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EVALUATION OF CARDIAC MASSES: THE ROLE OF CARDIOVASCULAR MAGNETIC RESONANCE

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Introduction

Evaluation of known or suspected cardiac masses is a frequent and expanding indication for referral to the cardiac magnetic resonance (CMR) laboratory. Most patients will have undergone an initial echocardiogram that raised the suspicion of an abnormality. However, echocardiography suffers from several well-described limitations: restricted field of view; incomplete assessment of an invading cardiac mass due to an unfavorable patient body habitus;[1] and limited ability to perform tissue characterization.[1] The role of CMR in this setting is well established because of its ability to obtain a wide field of view, generate high contrast and spatial resolution, and perform multiplanar imaging, allowing precise demonstration and localization of a mass.[2]

In clinical practice, CMR serves several useful purposes. First, it is able to help discriminate between a true cardiac mass and a pseudomass. Second, tissue characterization by CMR can assist in generating a differential diagnosis, and can distinguish a cardiac neoplasm (which generally will require excision) from other conditions, such as intracardiac thrombus, lipomatous hypertrophy or benign lipomas (all of which generally do not require excision). Third, even when the etiology of a mass is known, CMR can provide useful information as to the extent of invasion into cardiac, as well as extracardiac structures, and associated findings. This article provides a general overview as to how CMR may be clinically useful to the practicing cardiovascular specialist.

Pseudomasses

It is important to recognize that there are a number of normal structures or abnormal lesions that are not true masses, but can mimic a cardiac or paracardiac mass. The most common of these is a right atrial pseudotumor produced by a prominent crista terminalis, which can appear as a right atrial mass on echocardiography. A prominent Chiari network or eustacian valve can also be mistaken as a right atrial mass; these can be easily visualized by CMR, and a true mass can be excluded (Figure 1A).[3]

Extracardiac structures that can simulate cardiac pathology include a large hiatal hernia that can produce significant displacement of the atria. Additionally, the heterogeneous internal architecture in a hiatal hernia can lead to significant confusion on echocardiography.[4] CMR is readily able to discern a hiatal hernia from a true extracardiac mass (Figure 1B).

Tissue Characterization by CMR

Potentially the most robust feature of CMR in the evaluation of cardiac or paracardiac masses is its ability to characterize the composition of abnormal tissue better than any other imaging modality.[5] This is accomplished by imaging the mass using a variety of different MRI pulse sequences that are designed to allow examination for a vast number of different biological properties (e.g., T1 weighted, T2 weighted, perfusion or delayed contrast enhancement). These biologic properties can be analyzed to help narrow down a differential diagnosis as to the etiology of the mass, as shown in Table 1. One will notice that there may be significant overlap between biologic properties of different neoplasms, and therefore, decisions regarding chemotherapy or radiation treatment can generally not be made purely on the basis of CMR findings, but typically require a tissue diagnosis. There are, however,

