7
Imaging Cardiac Tumors

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Key Points

- Cardiac neoplasms are rare
- Metastases are the most common intracardiac tumors
- Cross-sectional imaging can help in localization and characterization of cardiac tumors
- Most literature on the role of imaging in cardiac tumors comes from retrospective case series and review articles
- Transthoracic echocardiography is generally sufficient for detecting cardiac masses and should be the first line of imaging
- MRI is critical for localization and characterization of cardiac tumors
- CT can be used in patients in whom MRI cannot be performed, such as those with pacemakers, metallic foreign body in the eyes, ferromagnetic or electronically operated stapedial implants and cerebral hemostatic clips
- CT is also the best imaging test for detecting calcification
- Imaging is also helpful in follow-up for recurrence or residual lesions

1 Introduction

Imaging of the heart has evolved rapidly in the last decade. With rapid advances in computed tomography (CT) and the introduction of multislice CT, it is possible to image the heart in less than a 10 seconds breath-hold [1]. Magnetic resonance imaging (MRI) of the heart has evolved to become the imaging modality of choice for evaluation of pericardial and cardiac tumors [2]. This chapter addresses the role of cross-sectional imaging, particularly CT and MRI, for evaluation of cardiac tumors.

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2 Classification of Tumors of the Heart

For sake of simplicity, we have classified tumors of the heart into those arising from the pericardium and those from the heart itself.

Metastatic tumors represent the most common tumors of the pericardium [3]. Most frequently, metastases occur from lung or breast cancers, lymphoma and leukemia. Autopsy studies suggest that almost one-quarter of all patients dying from cancer have pericardial metastasis. Primary neoplasms of the pericardium are exceedingly rare [4]. The most common primary pericardial tumor is the primary pericardial mesothelioma. Other primary neoplasms include benign and malignant teratoma, pheochromocytoma, angiosarcoma and fibrosarcoma [5].

Metastases are also the most common cardiac neoplasms, and may involve the heart via direct extension from local juxta-cardiac malignancies, lymphatic spread, venous extension or hematogenous spread [6]. Primary cardiac tumors are uncommon and include benign and malignant neoplasms originating from the myocardium, endocardium or cardiac valves [7-9]. Myxoma, lipoma, papillary fibroelastoma, fibroma, rhabdomyoma and hemangioma are some of the benign cardiac tumors. Malignant cardiac tumors include angiosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, intracardiac lymphoma and chondrosarcoma.

Thomas-de-Montpreville, et al. have proposed that, irrespective of patients' age, cardiac tumors may be classified as congenital tumors with spontaneous non-progressive or regressive lesions possibly needing surgery for mass effect, acquired benign tumors needing surgery for risk of thromboembolism and, finally, the remaining primary and secondary neoplasms with globally poor prognosis, but with some indications for resection nevertheless [10].

3 Clinical Presentation

Cardiac tumors are rare. Several autopsy series in unselected patients have a reported incidence between 0.0017 percent and 0.19 percent [11]. The first antemortem recognition of a intracardiac tumor was reported in 1934 although the first surgical excision of an intracardiac tumor, a left atrial myxoma, did not happen until 1955. Dr. Crafoord worked with Viking Olov Bjork and Ake Senning to improve the heart-lung machine and used it to perform the successful resection of myxoma of the left atrium, with long-term survival [12].

With remarkable growth in the use of cross-sectional imaging, particularly in CT and MRI, cardiac masses may be picked up in asymptomatic patients. However, patients with primary cardiac tumors may present with one or more symptoms from the classic triad of symptoms related to right or left ventricular outflow or inflow obstruction (such as those of congestive heart failure, atypical chest pain or palpitation), symptoms and signs of systemic thrombo-embolization (from stroke), and non-specific constitutional symptoms [5]. Clinical presentation with intracardiac obstruction and thrombo-embolization occurs in only about half of the cases [8]. In

general, signs and symptoms depend on the location of the tumor and its spread. Pericardial location or spread may be marked by pericardial pain, effusion, tamponade, constriction or predominantly atrial arrhythmias. Myocardial location or involvement may be associated with arrhythmias, conduction disturbances, heart blocks, congestive heart failure and EKG changes. Coronary involvement may present as angina pectoris or with myocardial infarction. Endocardial tumors may lead to valve obstruction, valve damage, thromboembolism and constitutional symptoms.

Treatment of intra-cardiac metastases is generally dictated by the status and extent of the primary tumor. For benign tumors surgery is the treatment of choice, whereas chemotherapy is indicated in the presence of unresectable or widespread malignancies [5].

4 Differential Diagnosis from Non-Tumorous Masses

One important consideration with imaging of a suspected cardiac or pericardial tumor is the possibility of a normal cardiac structure or a non-neoplastic mass lesion mimicking neoplasm [13].

In the pericardium lesions such as hematoma, abscess, pleuropericardialcysts, and hydatid cysts may present as masses and confound the diagnosis. Often, cross-sectional imaging modalities such as MRI (preferably) and CT scanning (as an alternative to MRI) can help in differentiating these lesions from solid pericardial masses or tumors involving the pericardium.

