium (curved white arrow), inferior left ventricle (solid straight white arrow), and right ventricle (open arrow). Contrast material is seen in the residual left ventricular lumen (straight black arrow), residual left atrial chamber (curved black arrow), and ascending aorta (arrowhead). **(d, e)** On axial **(d)** and coronal **(e)** T1-weighted MR images obtained 1 year later after chemotherapy, the mass (arrow) appears slightly smaller and is isointense relative to myocardium.



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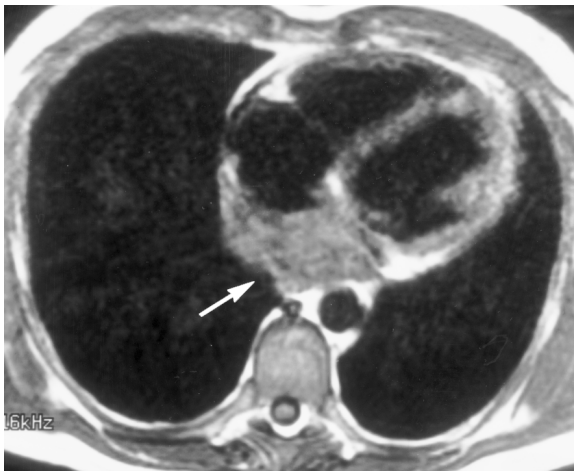

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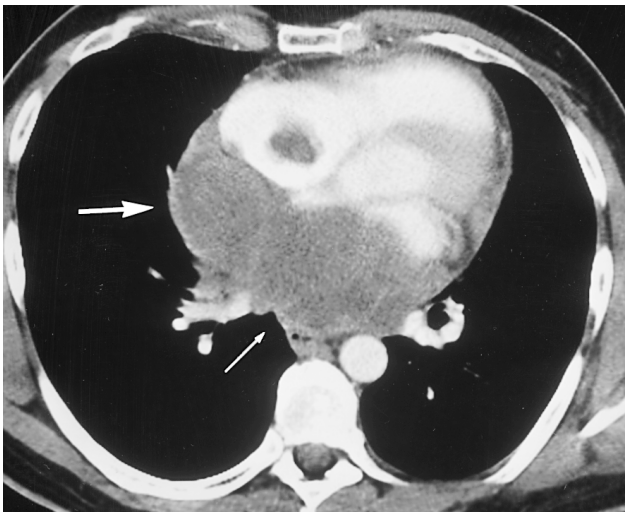

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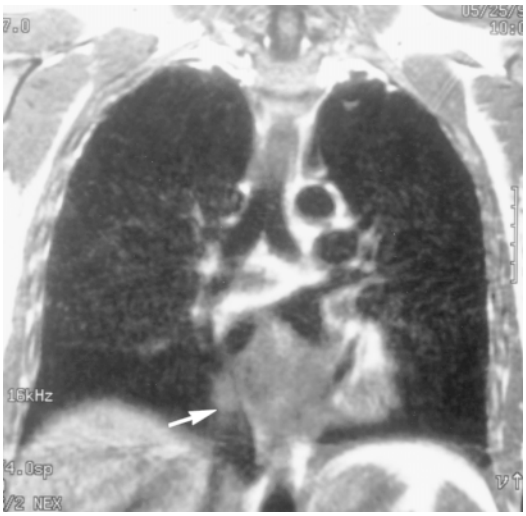

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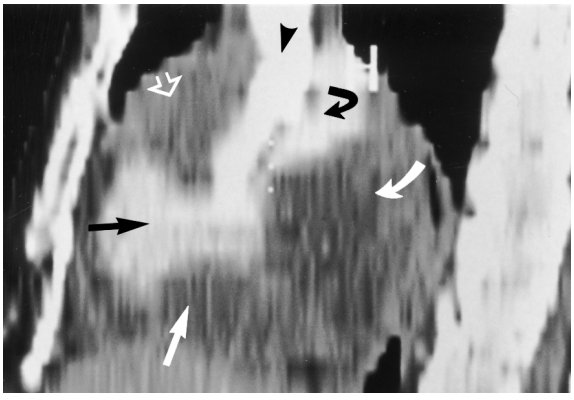

