

CT and MR Imaging of Benign Primary Cardiac Neoplasms with Echocardiographic Correlation¹

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LEARNING OBJECTIVES

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

- Recognize the clinical findings associated with selected benign cardiac neoplasms as well as the typical location and morphology of each tumor type.
- Relate the gross pathology of each tumor type to its echocardiographic, CT, and MR imaging appearances.
- Analyze the strengths and weaknesses of echocardiography, CT, and MR imaging in the evaluation of each tumor type.

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Benign primary cardiac neoplasms are rare but may cause significant morbidity and mortality. However, they are usually treatable and can often be diagnosed with echocardiography, computed tomography (CT), or magnetic resonance (MR) imaging. Myxomas typically arise from the interatrial septum from a narrow base of attachment. Fibroelastomas are easily detected at echocardiography as small, mobile masses attached to valves by a short pedicle. Cardiac fibromas manifest as a large, noncontractile, solid mass in a ventricular wall at echocardiography and as a homogeneous mass with soft-tissue attenuation at CT. They are usually homogeneous and hypointense on T2-weighted MR images and isointense relative to muscle on T1-weighted images. Paragangliomas usually appear as large, echogenic left atrial masses at echocardiography and as circumscribed, heterogeneous masses with low attenuation at CT. These tumors are usually markedly hyperintense on T2-weighted MR images and iso- or hypointense relative to myocardium on T1-weighted images. Cardiac lipomas manifest at CT as homogeneous, low-attenuation masses in a cardiac chamber or in the pericardial space and demonstrate homogeneous increased signal intensity that decreases with fat-saturated sequences at T1-weighted MR imaging. Cardiac lymphangiomas manifest as cystic masses at echocardiography and typically demonstrate increased signal intensity at T1- and T2-weighted MR imaging. Familiarity with these imaging features and with the relative effectiveness of these modalities is essential for prompt diagnosis and effective treatment.

Abbreviations: AFIP = Armed Forces Institute of Pathology, MIBG = metaiodobenzylguanidine

Index terms: Fibroma and fibromatosis, 52.313 • Heart, CT, 52.1211, 53.1211 • Heart, MR, 52.12141, 53.12141 • Heart, neoplasms, 52.31, 53.31 • Heart, US, 52.1298, 53.1298 • Lipoma, 52.312 • Lymphangioma, 52.3142 • Myxoma, 52.311 • Paraganglioma, 52.3163

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Introduction

Primary cardiac neoplasms are rare, occurring in only 0.001%–0.03% of patients in autopsy series (1). In all age groups, benign primary cardiac neoplasms are more common than malignant ones (1). Although histologically benign in the sense that they do not metastasize, benign cardiac neoplasms may lead to significant morbidity and mortality by affecting blood flow and causing arrhythmias and emboli (1). Before the advent of cross-sectional imaging, these neoplasms were frequently diagnosed only at autopsy. Nowadays, they can be detected in living patients and are usually treated successfully (2).

Echocardiography is the primary modality for imaging intracardiac disease. It provides high-resolution, real-time images, whose quality has further improved with the introduction of new ultrasonographic (US) imaging techniques such as tissue harmonics (3). However, as image acquisition with computed tomography (CT) and magnetic resonance (MR) imaging has steadily become faster, these modalities have played an increasingly important role in the evaluation of cardiac neoplasms. Although spatial and temporal resolution are far lower with these modalities than with echocardiography, the soft-tissue contrast of both CT and MR imaging is superior to that of echocardiography, and both modalities allow imaging of the entire mediastinum and evaluation of the extracardiac extent of disease. Unlike MR imaging, CT is capable of helping detect calcification, which is an important variable in the differential diagnosis of cardiac neoplasms. In addition, CT is faster, easier to perform, and generally has more reliable image quality. MR imaging has better soft-tissue contrast than CT and allows much greater flexibility in the selection of imaging planes.

We reviewed the pathology records of patients who underwent surgical excision of benign cardiac neoplasms at our institution and selected those cases for which CT or MR images were available. We found 20 patients from the past 10 years with six tumor types: myxoma, papillary fibroelastoma, fibroma, paraganglioma, lipoma, and lymphangioma. Some of these cases were previously reported (4,5). In this article, we present selected images from these examinations, along with correlative echocardiographic images, and review the literature for each tumor type. The clinical, morphologic, and imaging features of each tumor type are summarized in the Table.

Myxoma

Cardiac myxoma is a gelatinous tumor that mimics primitive mesenchyme and is histologically distinct from extracardiac soft-tissue myxomas (1,6). The pathogenesis of cardiac myxoma is not understood. However, some authors suggest that myxomas arise from embryologic rests that become trapped during the septation phase of cardiac development (7,8). Myxomas are the most common primary cardiac neoplasm, accounting for about 50% of all primary cardiac tumors (1). They are slightly more common in women and typically appear in adulthood, with 90% of patients being between the ages of 30 and 60 years (1,9,10).

Although most cardiac myxomas are sporadic, they may be part of a syndrome known as Carney complex, an autosomal dominant syndrome of cardiac myxomas and a variety of hyperpigmented skin lesions (eg, lentigines, ephelides, blue nevi) (11,12). Carney complex is also associated with the development of extracardiac tumors such as pituitary adenomas, breast fibroadenomas, and psammomatous melanotic schwannomas. Endocrine abnormalities may also occur independent from effects of tumors (13). Patients with myxomas associated with Carney complex are typically younger than those with sporadic myxomas (8, 13). The former neoplasms are also more likely to occur outside the left atrium, be multifocal, and recur after resection (8,13,14). About 7% of myxomas occur as part of Carney complex (8, 13), although this only amounts to about 150 cases worldwide (15). Although the names are similar, Carney complex is not related to the Carney triad, which is the association of pulmonary hamartomas, extraadrenal paragangliomas, and gastric leiomyosarcomas (16).

Patients with cardiac myxomas often present with signs and symptoms of central nervous system or peripheral embolization, which may be caused by tumor fragments or accumulated thrombus (8). Patients also often present with systemic signs and symptoms such as fatigue, arthralgias, weight loss, and anemia, although the reason myxomas cause these problems is not known (8). The tumors may also lead to hemodynamic obstruction. Because myxomas occur most frequently in the left atrium, they can mimic mitral stenosis at clinical examination. Right atrial tumors may mimic tricuspid stenosis. The degree of obstruction can vary with body position (8).

The gross appearance of cardiac myxomas is variable. The tumors may be lobular and smooth or may have villous extensions (1,8). They frequently have organized thrombi on the surface.

Features of Benign Primary Cardiac Neoplasms

Type of Tumor (Prevalence)	Patient Age at Presentation	Associated Syndromes	Most Common Location	Typical Morphologic Characteristics	Echocardiographic Features	CT Features	MR Imaging Features*
Myxoma (50% of all primary cardiac neoplasms)	30–60 y (younger if associated with Carney complex)	Carney complex	Interatrial septum at fossa ovalis, left atrium more common than right	Gelatinous, attached to stalk; calcification common; hemorrhage or necrosis common	Mobile tumor, narrow stalk	Heterogeneous, low attenuation	Heterogeneous, bright on T2WI; heterogeneous enhancement
Papillary fibroelastoma (uncertain)	Middle-aged, elderly	None	Cardiac valves	Small (<1 cm), frond-like, narrow stalk; calcification rare (<i>n</i> = 2); no hemorrhage or necrosis	“Shimmering” edges	Usually not seen	Usually not seen
Fibroma (100 cases since 1976)	Infants, children, young adults	Gorlin syndrome	Ventricles	Large, intramural; calcification common; no hemorrhage or necrosis	Intramural, calcified	Low attenuation, calcified	Isointense on T1WI, dark on T2WI; usually little to no enhancement
Parangangioma (less than 50 cases)	Young adults	Many possible, but almost always sporadic	Left atrium, coronary arteries, aortic root	Broad-based, infiltrative or circumscribed; calcification rare (<i>n</i> = 1); hemorrhage or necrosis common	Echogenic, relatively immobile	Low attenuation	Typically isointense or heterogeneous on T1WI, very bright on T2WI; marked enhancement
Lipoma (approximately 60 cases)	Variable	A few cases associated with tuberosous sclerosis	Pericardial space or any cardiac chamber	Very large, broad-based; no calcification, hemorrhage, or necrosis	Usually hypoechoic in the pericardial space, echogenic in a cardiac chamber	Homogeneous fat attenuation (low attenuation)	Homogeneous fat signal intensity (increased T1); no enhancement
Lymphangioma (nine cases)	Infants, children	None, but may be associated with other lymphangiomias	Pericardial space	Large, cystic; no calcification, hemorrhage, or necrosis	Heterogeneous, septated, hypoechoic	Heterogeneous, septated, low attenuation	Heterogeneous, septated, bright on T2WI; heterogeneous enhancement

*T1W1 = T1-weighted images, T2W1 = T2-weighted images.