In the heart intracardiac thrombus, septal aneurysm, crista terminalis, prominent trabeculae or papillary muscles, hydatid cyst, abscess, vegetations, benign lipomatous hypertrophy of the interatrial septum and aneurysms can be misinterpreted as cardiac neoplasms. Most often, echocardiography, transthoracic and/or transesophageal, can help in this differential diagnosis, particularly with reference to a left atrial thrombus. MRI and CT can also help in making a decision regarding characterization of such non-neoplastic cardiac masses. MRI is particularly useful in accurate characterization of ventricular thrombi, as thrombi usually do not show contrast enhancement after gadolinium administration on first pass or delayed MRI [14]. Post-contrast MRI in such cases can also show the underlying myocardial scar and wall motion abnormality, and help in differential diagnosis from cardiac tumor. However, at times, artifacts from slow flowing blood may also simulate cardiac masses on MRI.

5 Imaging of Cardiac Tumors

Cross-sectional imaging techniques such as echocardiography, CT and MRI can help in characterizing some tumors based on the patient's age, medical history, location of the tumor, tumor extension, morphology and mobility, attenuation value or signal intensity, and contrast enhancement pattern [15-20].

Cross sectional imaging techniques such as echocardiography, CT and MRI provide information about the size, shape, location, vascularity and mobility of cardiac tumors along with their relation and local extension to adjoining cardiac and non-cardiac thoracic structures. CT and MRI can also aid in the detection and evaluation of primary extracardiac malignancy with metastasis to the heart or pericardium. Additionally, MRI helps in tissue characterization of the cardiac tumors and its differentiation from thrombi and normal structures simulating a tumor.

5.1 Chest Radiography and Fluoroscopy

Chest radiography is limited as an evaluation tool for cardiac tumors, as chest radiographs may be completely normal in many cases. However, chest radiographs may show a lung mass silhouetting the cardiac border and provide a clue towards a possible malignant lung mass invading the pericardium. Although not as sensitive as CT, radiographs can depict lung, lymph nodal or bone metastases of breast, lymphoma and melanoma, and corroborate with a diagnosis of metastatic deposit to the heart. In addition, tumors presenting with left-sided obstructive physiology can be assessed for pulmonary venous hypertension (redistribution of pulmonary vascularity, interstitial or alveolar edema), and those presenting with right-sided obstructive physiology can be assessed for signs of systemic venous hypertension (superior vena cava and azygos vein enlargement) on chest radiography.

Barium swallow with fluoroscopy provides valuable information about suspected esophageal cancer invading the pericardium. It is relatively contraindicated, however, in the presence of suspected tracheo-esophageal fistula or broncho-esophageal fistula secondary to tumoral extension.

5.2 Catheter Angiography

Catheter angiography has a limited role in imaging of patients with suspected cardiac tumors and has been replaced by echocardiography, MRI and CT in most instances. Major limitations of the technique include inability to directly visualize or characterize the tumor, risk of tumor embolization and the possibility of missing small or intramural tumors. Since tumors are not directly visualized on catheter angiography, it is generally not possible to differentiate non-neoplastic cardiac lesions, such as thrombi and cysts, from neoplasms. On the other hand, catheter angiography can provide information on the vascular supply to the tumor and the status of coronary circulation. Some authors suggest that coronary angiography may be done when patients are older than 40 years of age, have risk factors for coronary artery disease or suspected involvement of the coronary arteries [11]. The most important role of cardiac catheterization in cardiac tumors is that it may be used for performing biopsy.

5.3 Echocardiography

Transthoracic echocardiography is an important evaluating modality for cardiac tumors. It is a widely available and relatively inexpensive way of evaluating cardiac tumors in an accurate, real-time manner. Generally, in most cases, information about tumor size, location, extent, mobility and its relation to adjacent structures can be obtained with echocardiography [21]. Additionally, cardiac valves and functions can also be assessed. Transesophageal echocardiography can help in evaluating cardiac tumors, especially those in the left atrium.

Limitations of transthoracic echocardiography include the lack of tumor characterization and suboptimal quality in large patients, and in those with chronic obstructive pulmonary disease. Evaluation of pericardial tumors and other intrathoracic tumors involving the heart or the pericardium is also limited with echocardiography.

5.4 Magnetic Resonance Imaging

MRI has established a unique position in imaging of the heart and pericardium. It is often performed to confirm a mass seen on echocardiography and provide further information about the mass. MRI helps in accurate evaluation of cardiac and pericardial tumors for their size, morphology, location, extent and relation to important structures such as valves, septum and ventricular outflow and inflow tracts. Extension into the myocardium and pericardium and involvement of lung and mediastinum can be better assessed with MRI [22, 23]. Of all the cross-sectional imaging modalities for the heart, MRI is the most valuable in terms of characterizing cardiac masses as solid or cystic, mobile or immobile, thrombus or fat-containing masses. MRI can specifically characterize benign tumors of the heart such as myxoma, lipoma, fibroma and hemangioma [24].

In addition, CINE MRI pulse sequences provide information on the mobility of the mass during the cardiac cycle. MRI also aids in the evaluation of cardiac function, in the presence of obstructive cardiac tumors. The ability to generate direct images within any cardiac plane enables interpreting physicians to evaluate the relationship between tumor and adjacent contiguous and non-contiguous cardiac and non-cardiac structures. The absence of ionizing radiation with MRI makes it more suitable for follow-up of patients with cardiac tumors, particularly for young patients with benign tumors. Contrary to echocardiography, MRI provides a much wider field of view, and is not limited by acoustic windows or large body habitus.

Massachusetts General Hospital's MRI protocol for imaging of patients with known or suspected cardiac tumors is summarized in Table 7.1.

