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Evaluation of cardiac tumors with magnetic resonance imaging

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Abstract Primary cardiac neoplasms are rare, and are more commonly benign than malignant. However, metastases are by far the most common cardiac neoplasms. MRI allows evaluation of myocardial infiltration, pericardial involvement and/or extracardiac extension. MRI overcomes the usual limitations of echocardiography and assesses more accurately changes in cardiac function. Specific tumoral characterization is only possible in cases of myxoma, lipoma, fibroma and hemangioma. Suggestive features of malignancy are right side location, extracardiac extension, inhomogeneity in signal intensity of the tumor and pericardial effusion. The use of intravenous contrast material improves tumor characterization and depiction of tumor borders. MRI also allows differentiation of tumor from other nontumoral masses such as intracavitary tumors or fibromuscular elements of the posterior wall of the right atrium.

Keywords Heart · MR, heart · Neoplasms, heart · Abnormalities, tissue characterization

Introduction

Metastases are by far the most common cardiac neoplasms and primary cardiac neoplasms are rare [1]. Imaging, traditionally performed with sonography, allows detection and analysis of their functional effect [2]. However, dependence on an adequate acoustic window and suboptimal visualization of extracardiac extension, limits evaluation. Although EKG-gated multislice computed tomography (CT) will undoubtedly have an important role in the evaluation of cardiac masses, magnetic resonance imaging (MRI) is presently the modality of choice in evaluating cardiac tumors. Excellent contrast resolution and multiplanar capability allow optimal evaluation of myocardial infiltration, pericardial involvement and/or extracardiac extension [3].

The use of MRI in the evaluation of cardiac tumors is reviewed in this article, and the features typically asso-

Table 1 Clinical and MRI features of primary cardiac tumors

	Location	Population	T1-weighted	T2-weighted	Postcontrast	Cine-MRI	Other data
Myxoma	Interauricular septum	Female, 30–60 years	Isointense, heterogeneous	Hyperintense, heterogeneous	Low to high enhancement	Low signal	Mean size 5.7 cm, hemorrhage, calcification
Papillary fibroelastoma	Left-sided valves	80	Isointense	Hypointense	Not published	Turbulent flow	Mean size 1 cm
Lipoma	Any	Adults	Hyperintense	Hyperintense	None		Suppression with fat saturation techniques
Rhabdomyoma	Left ventricle	Children	Iso- or hyperintense	Slightly hyperintense	Strong	Noncontractile areas	Mean size 4 cm, multiplicity
Fibroma	Left ventricle	Children	Iso- or hyperintense	Hypointense	Variable		Mean size 5 cm, calcification
Hemangioma	Any	Variable	Isointense	Hyperintense, heterogeneous	Strong, heterogeneous		Small calcifications
Paraganglioma	Left atrium	30–40 years	Iso- or hypointense	Hyperintense	Strong		Paraneoplastic catecholamine syndrome
Intravenous leiomyomatosis	Right atrium	Female, 35–50 years	Isointense	Isointense	Heterogeneous	Mobile mass	Origin in IVC
Bronchogenic cyst	Interauricular septum	Adults	Hypointense	Hyperintense	None		Differential diagnosis hydatid cyst
Angiosarcoma	Right atrium	Males, 30–50 years	Isointense, with hyperintense areas	Isointense, heterogeneous	Strong	Hypointense foci	Hemorrhage, possible pericardial origin
Undifferentiated sarcoma	Left atrium	Variable	Isointense	Isointense	Nonspecific		Possible pericardial origin, infiltrative or mass-like appearance
Rhabdomyosarcoma	Any	Children	Isointense	Isointense, heterogeneous	Central non-enhancing areas		Necrosis
Osteosarcoma	Left atrium	Variable	Hyperintense	Hyperintense	Nonspecific		Calcifications
Malignant fibrous histiocytoma	Left atrium	Females, 30–40 years	Isointense	Hyperintense, heterogeneous	Nonspecific		Pulmonary veins involvement
Leiomyosarcoma	Left atrium	Variable	Isointense	Hyperintense	Nonspecific		Pulmonary veins and mitral valve involvement
Fibrosarcoma	Left atrium	Variable	Isointense, heterogeneous	Hyperintense	Central non-enhancing areas	Possible pericardial origin	Necrosis
Liposarcoma	Left atrium	Variable	Not published	Not published	Not published		Possible pericardial origin, little intratumoral macroscopic fat
Lymphoma	Right atrium	Immunocompromised patients	Hypo- or isointense	Hyperintense	Variable		No necrosis, possible pericardial origin, rare intracavitary

ciated with the different neoplasms and the clinical importance of this imaging technique are emphasized.