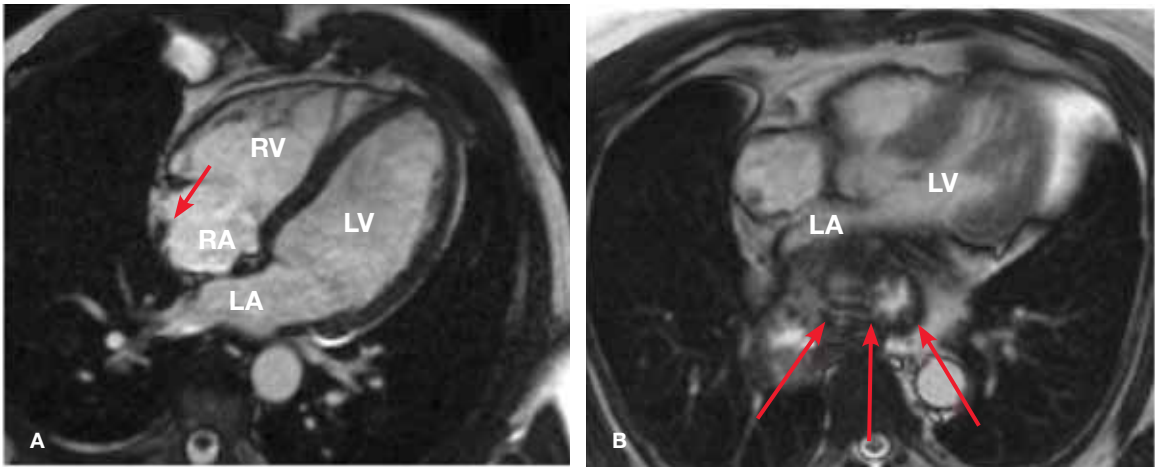

Figure 1. Typical cardiac pseudomasses that are easily identified on CMR. Frame A represents a 4-chamber view displaying a prominent crista terminalis (red arrow) — a normal structure in the right atrium that can appear as a cardiac mass on echocardiography. Frame B represents an axial image in the chest that demonstrates a large hiatal hernia (red arrows), which is compressing upon the left atrium. This can create the appearance of a left atrial mass on echocardiography. RV: right ventricle; RA: right atrium; LV: left ventricle; LA: left atrium.

Table 1. CMR characteristics of cardiac masses

Cardic Mass	T1 Weighted	T2 Weighted	Post Contrast
Myxoma	Isointense, heterogeneous	Hyperintense, heterogeneous	Heterogeneous enhancement
Papillary fibroelastoma	Isointense	Hyperintense	Hyperintense
Rhabdomyoma	Iso- or hyperintense	Slightly hyperintense	Hyperintense
Fibroma	Iso- or hyperintense	Hypointense	Hyperintense
Hemangioma	Isointense	Hyperintense, heterogeneous	Hyperintense or heterogeneous
Paraganglioma	Iso- or hypointense	Hyperintense	Hyperintense
Intravenous leiomyomatosis	Isointense	Isointense	Heterogeneous
Bronchogenic cyst	Hypointense	Hyperintense	None
Angiosarcoma	Isointense, with hyperintense areas	Iso- or hyperintense	Hyperintense
Undifferentiated sarcoma	Isointense	Isointense	Nonspecific
Rhabdomyosarcoma	Isointense	Isointense, heterogeneous	Central nonenhancing areas
Osteosarcoma	Hyperintense	Hyperintense	Nonspecific
Malignant fibrous histiocytoma	Isointense	Hyperintense, heterogeneous	Nonspecific
Leiomyosarcoma	Isointense	Hyperintense	Nonspecific
Fibrosarcoma	Isointense, heterogeneous	Hyperintense	Central nonenhancing areas
Lymphoma	Hypo- or isointense	Hyperintense	Variable