The presence of cardiac pacemakers, metallic foreign body in the eyes, ferromagnetic or electronically operated stapedial implants and cerebral aneurysm clips are contraindications for MRI. In such circumstances, CT may provide some of the required information [25].

Table 7.1 MRI protocol for imaging of patients with known or suspected cardiac tumor used at Massachusetts General Hospital. To focus the study on the location of the mass, all relevant prior studies (CT, echocardiography, cardiac catheterization) are checked prior to the MRI. (Key: SSFP, single shot free precession; ETL, echo train length; SE, spin echo; PD, proton density; MDE, myocardial delayed enhancement; TE, echo time; FSE, fast spin echo; TI, inversion time)

Pulse-Sequence	Orientation	Comments
3-plane localizer	Axial, Coronal, Saggital	
Sequential 2D SSFP localizer	Axial	Entire chest in one breath hold
Cine SSFP	4-chamber	6 to 8 slices to cover left ventricle (varies according to tumor location)
Cine SSFP	Through mass	To cover mass if visible on prior sequences
Double IR T2 FSE	In best plane visualizing mass	4 to 5 slices; TE ~ 100-120; ETL ~ 20
PD FSE with fat sat	Use orthogonal view (2 or 4 chamber view)	
T1 SE	Axial (may change orientation depending on mass location)	
T1 SE post double dose of gadolinium (0.2 mmol/kg)	Same view as prior sequence	Look for enhancement of the mass
3D MDE (2D MDE optional)	Short axis	Determine appropriate TI time at base, repeat at apex and through mass

Gadolinium-based MRI contrast agents are relatively contraindicated in patients with advanced renal insufficiency, due to risk of nephrogenic systemic fibrosis.

5.5 CT

In the past five years, multi-detector-row CT scanning has emerged as one of the most promising modalities for non-invasive imaging of the heart and coronary vasculature [26-29]. Cardiac CT examinations are done with electrocardiographic (EKG) gating within a single-breath-hold duration [30]. Iodinated contrast agents are used to opacify the cardiac chambers and coronary arteries. EKG-gated cardiac CT studies allow evaluation of cardiac chambers and coronary vessels. CT is the investigation of choice for evaluation of juxta-cardiac malignancies invading or metastasizing to the heart or the pericardium. In patients who cannot undergo MRI due to any reason as discussed above, CT may be used to assess suspected cardiac tumors. CT is also the best imaging technique for evaluation of cardiac calcifications and may be used as a problem-solving technique (e.g., cardiac masses versus tumoral mitral annular calcification).

Although CT does provide superior spatial resolution and faster scanning, the information on tumor characterization is limited, compared to MRI. The functional cardiac information obtained from MRI is also superior to CT in terms of cardiac output and flow mapping. It is important to remember that cardiac CT studies are done with iodinated contrast agents, which are nephrotoxic in the presence of prior or existent renal dysfunction. Patients with compromised renal function secondary to widespread malignant disease, co-existing diabetes mellitus and chemotherapy may be more vulnerable to the contrast media induced-nephropathy. In general, estimated glomerular filtration rate (eGFR) is a better index of the risk of contrast-induced nephropathy than serum creatinine. Proper hydration prior to administration of the contrast injection may help in decreasing the risk of nephropathy in these patients.

6 Level of Evidence for Imaging of Cardiac Tumors

Neoplastic cardiac masses are rare entities and that may explain the lack of prospective randomized controlled trials for imaging of cardiac neoplasms. Most published studies are indeed retrospective reviews of imaging findings, case series or case reports of rare cardiac tumors [2-24, 31, 32].

7 Malignant Cardiac Tumors

Cardiac masses are more likely to be malignant if they are present on the right side of the heart, and have extracardiac extension, inhomogeneous signal intensity and associated pericardial effusion [24, 31]. In addition, evidence of a wide mural attachment, destruction of the cardiac chamber wall, invasion of the pericardium, pulmonary arteries, vein, or vena cava, involvement of two cardiac chambers and presence of multiple lesions also favor malignancy. Imaging of malignant cardiac tumors can help in detection and localization of the lesions, in assessment of their relation to adjoining critical structures, local invasion and distant metastases. The ultimate diagnosis about the exact type of malignancy frequently requires invasive procedures such as biopsy or surgery.

7.1 Metastases

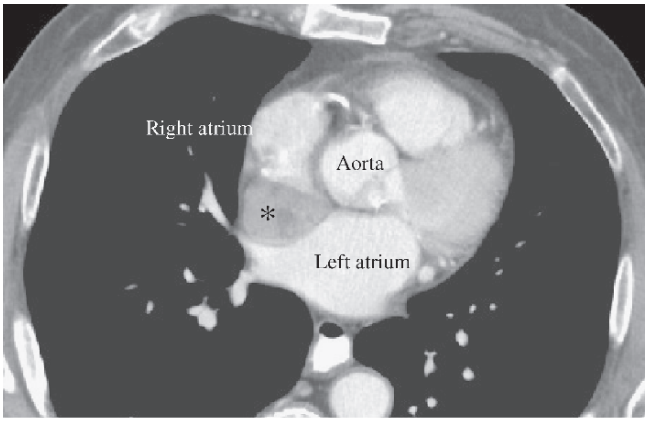
The reported incidence of cardiac metastases in different published series varies widely [11, 33]. Cardiac metastases are about 100- to 1,000-fold more common than primary cardiac tumors [34]. Cardiac metastases most frequently originate from the lungs, hemopoietic system, breast and gastrointestinal and genitourinary tracts [35]. Almost 50 percent of patients who die from melanoma have cardiac

metastases at autopsy [11]. At autopsy, almost 40 percent of patients dying from hematogenous tumors (leukaemia > lymphoma) have cardiac involvement [34].

