Cardiac Tumors: Optimal Cardiac MR Sequences and Spectrum of Imaging Appearances

David H. O’Donnell1
Suhny Abbara2,3
Vithaya Chaithiraphan2
Kibar Yared2,3
Ronan P. Killeen1
Ricardo C. Cury2,3,4
Jonathan D. Dodd1,2,3

OBJECTIVE. This article reviews the optimal cardiac MRI sequences for and the spectrum of imaging appearances of cardiac tumors.

CONCLUSION. Recent technologic advances in cardiac MRI have resulted in the rapid acquisition of images of the heart with high spatial and temporal resolution and excellent myocardial tissue characterization. Cardiac MRI provides optimal assessment of the location, functional characteristics, and soft-tissue features of cardiac tumors, allowing accurate differentiation of benign and malignant lesions.

Recent technologic advances have allowed cardiac MRI to enter mainstream imaging practice for many disease entities [1, 2]. The development of increasing magnet strengths and surface coil channels, rapid k-space sampling and postprocessing techniques, and sophisticated myocardial soft-tissue characterization sequences have made cardiac MRI a powerful tool in the workup of many complex cardiac conditions [3]. Cardiac tumors are one such entity that has become highly suited to the application of cardiac MRI [4, 5]. Their overall frequency—both primary and metastatic—occurs with an estimated prevalence of 0.002–0.3% at autopsy and 0.15% in echocardiographic series [6]. Cardiac neoplasms may be categorized into primary and secondary tumors. The incidence of all primary cardiac tumors is estimated from unselected autopsy reports to be between 0.002% and 0.19% [7]. Myxomas are the most common benign primary cardiac tumors found in all surgical reports. Primary benign tumors have a good prognosis and a relatively low mortality rate from surgery (≈ 1%) [8]. Clinical symptoms are typically related to the size and location of the cardiac tumor, which may alter cardiac hemodynamics, cause embolization, or trigger cardiac arrhythmias [9]. Interleukin-6 secretion has been strongly implicated in causing many of the systemic symptoms and signs [10].

Approximately 25% of primary cardiac tumors are malignant [5, 11]. They may be clinically challenging to diagnose because of non-specific symptoms and are usually at an advanced stage when detected [12]. With sophisticated cardiac MRI, early detection and management of these tumors is improving, although cure is rare because of tumor inoperability and high recurrence rates [13]. Angiosarcomas and undifferentiated sarcomas are the two most common primary malignant cardiac tumors. Other cardiac malignancies include rhabdomyosarcoma, osteosarcoma, leiomyosarcoma, fibrosarcoma, liposarcoma, and cardiac lymphoma. Metastases are much more common than primary cardiac tumors in the adult population [13]. Autopsy studies in patients with a known primary cancer reveal a prevalence of cardiac metastases of 9.7–10.7%.

Transesophageal echocardiography is the mainstay imaging technique for cardiac tumor detection [6]. Although generally robust, it carries several well-described limitations, including operator dependence, restricted field of view (especially in patients with a large body habitus), and occasional limited imaging of the right heart chambers. Transesophageal echocardiography improves image quality considerably but is more invasive and carries a restricted field of view, with limited views of the aortic arch, inferior vena cava, and left ventricular apex. Recent developments in 3D imaging have further enhanced its role in the evaluation of cardiac masses [14]. More recently, cardiac MDCT has been developed, primarily as a tool for the evaluation of the coronary arteries [15]. Several CT technologic advances—submillimeter
detector arrays, increased rows of detectors, half-scan postprocessing algorithms, ECG gating—have resulted in improved imaging of cardiac structures, including cardiac masses [16]. Current limitations of the technique include a significant radiation dose (8–14 mSv), and lower temporal resolution compared with echocardiography and cardiac MRI. Neither echocardiography nor MDCT has as good soft-tissue contrast resolution as cardiac MRI. Thus, several consensus statements now include cardiac MRI as a primary imaging technique in the workup of cardiac tumors [1, 4].

In this article, we describe the optimal cardiac MRI protocols and provide detailed descriptions of each sequence that allow the most favorable characterization of cardiac tumors. We also illustrate the spectrum of cardiac MRI appearances of the most common benign and malignant cardiac tumors.