Tumor characterization

Although the signal intensity characteristics of benign and malignant tumors are often similar [4], MRI can be diagnostic in patients with cardiac lipomas, fibromas and angiomas [5]. MRI is also useful in determining cystic, necrotic, lipomatous and hemorrhagic components within a mass. The presence of tumor signal heterogeneity, necrosis, wide point of attachment, involvement of more than one chamber, extracardiac extension and hemorrhagic pericardial effusion are suspicious for malignancy [1, 5–7]. In a recent study the sensitivity and specificity for malignancy of these manifestations were not optimal with the more accurate predictors of malignancy being right-sided location, inhomogeneity of tumor tissue and presence of a pericardial effusion [8]. Additionally, the use of intravenous contrast agent has been reported to improve differentiation between benign and malignant tumors [9]. Table 1 shows the most important clinical and MRI features of primary cardiac tumors.

Benign cardiac tumors

Primary benign cardiac neoplasms, although rare, are more common than primary malignant cardiac neoplasms [10]. Clinical symptoms are typically related to the location of the cardiac tumor and include arrhythmias, emboli, obstruction of blood flow, and alteration of normal myocardial contractility [1, 11]. MRI is useful not only in detection and diagnosis, but also in providing information that can assist surgical treatment.

Myxoma

Myxomas are the most common cardiac tumors and account for 25–50% of all primary cardiac tumors [1, 10]. They usually occur in adults of 30–60 years of age [11] and typically arise from the interatrial septum or the fossa ovalis, although they can arise from any endocardial area [12]. They are usually located in the left atrium (75%) or right atrium (20%) and rarely in the ventricles [11]. Uncommonly, growth occurs through the fossa ovalis resulting in tumor into both atria [12]. Myxomas rarely can show malignant changes [13].

Myxomas typically manifest as intracardiac masses attached to the endocardium by a narrow pedicle although broad-based and nonmobile masses have been reported [11, 12]. The masses are usually isointense to the myocardium on T1-weighted images, hyperintense on T2-weighted images and show heterogeneous contrast enhancement (Fig. 1) [1, 14, 15]. Less commonly myxomas can be heterogeneous on both T1- and T2-weighted images due to the presence of calcification, hemorrhage or necrosis [11, 15]. Cine MRI reveals low signal intensity mobile masses, and usually allows better visualization of the endocardial attachment than T1- and T2-weighted images [15].

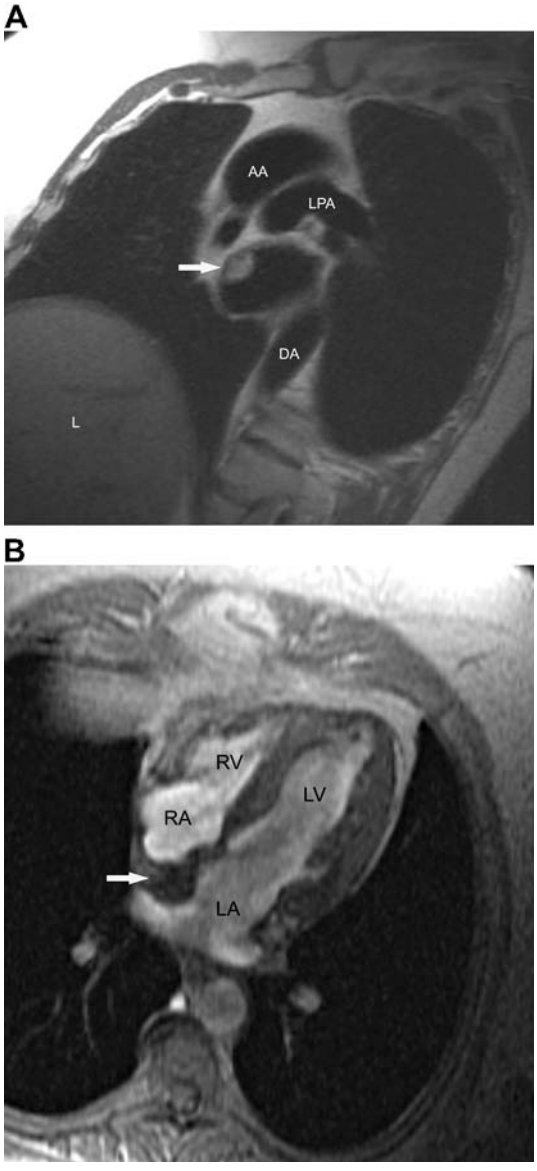

Fig. 1 Cardiac myxoma. **A** Left anterior oblique double inversion recovery FSE image shows small mass in right atrium (*arrow*). *AA* ascending aorta, *DA* descending aorta, *LPA* left main pulmonary artery, *L* liver. **B** Four-chamber steady-state free precession (SSFP–FIESTA) image shows origin from interatrial septum. Origin from interatrial septum is typical of cardiac myxomas. *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