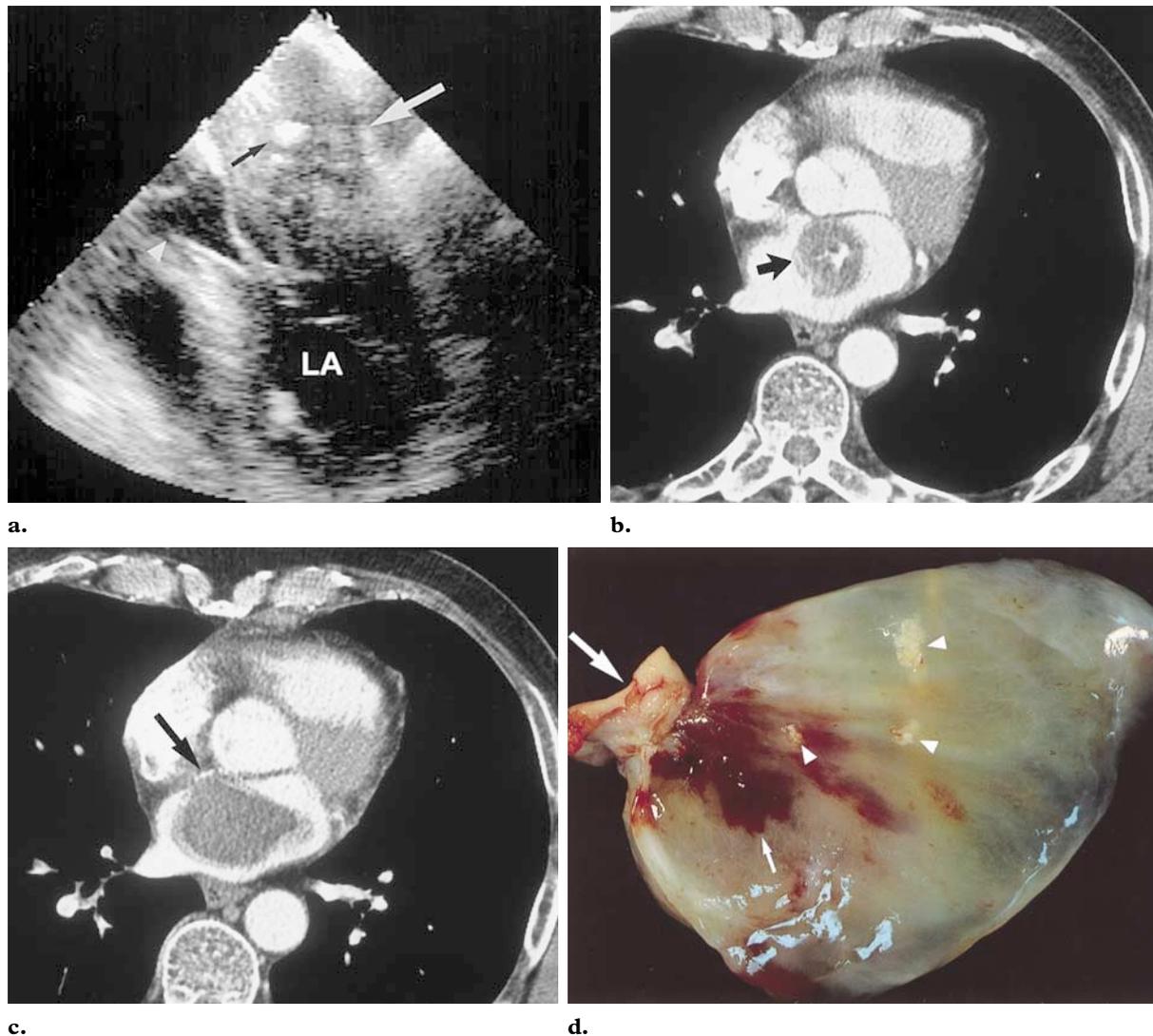


Figure 1. Left atrial myxoma in a 60-year-old man. **(a)** Transverse transesophageal echocardiogram obtained during diastole shows a large, inhomogeneous mass in the left atrium (LA) (white arrow) with focal areas of calcification (black arrow). The mass fills much of the visualized left atrium. The left ventricular outflow track (arrowhead) is seen on the left side of the image. **(b)** Contrast material-enhanced electron beam CT scan obtained at the level of the aortic root shows the left atrial mass with prominent calcification (arrow). **(c)** On a contrast-enhanced electron beam CT scan obtained at a slightly lower level, the mass demonstrates a narrow base of attachment to the interatrial septum (arrow). **(d)** Photograph of the resected specimen shows the myxoma with a narrow pedicle (large arrow). Marked myxoid change, focal hemorrhage (small arrow), and focal calcifications (arrowheads) are also seen.

Internally, myxomas are heterogeneous and frequently contain cysts, necrosis, and hemorrhage (1,10,17). In a large surgical series, calcification was present in 16% of tumors (10). Myxomas may arise from a broad base but frequently arise from a narrow stalk, a feature that is also found in papillary fibroelastomas but is very unusual in other cardiac tumors (1).

Over 90% of myxomas are solitary, intracavitary, and atrial in location (1,10). Multiple myxomas are rare and are usually associated with Carney complex (1). As mentioned previously, myxomas have a predilection for the interatrial

septum. More specifically, they tend to arise from the fossa ovalis, a depression representing the remains of the fetal foramen ovale. In a large series from the Armed Forces Institute of Pathology (AFIP), 78% of myxomas arose from this site (1). Myxomas occur more commonly in the left atrium (75%–80% of all cardiac myxomas) than in the right (10%–20%) (1,9) but may occasionally grow through the fossa ovalis and into both atria (1,8).

At echocardiography, the characteristic narrow stalk is the most important distinguishing feature of a myxoma, followed by tumor mobility and distensibility (18,19). When these features are seen, myxoma can be diagnosed with a high degree of