About 90 percent of patients with cardiac metastases are clinically silent. In a patient with known malignancy development of tachycardia, arrhythmias, cardiomegaly or congestive cardiac failure, cardiac metastases must be suspected. Two-thirds of cardiac metastases occur in the pericardium, one-third in the myocardium and about 5 percent in the endocardium. Most patients with pericardial metastasis clinically present with a pericardial effusion or pericarditis. Pathologically, pericardial metastases may appear as massive infiltration of the pericardium, fibrinohemorrhagic pericarditis, pericardial infiltrate, direct invasion of pericardium or one or more pericardial nodules and masses. MRI provides the most comprehensive information about pericardial tumors, including metastases. In patients with pericardial invasion from tumors originating in contiguous structures such as lungs, mediastinum and esophagus, obliteration of the fat plane between the tumor and pericardium, pericardial enhancement and effusion are suggestive of invasion on MRI and CT. In cases of transvenous extension of tumor thrombi, for example with renal or hepatocellular cancer extending into the inferior vena cava, or lung cancer extending into the superior vena cava and pulmonary veins, both MRI and CT can provide critical information regarding the presence and extent of tumor thrombi in the veins as well as into the cardiac chambers (Fig. 7.1). In addition, both modalities allow differentiation between thrombus and tumor, as only the latter shows contrast enhancement on either imaging modality.

On the other hand, endocardial metastases can present as multiple small endocardial lesions, a large intracardiac cavitary mass, massive neoplastic thrombosis or those invading the coronary veins or arteries. The right atrium and ventricles tend to be more commonly involved with endocardial metastases [33]. These lesions have no specific CT or MRI features to differentiate them from primary malignant tumors of the heart. On MRI they have non-specific characteristics, such as low T1 signal intensity and high T2 signal intensity, with variable contrast enhancement [22]. A notable exception is melanoma metastases, which have high signal intensity on both T1- and T2-weighted MR images. Also, in patients with carcinoid syndrome, tricuspid valve disease can be evaluated with echocardiography and MRI [36, 37]. Echocardiography demonstrates specific tricuspid valve abnormalities such as thickening, shortening and decreased mobility of regurgitant leaflets, and thickening and doming in the presence of a stenotic valve [36]. MRI has a distinct advantage over CT, as it can image valve motion and valvular dysfunction [37]. This is particularly true for right-sided valves which are difficult to assess with CT. Limited valvular functional evaluation by CT is possible for the aortic and mitral valves [38, 39].

There are no specific appearances of metastases or direct extension to the heart; malignant tumors, in general, have low signal intensity on T1-weighted images and high signal intensity on T2-weighted images with varying degrees of enhancement after contrast material administration [22]. The only exception, again, is metastatic melanoma, which may be bright on both T1- and T2-weighted images due to the presence of large amounts of paramagnetic melanin.



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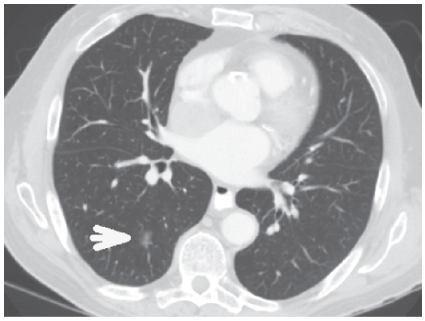


(b)

Fig. 7.1 80-year-old man with widespread metastases from malignant melanoma. Contrast-enhanced CT images show a large, enhancing, broad–based, well-defined mass (*) representing a metastatic deposit in the right atrium (**1a**, mediastinal window), and a lung metastasis (arrow) in the right lower lobe (**1b**, lung window)

7.2 Angiosarcoma

Angiosarcomas are the most common primary malignant tumors of the heart, and second most common primary cardiac tumor after myxoma (the third most common cardiac tumor is lipoma), constituting slightly more than one-third of all such tumors. These tumors present with non-specific clinical signs and symptoms, typically in the fifth and sixth decade of life, with men being two times more likely to have the tumor, compared with women, and the right atrium being the preferential location. Almost 50 percent to 90 percent of patients with angiosarcoma develop metastases to lungs, brain, bone and colon, which are quite frequently present at the time of presentation [5, 11]. Pathologically, these tumors are poorly defined, often hemorrhagic, aggressive lesions that invade contiguous structures such as vena cava

and tricuspid valve. Microscopic examination of angiosarcoma shows atypical mesenchymal cells lining the anastomotic vascular spaces. Two morphologic types of angiosarcomas have been described: a well-defined exophytic mass-like lesion of the right atrium and a diffusely infiltrative type of lesion extending along the pericardium [16].