Papillary fibroelastoma

In surgical series, papillary fibroelastomas account for 10% of all primary cardiac tumors [7]. Histologically, papillary fibroelastomas are avascular papillomas lined by endothelium [11]. Although they can originate from any endocardial surface, they are typically in the left chambers attached to the cardiac valves (>90% of cases) [11]. They are usually solitary and less than 20 mm in diameter. Most are incidentally detected and sonography can suggest the diagnosis when they arise from a valve [16, 17]. MRI can be helpful when the origin is not typical [10]. On T2-weighted images, they manifest as small hypointense masses [7]. The smaller papillary fibroelastomas may not be visualized on routine imaging sequences [11], but cine MRI can improve detection due to the presence of peri-tumoral turbulent blood flow [18].

Lipoma

Cardiac lipomas are encapsulated tumors that usually originate from the epicardium, although myocardial or endocardial origins have been reported [7]. They occur across a wide age range and patients are typically asymptomatic [11]. MRI is diagnostic as tumor signal intensity is parallel to fat on all sequences [7, 11]. Cardiac lipomas need to be differentiated from lipomatous hypertrophy of the interatrial septum. Lipomatous hypertrophy of the interatrial septum is a non-neoplastic abnormality that manifests as fat in the atrial septum that exceeds 2 cm in transverse diameter and typically spares the fossa ovalis (Fig. 2) [10, 11].

Rhabdomyoma

Cardiac rhabdomyomas are myocardial hamartomas and up to 50% occur in association with tuberous sclerosis [7]. Most are diagnosed by sonography in the prenatal period and are most frequently located in the myocardium of the ventricles [10]. Multiplicity is common, and the size of the tumors is variable [10]. Rhabdomyomas tend to disappear spontaneously although, occasionally, resection is necessary to treat arrhythmias or left ventricular outflow obstruction [7]. Multiple rhabdomyomas smaller than 1 mm (rhabdomyomatosis) can manifest as diffuse myocardial thickening [7]. Rhabdomyomas are iso- or hyperintense to myocardium on T1-weighted images and slightly hyperintense on T2-weighted images [19, 20]. The borders of the tumor are generally better depicted on proton density-weighted images and postcontrast images [15]. Myocardial tagging sequences allow identification as noncontractile myocardium [21].

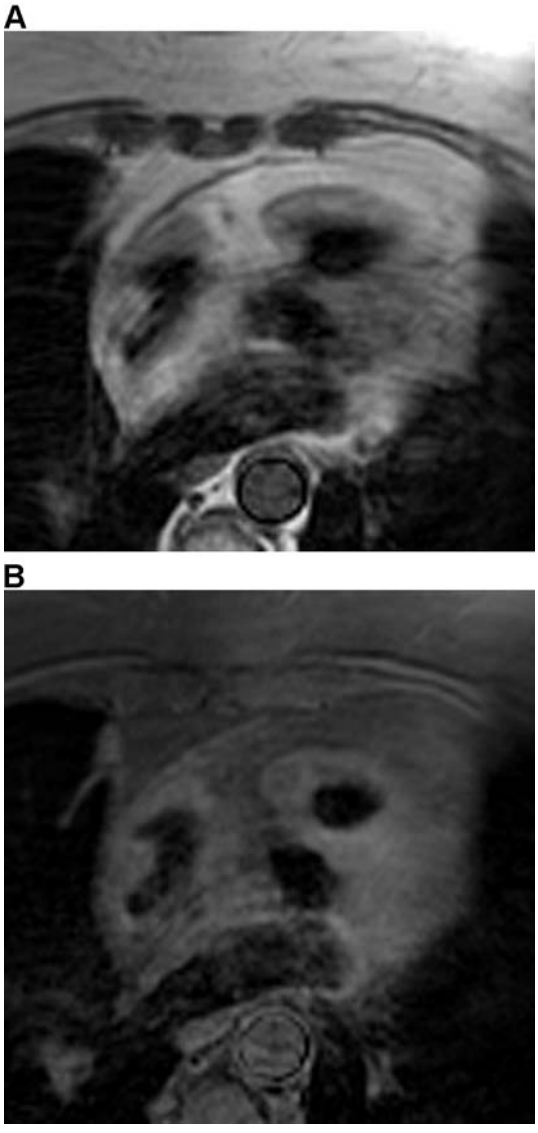

Fig. 2 Lipomatous hypertrophy of the interatrial septum. Axial T1 EPI (**A**) and axial fat-suppressed T1 EPI (**B**) show enlargement of the interatrial septum. Note that the signal characteristics of the septum are identical to those of fat in both sequences. The phase ghosting artifact is due to poor breath-holding