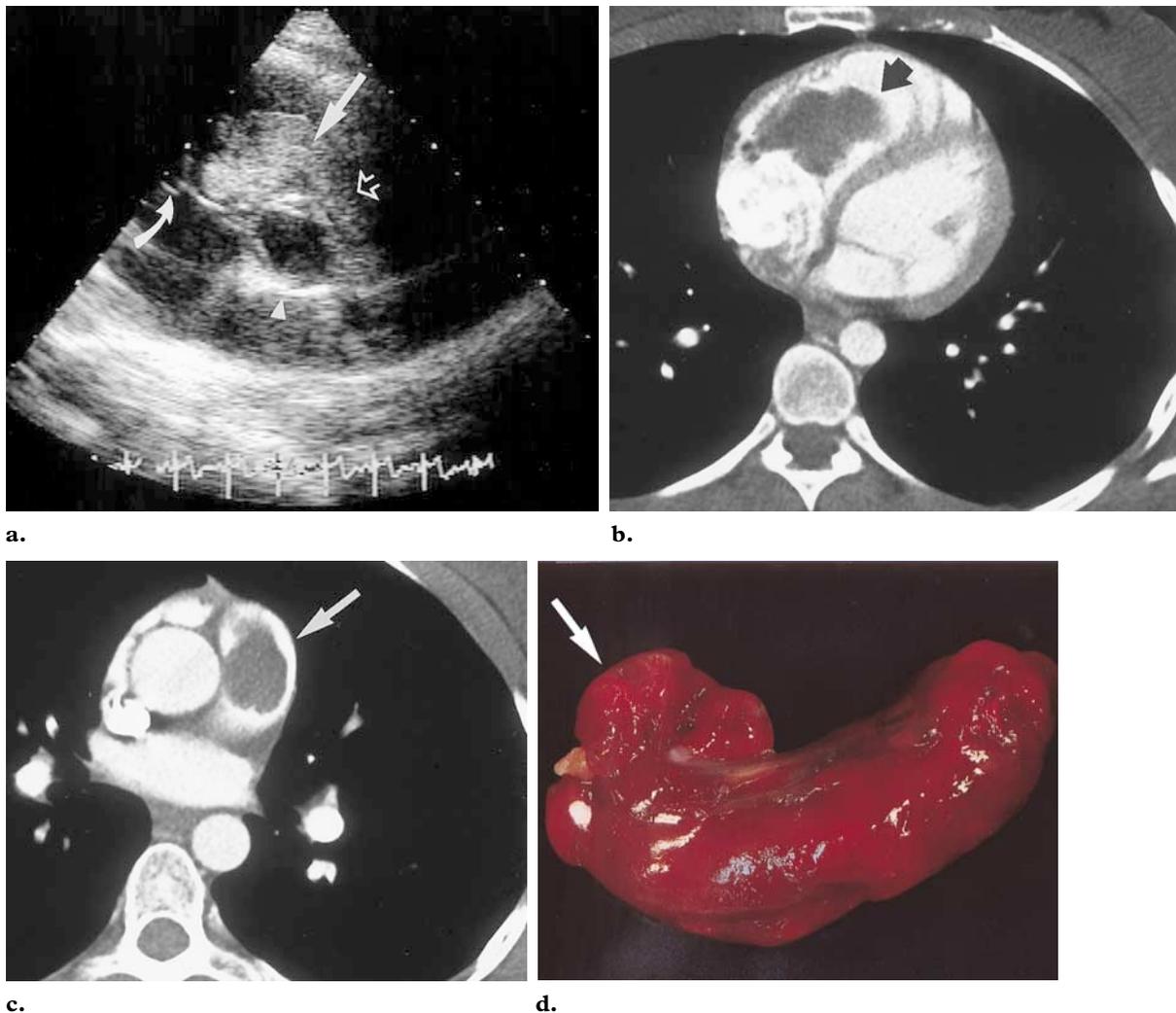


Figure 2. Right ventricular myxoma in a 55-year-old man. **(a)** Parasternal transthoracic echocardiogram (short-axis view) obtained during systole shows a mass (straight solid arrow) that prolapses from the right ventricle into the main pulmonary artery (open arrow). Rib attenuation casts a shadow over the right side of the image. The tricuspid valve (curved arrow) and aortic root (arrowhead) are also seen. **(b, c)** Contrast-enhanced electron beam CT scans obtained at the level of the ventricles **(b)** and main pulmonary artery **(c)** show the low-attenuation mass in the right ventricle (arrow in **b**) with extension into the main pulmonary artery (arrow in **c**). **(d)** Photograph of the resected specimen shows the homogeneous, elongated myxoma with a pedicle at the base (arrow).

confidence (18), especially if the tumor is also attached to the interatrial septum. Myxomas demonstrate variable internal echocardiographic features. They may be homogeneous or may have central areas of hyperlucency representing hemorrhage and necrosis (20,21). Echogenic foci of calcification may also be detected. Broad-based, nonmobile myxomas may also occur, but they are indeterminate at echocardiography.

The thin, delicate stalk of myxomas usually cannot be defined at CT or MR imaging, although a generally narrow base of attachment can be seen. This narrow base of attachment is suggestive of but probably not definitive for a diagnosis of myxoma. Because tumor mobility and distensibility also cannot be depicted, tumor lo-

cation is the other important distinguishing feature. A mass that arises from the interatrial septum from a narrow base of attachment may be identified as a cardiac myxoma with a high degree of confidence (Fig 1) (22). Other CT and MR imaging findings are variable and reflect gross pathologic features. Because of their gelatinous nature, myxomas usually have heterogeneous low attenuation at CT. Calcification is frequently seen (22). CT may occasionally show myxomas prolapsing through cardiac valves (Fig 2), which is a common finding at echocardiography. Myxomas tend to have markedly increased signal intensity on T2-weighted MR images (23, 24). However, they may have areas of decreased