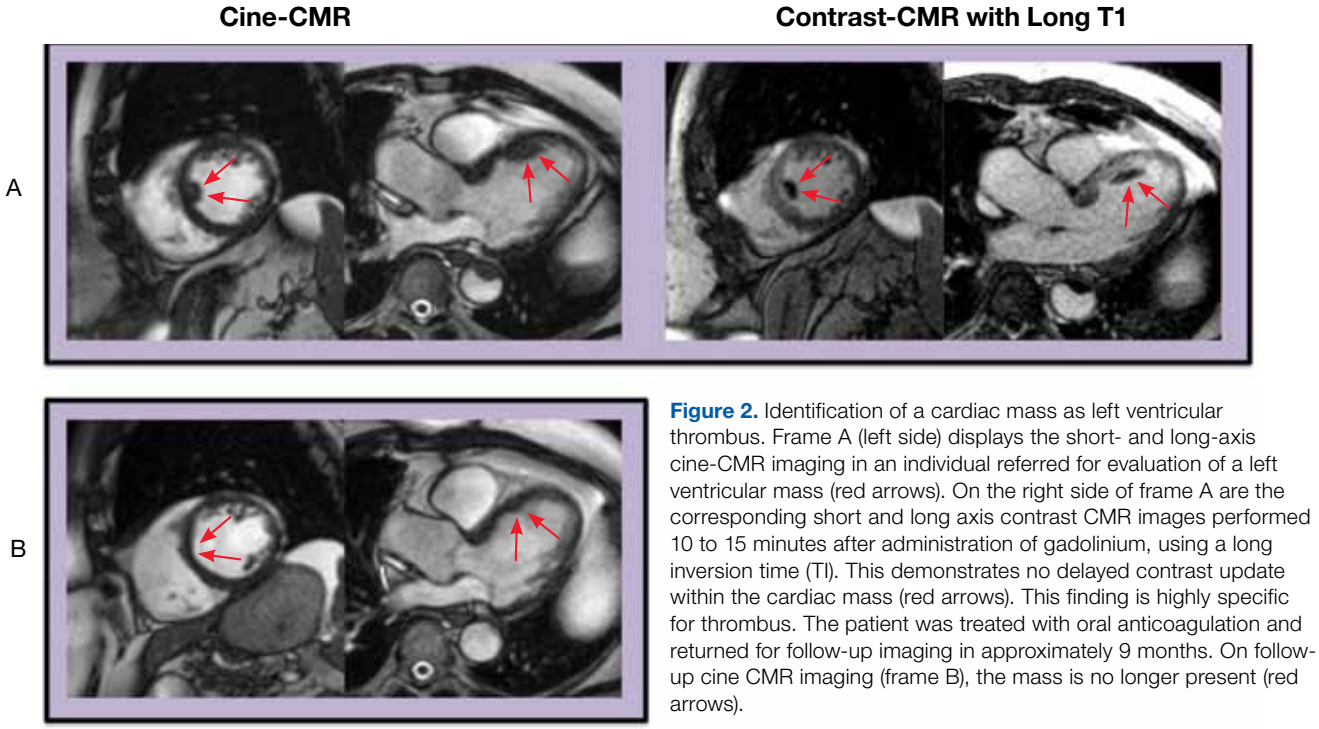

Figure 2. Identification of a cardiac mass as left ventricular thrombus. Frame A (left side) displays the short- and long-axis cine-CMR imaging in an individual referred for evaluation of a left ventricular mass (red arrows). On the right side of frame A are the corresponding short and long axis contrast CMR images performed 10 to 15 minutes after administration of gadolinium, using a long inversion time (TI). This demonstrates no delayed contrast update within the cardiac mass (red arrows). This finding is highly specific for thrombus. The patient was treated with oral anticoagulation and returned for follow-up imaging in approximately 9 months. On follow-up cine CMR imaging (frame B), the mass is no longer present (red arrows).

a number of cardiac masses that can be definitively diagnosed by CMR and management decisions made without the need for an invasive procedure. The most commonly encountered of these are intracardiac thrombus, benign lipomatous hypertophy of the interatrial septum, and benign intracardiac lipoma.

Thrombus. Although not a neoplasm, thrombus is actually the most common intracardiac mass.[3,6] It tends to occur most commonly in the left atrial appendage, but can occur within any cardiac chamber. Left atrial appendage thrombus is most commonly associated with atrial fibrillation, atrial dilation and mitral valve disease.[2] Ventricular thrombus usually occurs in the setting of cardiomyopathy, most commonly ischemic in nature. The formation of a ventricular aneurysm, or simply decreased contractility predisposed to sluggish blood flow, and can be a substrate for thrombus formation. Denudation of the endothelium from prior infarction is also a likely contributor to thrombus formation. Right atrial thrombi can develop adjacent to long-standing central venous catheters, or be "thrombi in transit" from embolization of lower-extremity thrombi.