Fibroma

Fibromas, rare congenital tumors usually diagnosed in children under 1 year of age, have been described in adults [10]. They are fibrous masses typically located in the myocardium, most commonly in the ventricular septum or the left ventricle free wall [10]. Calcification is common, and is an important feature in differentiating fibromas from rhabdomyomas [15]. Surgical resection is usually necessary, as they can cause arrhythmias or sudden death [10]. Fibromas have a characteristic appearance on MRI. They

are iso- to slightly hyperintense compared to myocardium on T1-weighted images and hypointense on T2-weighted images due to their fibrous composition and presence of calcification [7, 15]. Fibromas have variable patterns of enhancement [7, 11, 15]. However, postcontrast images can be useful in evaluation because enhancement of compressed peritumoral myocardium allows an assessment of the tumor borders [9, 22].

Hemangioma

Cardiac hemangiomas account for 5–10% of benign cardiac tumors [7]. They are classified according to the size of their vascular channels into capillary, cavernous or venous hemangiomas [1]. They occur in any cardiac location, including the pericardium [23]. They are isointense compared to myocardium on T1-weighted images and hyperintense on T2-weighted images (Fig. 3A) [1, 7]. Cardiac hemangiomas enhance intensely with contrast and (Fig. 3B) this can be inhomogeneous because of interspersed calcification and fibrous septa within the masses [24].

Paraganglioma

Cardiac paragangliomas are rare tumors that originate from neuroendocrine cells that are usually located in the left atrium [11]. They occur more commonly in young adults and patients are typically symptomatic due to catecholamine overproduction. Cardiac paragangliomas are typically iso- or hypointense to myocardium on T1-weighted images and hyperintense on T2-weighted images [25]. Other features, such as hyperintensity on T1-weighted images (related to intratumoral hemorrhage) [11], or hypointensity on T2-weighted sequences [26], have been reported. Cardiac paragangliomas are hypervascular tumors and accordingly enhance with contrast [25].

Other benign cardiac tumors

Intravenous leiomyomas, benign smooth muscle tumors originating either from a uterine myoma or from a vessel wall, can extend into the heart [27]. They are rare neoplasms that usually occur in premenopausal women and usually manifest as a mass in the IVC that extends into the