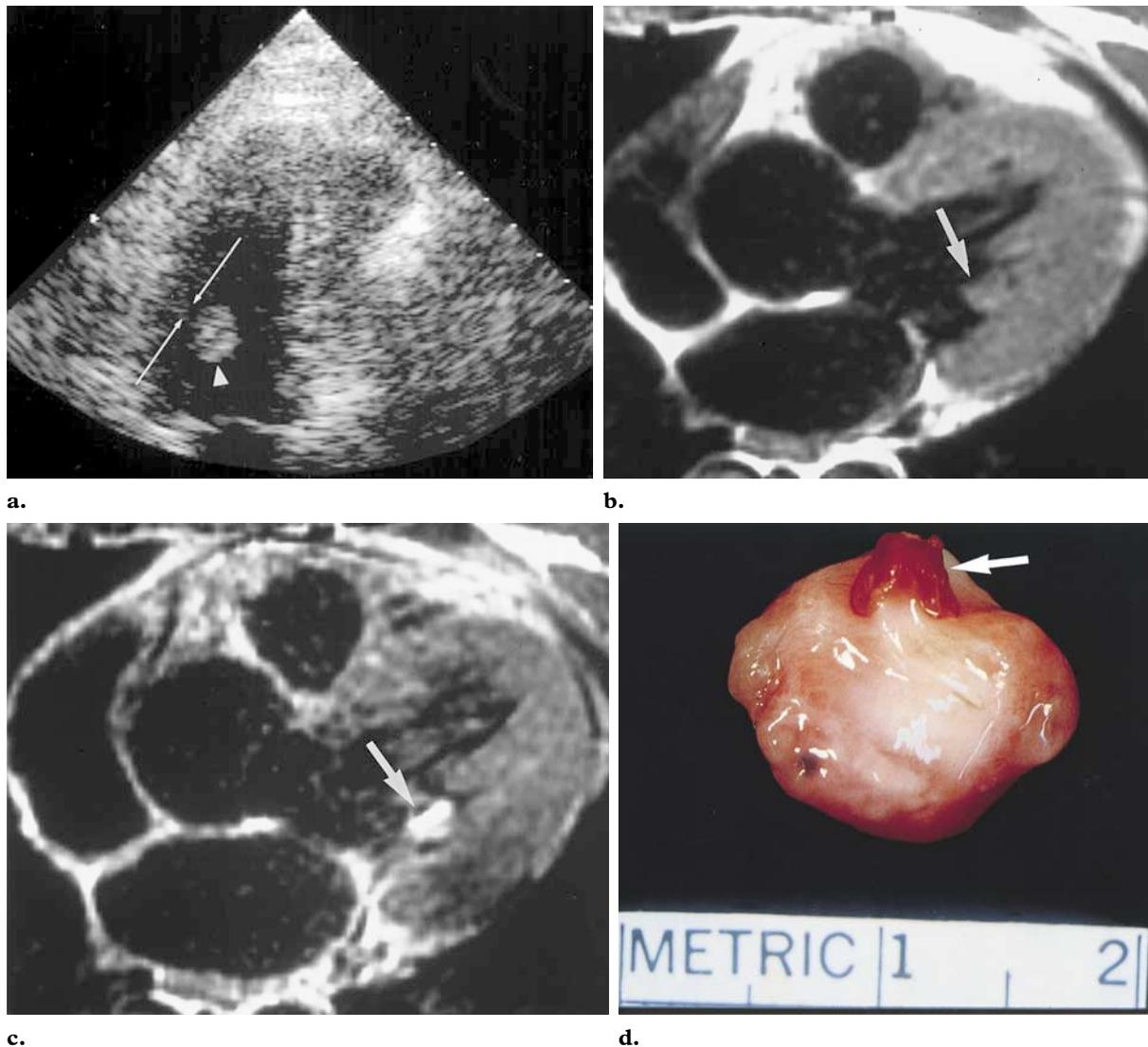


Figure 3. Left ventricular myxoma in a 41-year-old man. **(a)** Transesophageal echocardiogram (four-chamber view) centered on the left ventricle shows a pedunculated mass (arrowhead) with a faintly visible stalk arising from the lateral wall of the left ventricle (arrows). **(b, c)** Unenhanced **(b)** and contrast-enhanced **(c)** T1-weighted MR images demonstrate the small mass attached to the lateral wall of the left ventricle (arrow). The mass shows marked enhancement in **c**. **(d)** Photograph of the resected specimen shows the homogeneous, polypoid myxoma with a narrow pedicle (arrow). Scale is in centimeters.

signal intensity due to the presence of calcification or to magnetic susceptibility artifacts caused by hemosiderin (17,23,24). Contrast material enhancement in myxomas is usually heterogeneous, which also likely reflects the presence of necrotic areas within the tumor (23,25), but intense enhancement may be seen (Fig 3).

In adults, the primary differential diagnosis for a solitary, intracavitary cardiac mass includes myxoma, thrombus, and nonmyxomatous neoplasms, most of which are malignant. As stated previously, a cardiac mass with a narrow base of attachment to the interatrial septum almost certainly represents a myxoma.

Papillary Fibroelastoma

Papillary fibroelastoma is a collection of avascular fronds of dense connective tissue lined by endothelium (1). As with myxoma, the cause of fibroelastoma is not clear. Some authors believe that fibroelastoma is a reactive process, whereas others believe it is a hamartoma (1,26). Synonyms include fibroelastic papilloma, papilloma of valves, giant Lambl excrescence, myxofibroma, myxoma of valves, hyaline fibroma, and fibroma of valves (1).

The reported prevalence of papillary fibroelastoma varies. Because they are usually asymptomatic, papillary fibroelastomas are likely underrepresented in surgical series, and there have been several large surgical series in which they did not appear at all (1). However, in surgical series at

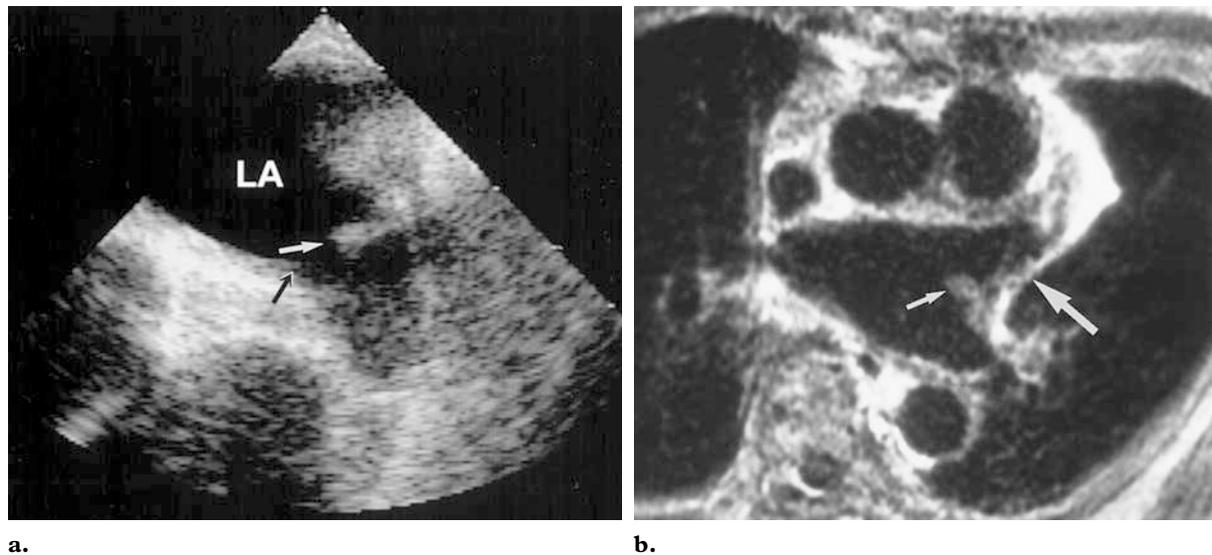


Figure 4. Left atrial papillary fibroelastoma in a 66-year-old woman. **(a)** Transverse transesophageal echocardiogram shows a small, pedunculated mass (white arrow) arising from the lateral wall of the left atrium (LA) near the mouth of the left atrial appendage (black arrow). **(b)** T1-weighted MR image reveals the mass in the same position (small arrow) (cf **a**). Large arrow indicates the left atrial appendage.

the Mayo Clinic (10) and the AFIP (1), they represented 10% of all primary cardiac tumors and were the second most common benign primary cardiac neoplasm after myxoma. When symptoms are present, they are usually related to embolization from thrombi that collect on the tumors (27). Right-sided fibroelastomas are usually asymptomatic (28,29). Fibroelastomas are equally common in men and women, with the mean age at presentation being about 60 years (1).

At gross examination, the tumors are small (usually less than 1 cm in diameter) (30), although they have been reported as large as 5 cm (1). They are composed of delicate, frondlike excrescences attached to the endocardium by a short pedicle and are often likened to a sea anemone (1). Over 90% occur on valve surfaces, making them by far the most common primary cardiac tumor to occur on valves (31). They are slightly more common on the aortic (29%) and mitral (25%) valves than on the pulmonary (13%) and tricuspid (17%) valves (27), although this reported predilection may be due to the increased prevalence of symptoms associated with left-sided fibroelastomas. About 16% of the tumors arise from nonvalvular surfaces (1). Calcification rarely occurs, having been reported in only two cases (32). The tumors are usually isolated, although in one series, three of 17 patients with histologically proved papillary fibroelastoma had multiple tumors (33).

Fibroelastomas are easily detected at echocardiography, where they appear as small, mobile masses attached to valves by a short pedicle.

They may appear as elongated strandlike projections or may have a well-defined “head” (33). Fibroelastomas have a stippled edge with a “shimmer” or “vibration” at the interface of the tumor with the surrounding blood (33). This has been reported as a characteristic feature of fibroelastomas, helping distinguish them from the more amorphous thrombi (33).

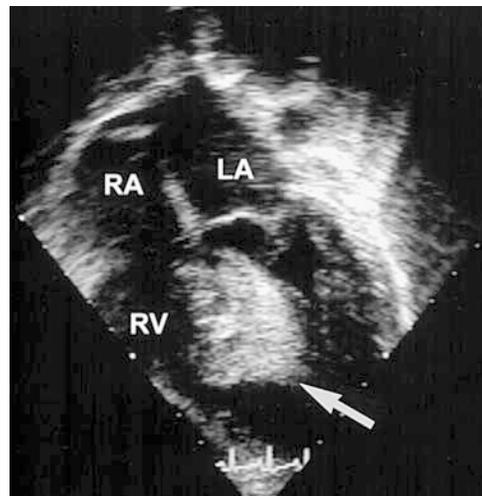
Although fibroelastomas are well visualized at echocardiography, they are usually not seen at CT or MR imaging because they are small and are attached to moving valves. To our knowledge, there have been no reported cases of a fibroelastoma detected at CT and only one report of a fibroelastoma seen at MR imaging (34). In that case, the fibroelastoma was a 1.2-cm-diameter mass seen on a tricuspid valve on cine images. With current technology, CT and MR imaging will probably help detect only exceptionally large fibroelastomas or the atypical fibroelastomas that occur away from the valves (Fig 4).