Benign Versus Malignant Cardiac Tumors: Useful Differentiating Features

Certain cardiac MRI features allow differentiation between benign and malignant cardiac masses (Table 1). These include border, size, location, calcification, and pericardial effusion. The border of most benign cardiac tumors is smooth and well-defined, with no irregularity or infiltration [5]. Malignant cardiac tumors tend to have more lobular, ill-defined, and invasive borders, and may be identified invading the myocardium, pericardium, and adjacent extracardiac structures [11]. Some benign tumors (myxomas) have a thin pedicle attaching their border to the myocardium, whereas malignant tumors are usually broad-based [17]. Most benign tumors are small in comparison with malignant tumors, which are usually of considerable size, sometimes almost filling an entire chamber [17]. As a general rule, most cardiac tumors involving the right atrial wall should be considered suspicious for malignancy; the right atrial wall is an uncommon location for benign tumors [11]. Large foci of calcification are a characteristic hallmark of cardiac osteosarcoma. Some benign tumors (myxomas, fibromas) may contain small foci of calcification. A pericardial effusion in the presence of a cardiac mass should be considered suspicious for malignancy.

Differentiating Cardiac Tumors From Thrombus

A common differential diagnosis for cardiac tumors is intracardiac thrombus. The detection of thrombus is important to avoid embolic events and to provide a basis for anticoagulation. Echocardiography has been

### TABLE 1: Cardiac MRI Findings of Cardiac Tumors

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Morphology</th>
<th>Most Common Location</th>
<th>Imaging Sequence</th>
<th>T2-Weighted</th>
<th>Fat Saturation</th>
<th>T1-Weighted</th>
<th>Delayed Enhancement</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Benign</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Myxoma</td>
<td>Lobular, pedicle</td>
<td>Left atrium, intracavitary, interatrial septum</td>
<td>Steady-State</td>
<td>Usually</td>
<td>High</td>
<td>No change</td>
<td>Isointense (moderate)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Free Precession</td>
<td>mobile</td>
<td></td>
<td></td>
<td>(heterogeneous)</td>
</tr>
<tr>
<td>Lipoma</td>
<td>Smooth</td>
<td>Any chamber, intramyocardial or intracavitary</td>
<td>Low</td>
<td>May be</td>
<td>Low</td>
<td>Signal dropout</td>
<td>High / minimal</td>
</tr>
<tr>
<td>Papillary</td>
<td>Smooth, pedicle</td>
<td>Valvular</td>
<td>Usually mobile</td>
<td>High</td>
<td>No change</td>
<td>Isointense / high</td>
<td>High</td>
</tr>
<tr>
<td>Fibroelastoma</td>
<td>Smooth, pedicle</td>
<td>Valvular</td>
<td>Usually mobile</td>
<td>High</td>
<td>No change</td>
<td>Isointense / high</td>
<td>High</td>
</tr>
<tr>
<td>Fibroma</td>
<td>Smooth, calcification</td>
<td>Ventricles, intramyocardial</td>
<td>Intramyocardial</td>
<td>Low</td>
<td>No change</td>
<td>Isointense / minimal</td>
<td>Intense</td>
</tr>
<tr>
<td>Rhabdomyoma</td>
<td>Smooth, broad base</td>
<td>Ventricles, intramyocardial</td>
<td>Intramyocardial</td>
<td>High</td>
<td>No change</td>
<td>Isointense / moderate</td>
<td>High</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>Lobular, broad base</td>
<td>Any chamber, intracavitary</td>
<td>Usually mobile</td>
<td>High</td>
<td>No change</td>
<td>Isointense / intense</td>
<td>None</td>
</tr>
<tr>
<td><strong>Malignant</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Angiosarcoma</td>
<td>Lobular, broad base</td>
<td>Right atrium</td>
<td>Intramyocardial</td>
<td>Heterogeneous</td>
<td>No change</td>
<td>Isointense / heterogeneous</td>
<td>Heterogeneous</td>
</tr>
<tr>
<td>Undifferentiated</td>
<td>Lobular, broad base</td>
<td>Left atrium</td>
<td>Intramyocardial,</td>
<td>Isointense</td>
<td>No change</td>
<td>Low</td>
<td>—</td>
</tr>
<tr>
<td>Sarcoma</td>
<td></td>
<td>involves valves</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Leiomyosarcoma</td>
<td>Lobular, broad base</td>
<td>Left atrium, posterior wall</td>
<td>Intramyocardial</td>
<td>High</td>
<td>No change</td>
<td>Isointense / high</td>
<td>—</td>
</tr>
<tr>
<td>Fibrosarcoma</td>
<td>Lobular, broad base</td>
<td>Left atrium, pericardium</td>
<td>Intramyocardial</td>
<td>—</td>
<td>No change</td>
<td>Isointense / —</td>
<td>—</td>
</tr>
<tr>
<td>Liposarcoma</td>
<td>Lobular, broad base</td>
<td>Left or right atrium, pericardium</td>
<td>Intramyocardial</td>
<td>—</td>
<td>Signal dropout</td>
<td>Isointense / —</td>
<td>—</td>
</tr>
<tr>
<td>Osteosarcoma</td>
<td>Lobular, calcification</td>
<td>Left atrium</td>
<td>Intramyocardial</td>
<td>High</td>
<td>No change</td>
<td>Isointense / —</td>
<td>—</td>
</tr>
<tr>
<td>Lymphoma</td>
<td>Lobular, pericardial</td>
<td>Right atrium</td>
<td>Intramyocardial</td>
<td>High</td>
<td>No change</td>
<td>Isointense / variable</td>
<td>High</td>
</tr>
<tr>
<td>Metastases</td>
<td>Smooth, lobular</td>
<td>Variable, pericardial effusion</td>
<td>Intramyocardial</td>
<td>High</td>
<td>No change</td>
<td>Low</td>
<td>High</td>
</tr>
</tbody>
</table>