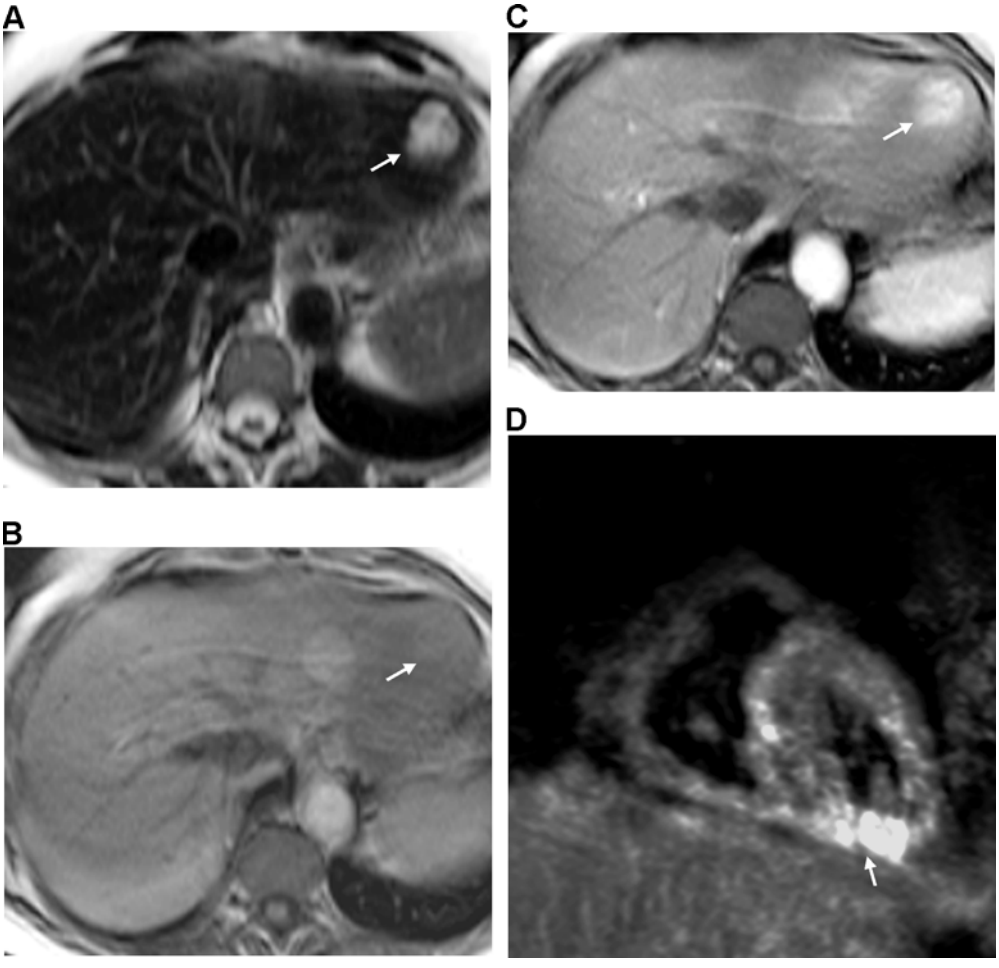

Fig. 3 Cardiac hemangioma. **A** Axial T2-weighted TSE image shows a hyperintense intramyocardial mass at the apex of the left ventricle. **B**, **C** Axial T1-weighted GE images before and after contrast administration show the mass as isointense to myocardium in the precontrast image with rapid and intense enhancement in the postcontrast image in arterial phase. **D** Coronal fat-suppressed T1-weighted TSE image shows intense enhancement of the mass

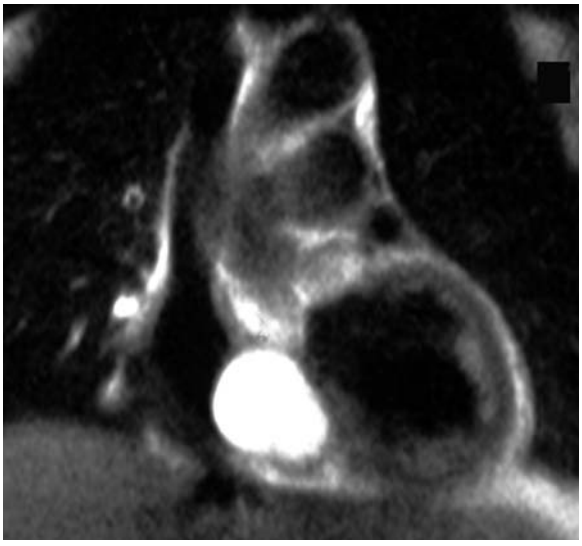

Fig. 4 Cardiac bronchogenic cyst. Coronal T2-weighted TSE image shows a well-defined hyperintense mass in the interatrial septum. Surgical resection revealed a bronchogenic cyst

right atrium [5]. Intravenous/intracardiac leiomyomas are isointense to muscle on all pulse sequences [5]. MRI, due to its large field of view, can allow accurate localization and determination of origin [27].

Cardiac bronchogenic cysts are rare, but can manifest as cystic masses in the interatrial septum (Fig. 4) [14]. Differential diagnosis includes a hydatid cyst, although these cystic masses tend to have a solid component and can be calcified [28].

Malignant cardiac tumors

Metastases are the most common cardiac malignancy and primary malignant cardiac tumors are rare [5, 29]. The most common primary cardiac malignancies are sarcomas and lymphomas [7]. Malignant cardiac tumors have a higher incidence in the right chambers and frequently show extracardiac extension [8]. MRI is the modality of choice in the evaluation of these tumors as it is superior to sonography in the evaluation of the right chambers of the heart and in the assessment of extracardiac extension [15, 30].

Sarcomas

Sarcomas are the most common primary cardiac malignancy and the second most common primary cardiac tumor after myxoma [7]. They are rare mesenchymal tumors and the most common subtype is an angiosarcoma [6]. With the exception of rhabdomyosarcomas, which usually occur in infants, they typically occur in adults and present with cardiopulmonary symptoms [7].

Angiosarcoma

Angiosarcomas usually occur in middle-aged men [6]. They are generally located in the right atrium, although they can extend to the pericardium or have a pericardial origin in patients with Kaposi's sarcoma [6]. The appearance of angiosarcomas on MRI is suggestive of the diagnosis. The masses have marked signal heterogeneity on T1- and T2-weighted images with hyperintense foci corresponding to intratumoral hemorrhage (Fig. 5) [1, 6, 15]. On GE sequences, hemorrhagic foci manifest as areas of low signal intensity. There are also flow voids within the mass on both T1- and T2-weighted images, which are hyperintense on GE, due to vascular structures [1]. This feature has been described as a "cauliflower" appearance [1, 15]. Angiosarcomas typically show diffuse and intense contrast enhancement that has been described as a "sun-ray" appearance [15].

Undifferentiated sarcoma

This group of cardiac sarcomas have no specific histological features. Their incidence has decreased with the advent of immunohistochemical staining and electron microscopy. They are usually located in the left atrium [6] and have no specific imaging features, appearing either as focal polypoid myocardial masses [31] or as infiltrating intracardiac lesions [32].