Both CT and MRI reflect the pathologic features of angiosarcomas. On MRI, the tumor has a heterogeneous T1 signal intensity due to interspersed regions with solid tumor tissue, necrosis and hemorrhage (methemoglobin). Heterogeneous contrast uptake in the tumor is common with marked peripheral surface enhancement and little to no central enhancement corresponding to an area of necrosis. In addition, both MRI and CT can show the invasion of adjoining structures, including the pericardium and great vessels. Pericardial involvement may be depicted as discrete enhancing mass or masses, or effusion. Since lungs are the most frequent sites of metastases from cardiac angiosarcoma, a chest CT may be included in the management. There is little literature on use of fluorine-18 fluordeoxyglucose (FDG) positron emission tomography (PET) and PET-CT for evaluation of right atrial angiosarcoma, its local recurrence and detection of metastatic lesions [40, 41].

7.3 Rhabdomyosarcoma

There are two types of rhabdomyosarcoma: the embryonal type – the most common primary malignant tumor of the heart in children – and the adult type, which is more pleomorphic and less common. Contrary to other primary cardiac malignancies, these tumors have equal prevalence on either side of the heart, and are more likely to involve the cardiac valves. The epicenter of these tumors lies in the myocardium, although they frequently involve the pericardium. They frequently become quite bulky in size, measuring up to 10 cm.

On MRI the tumor is isointense to the myocardium on T1-weighted sequences with relatively homogeneous contrast enhancement, although an area of necrosis within the tumor may give rise to a more heterogeneous enhancement pattern.

7.4 Other Sarcomas: Undifferentiated Sarcoma, Osteosarcoma, Fibrosarcoma, Liposarcoma and Leiomyosarcoma

In the older literature undifferentiated sarcoma was reported to be the most common sarcoma of the heart [16]. The diagnosis of undifferentiated pleomorphic sarcoma is made after excluding other sarcomas, with appropriate use of tissue sampling and other ancillary diagnostic techniques (Fig. 7.2). In the heart the most common location of origin of the undifferentiated sarcoma, osteosarcoma or leiomyosarcoma is the left atrium, and they present most frequently with pulmonary congestion. The most common site for liposarcoma is an atrial chamber without any side preference. Liposarcoma and fibrosarcoma can also originate directly from the pericardium and invade the underlying myocardium.

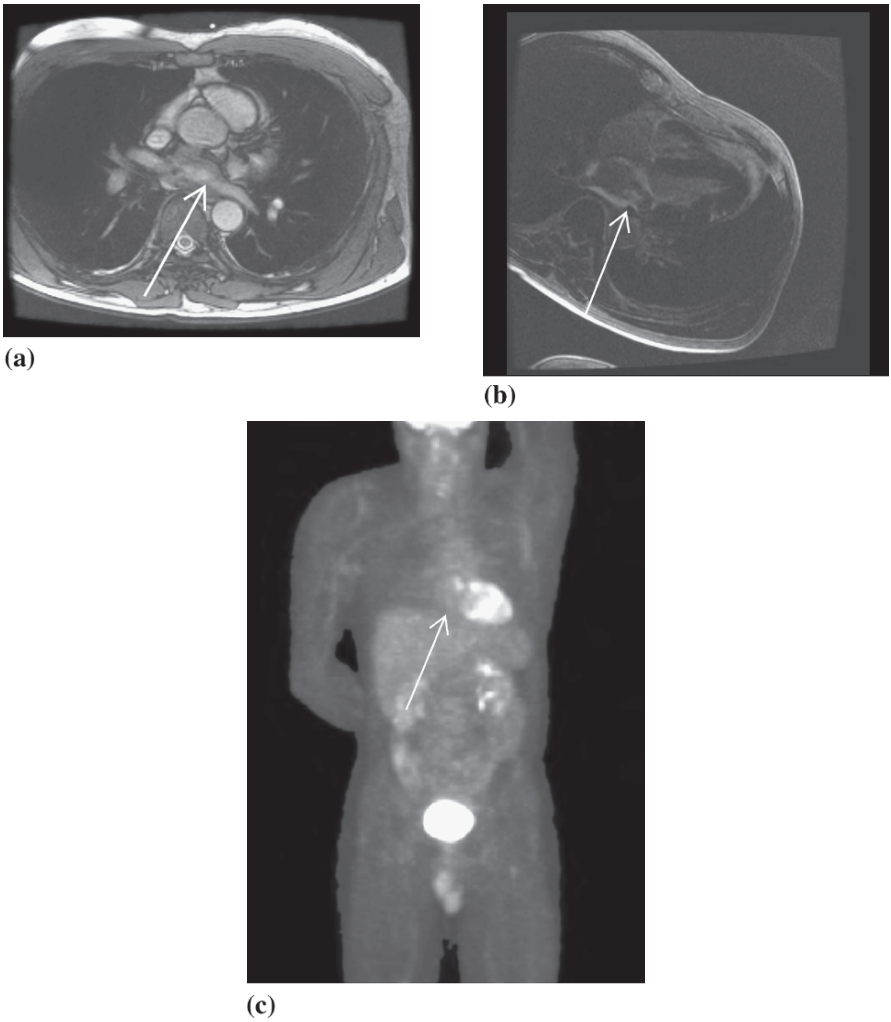


Fig. 7.2 47-year-old man with pathologically proven spindle cell sarcoma of the left atrium. T2- (**2a**) and post-contrast T1- (**2b**) weighted MR images show an ill-defined infiltrating mass in the superior portion of the left atrium involving the ostium of left superior pulmonary vein (arrow). A three-dimensional projection PET image (**2c**) shows an area of increased glucose uptake in the area corresponding to left atrium (arrow). (Note: the left ventricle shows variable uptake under normal circumstances). (Ao, aorta; LA, left atrium; RVOT, right ventricle outflow tract)

These five sarcomas do not have any specific gross pathology features and may show occasional areas of hemorrhage. Whereas calcification on CT may help in suggesting the diagnosis of osteosarcoma, identification of macroscopic fat in an invasive or aggressive tumor on CT or MRI may help confirm a diagnosis of liposarcoma. The role of imaging, however, is not to diagnose the specific cell type of these tumors, but to help in determining the anatomical limits of the tumors (local spread to cardiac, pericardial and mediastinal structures, and distant metastases),

their functional implications on the heart (valvular motion, venous return, cardiac output) and follow-up after surgery or chemotherapy.