CMR is significantly more sensitive and specific than echocardiography for detecting ventricular or atrial thrombi, with studies demonstrating an approximately twofold increase in sensitivity compared to echocardiography.[3] This sensitivity is significantly improved by the administration of intravenous contrast material. Pre-contrast cine imaging often will fail to detect ventricular thrombi that clearly are seen as low-signal intensity foci, following the administration of contrast. In addition, post-contrast delayed-enhancement inversion recovery images with a long inversion time are exquisitely sensitive for the detection of even small thrombi.[7] In this instance, the long inversion time allows recovery of signal by virtually all tissues except thrombus, which remains low in signal intensity and therefore dark on imaging (Figure 2). At the Methodist DeBakey Heart & Vascular Center, CMR diagnosis of an intracardiac mass as thrombus mitigates the need for additional invasive procedures, and patients are simply followed with serial imaging after an appropriate period of anticoagulation.

Lipomatous Hypertrophy of the Interatrial Septum. Lipomatous hypertrophy of the interatrial septum is not a true neoplasm, and it is not truly hypertrophy of the adipocytes. Rather, it represents a nonencapsulated hyperplasia of otherwise normal fatty cells within the interatrial septum. This diagnosis is based on the finding of fatty deposits in the interatrial septum, resulting in a diameter exceeding 2 cm in transverse dimension.

The exact etiology is unknown, but it appears to be associated with obesity and advanced age. The average age at diagnosis is approximately 69 years, and there appears to be a slight male predominance.[3] It is said to be associated with tachyarrhythmias, predominantly atrial in origin. The exact incidence of this disorder is

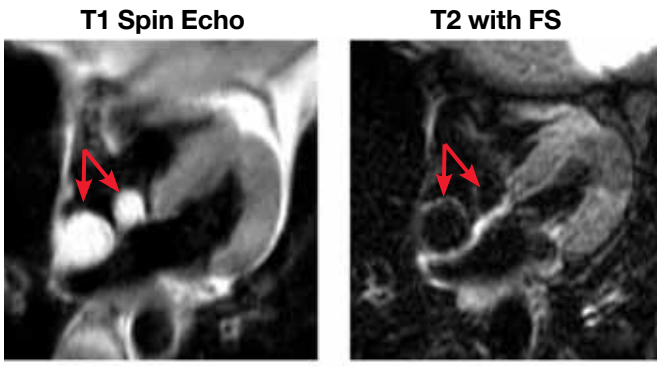

Figure 3. Lipomatous hypertrophy of the interatrial septum. Tissue characteristics of lipomatous hypertrophy are highly specific for diagnosis of this condition (see text for details). FS: fat saturation.

difficult to discern, as some series do not separate this disorder from lipomas.[3] However, it increasingly is recognized based on echocardiographic imaging and CMR.

CMR is quite specific in this disorder. Thickening of the interatrial septum to a diameter greater than 2 cm is noted, and sparing of the fossa ovalis is apparent. This often results in a dumbbell- or barbell-like appearance (Figure 3). The fatty hyperplasia results in high signal on T1-weighted images through the interatrial septum. The addition of fat saturation to the imaging sequences results in signal dropout, confirming the fatty nature of these lesions (Figure 3). In addition, cine imaging with SSFP sequences results in a characteristic chemical shift artifact at the interface between the fatty portions of the septum and the remainder of the myocardium. As CMR is highly specific for diagnosis and since the condition is benign, no further evaluation is generally necessary.