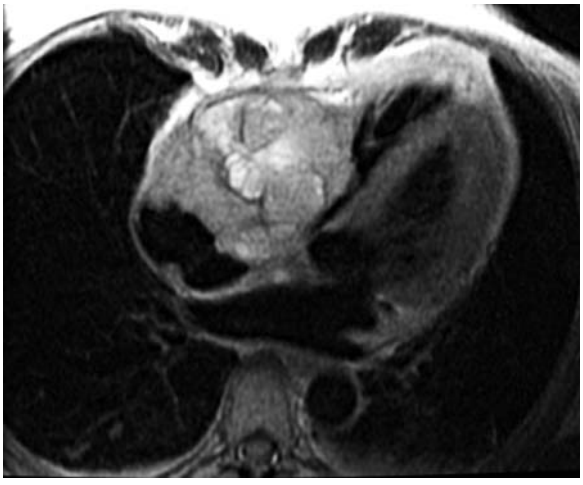

Fig. 5 Intraatrial angiosarcoma. Four-chamber double-inversion recovery FSE image shows a heterogeneous mass on the free wall of the right atrium that contains hyperintense foci corresponding to intratumoral hemorrhage. Note intracavitary extension of the mass and pericardial involvement

Rhabdomyosarcoma

Cardiac rhabdomyosarcomas usually occur in young patients, without any chamber predilection [6]. They are typically located in the myocardium, and have a tendency to arise from the valves [30]. Large areas of necrosis within the tumor and secondary involvement of the pericardium are common findings [6].

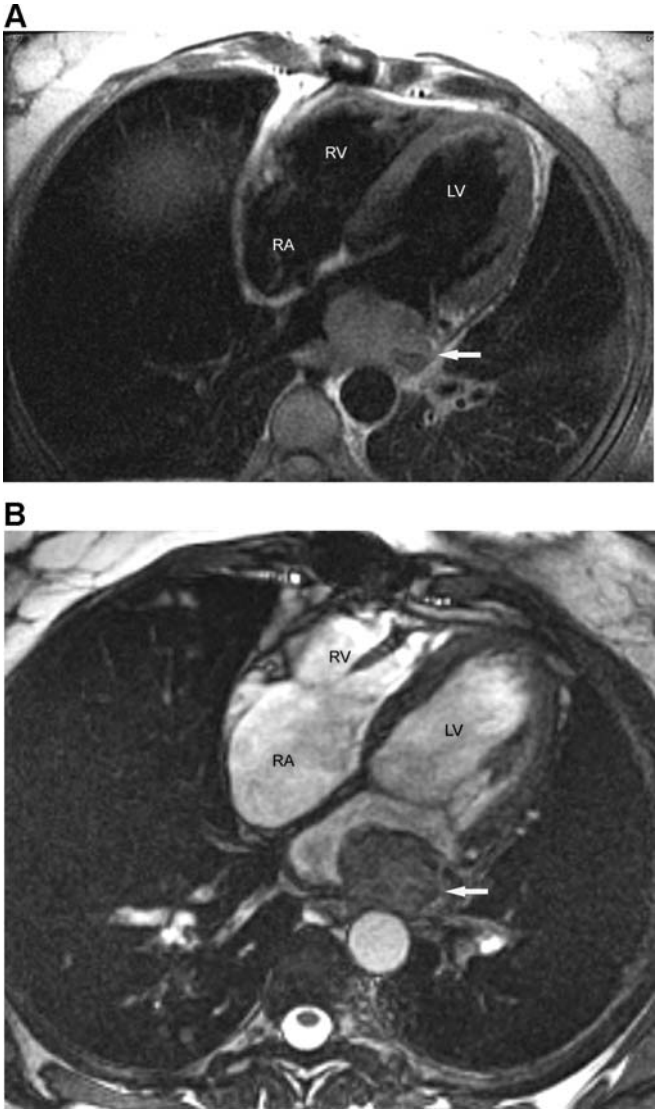

Fig. 6 Malignant fibrous histiocytoma. **A** Four-chamber double-inversion recovery FSE image shows a mass in the left atrium at the insertion of inferior left pulmonary vein (arrow). **B** Four-chamber steady-state free precession (SSFP–FIESTA) image during diastole confirms the origin from the posterior wall of atrium (*arrow*). Surgical resection revealed extension into left inferior pulmonary vein. *LV* left ventricle, *RA* right atrium, *RV* right ventricle (courtesy of Dr. Bradley S. Sabloff, University of Texas M.D. Anderson Cancer Center)

Osteosarcoma

Primary cardiac osteosarcomas are commonly located in the left atrium in contradistinction to metastatic osteosarcomas that usually involve the right atrium. CT is very useful in evaluation because it typically shows calcification [10]. The differential includes myxoma, and MRI generally allows confident differentiation of these tumors.