Note—Dash (—) indicates not documented.
the mainstay for thrombus detection, but its prevalence varies considerably among echocardiographic studies because of modest image reproducibility and poorer spatial and soft-tissue resolution than cardiac MRI [18, 19]. Contrast-enhanced cardiac MRI allows differentiation between thrombus and surrounding myocardium because thrombus is avascular and hence is characterized by an absence of contrast uptake. Rarely, large chronic thrombi may enhance peripherally, and these cases can be diagnostically challenging [20].

Barkhausen et al. [21] studied the use of contrast-enhanced cardiac MRI for the detection of cardiac thrombus. In 24 patients with known or suspected thrombus, a 2D breathhold recovery turbo FLASH sequence was performed immediately after gadolinium injection, which detected 15 thrombi compared with only 12 by echocardiography. Thrombi appeared as low-signal-intensity filling defects in the cavity. Delayed enhanced cardiac MRI sequences also allow useful differentiation between tumor and thrombus [22]. Wein- saft et al. [23] recently used delayed enhanced cardiac MRI sequences for the detection of cardiac thrombus in 784 consecutive at-risk patients with systolic dysfunction [23]. Delayed enhanced cardiac MRI detected thrombus in 7% (n = 55) of patients compared with cine cardiac MRI in 4.7% (37 patients; p < 0.005). Delayed enhanced cardiac MRI also proved to be an excellent reference standard, providing 100% detection among five patients with thrombus verified by pathology. A further advantage is the detection of myocardial scars, which are an independent risk factor for thrombus. Thus, delayed enhanced sequences on cardiac MRI appear to be most useful for thrombus detection.

Basic Cardiac MRI Protocols for Assessment of Cardiac Masses

The cardiac MRI protocol used in tumor imaging should be specifically tailored to the mass. In our experience, it is important that the radiologist be available to review the initial images; otherwise, generic image protocols and image planes may be applied that may not optimally depict or characterize the mass. Many cardiac MRI sequences are available, but all protocols follow a basic set of generic sequences [24]. The basic protocol for cardiac tumors attempts to depict the location, size, and extent of the mass. The following section outlines the general basic set of sequences for cardiac MRI tumor evaluation. We have used technical parameters derived from a Magnetom Avanto 1.5-T scanner (Siemens Medical Solutions).

Fig. 1—Examples of sequences used for cardiac tumor assessment on cardiac MRI.

A, 52-year-old man with progressive dyspnea. Axial fast spin-echo image quickly allows identification of mass (arrow), enabling subsequent sequences to be targeted to left atrium in this case. Left atrial myxoma was proven at surgery.

B, 43-year-old woman with previously resected cutaneous malignant melanoma. Two-chamber (left), four-chamber (middle), short-axis (right) steady-state free precession images. Tumor (arrows) is accurately located using all three image planes. Presumed diagnosis is malignant melanoma metastases. (See also Fig. S1, cine images, in supplemental data at www.ajronline.org.)
through the center of the left ventricular cavity and the right ventricular costophrenic angle using similar technical parameters to the two-chamber sequence (Fig. 1C and Fig. S2, cardiac MRI at www.ajronline.org).