Lipoma. Cardiac lipomas are benign neoplasms composed of encapsulated mature adipose tissue, similar to extracardiac lipomas.[2, 3] There does not appear to be any sex predilection. They are usually discovered in adulthood but can occur at any age. Most originate along the epicardial surface of the heart, although myocardial or endocardial origins have been reported and can occasionally protrude into cardiac cavities.[2]

Most lipomas do not cause any symptoms, but occasionally can lead to dyspnea if there is obstruction of blood flow, and/or to arrhythmias if there is involvement of the cardiac conduction system.[8-10] In the absence of symptoms, most cardiac lipomas do not warrant surgical excision and, as such, appropriate noninvasive diagnosis is very important. Given their fatty nature, cardiac lipomas are high in signal intensity on T1-weighted sequences, and they show evidence of signal dropout on fat saturation sequences. CMR therefore provides a specific diagnosis of these lesions and can obviate the need for unnecessary cardiac surgery in patients with this condition.

Other Primary Benign Cardiac Tumors. Cardiac myxoma represents approximately 50% of all primary benign cardiac tumors. Myxomas occur predominantly in adulthood and do not tend to recur after complete excision, except when they are part of a familial myxoma syndrome.[11] Myxomas are always intracavitary masses and are most commonly located in the left atrium, usually attached to the fossa ovalis; a small number originate in the right atrium. Ventricular origin for a myxoma is very rare (< 2%).[3] CMR features of myxoma are described in Table 1 and an example is shown in Figure 4.

Papillary fibroelastomas are benign avascular papillomas of the endocardium, and are similar to Lambl's excrescences both histologically and in their predilection for cardiac valves; they differ in that they tend to be larger in size and away from the site of valve closure (whereas Lambl's excrescences, are by definition, at the sites of valve closure).[12] These benign lesions are usually detected incidentally by echocardiography that is performed for another indication. They rarely require excision except in instances where there are neurological symptoms, presumably on an emboli basis.[3] The

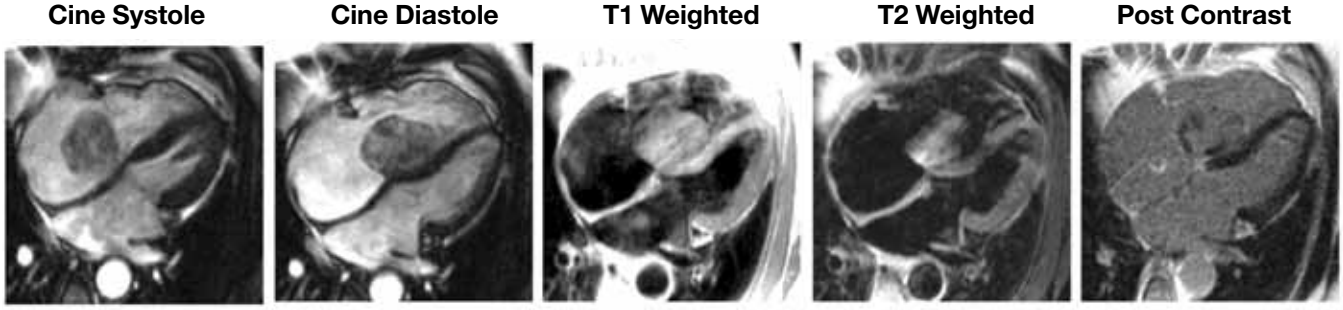

Figure 4. Example of a right atrial myxoma. Tumor is attached to the atrial septum (cine systole) but prolapses through the tricuspid valve in diastole. The mass is isointense to myocardium on T1-weighted imaging, hyperintense to mycardium on T2-weighted imaging, and demonstrates heterogeneous uptake on post-contrast imaging.