Other sarcomas

All of these tumors are very rare, typically arise in the left atrium and generally have a non-specific appearance on MR imaging [6]. Malignant fibrous histiocytomas occur in middle-aged women, have a predilection for the posterior wall of the left atrium and typically involve the pulmonary veins (Fig. 6) [30]. Leiomyosarcomas tend to invade the pulmonary veins or the mitral valve and rarely are calcified [6]. Fibrosarcomas can have intratumoral necrosis and a heterogeneous appearance on T1-weighted images [30]. Liposarcomas are extremely rare, can be locally invasive with pericardial or valvular involvement and rarely contain intratumoral macroscopic fat [6].

Lymphoma

Primary cardiac lymphomas are rare, and much less frequent than secondary cardiac involvement by systemic lymphomas [7]. They are usually aggressive B-cell lym-

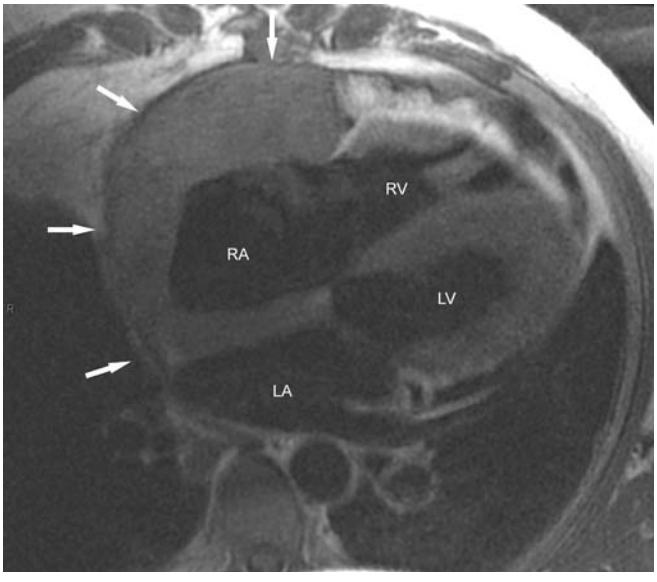

Fig. 7 Primary cardiac B-type lymphoma. Four-chamber double-inversion recovery FSE image shows mass isointense to the myocardium diffusely involving the wall of the right atrium and the interatrial septum (*arrows*). *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

phomas and there is a higher incidence in immunocompromised patients [6]. They usually arise in the right atrium and an associated pericardial effusion is common [7]. Features that can be useful in suggesting the diagnosis and differentiating these tumors from cardiac sarcomas are the absence of necrosis and less frequent involvement of the cardiac valves and intracavitary extension [10]. The masses are hypo- or isointense compared to the myocardium on T1-weighted images (Fig. 7) and iso- or hyperintense on T2-weighted images [6, 7]. Contrast enhancement varies from minimal to intense and homogeneous to heterogeneous [6].

Metastatic heart disease

Metastases to the heart and pericardium are 20–40 times more frequent than primary cardiac malignancies [30]. They occur late in the course of a malignancy and 30% of patients have symptoms related to impairment of cardiac function [29]. Cardiac metastases occur via lymphatic, hematogenous, direct/contiguous or transvenous extension [29, 30]. Lymphatic extension, particularly frequent in lung, breast and hematological malignancies, is the most common pathway that results in cardiac involvement [30] (Fig. 8A). Multiple microscopic epicardial implants follow a retrograde route through mediastinal lymphatics to the heart and typically manifest as a pericardial effusion. Although MRI allows the detection of small pericardial effusions and can indicate that the effusion is hemorrhagic, pericardiocentesis is required for diagnosis. Hematogenous myocardial or epicardial implants usually reach the heart via the coronary arteries, although dissemination through the IVC is also possible (Fig. 8B) [29, 30]. Direct extension is typical of primary lung carcinomas, but can occur with breast and esophageal carcinomas (Fig. 8C),