Fibroma

Cardiac fibroma is a collection of fibroblasts interspersed among large amounts of collagen (1). Some authors believe that this tumor is a hamartoma rather than a true neoplasm (1). Cardiac fibroma has also been referred to as fibromatosis, fibrous hamartoma, and fibroelastic hamartoma (1).

Fibroma is a tumor that primarily affects children and in most cases is detected in infants or in utero (1,35). At autopsy, fibromas are the second most common benign primary cardiac tumor in

Figure 5. Left ventricular fibroma in a 7-year-old boy. **(a)** Transesophageal echocardiogram (four-chamber view oriented with the apex downward) obtained during systole shows a large, homogeneous mass arising from the interventricular septum and protruding into the left ventricular cavity (arrow). *LA* = left atrium, *RA* = right atrium, *RV* = right ventricle. **(b)** T1-weighted MR image shows diffuse thickening of the interventricular septum, left ventricular apex, and lateral wall. **(c)** Photograph of the resected specimen shows the dense, firm, white fibroma with a whorled cut surface. Scale is in centimeters.



a.



b.



c.

children after rhabdomyoma (1). Fibromas are rare, with only about 100 cases reported since 1976 (1).

Cardiac fibromas are often associated with arrhythmias (1,35) and are the second most common primary cardiac tumor associated with sudden death after endodermal heterotopia of the atrioventricular node (36). They may also accompany heart failure (1). About one-third of cardiac fibromas are asymptomatic (1) and may be discovered incidentally due to electrocardiographic abnormalities (37), heart murmurs (38), or chest x-ray abnormalities (39).

There is an increased prevalence of cardiac fibromas in Gorlin syndrome, also known as nevoid basal cell carcinoma syndrome (1). This is an autosomal dominant condition associated with basal cell carcinomas, odontogenic keratocysts of the mandible, skeletal anomalies, and a tendency toward neoplastic growth in several organ systems (40–42).

At gross examination, the tumors are firm, white, fibrous masses that arise within the myocardial wall (Fig 5) (1). Unlike many primary cardiac tumors, fibromas usually have no foci of cystic change, hemorrhage, or necrosis (1,43,44). Cardiac fibromas almost always arise in the ventricles, most commonly in the ventricular septum and left ventricular free wall (1). They are usually large, with a mean diameter of about 5 cm (1), and may obliterate the ventricular cavity (41). The tumors may be circumscribed at gross examination, although not truly encapsulated (10), or may be infiltrating (1). In all but one reported case, the tumors were single (1). Dystrophic calcification is common (10,45).

At echocardiography, cardiac fibroma appears as a large, noncontractile, solid mass in a ventricular wall. The tumor may be nodular and discrete (46) or may mimic focal hypertrophic cardiomyopathy (45,47) or hypertrophy of the ventricular septum (4).

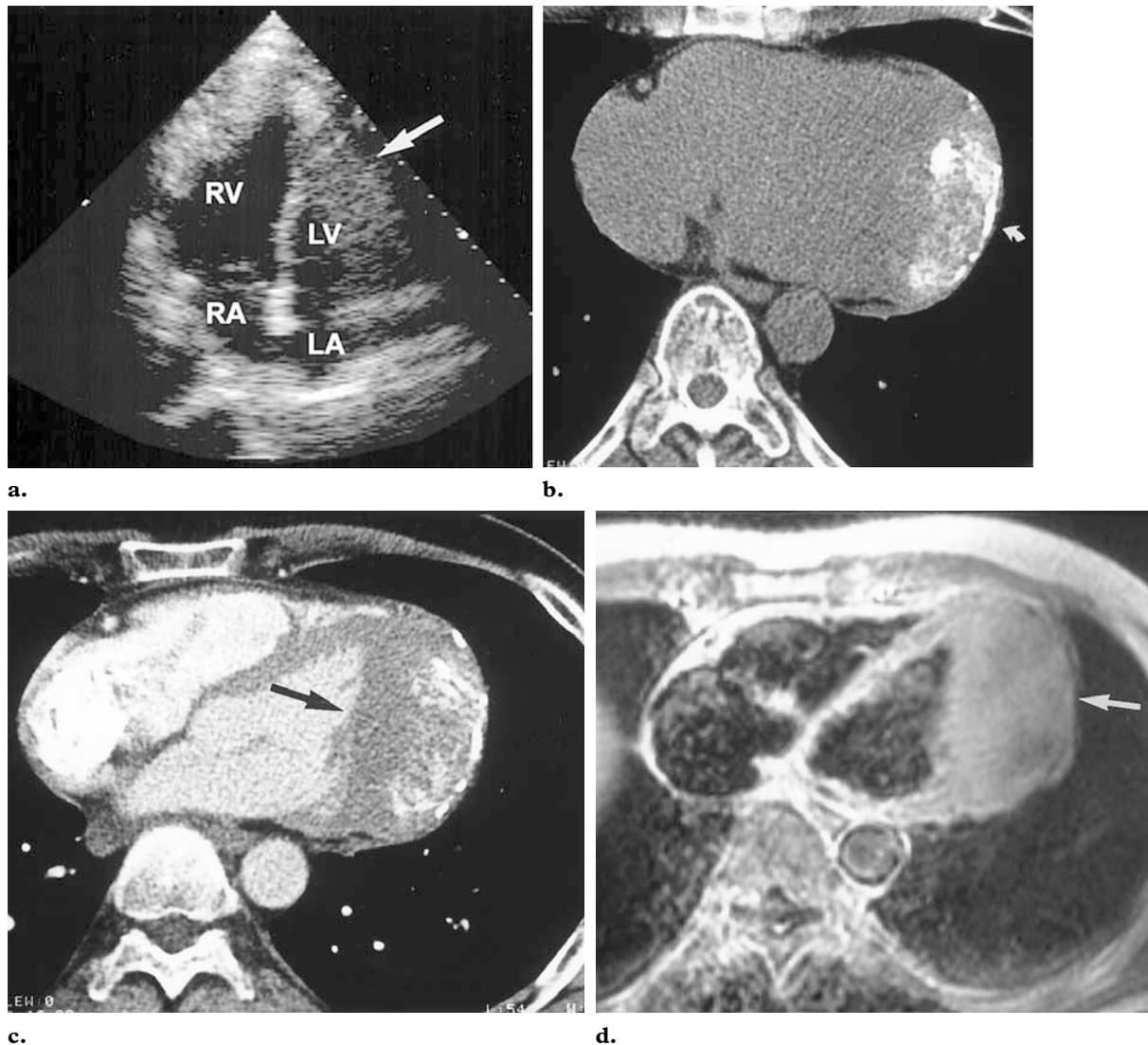


Figure 6. Left ventricular fibroma in a 55-year-old man. **(a)** Transesophageal echocardiogram (apical four-chamber view) demonstrates a large, homogeneous mass arising from the lateral wall of the left ventricle (*LV*) (arrow). *LA* = left atrium, *RA* = right atrium, *RV* = right ventricle. (Reprinted, with permission, from reference 4.) **(b)** Unenhanced electron beam CT scan shows coarse calcifications in the region of the lateral wall of the left ventricle (arrow). **(c)** On a contrast-enhanced CT scan, the calcifications are seen within the large, low-attenuation mass, which diffusely replaces the lateral wall (arrow). **(d)** T1-weighted MR image shows the homogeneous, low-signal-intensity mass in the lateral wall of the left ventricle (arrow).