Five: A short-axis sequence of SSFP acquisitions is acquired along an image plane perpendicular to the interventricular septum, passing through both ventricles, and sometimes the atria, using similar technical parameters to the two-chamber sequence. Two-chamber, four-chamber, and short-axis stack SSFP sequences should be targeted specifically to encompass the tumor mass. Slice thickness and gap can be reduced (5- to 6-mm slice thickness, 0% slice gap) to obtain high-spatial-resolution images of the tumor. These sequences are particularly useful for evaluating tumors that are mobile, such as atrial myxomas (Fig. 1B and Fig. S1, cine CT, at www.ajronline.org).

Six: T2-weighted triple-inversion recovery images to assess for edema or necrosis in the mass are acquired. These are breath-hold sequences with generally 3–6 slices through the mass to assess for high signal (2,000/65; repletion time of two R-R intervals; inversion time (TI), 150 milliseconds; section thickness, 8–10 mm; matrix, 256 × 256; echo-train length, 20–32 (Fig. 1C).

Seven: T1-weighted FSE sequence before and approximately 15 seconds after the IV injection of 0.1 mmol/kg to acquire 5–8 identical transverse sections encompassing the mass (1,000/25; repletion time of one R-R interval, 8-mm section thickness, 256 × 256 matrix (Figs. 1D and 1E). An additional T1-weighted FSE saturation section across the superior vena cava (SVC) and the inferior vena cava (IVC) is often useful to reduce the signal from slowly flowing blood, although this does increase the time required.

Eight: Delayed enhanced images are acquired 7–10 minutes after contrast injection (Fig. 1E). Most centers use a bolus of 20 mL of 0.1 mmol/kg of gadolinium injected via an arm vein, either as a hand injection or by infusion pump. This should be followed by a saline bolus chaser of 20 mL. Certain vendors offer a scout T1-weighted TI sequence (24/1.1; flip angle, 50°; one slice repeated 41 times; slice thickness, 8.0 mm), which allows the optimal TI to be identified. This is then followed by the formal delayed enhanced sequence, which is a segmented, 2D double-inversion gradient-echo sequence (700/3.4; trigger pulse, 2; flip angle, 25°; average number of slices, 8–12; slice thickness, 6–8 mm; gap, 20%). The TI will vary from patient to patient and must be identified on an individual basis. If the myocardium has a “train track” appearance, then the TI is usually too early; if it has a confluent gray appearance, then it is too late. The normal TI is generally approximately 300 milliseconds, depending on the cardiac output and the time after contrast injection. The TI is adjusted as the scan progresses to allow contrast washout of the normal myocardium; the nulling time will change as this is happening. Increasing the time by 10 milliseconds every 1–3 slices is usually sufficient. Incorrect times on T1-weighted sequences will result in improperly nulled myocardium. If the TI is too high, the tumor may not be apparent in the gray myocardium. This sequence evolved primarily for the detection of myocardial infarction and has only relatively recently been applied to the imaging of cardiac masses [25, 26]. Thus, for several tumors, there are no documented appearances on delayed enhanced sequences (Table 1). In the original pathophysiologic model of acute myocardial infarction, cellular breakdown was considered an important mechanism whereby gadolinium (normally an extracellular contrast agent) could enter cells to become intracellular, resulting in an increased concentration in the necrotic myocardium [27]. In other conditions such as acute myocarditis, increased tissue edema resulting in increased extracellular space allows gadolinium to pool extracellularly, thus increasing its concentration in edematous myocardium [28]. A combination of both factors probably accounts for the delayed enhancement on cardiac MRI seen in cardiac tumors because they often have elements of necrosis and fibrosis. It is likely that local myocardial hyperemia in response to the tumor also allows an increased delivery of contrast agent, similar to other acute myocardial conditions.