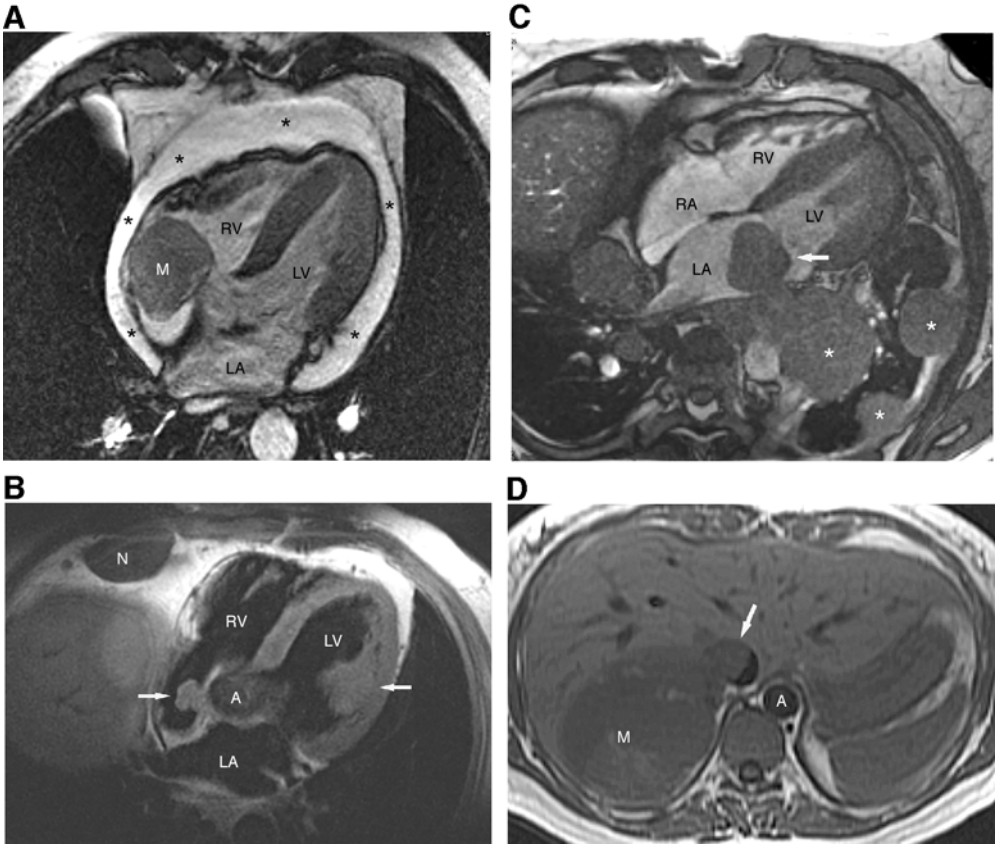

Fig. 8 Mechanisms of spread of metastases to the heart. **A** Four-chamber steady-state free precession (SSFP–FIESTA) image shows malignant pericardial effusion (*asterisks*) in a patient with melanoma. Note hematogenous metastasis and thrombus in the right atrium (*M*) (courtesy of Dr. Gregory W. Gladish, University of Texas M.D. Anderson Cancer Center). **B** Four-chamber double-inversion recovery FSE image shows masses (*arrows*) involving the wall of the right atrium and the left ventricle representing hematogenous melanoma metastases. Note right paracardiac lymph node metastasis (*N*). (courtesy of Dr. Mylene T. Truong, University of Texas M.D. Anderson Cancer Center). **C** Four-chamber steady-state free precession (SSFP–FIESTA) image shows primitive neuroectodermal tumor (PNET) of kidney metastatic to pleura (*asterisks*) with direct invasion of the posterior wall of the left atrium (*arrow*). **D** Axial T1-weighted image shows large adrenocortical malignancy (*M*) extending into inferior vena cava (*arrow*). Superiorly the mass extended into the right atrium (not shown) (courtesy of Dr. Eric P. Tamm, University of Texas M.D. Anderson Cancer Center). *A* aorta, *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

thymomas, and lymphomas [29, 30]. On MRI, obliteration of the fat plane between the tumor and pericardium is suggestive of invasion [30]. Transvenous cardiac extension via the IVC is typical of renal cell carcinomas but extension can occur via the suprahepatic veins and the IVC with hepatocellular carcinomas, cholangiocarcinomas and adrenal carcinomas (Fig. 8D) [29]. The pulmonary veins and SVC are potential pathways for intracardiac extension in patients with primary lung carcinomas. MRI allows delineation of tumor extension and depicts the extent of the tumor/thrombus in the atrium, a factor important in surgical planning. In this regard, postcontrast images can be useful in differentiating tumor from thrombus [30].

The heart can also be involved in paraneoplastic syndromes, most commonly in carcinoid heart disease which usually presents as right-sided valvular heart disease. MRI allows its accurate evaluation [33].

Conclusions

MRI allows comprehensive evaluation of cardiac tumors and often allows a diagnosis of myxoma, fibroma, lipoma and hemangioma to be suggested. Postcontrast sequences can improve tumor characterization and allow a better depiction of tumor borders. MRI has an important role in defining the intracardiac extent of malignancies, such as sarcomas and lymphomas, and in evaluating the presence and extent of extracardiac extension.

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