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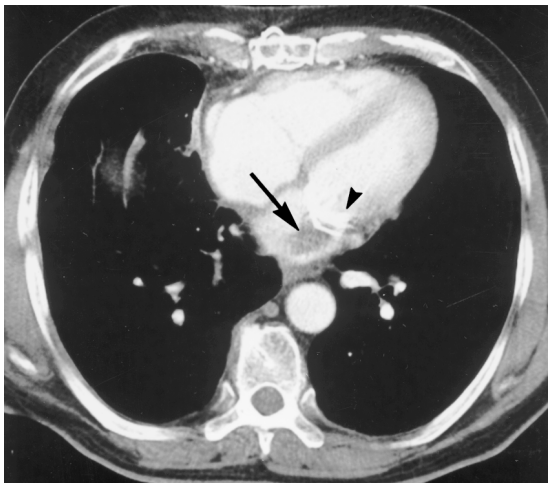


Figure 8. Recurrent liposarcoma in a 41-year-old man. Contrast-enhanced electron beam CT scan shows a broad-based soft-tissue mass arising from the posterior wall of the left atrium (arrow). A mitral valve prosthesis from a previous resection is also seen (arrowhead).

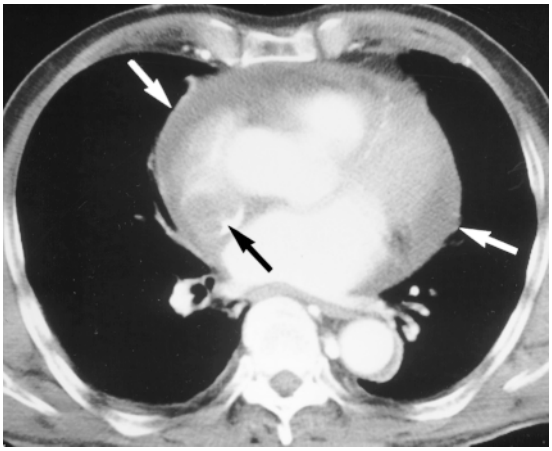


Figure 9. Primary cardiac lymphoma in a 75-year-old man. Contrast-enhanced CT scan of the chest shows a low-attenuation mass in the superior right atrium (black arrow) and a large pericardial effusion (white arrows). Pericardial biopsy revealed follicular mixed type B-cell lymphoma.

■ LIPOSARCOMA

Primary cardiac liposarcoma is a malignant mesenchymal neoplasm that contains lipoblasts. These tumors are extremely rare: In the most recent AFIP series (1), only two of 145 malignant primary cardiac sarcomas were liposarcomas. Only 18 such tumors have been reported in the literature (76).

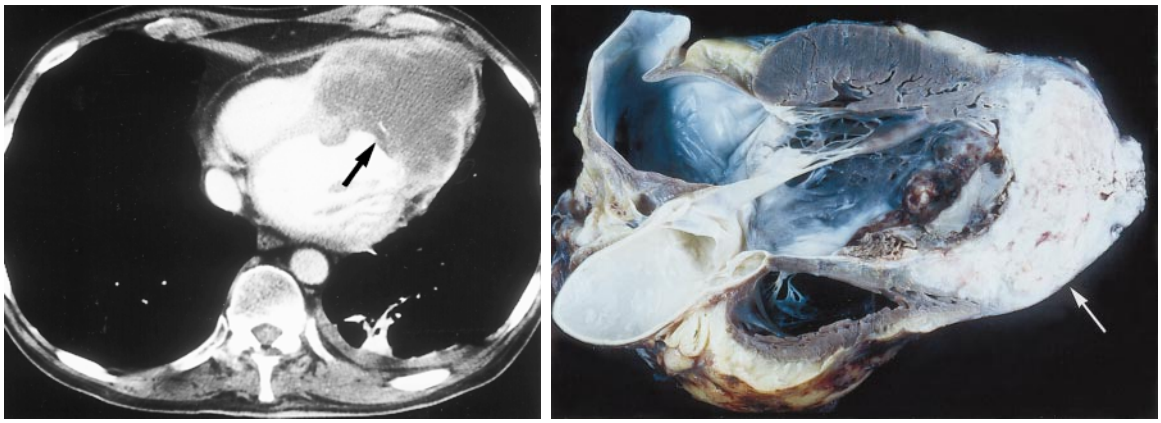
Affected patients most often present with shortness of breath or arrhythmia (1); however, some patients have presented with pericardial tamponade (77), generalized systemic signs such as fever and weight loss (78), and metastases (79). There appears to be no sex predilection (79).