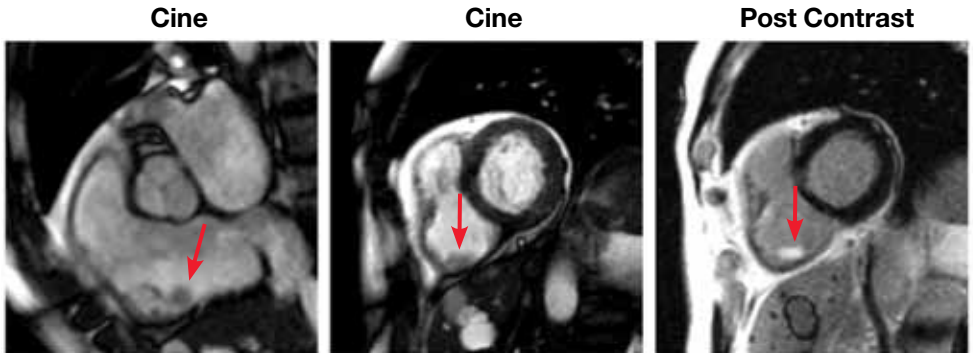

Figure 5. Papillary fibroelastoma of the posterior leaflet of the tricuspid valve (see arrow). Post-contrast imaging demonstrates intense contrast uptake by the mass.

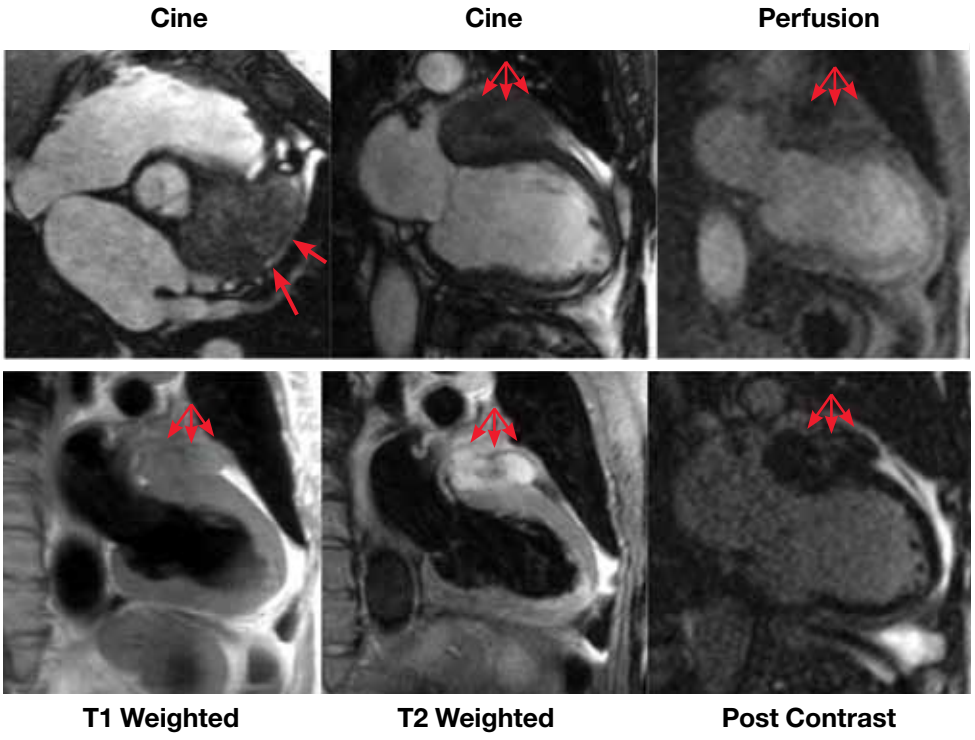

Figure 6. Paraganglioma originating at the aortic root and extending along the anterior wall of the LV in the atrioventricular sulcus. Perfusion imaging demonstrates first-pass perfusion comparable to myocardium. The mass is isointense to myocardium on T1-weighted imaging, hyperintense to myocardium on T2-weighted imaging, and demonstrates heterogeneous uptake on post-contrast imaging. LV: left ventricle.

CMR characteristics of these lesions are described in Table 1 and an example is shown in Figure 5.