7.5 Primary Cardiac Lymphoma

Up to 20 percent of patients with widely disseminated malignant lymphoma have cardiac or pericardial involvement on autopsy [42]. Primary cardiac lymphomas are rare in immunocompetent patients, and represent 1.3 percent of all cardiac tumors [42]. However, its incidence is increasing due to acquired immunodeficiency syndrome (AIDS) and patients who have received transplantations [11]. There is some controversy regarding the definition of primary cardiac lymphoma. Some require complete absence of lymphoma outside the pericardium on autopsy, while others assert that, for diagnosis, the bulk of tumor must be within the pericardium or cardiac symptoms must be present at the time of initial presentation [43]. Imaging, particularly CT and MRI, plays a vital role in staging of the tumor, as well as defining the extent of spread of the tumor. These tumors are more common on the right side of the heart, with right ventricle and right atrium being the most common sites of origin. They frequently invade the pericardium. Both MRI and CT have a distinct advantage over echocardiography due to a wider field of view and the ability to image tumor extension into the pericardium, or the great vessels. On CT and MRI, primary cardiac lymphomas have non-specific features and cannot be differentiated from other malignant tumors of the heart. Like most malignant neoplasms of the heart, lymphomas are also isointense to myocardium on T1-weighted images, and heterogeneous on T2-weighted images with heterogeneous contrast enhancement.

7.6 Pericardial Mesothelioma

Primary pericardial mesothelioma is a rare malignancy which is often lethal. Most patients are males between 30 to 50 years of age [32]. A primary pericardial mesothelioma originates from mesothelial cells of the pericardium. MRI is the imaging modality of choice for demonstrating the nature and, more importantly, the extent of the tumor, and the infiltration into the cardiac wall and great vessels [44]. Most often, this tumor forms multiple coalescing masses in the pericardium, which are isointense to the myocardium on T1-weighted images and heterogeneous on T2-weighted images, with marked contrast enhancement [22]. MRI also depicts the presence of constriction of the pericardium, secondary to the malignancy [45]. These findings can help guide the surgical resection, which is more likely to be successful in the presence of a localized tumor.

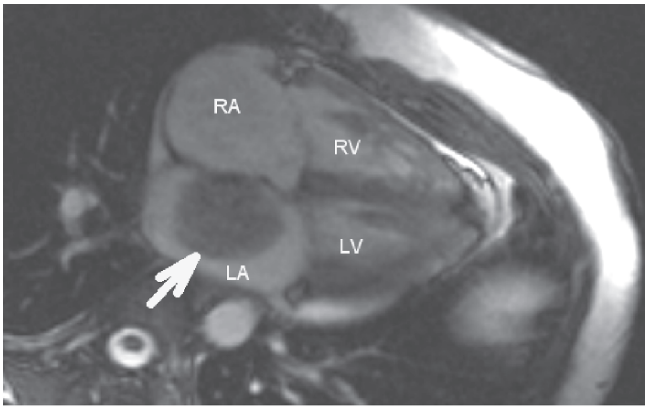
8 Benign Cardiac Tumors

Most primary cardiac tumors are benign. In contrast to the malignant tumors of the heart, benign tumors tend to have better prognosis. Surgery is the mainstay of treatment for benign tumors, whereas chemotherapy is the preferred treatment for most

malignant cardiac tumors due to the usual presence of distant metastases at presentation and the extent of local invasion. The benign tumors tend to be more frequent on the left side, particularly from the interatrial septum or roof of left atrium, although they can occur in any cardiac chamber. Likewise, a mobile or pedunculated tumor is also more likely to represent a benign tumor such as a myxoma (Fig. 7.3) or papillary fibroelastoma (Fig. 7.4). On the other hand, a broad based lesion may be a benign or malignant tumor.

Clinically the signs and symptoms of benign tumors depend on their location. Some benign tumors are related to syndromes including tuberous sclerosis (rhabdomyoma), Gorlin syndrome (fibroma) and Carney's complex (cardiac myxomas, endocrine hyperfunction and areas of skin pigmentation).