O’Donnell et al.
Benign Cardiac Tumors

Cardiac Myxoma

Myxomas account for 25–50% of all primary cardiac tumors [29]. They usually occur in the third to sixth decade of life. The size varies from 1 to 15 cm. The most common locations are the left atrium (60–75%), the right atrium (20–28%) and, rarely, in both atria or ventricles [29]. Typically, myxomas arise near the fossa ovalis of the interatrial septum, although some arise from atrial free walls and, rarely, from atroventricular valve leaflets. Their contours are generally round or oval, sometimes lobulated with a smooth surface and a narrow pedicle. However, a frondlike appearance and broad-based features have been reported. Myxomas are highly mobile and frequently protrude into the respective atroventricular valve during diastole, potentially causing obstruction. Systemic or pulmonary embolization may be encountered, depending on the location of the myxoma [5]. Hemorrhage, fibrosis, and calcification are frequently identified in the tumor [30]. Recurrence after resection has been reported, especially in the familial type [30]. Most cases are sporadic, but if multiple tumors are present they may be part of a syndrome known as Carney’s triad [31]. This autosomal dominant syndrome consists of multiple cardiac myxomas, a variety of pigmented skin lesions such as blue nevi, extracardiac tumors including pigmented adenomas, breast fibroadenomas, and melanotic schwannomas. Patients with Carney’s triad are usually younger than patients with sporadic myxomas, are more likely to have myxomas outside the left atrium, and experience recurrence after resection.

On cardiac MR images, myxomas typically have a heterogeneous appearance with intermediate signal intensity on T1-weighted and higher signal intensity on T2-weighted FSE sequences (Table 1) (Figs. 1D and 2, and Fig. S2, cine CT, at www.ajronline.org) [32]. They may show high signal intensity if there are areas of subacute or chronic hemorrhage in the tumors. Cine SSFP sequences are useful to assess the mobility of the tumors and their attachment. Attachment may be impossible to identify in very large masses that occupy the entire cavity. Myxomas show moderate heterogeneous enhancement after gadolinium injection [33].

Lipoma and Lipomatous Hypertrophy of the Interatrial Septum

The two most common fat-containing lesions in the heart are cardiac lipomas and lipomatous hypertrophy of the interatrial septum. True cardiac lipomas are encapsulated, contain neoplastic fat cells, and occur at a young age. Lipomas constitute 8–12% of primary cardiac tumors. Half of lipomas arise from the subendocardial layer and are relatively small, whereas the other half are from either subepicardial or mid-myocardial layers [7]. If the tumor is large enough, it may cause symptoms from disturbed cardiac hemodynamics; there have been reports of sudden death from compressive effects on either the coronary circulation or the cardiac nerve conduction system [34].

Lipomatous hypertrophy of the interatrial septum is often found in older and overweight patients [35] (Fig. 3). Unlike true lipomas, they are not encapsulated and contain lipoblasts and mature fat cells. Because of the presence of brown fat, this entity may show increased radiotracer uptake on PET/CT scans [36]. The bilobed, dumbbell-shaped fatty mass must be greater than 20 mm in thickness and characteristically spares the fossa ovalis [34]. The lesion is rarely associated with symptoms although atrial arrhythmias are reported [37].

Lipomas usually have a homogeneous signal on cardiac MR images (Table 1 and Fig. 4). They have the same signal intensity as fat tissue—that is, high intensity on T1 and lower intensity T2-weighted FSE images. An invaluable sequence if this type of tumor is suspected is pre- and post-fat-saturated T1-weighted FSE
sequences, in which signal dropout is identified in the mass on the fat-saturation sequence, confirming a fat-containing lesion.

**Papillary Fibroelastoma**

Cardiac papillary fibroelastomas arise from endocardium, 80% of which are found on aortic or mitral valves [5]. Histopathologic features are of a papillary growth covered by a single layer of hypertrophied endothelium. The stroma is composed of a central dense amorphous fibrous core with an outer layer of loose connective tissue. Autopsy series report the incidence of cardiac papillary fibroelastomas to be 2–33%, making them the second most common primary benign cardiac tumor. From one comprehensive review, tumor size varied from 2 to 70 mm, with a mean diameter of approximately 10 mm [38]. Clinical presentations usually result from systemic embolization of the tumor or thrombi on the tumor surface [39]. Incidental detection is also common because many patients are asymptomatic. Most cases are diagnosed by echocardiography, and cardiac MRI is rarely needed except in atypical or inconclusive cases [40]. Surgical removal is recommended in symptomatic patients and in asymptomatic patients with mobile, left-sided tumors [35].