At CT, fibromas manifest as homogeneous masses with soft-tissue attenuation that may be either sharply margined or infiltrative. Calcification is often seen (Fig 6) (44). Because of their dense, fibrous nature, the tumors are usually homogeneous and hypointense on T2-weighted MR images (48–50) and isointense relative to muscle on T1-weighted images (Figs 5, 6) (41,50–52). For the same reason, the tumors often demonstrate little or no contrast material enhancement (4,44,50). However, several other patterns of enhancement have been reported, including both homogeneous enhancement (44,47) and heterogeneous enhancement (44). One case showed a peripheral rim of enhancement with a central, nonenhancing area thought to represent a more dense fibrous core (53).

In infants, the primary differential diagnosis for an intramural mass is rhabdomyoma. If there are multiple masses or signs of tuberous sclerosis, rhabdomyoma can be confidently diagnosed (35). If the tumor is solitary, ventricular, and calcified, the diagnosis of fibroma can be made from the radiologic findings (35). Rhabdomyosarcoma is a much less common cardiac tumor in children. This rare, malignant tumor does not calcify and occurs in all cardiac chambers with equal frequency (54). It is frequently cystic or necrotic and may be seen invading pulmonary veins, the pericardial space, or other adjacent structures (54).

Paraganglioma

Paraganglioma is a tumor that arises from paraganglia, the clusters of neuroendocrine cells widely distributed throughout the body, including the adrenal medulla, carotid, vagal and paraaortic bodies, and groups of cells associated with sympathetic ganglia. There have been a few reports of metastatic spread of cardiac paragangliomas (25,55–58), but because these tumors are usually benign, they are included in this discussion. Paraganglioma has sometimes been called pheochromocytoma or chemodectoma depending on whether the tumor was functioning or nonfunctioning. The authors at the AFIP recommend the terms *functioning paraganglioma* and *nonfunctioning paraganglioma* (1).

Cardiac paragangliomas are very rare. There have been fewer than 50 reported cases in the medical literature written in English (56), although some of these were tumors of the great vessels and not truly cardiac paragangliomas (59–63). Patients range in age from the early teens (55) to the mid-60s (1) but typically are young adults in their 30s and 40s (64). Most patients present with hypertension and biochemical evidence of catecholamine overproduction (64). The tumor is usually localized to the heart with nuclear medicine imaging with iodine-131 or -123 metaiodobenzylguanidine (MIBG) (Fig 7) (55). MIBG, a norepinephrine analogue, is widely used for localizing extraadrenal paragangliomas (65). Because the tumors are highly vascular and tend to involve the coronary arteries, surgical resection is often difficult (64,66), and there have been several deaths from intraoperative or postoperative hemorrhage (64,67).

Although there are several familial syndromes associated with adrenal and extraadrenal paragangliomas, nearly all cases of cardiac paraganglioma are sporadic. In our review, we found only one case of a cardiac paraganglioma associated with a syndrome (Carney triad) (57). There have been several cases of cardiac paraganglioma with synchronous paragangliomas elsewhere in the body (68–72) and one case involving a patient with a family history of paragangliomas (73). However, to our knowledge, there have been no cases in which cardiac pheochromocytoma occurred with documented multiple endocrine neoplasia, neurofibromatosis, or von Hippel–Lindau syndrome.

At gross examination, the tumors are soft, fleshy, and usually tan or brown (1,64). They are generally 3–8 cm in diameter (57), although 15-cm tumors have been reported (1). The tumors may be encapsulated (74,75), but they may also

be infiltrative and difficult to “shell out” at surgery (64,76,77). Necrosis is common and was present in seven of 12 patients in one series (57). Calcification rarely occurs, having been reported in only one case to our knowledge (57).

The tumors occur in the distribution of the cardiac paraganglia. Most arise from visceral paraganglia in the left atrium (1,59,78), most frequently in the posterior wall of the left atrium or the left atrial roof (60,64,79). However, they may also arise from the interatrial septum (64,79–82) or from paraganglia along the coronary arteries (59,60,64,66,78,80,83).

At echocardiography, paragangliomas usually appear as large, echogenic left atrial masses. There have been cases of primary cardiac paragangliomas arising from the interatrial septum that have been mistaken for myxomas (75). Unlike myxomas, however, paragangliomas have a broad base of attachment and, although they are soft, are firmer than myxomas (74,75). Compression of adjacent structures such as the superior vena cava may be seen (74,75). Transesophageal echocardiography may demonstrate encasement of the coronary arteries (78).

Because they are usually localized to the mediastinum with nuclear medicine imaging and not on the basis of cardiac symptoms, paragangliomas are more often imaged with CT or MR imaging than are other cardiac neoplasms (57,84). At CT, they usually appear as circumscribed, heterogeneous masses with low attenuation (57, 79,85). However, extracardiac extension (56) and ill-defined, infiltrative borders (55) may be seen. At MR imaging, primary cardiac paragangliomas usually have markedly increased signal intensity on T2-weighted images, similar to abdominal paragangliomas (57), although at least one case has been reported in which the tumor was hypointense on a T2-weighted image (86). The tumors are usually iso- or hypointense relative to myocardium on T1-weighted images (25, 57). Increased signal intensity on T1-weighted images has been reported (87), presumably due to hemorrhage within the tumor. Because of their vascularity, paragangliomas demonstrate intense contrast material enhancement. However, enhancement is often heterogeneous, with central nonenhancing areas due to tumor necrosis (25, 57,88).

In most cases, the diagnosis of paraganglioma can be made on the basis of clinical information, which is fortunate because the imaging features of the tumor are nonspecific. The predominantly left atrial location and broad base of attachment that are seen in paraganglioma may also be seen in primary cardiac sarcomas (54).