Cross-sectional imaging is important for tumor detection, as well as treatment planning. Table 7.2 summarizes the types of benign cardiac tumors along with site



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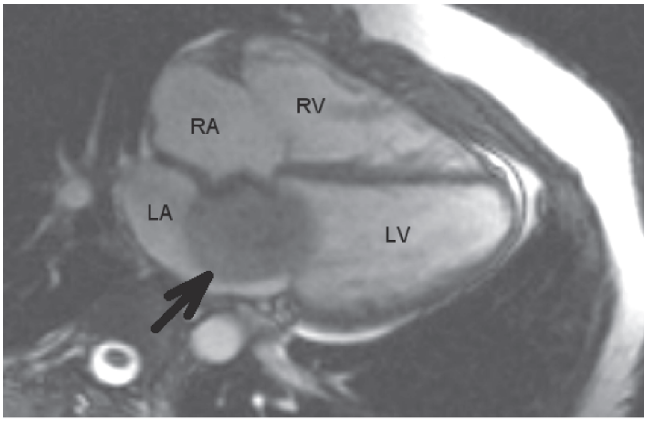

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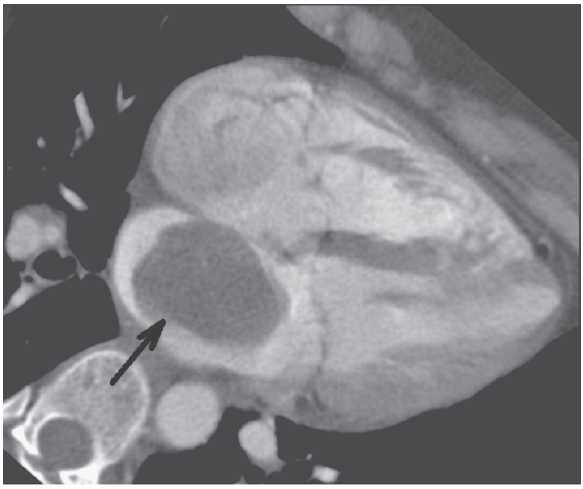

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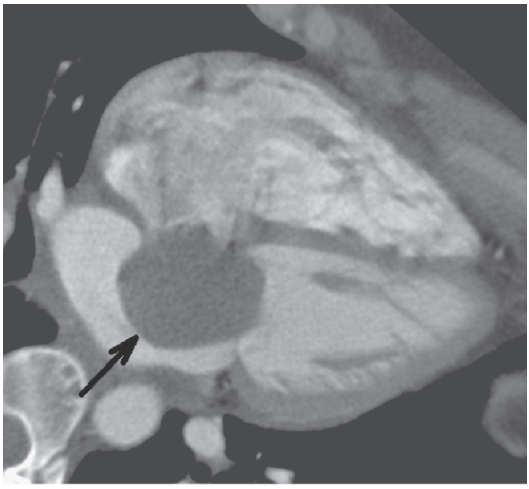

(d)

Fig. 7.3 46-year-old woman presented with worsening of shortness of breath with histopathologically proven left atrial myxoma. Cine-SSFP (steady state free precession) MR images (**3a**, ventricular systole; **3b**, diastole) and contrast–enhanced, cardiac-gated CT (**3c**, ventricular systole; **3d**, diastole) in four-chamber plane demonstrates a fairly large left atrial mass (arrow) arising from the interatrial septum and prolapsing into the mitral valve annulus in diastole (LA, left atrium; RV, right ventricle; RA, right atrium; LV, left ventricle)

preponderance, and most frequent pathologic manifestations [4, 5, 7, 8, 11]. Imaging features of these tumors are described in Table 7.3 [4, 16, 19, 22, 32, 46, 47].

Cardiac teratomas are rare primary tumors that occur most frequently in infants and children. Generally, cardiac teratoma originates from the pericardium and