On cardiac MR images, papillary fibroelastoma typically appears as a small, round, homogeneous mass, sometimes with a small pedicle attached, usually to valvular structures (Table 1 and Fig. 5). Signal intensity is intermediate on T1- and hyperintense on T2-weighted FSE images. Fat-saturation sequences differentiate fibroelastomas from lipomas; the former do not show any signal dropout. Although lipomas are reported on valves, they rarely occur in this location [41]. Cine SSFP cardiac MRI may show a highly mobile, hypointense mass with turbulent flow signal surrounding it.

**Cardiac Fibroma**

Cardiac fibromas are the second most common congenital tumor; they are usually detected in infants and children and occasionally in young adults. Cardiac fibromas have a rare association with polyposis syndromes such as familial adenomatous polyposis and Gardner’s syndrome [42]. Typically, cardiac fibromas arise from the interventricular septum and affect the left ventricle more often than the right [43]. Those associated with polyposis syndromes appear to occur more commonly in the atria. Pathologic features are similar to those of fibromas found elsewhere in the body. Patients may present with heart failure, chest pain, arrhythmia, and sudden cardiac arrest. Because of the risk of sudden death, surgical removal is recommended, even in asymptomatic patients [32, 38].

Morphologic features include typically solitary, well-defined masses in the myocardium, usually involving the interventricular septum, often with central calcification, which helps differentiate them from rhabdomyomas. MRI findings of the tumor are characteristic, with inhomogeneous isointense or slightly hyperintense signal on T1-weighted and hypointense signal on T2-weighted FSE images compared with myocardium [32] (Table 1). A disadvantage of cardiac MRI over CT is its inability to directly visualize calcium, which may be represented by low signal areas in the tumor. If doubt exists about the nature of these tumors, cardiac MDCT may elucidate the features. There is slight or no enhancement with gadolinium injection compared with sur-
rounding myocardium because of the minimal vascular supply or cardiac fibromas (Fig. 6). After gadolinium administration, T1-weighted images may show central hypointense areas with a well-demarcated isointense surrounding shell [44].

**Rhabdomyoma**

Rhabdomyomas are the most common primary cardiac tumors in children; they are usually diagnosed in neonates [45]. Typical findings include a single or multiple masses arising in the ventricular myocardium. Although many cases are sporadic, rhabdomyomas have a strong association with tuberous sclerosis, which should be considered if multiple tumors are present. Up to 50% of index tuberous sclerosis cases will manifest this cardiac tumor. Most cardiac rhabdomyomas regress spontaneously, and surgery is therefore not routinely required unless patients develop significant symptoms from severe arrhythmias or heart failure [46].

Echocardiography is usually adequate for making the diagnosis, but cardiac MRI is useful in atypical presentations or when surgical removal is planned. Rhabdomyomas typically have a solid and homogeneous appearance that may be hypointense to myocardium on T1-weighted and slightly hyperintense on T2-weighted FSE images [47]. They typically show minimal or no enhancement with gadolinium (Table 1).

**Cardiac Hemangioma**

Cardiac hemangiomas represent 5–10% of primary benign cardiac tumors [47] and may originate from endocardium, myocardium, or epicardium. They are usually found in the ventricles, although they may occur in multiple locations. On cardiac MRI, hemangiomas have isointense or increased intensity on T1-weighted FSE images because of slow blood flow, and they are hyperintense on T2-weighted FSE images (Table 1). After gadolinium administration, images typically show intense enhancement because of the vascular nature of these tumors but the findings may be heterogeneous because of calcification and fibrous septa. A variant of hemangiomas, cavernous hemangiomas, have very slow flow and may not show significant enhancement.

**Other Benign Tumors**

Cardiac paragangliomas are extremely rare tumors arising from cardiac paraganglial (chromaffin) cells [48]. Patients may experience symptoms of pheochromocytoma because tumors may secrete active catecholamines. IV leiomyomatosis is a rare benign tumor of the smooth muscle that may extend into the right side of the heart via the IVC [49].

**Primary Malignant Tumors of the Heart**

**Angiosarcoma**

Angiosarcomas are the most common cardiac sarcomas in surgical studies [9]. They occur more commonly in middle-aged men and almost exclusively arise in the right atrium. They frequently show pericardial involvement. Other cardiac sarcomas occur mainly in the left side of the heart and are often mistaken for left atrial myxoma.