Primary cardiac liposarcomas tend to arise in one of the atria (Fig 8) (79) but have also been discovered in both ventricles and in the pericardium (76,80–82). Pericardial involvement may take the form of effusion, tumor nodules, or irregular masses (83). There have been several reports of involvement of cardiac valves (79, 81). Primary cardiac liposarcomas may involve the pericardium as effusion, tumor nodules (84), or an irregular mass arising primarily in the pericardium (77). At gross examination, the tumors are usually large and multilobulated with areas of necrosis and hemorrhage (81–83,85,86). Primary cardiac liposarcomas have little or no macroscopic fat and do not resemble benign lipomas (76–86).

■ LYMPHOMA

The definition of primary cardiac lymphoma has been debated but currently includes lymphoma that is mostly confined to the heart or pericardium (1). This distinguishes primary cardiac lymphoma from the more common cardiac spread of non-Hodgkin lymphoma (24% in an autopsy series) (87). Almost all primary cardiac lymphomas are aggressive B-cell lymphomas (88–90).

Several studies cite shortness of breath as the most common clinical manifestation (88–91), but arrhythmia (92,93), superior vena cava obstruction (94), cardiac tamponade (90), chest pain (95), epigastric pain (88), and vague systemic complaints (96) have also been reported. There is an increased prevalence of primary cardiac lymphoma in immunocompromised patients, who are more prone to develop all forms of extranodal non-Hodgkin lymphoma (97).



a. **b.**

Figure 10. Primary cardiac lymphoma in a 65-year-old man. **(a)** Contrast-enhanced CT scan shows a large left ventricular mass (arrow). **(b)** Photograph of an autopsy specimen shows cardiac lymphoma in the left ventricle (arrow), which is partially necrotic and has intramural and intraluminal tumor growth.

The diagnosis can be made from analysis of pericardial fluid (90,92,98), although direct biopsy is often necessary (89). Although primary cardiac lymphoma is rare, it is important to maintain a clinical suspicion for the tumor because early treatment with chemotherapy appears to be effective (83,89,99).

At gross examination, primary cardiac lymphoma often appears as multiple masses of firm, white nodules like those typically seen in extracardiac lymphoma. Primary cardiac lymphoma is less likely than sarcomas to have necrosis, involve the valves, and extend into heart chambers (1). The tumor most commonly arises in the right side of the heart, with the right atrium being reported as the most common site (88–90), but may involve any chamber (89,90). Pericardial effusions are often seen and are usually large (Fig 9) (1).

Pericardial effusion may be the only finding at CT or MR imaging (89). In other cases, there may be only pericardial thickening (99). In cases in which a mass was seen at CT or MR imaging, variable morphologic types have been reported. Either circumscribed polypoid masses (94) or ill-defined infiltrative lesions may be seen (Fig 10) (95,96,100).

The tumors are often dark on T1-weighted MR images and bright on T2-weighted images (96), but isointensity relative to cardiac muscle on both T1- and T2-weighted images has been reported (94,95). Contrast material enhancement may be homogeneous (94), heterogeneous (95,96,101), or minimal (89).

■ CONCLUSIONS

Important features of primary cardiac malignancies are clearly depicted at CT and MR imaging. Both modalities can demonstrate whether a tumor arises from the atrial septum, the width of the base of attachment, infiltrative growth, and extracardiac extent, criteria that can help distinguish benign from malignant neoplasms. Although all primary cardiac malignancies have variable appearances, they tend to differ with respect to chamber of origin, hemorrhage, calcification, and pericardial and valvular involvement. Taken together with a patient's clinical history (eg, age, immune status), CT and MR imaging findings may suggest the tumor type. Tumor differentiation can be clinically significant, especially in cases of cardiac lymphoma that may respond to chemotherapy. Primary cardiac malignancies are rare and will always present a diagnostic dilemma. However, as practitioners gain experience with the noninvasive evaluation of primary cardiac malignancies, these tumors should become less of a diagnostic challenge.

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