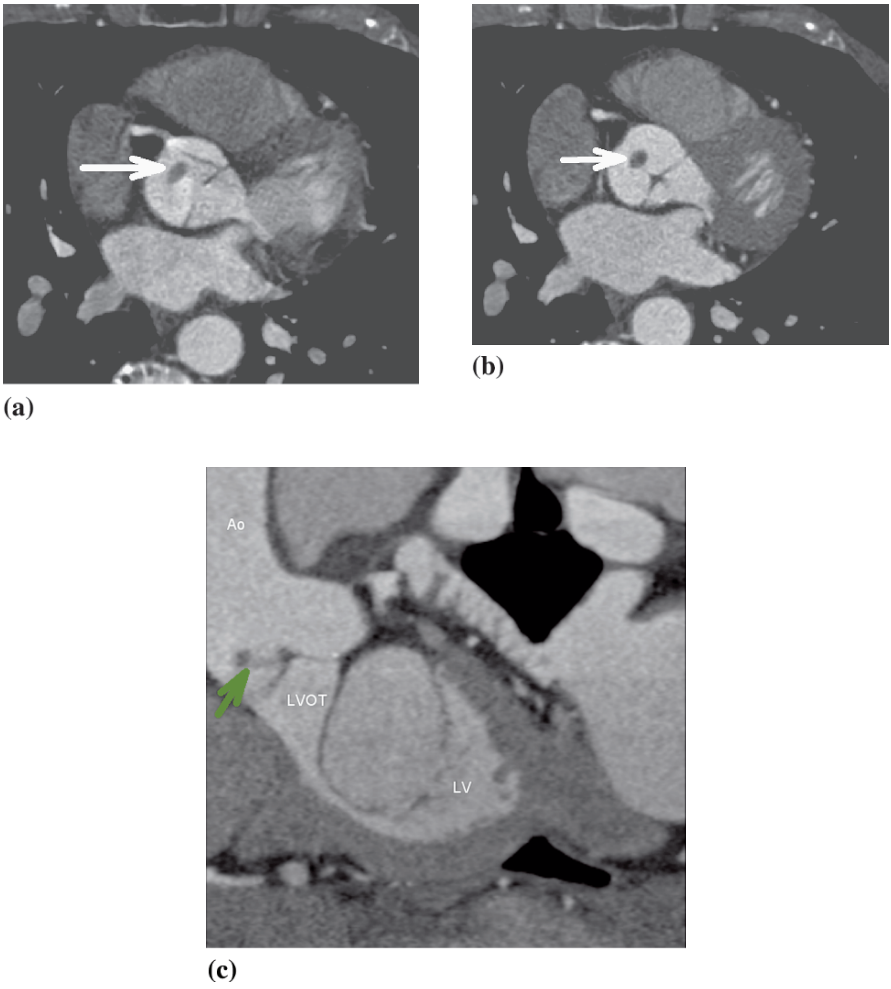

Fig. 7.4 60-year-old woman presented with transient neurologic deficit. A small pedunculated mass arising from the aortic side (contrary to vegetations, which occur on ventricular surface) of the non-coronary cusp of the aortic valve (**4a**, sytole; **4b**, diastole; **4c**, LVOT in systole) was detected on EKG-gated cardiac CT angiography. This mass was resected and turned out to be a papillary fibroelastoma (Ao, aorta; LVOT, left ventricle outflow tract)

intracardiac location is very rare [32]. As these tumors are rarely malignant, surgery is generally curative despite the frequently bulky size of the tumor. Imaging helps in detection, localization and surgical planning.

Thus, echocardiography and MRI are the mainstay imaging modalities for evaluation of benign cardiac tumors. They provide information on size, location, extent and important relations of the tumors. In patients with multiple rhabdomyomas, echocardiography can help in follow-up of the tumors for progression or regression.

Table 7.2 Pathologic features, age of presentation and most common locations of benign cardiac tumors. Figures in the parenthesis suggest mean size of the lesion [4, 5, 7, 8, 11]

Type	Age at Presentation	Most common location	Typical gross features	Calcification	Hemorrhage and necrosis	Cell type
Myxoma (5.7 cm)	30-60 years	Interatrial septum 75 percent left atrium 15-20 percent right atrium	Soft, gelatinous or firm smooth, bosselated surface	Common	Common	Myxoma cells
Papillary fibroelastoma (<1 cm)	Middle-aged, elderly	Aortic and mitral valvular endocardium	Resembles a fuzzy ball attached to short pedicle	Rare	Rare	Hyperplastic endothelium
Lipoma	Any	Any	Broad based tumor Capsulated	Rare	Rare	Adipose tissue and muscles
Rhabdomyoma (4 cm)	Children	Left ventricle	Multiple mural masses	Rare	Rare	Enlarged vacuolated cells
Fibroma (5 cm)	Children, young adults	Left ventricle	Circumscribed firm tumors	Common	Rare	Spindle cells (fibroblasts)
Paraganglioma	30-40 years	Left atrium	Broad based infiltrative or circumscribed lesion	Rare	Common	Chromaffin cells
Lymphangioma	Children	Pericardial space	Multilocular cystic tumor	Rare	Rare	Endothelium lined thin wall spaces

Table 7.3 Imaging features of benign cardiac tumors ([#] T2 bright relative to the myocardium) [4, 16, 19, 22, 32, 46, 47]

Type	Echocardiography	CT and MRI
Myxoma	Broad based myxoma- non-specific	Narrow base supports diagnosis of myxoma
	Narrow stalk, mobility and distensibility - are diagnostic	Heterogeneous on T1-weighted MR, Bright on T2[#], and shows heteroegeneous contrast enhancement
Papillary fibroelastoma	Small, mobile, valvular masses with stalk (<1 cm)	May not be apparent.
	Stippled edge with "shimmer" at interface with blood	Small pedunculated tumor attached to the valves
Lipoma	Pericardial – Uniformly echogenic or hypoechoic or with hypoechoic areas; Intracardiac – hyperechoic	Permit unequivocal diagnosis of fat and lipoma when capsule is identified (lipomatous hypertrophy in atrial septum is dumb-bell shape from sparing of fossa ovale)
Rhabdomyoma	Homogeneous masses with non-contractile myocardium	Hyperintense on T1 and T2 images, strong enhancement
		Cine MR or CT: Non-contractile area
Fibroma	Large, noncontractile, solid mass in a ventricular wall	CT: Low attenuation lesion with dystrophic calcification
		MRI: Hypointense on T2, isointense on T1 images
		Calcification may appear as hypointense T1 and T2 core
		Either: Little or no contrast enhancement
Paraganglioma	Echogenic mass, immobile	CT: Low attenuation mass, strong enhancement
		MRI: Very bright on T2 images, marked enhancement
Lymphangioma	Heterogeneous, internal septae, hypoechoic intrapericardial mass	CT: Low attenuation heterogeneous mass, ± internal septae
		MRI: T2 bright lesion with septations and heterogeneous enhancement

The degree of cardiac functional impairment can also be judged on echocardiography and MRI.

Summary

Cross-sectional imaging techniques such as echocardiography and MRI provide important information about cardiac masses. Echocardiography can provide information about cardiac tumors with regard to size, location and response to treatment. MRI is helpful in determining tumor extent, mobility, location, local invasion, relationship with critical cardiac and non-cardiac structures and tumor characterization. For pericardial tumors and metastatic lesions, MRI appears to be the imaging modality of choice.

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