There are two main morphologic types. The first is a well-defined mass protruding into the chamber, usually the right atrium, and sparing the interatrial septum (Fig. 7 and Fig. S3, cine CT, at www.ajronline.org). The second is a more diffusely infiltrative mass extending into
the right ventricle and invading through the pericardium with pericardial thickening or hemorrhagic effusion [50] (Fig. 8). Because of the location of angiosarcomas, patients often present with right-sided heart failure or tamponade. Prognosis is poor because metastases have usually occurred at the time of diagnosis (66–89%). Because of the propensity of angiosarcomas for necrosis and hemorrhage, they typically have heterogeneous signal on cardiac MR images [11] (Table 1). On T1-weighted FSE sequences, tumors are typically low-signal, and T2-weighted FSE images typically show increased signal and central areas of hyperintensity, consistent with hemorrhage and necrosis, and areas of moderate signal intensity in more peripheral regions [51]. Because of their high vascularity, strong signal enhancement is seen after the administration of IV gadolinium.

Undifferentiated Sarcoma

Most undifferentiated sarcomas are found in the left atrium [9] (Fig. 9). They may appear as a discrete mass or irregular and infiltrative with necrosis and hemorrhage. On cardiac MR images, tumors usually appear isointense compared with myocardium on FSE images [11] (Table 1). In recent reports, the prevalence has been found to account for approximately 24% of primary cardiac malignancies [45]. Undifferentiated sarcomas have a similarly poor prognosis as angiosarcomas in adults.

Leiomyosarcoma

Leiomyosarcomas represent about 8% of cardiac sarcomas. They usually affect patients during the fourth decade. The most common site of the tumor is the posterior wall of the left atrium, and invasion of the pulmonary veins is frequently seen [52] (Fig. 10). They are usually sessile, lobulated, irregular masses and may be multiple in 30% of cases. Some may originate from the IVC, which can cause the symptoms of right-sided heart failure. MRI findings are nonspecific, with tumors showing intermediate signal on T1-weighted and increased signal intensity on T2-weighted FSE images [53] (Table 1).

Fibrosarcoma

These rare tumors represent about 5% of cardiac sarcomas in surgical series. Like osteosarcoma and leiomyosarcoma, fibrosarcoma tends to occur in the left atrium and usually manifests as congestive heart failure. Prognosis is also similar to other cardiac sarcomas. MRI findings are nonspecific, with a heterogeneous signal intensity on T1-weighted FSE images (Table 1).

Liposarcoma

Primary cardiac liposarcoma is extremely rare, accounting for fewer than 1% of cardiac sarcomas [54]. It may occur in any cardiac chamber and may show pericardial and valvular involvement. Primary cardiac liposarcomas are generally large, with areas of necrosis and hemorrhage. Liposarcomas may have little or no macroscopic fat and do not resemble benign lipomas. They appear as masses with heterogeneous high signal intensity on T1-weighted FSE images and show mild gadolinium enhancement [55] (Table 1).

Extraskeletal Cardiac Osteosarcoma

Cardiac osteosarcomas make up about 3–9% of primary malignant cardiac tumors [11]. They commonly present with pulmonary congestion because they are usually centered in the left atrium, unlike metastatic cardiac osteosarcoma (Fig. 11). They commonly contain large foci of calcification, which is their most distinguishing noninvasive imaging feature (Table 1). This may be more difficult to appreciate on cardiac MRI than CT, which can provide complementary information if intratumoral calcification is suspected. Other useful features include a broad base of attachment and aggressive invasion of local vascular structures such as the pulmonary veins. Histologically, they may be predominantly osteoblastic or may have chondroblastic or fibroblastic features in addition. All subtypes have a poor prognosis.

Primary Cardiac Lymphoma

Cardiac involvement of disseminated non-Hodgkin’s lymphoma is much more common than primary cardiac lymphoma. Almost all primary cardiac lymphomas are aggressive B-cell lymphomas, most commonly occurring in immunocompromised patients. Early diagnosis and prompt treatment with chemotherapy may improve prognosis [56]. The tumors usually arise from the right side of the heart, often the right atrium, and are accompanied by a large pericardial effusion. Occasionally, a pericardial effusion may be the only finding on MRI. Generally, tumors are large at presentation, with either diffuse infiltration of the right ventricle or multiple nodules. They may appear isointense or hypointense relative to myocardium on both T1- and T2-weighted FSE sequences and show heterogeneous enhancement after administration of gadolinium [57, 58] (Table 1, Fig. 12, and Fig. S4, cine CT, at www.ajronline.org). The FSE axial images of the thorax (Fig. 10) show homogeneous enhancement of mass (arrow) by mass. B, Four-chamber T1-weighted image after gadolinium administration shows enhancement of mass (arrow), excluding acute thrombus. Diagnosis was presumed leiomyosarcoma.
MRI of Cardiac Tumors