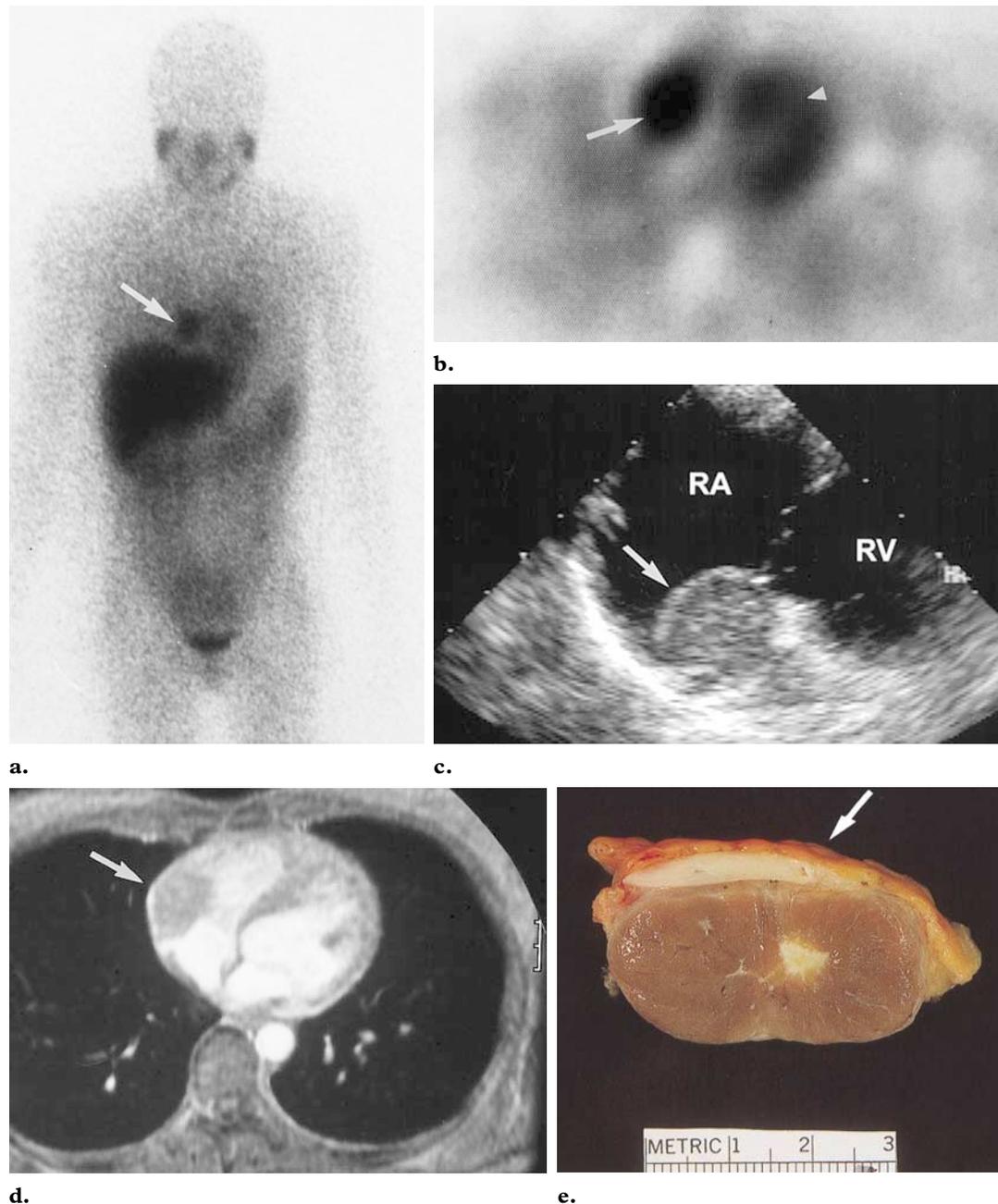


Figure 7. Right atrial paraganglioma in a 27-year-old woman. **(a)** Whole-body I-123 MIBG scan shows an abnormal focus of increased uptake in the right side of the mediastinum (arrow). Uptake in the liver, left side of the heart, gastrointestinal tract, salivary glands, and bladder is normal. **(b)** Axial single-photon-emission tomogram through the chest shows an abnormal focus of increased uptake in the right side of the heart (arrow). Uptake in the left ventricular myocardium is normal (arrowhead). **(c)** Transverse transesophageal echocardiogram obtained during systole shows a homogeneous, broad-based mass in the posterolateral right atrium (*RA*) adjacent to the tricuspid valve (arrow). *RV* = right ventricle. **(d)** Cine phase-contrast MR image shows the broad-based mass in the right atrium (arrow). **(e)** Photograph of the resected specimen shows the circumscribed, characteristically tan-brown paraganglioma with a central scar. A portion of the excised right atrial wall is included in the specimen (arrow). Scale is in centimeters.

Lipoma

Primary cardiac lipomas are benign neoplasms composed of mature adipose tissue and are histologically similar to extracardiac soft-tissue lipomas (1). The number of reported cases is not clear because some series do not differentiate between lipoma and lipomatous hypertrophy of the atrial septum (1), which is not a true neoplasm and will be discussed later. There have probably been about 60 reported cases of primary cardiac lipoma (1).

The tumors occur across a wide age range (1). They are soft and may grow to a large size without causing symptoms. In asymptomatic patients, the tumors may be found incidentally because of a chest x-ray abnormality (89–91) or heart murmur (92). There has been at least one case of a large, asymptomatic cardiac lipoma that grew over several years and eventually caused shortness of breath (90). Intracavitary lipomas may cause obstruction with related symptoms (93). Lipomas in the pericardial space may compress the ventricles (1,94) or may cause shortness of breath by displacing the lungs without affecting left ventricular function (90,95,96). Cardiac lipomas have been associated with a variety of arrhythmias including atrial fibrillation (91), ventricular tachycardia (97), and atrioventricular block (98). There have been a few reported cases of children with tuberous sclerosis with multiple cardiac lipomas; generally, however, cardiac lipomas do not occur as part of a syndrome (1). The tumors are usually easily resected (1,93), although there have been reports of lipomas infiltrating the coronary arteries, requiring complex surgery (1) or making the tumor unresectable (89,97).

At gross examination, lipomas are encapsulated, homogeneous fatty tumors (1). They frequently arise from the epicardial surface, usually from a broad pedicle, and grow into the pericardial space (1,90,94–96,99). They may also arise from the endocardium and grow as broad-based, pedunculated masses into any of the cardiac chambers (1,10,89,91–93,98,100,101). Several lipomas have been reported arising from the interatrial septum (91,98,101). These tumors are often very large by the time they come to clinical attention and have weighed as much as 4,800 g (90).

The echocardiographic appearance of lipomas varies with their location. Lipomas in the pericardial space may be completely hypoechoic (95), have hypoechoic regions (94,96), or be completely echogenic (97), whereas intracavitary lipomas are homogeneous and hyperechoic (92,

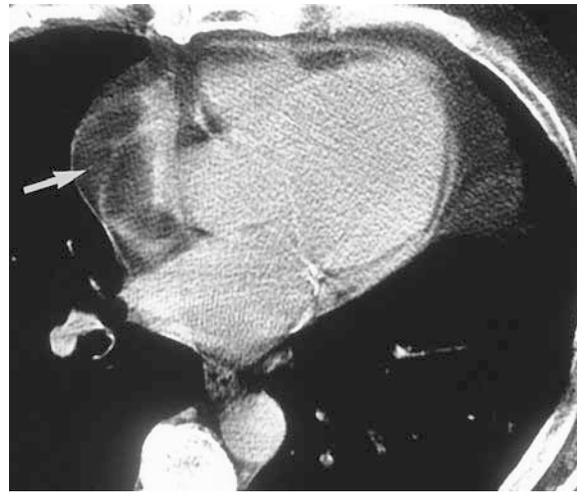
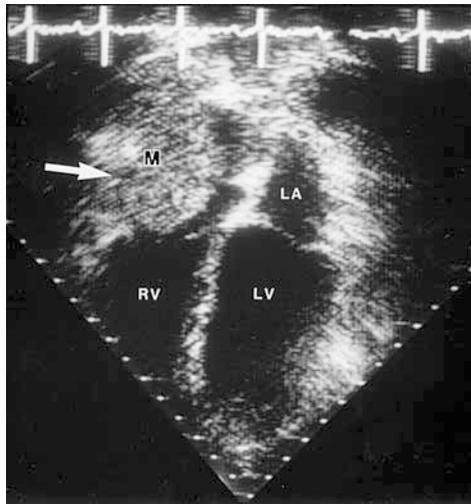


Figure 8. Right atrial lipoma in a 60-year-old man. Unenhanced CT scan shows a fat-attenuation mass filling most of the right atrium (arrow).

101). The reason for this difference is not known. Lipomas arising from the interatrial septum have been mistaken for myxomas (101); however, lipomas have a broad base of attachment (98,101) and are not as mobile as myxomas (101). Although lipomas can be differentiated from typical myxomas, they cannot be distinguished from other cardiac masses with similar morphologic characteristics at echocardiography.

Both CT and MR imaging can help identify fat with a high degree of specificity and can therefore be used to diagnose cardiac lipomas without equivocation. For this reason, reports of cardiac lipomas over the past 10–15 years have tended to emphasize the role of CT and MR imaging and go into detail about the imaging features of these tumors. At CT, cardiac lipomas appear as homogeneous, low-attenuation masses either in a cardiac chamber or in the pericardial space (Fig 8) (90,92–94,97,98,101). At MR imaging, lipomas have homogeneous increased signal intensity on T1-weighted images (Fig 9) that decreases with fat-saturated sequences. They may have a few thin septations (89) but no soft-tissue component (49,90,91,92,94,95,101). Like soft-tissue lipomas, cardiac lipomas do not enhance with the administration of contrast material.