Cardiac fibromas are fairly uncommon tumors that are congenital in origin and represent a discrete focal mass of collagen and fibroblasts. Because of their congenital origin, they primarily occur in children. Approximately one-third of patients present with arrhythmias, one-third with heart failure or cyanosis, and one-third are detected incidentally.[13] Their CMR features are described in Table 1; the key findings are that they demonstrate reduced signal on T2-weighted imaging (due to their limited water content) and demonstrate very high signal intensity on delayed enhancement imaging (due to their high collagen content).[14]

Paragangliomas are tumors originating from neuroendocrine cells, and typically present with symptoms of catecholamine excess (e.g., hypertension, tachyarrhythmias or heart failure). Their point of origin is typically in the atria, along the atrioventricular sulcus, or at the root of the great vessels. Imaging features are described in Table 1; key findings are of high signal on T2-weighted imaging, heterogeneous delayed hyperenhancement, and high vascularity on perfusion imaging (Figure 6).

Malignant Cardiac Tumors. Malignant tumors comprise approximately 25% of primary cardiac neoplasms.[3] Most of these are some form of sarcoma with the remainder being lymphomas. Imaging characteristics of malignant tumors are quite similar, with most lesions demonstrating invasion of surrounding

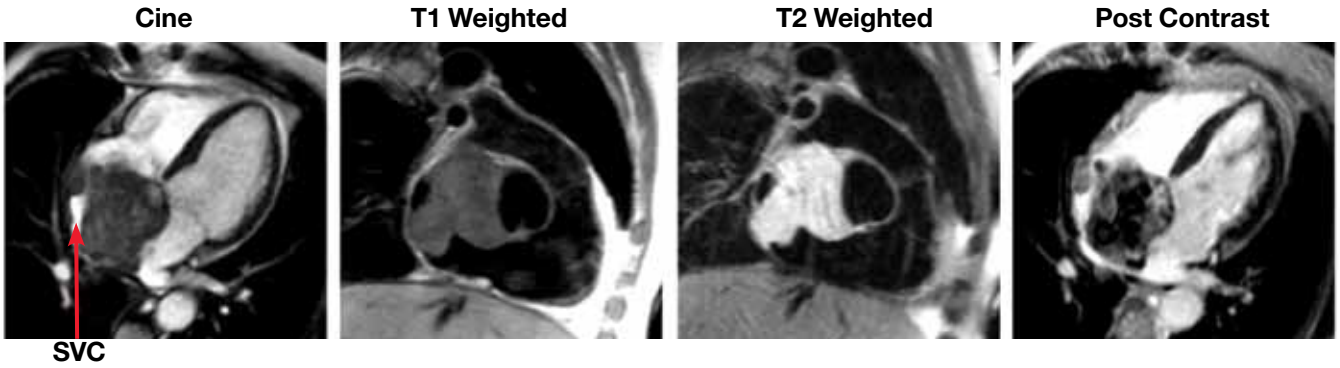

Figure 7. Right atrial angiosarcoma with invasion into the interatrial septum and partial obstruction of the SVC. SVC: superior vena cava.

structures and myocardium, poor border definition, and frequent coexisting pericardial effusion. While the exact etiology cannot be distinguished based on imaging characteristics, findings favoring malignancy usually can be detected.

Angiosarcoma is the most common form of cardiac sarcoma and accounts for approximately 40% of primary cardiac malignancies.[3] Angiosarcoma has a predilection for the right atrium with more than 90% originating at this location.[15] Location of origin is a key distinguishing feature different from most other forms of sarcomas (e.g., undifferentiated sarcomas, malignant fibrous histiocytoma, osteosarcoma, leiomyosarcoma, fibrosarcoma or rhabdomyosarcoma) that tend to arise in the left atrium.[16] As the imaging features of different forms of sarcomas are similar, specific differentiation can only be made by histology. See Figure 7 for an example of a right atrial angiosarcoma.

Primary cardiac lymphomas are distinct from systemic lymphoma with cardiac involvement. These neoplasms have increased prevalence in immunocompromised patients, but also can occur in immunocompetent patients. Clinical features are typically of dyspnea, arrhythmia, superior vena cava obstruction or cardiac tamponade due to frequent involvement in the pericardium resulting in pericardial effusions. Imaging features include frequent epicardial surface or pericardial involvement. Tissue characteristics are described in Table 1 and an example case is shown in Figure 8.