Cardiac Metastasis

Metastases are much more common in the adult population than primary cardiac tumors [13]. Most patients with cardiac metastases have no cardiac symptoms, and their diagnosis is generally made postmortem. Rarely, cardiac metastasis presents as the first clinical feature of malignancy. Cardiac metastasis may occur via one of four pathways: direct invasion, lymphatic extension, hematogenous spread, or transvenous extension [59]. Malignant pericardial effusion is the most common manifestation of metastasis, most likely through lymphatic spread or direct invasion from adjacent tissues (e.g., lungs, breasts, and lymph nodes). The most common metastases to the heart and pericardium are from lung cancer because of their proximity and the high prevalence of lung cancer itself [59]. The other common primary tumors that metastasize to the heart include tumors of the breast, kidney, and esophagus, as well as lymphoma, leukemia, and melanoma.

Myocardial metastases are often associated with melanoma and lymphoma and are suggestive of hematogenous spread. Renal cell carcinoma may spread via IVC tumor thrombus into the right atrium. Obliteration of a pulmonary vein may be a clue to intracardiac extension of bronchogenic carcinoma. Because metastases are rarely limited to the heart, evidence of a primary malignancy and metastases elsewhere strongly support the diagnosis of myocardial metastasis.

Most metastases show low signal intensity compared with myocardium on T1-weighted and high signal intensity on T2-weighted FSE images, with the exception of melanoma, which appears bright on T1-weighted FSE images because of the paramagnetic effects of

Fig. 11—54-year-old man with extraskeletal cardiac osteosarcoma. Four-chamber steady-state free precession image shows nodule thickening of floor of left atrium (arrows). Nodules were isointense with myocardium. Patient had multiple pulmonary nodules, biopsy of which showed metastatic osteosarcoma.

Fig. 12—54-year-old woman with chest pain, dyspnea, and palpitations. (See also Fig. S4, cine image, in supplemental data at www.ajronline.org.)

A, Four-chamber T1-weighted image shows extensive lobulated masses infiltrating right atrium, left ventricle, and right ventricle (arrows). Nodules were isointense with myocardium. Patient had multiple pulmonary nodules, biopsy of which showed metastatic osteosarcoma.

B, Four-chamber T1-weighted image after gadolinium shows enhancement of masses (arrows), consistent with tumor.

C, Short-axis steady-state free precession image shows extent of right atrium and infiltration of right atrioventricular groove (arrows). Note associated malignant pericardial effusion.

D, Short-axis T2-weighted image shows high signal in mass (arrows) that is characteristic of tumor. Cardiac lymphoma was proven at biopsy.

E, Short-axis delayed enhanced sequence after first cycle of chemotherapy shows extensive scarring of right ventricular free wall (straight arrow). Note some small foci of viable myocardium (curved arrow). During treatment, patient required multiple emergency cardioversions for ventricular tachycardia and fibrillation, probably originating from scarred right ventricular wall.
melanin (Table 1, Figs. 1 and 13, and Fig. S5, cardiac MRI, at www.ajronline.org). Almost all malignant lesions will show identifiable enhancement after gadolinium injection.

Conclusion

Recent technology advances have led to cardiac MRI entering the practice of mainstream cardiac imaging. It has become a particularly valuable tool in the workup of suspected cardiac tumors. These tumors are rare but, when present, can have devastating implications for patients, altering cardiac hemodynamics and acting as embolic or arrhythmia substrates. Some may be frankly malignant. Although echocardiography is the mainstay for tumor detection, optimal tumor characterization may be achieved with cardiac MRI, which provides an unrestricted field of view and superior soft-tissue depiction without ionizing radiation. Several cardiac MRI features have been described that are useful in differentiating benign from malignant tumors. Cardiac MRI is also accurate in differentiating tumor from cardiac thrombus. Although many cardiac MRI sequences exist, all follow a basic generic protocol, and, if followed correctly, allow optimum characterization of the anatomy and tissue features of cardiac tumors and enable accurate assessment of their functional impact.

References

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MRI of Cardiac Tumors