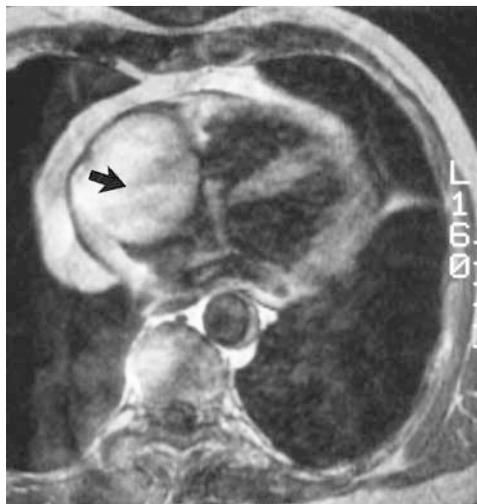
As stated previously, cases of lipomatous hypertrophy of the interatrial septum have been included in reports of cardiac lipomas. Lipomatous hypertrophy of the interatrial septum is defined as “any deposit of fat in the atrial septum at the level of the fossa ovalis which exceeds 2 cm in transverse dimension” (1). However, lipomatous hypertrophy of the interatrial septum is caused by an increase in the number of adipocytes, not



a.



c.



b.

Figure 9. Right atrial lipoma in a 72-year-old man. **(a)** Transesophageal echocardiogram (apical four-chamber view) shows a large mass (*M*) arising from the posterolateral wall of the right atrium (arrow). *LA* = left atrium, *LV* = left ventricle, *RV* = right ventricle. **(b)** T1-weighted MR image shows the circumscribed, broad-based mass with increased signal intensity (arrow). The mass appears similar to the mediastinal fat and fills most of the right atrium. **(c)** Intraoperative photograph shows the smooth lipoma filling the right atrium (arrow). (Figure 9 reprinted, with permission, from reference 5.)

hypertrophy, so that the term is actually a misnomer (1). The fatty infiltration spares the fossa ovalis, a characteristic feature that can be seen at echocardiography, CT, and MR imaging. Unlike cardiac lipoma, lipomatous hypertrophy of the interatrial septum is not encapsulated and is not a true neoplasm (1). It is associated with advanced age and obesity and is much more common than cardiac lipoma (1).

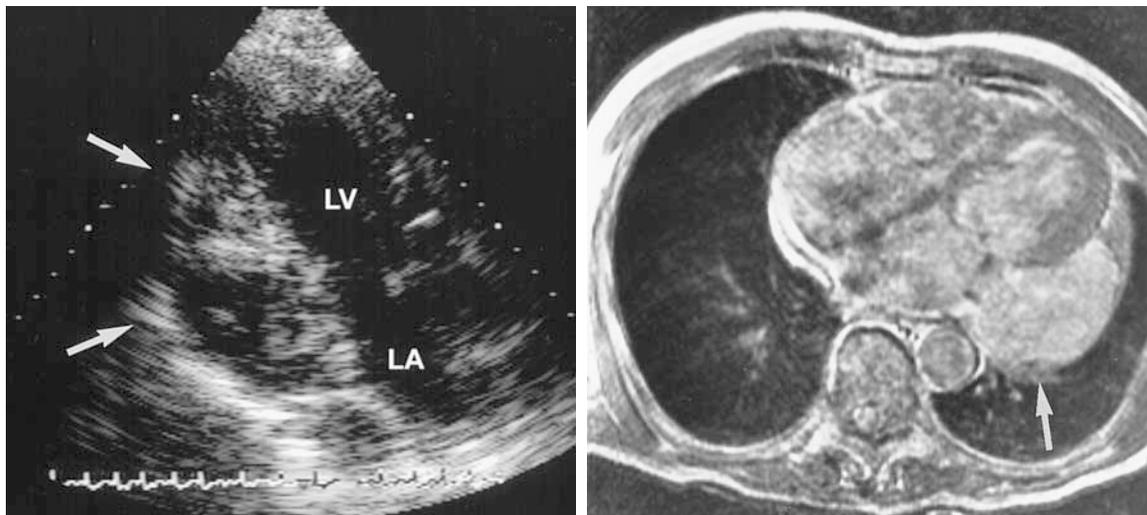
Primary cardiac hemangiomas are rare tumors that may grow into the pericardial space and may contain fat (1). However, like soft-tissue hemangiomas, these tumors are heterogeneous and demonstrate marked enhancement (102). Primary cardiac liposarcoma is a very rare malignant neoplasm that is large, aggressive, and composed predominantly of soft tissue and that has not been mistaken for benign lipoma to our knowledge (54).

Lymphangioma

Lymphangioma is a benign neoplasm composed of endothelial-lined, thin-walled spaces that contain lymph but are isolated from the lymphatic system (1). Lymphangiomas are histologically similar to hemangiomas, but the vascular spaces in hemangiomas contain blood rather than lymph (1). Lymphangiomas are sometimes called hygromas, especially when they occur in the neck.

Cardiac lymphangiomas are exceptionally rare. Nine cases have been reported (103), although some of these cases may be better classified as cardiac hemangiomas (104,105). More than half of the reported cases occurred in patients under 6 years of age (103,106,107). Two cases have been reported in newborns (103,106).

Four of the nine patients with cardiac lymphangioma presented with arrhythmias or palpitations (104,105,108). One of these four patients experienced cardiac arrest and died during cardiac catheterization (108). One patient, a neonate,



a.

Figure 10. Pericardial lymphangioma in a 76-year-old man. (a) Transesophageal echocardiogram (apical long-axis view) shows a heterogeneous extra-cardiac mass abutting or invading the inferolateral wall of the left ventricle (*LV*) and left atrium (*LA*) (arrows). (b, c) Axial (b) and coronal (c) T1-weighted MR images show the heterogeneous, lobulated mass in the pericardial space (arrows).

presented with cardiac tamponade immediately after birth (106). One patient presented with pain in his arms with exertion (107), although it is not clear whether this was related to ischemia or was an unrelated finding.

In none of the cases was cardiac lymphangioma associated with a characterized syndrome, but in three cases, patients with cardiac lymphangiomas had other tumors as well. A 10-year-old boy had subcutaneous, lymphvascular masses of the axilla and neck (107). A 10-month-old child had large lymphangiomas of the neck and mediastinum (107). A 43-year-old man had a right atrial lipoma and lipomatous infiltration of much of the myocardium (108). There has been one case of a cardiac lymphangioma associated with congenital malformation. In that case, a newborn with pericardial tamponade from a lymphangioma also had coarctation of the aorta (106), although it is not clear whether the coarctation was actually aortic hypoplasia caused by lack of left ventricular output or a truly separate lesion.

At gross examination, cardiac lymphangiomas may be soft and spongy (104,107) or firm and fibrous (108). Vascular spaces are often visible to the eye (107,108), and lymphatic fluid can sometimes be expressed (108). The tumors most commonly occur in the pericardial space, sometimes compressing adjacent structures (106,107). A chylous pericardial effusion may also be present

b.



c.

(106). There has been at least one case in which a lymphangioma was primarily myocardial (6).

At echocardiography, cardiac lymphangiomas appear as cystic masses (106,107). In one report, the tumor was described simply as echogenic (104). To our knowledge, there have been no reports regarding CT of cardiac lymphangiomas. At MR imaging, these tumors may have increased signal intensity on T1-weighted images (106), possibly due to the presence of fat in the stroma (Fig 10). The tumors will also demonstrate areas of increased signal intensity on T2-weighted images due to cystic spaces. There have been no reports concerning the enhancement pattern of cardiac lymphangiomas, but one would expect the pattern to be variable like that of soft-tissue lymphangiomas. Lymphangioma may be mistaken for a complex pericardial effusion. Primary cardiac hemangioma and primary cardiac teratoma are other rare, multiloculated neoplasms that arise in the pericardial space in children (1).

Conclusions

Benign primary cardiac neoplasms are rare but can be well characterized and often confidently diagnosed with echocardiography, CT, or MR imaging, either alone or in combination. These neoplasms may be a source of serious morbidity and mortality but are also usually treatable. It is therefore important to be familiar with the features of these tumors and to understand the strengths and limitations of the various modalities in this context.

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