Associated Findings

Even in patients with cardiac masses where a tissue diagnosis has been made, CMR plays a role in determining the full extent of tumor involvement and associated findings. This information is useful for the surgeon planning the excision procedure. In addition to determining which cardiac chambers are involved, CMR can provide several additional, useful pieces of information that are detailed in this section.

Pericardial Effusion. The presence of a pericardial effusion in association with a cardiac mass is highly suggestive of malignancy. In fact, pericardial effusion is probably the most common imaging manifestation of metastatic disease. Additionally, the presence of nodular implants on the pericardium should be viewed as highly suggestive of metastatic disease. Lastly, CMR imaging characteristics of pericardial effusion can be useful to ascertain whether an effusion is likely transudative, exudative or hemorrhagic.

Coronary Involvement. Cardiac masses that involve the epicardium of the heart can occasionally affect the coronary arteries, leading to direct coronary invasion that results in coronary stenosis and occasionally myocardial infarction. CMR is very useful in identify-

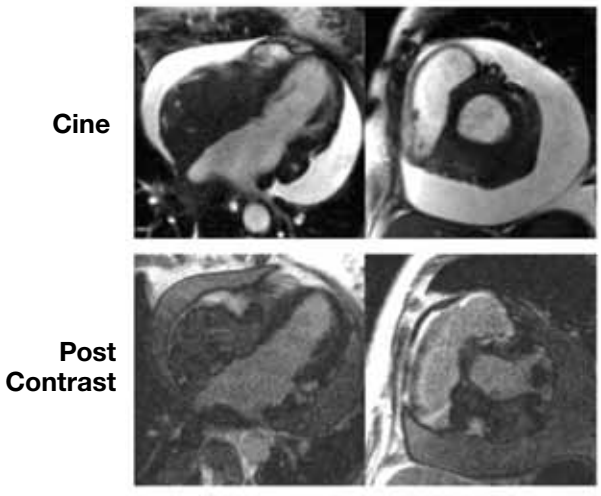

Figure 8. Patient with cardiac T-cell lymphoma. Note the typical features of tumor involvement, not only within the cardiac chamber cavities but also within the myocardium, and an associated large pericardial effusion.

ing coronary involvement, and delayed-enhancement CMR of the myocardium can identify associated myocardial infarction.[17] Identification of coronary involvement is useful in surgical planning, as these patients may require coronary excision and replacement with a free graft or an internal mammary graft.[18]

Cardiac Valve Involvement. Tumors of the heart can occasionally infiltrate into the cardiac valves. This can lead to the need to replace the affected valve either with a mechanical valve (in which case the patient will require chronic anticoagulation therapy) or a biologic valve (in which case the patient may require reoperation in 10 to 12 years). CMR can readily identify tumor infiltration into the valve leaflets or valve annulus. Even in patients who do not have valve involvement directly from tumor, cine and phase-contrast, CMR can identify coincident valve stenosis or regurgitation[19] that may warrant intervention at the time of cardiac surgery.

Conclusion

CMR has emerged as an extremely useful tool in evaluating known or suspected cardiac masses. In its ability to discriminate between true cardiac masses and pseudomasses, tissue characterization by CMR can distinguish a cardiac neoplasm (which generally will require excision) from other conditions, such as intracardiac thrombus, lipomatous hypertrophy or benign lipomas (which generally do not require excision). This can obviate the need for unnecessary surgical procedures. In the setting of biopsy-confirmed cardiac masses, CMR can assist with identifying the full extent of tumor involvement and associated features, such as pericardial effusion, coronary involvement, or cardiac valve involvement, and thus aid in surgical planning. This has led to its essential role in the evaluation and pre-surgical work up of cardiac masses.

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