MRI assessment of cardiac tumours: part 1, multiparametric imaging protocols and spectrum of appearances of histologically benign lesions

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Abstract: Cardiac magnetic resonance imaging (MRI) is the reference standard technique for assessment and characterization of a suspected cardiac tumour. It provides an unrestricted field of view, high temporal resolution and non-invasive tissue characterization based on multi-parametric assessment of the chemical micro-environment. MRI exploits differences in hydrogen proton density in conjunction with T1 and T2 relaxation properties of different tissues to help differentiation normal from abnormal and benign from malignant lesions. In this article we review specific cardiac MRI techniques, tumour protocol design and the appearance of the spectrum of histologically benign tumours.

Keywords: Magnetic resonance imaging (MRI); cardiac tumour; myxoma

Submitted Nov 18, 2014. Accepted for publication Nov 19, 2014.

doi: 10.3978/j.issn.2223-4292.2014.11.23

View this article at: http://dx.doi.org/10.3978/j.issn.2223-4292.2014.11.23

Introduction

Cardiac magnetic resonance imaging (MRI) is considered the reference standard technique for characterization of a suspected cardiac mass. It provides an unrestricted field of view, high temporal resolution (30-50 ms) and non-invasive tissue characterization based on multi-parametric assessment of the chemical micro-environment (1). As such MRI is extremely helpful for diagnosis, assessment of the functional impact of a lesion, treatment planning and post treatment follow-up (2). We present a two-part review of the role of cardiac MRI in the assessment of cardiac tumours. Part one focuses on specific cardiac MRI techniques, protocol design and the appearance of histologically benign tumours. Part two covers histologically malignant tumours, and also reviews the MRI appearance of potential tumour mimics.

Primary cardiac tumours are extremely rare with an estimated lifetime incidence across all age groups of 0.0017-0.02% (3,4). Among the primary tumours approximately 75% are benign with more than half of these being myxomas (5). Histologically malignant tumours are predominantly sarcomatous in nature (6-8). By comparison, metastatic involvement of the heart is 100-500 times more frequent than primary tumours (9). Cardiac tumours are frequently asymptomatic and often discovered incidentally during evaluation of an unrelated problem or physical finding. Symptoms and signs depend on the size and location of the tumour, haemodynamic effects (chamber obstruction or embolization) and interference with cardiac conduction (heart blocks or cardiac arrhythmias). Benign primary tumours are often amenable to complete surgical resection with low mortality and morbidity and most patients have a good prognosis if surgery is successful (10). Conversely the prognosis for patients with malignant cardiac sarcomas is dismal with a median survival of only 6-12 months (11), although some may enjoy long-term survival with a radical multidisciplinary approach (12).
Most malignant lesions cannot be completely resected but patients may gain symptomatic benefit from a debulking procedure.

Most cardiac masses are not amenable to percutaneous or catheter biopsy and imaging plays a crucial role in their evaluation. Transthoracic echocardiography (TTE) is usually the first line imaging technique but is often unable to provide complete evaluation due to its limited field of view and limited soft tissue resolution (13). Transesophageal echocardiography (TOE) improves image quality and provides more detailed assessment but also has limited tissue characterization abilities. Multi-detector computed tomography (MDCT) has an emerging role in the assessment of a cardiac mass due to its high spatial resolution (0.4-0.6 mm), short examination time (especially useful in patients with orthopnoea) and ability to definitively detect fat and calcification. Currently the main disadvantage of MDCT is a lack of inherent soft tissue contrast resolution (14,15). Cardiac MRI is considered to be the gold standard technique for assessing a suspected cardiac tumour owing to its unique ability to provide non-invasive tissue characterization in conjunction with high temporal resolution and lack of exposure to ionizing radiation (16,17). In day to day practice echocardiography, MDCT and MRI are frequently used complimentary to one another to provide as much information as possible concerning a suspected cardiac tumour.

**Cardiac MRI sequences**

A variety of MRI pulse sequences can be used to confirm the presence of a suspected cardiac mass and help assess its tissue composition and impact on adjacent structures including the valves and coronary arteries. MRI exploits differences in hydrogen proton density in conjunction with T1 and T2 relaxation properties of different tissues to help differentiation normal from abnormal and benign from malignant lesions. As malignant cells have a higher free intracellular water content and a greater degree of surrounding extracellular fluid due to interstitial oedema they usually have longer T1 and T2 relaxation times compared with benign pathology (15). Malignant tissue is also associated with a greater degree of neo-angiogenesis and hence first pass contrast enhancement. Necrotic tumour tissue is associated with interstitial expansion and the accumulation and delayed washout of gadolinium based contrast agents (2).

**Black blood prepared techniques**

“Black-blood” prepared static MRI images are used to help localise a suspected cardiac or juxta cardiac mass and to non-invasively assess its tissue composition. Black blood images are usually acquired using a double-inversion recovery fast spin-echo sequence whereby the initial 180° inversion pulse is non slice selective and followed by a slice-selective 180° pulse (18). This causes blood flowing into the slice to undergo only the first non-selective 180° inversion pulse giving it a black-blood appearance. To achieve T1-weighted images the time to repeat (TR) is set at around 1,000 ms; T2-weighted images require a TR of around 2,000 ms which doubles the acquisition time. The addition of a third slice selective 180° inversion pulse (triple inversion recovery) can be used to give fat saturation which is helpful for characterisation of a fat containing lesion (19).

**Bright blood prepared techniques**

Cine steady-state free precession (SSFP) is an un-enhanced fast gradient echo technique which uses a short TR (2-3 msec) and segmented k-space filling. SSFP enables cine cardiac imaging at high temporal resolution and at the same time gives excellent contrast definition between the blood pool and myocardium. SSFP has utility for assessing the mobility and attachment point of a mass as well as its dynamic relationship and impact upon the valve structures. SSFP should not be used for assessment of tissue composition of a cardiac mass as its tissue weighting is dependent on both T1 and T2 effects (2,20).

**First pass perfusion and delayed enhancement imaging**

First pass rest perfusion imaging using a T1-weighted gradient echo sequence and infusion of gadolinium chelate followed by 10-15 minutes delayed enhancement imaging (using a T1-weighted inversion recovery sequence) assesses lesion vascularity. First pass enhancement is typical of highly vascular tumours such as haemangiomia or angiosarcoma. The presence of late phase gadolinium enhancement (LGE) implies delayed washout from a lesion, usually as the result of extracellular space expansion or necrosis (21). LGE can be seen with both benign and malignant lesions. Benign tumours like fibroma usually display uniform LGE whereas tumours with a more heterogeneous composition like myxoma or angiosarcoma (containing a mixture of tumour
tissue, necrosis and foci of haemorrhage) usually show patchy LGE (22).

**MRI tumour protocol**

At our institution cardiac MRI is performed on a 1.5 Tesla scanner (Ingenia, Philips Healthcare, Best, The Netherlands). A summary of our institutional imaging protocol for a suspected cardiac mass is presented in Table 1. Black blood images in both the axial and coronal planes are obtained with both T1- and T2-weighting and then followed by fat-suppressed triple inversion recovery images. SSFP sequences are acquired as a contiguous axial plane stack from the diaphragm to aortic arch as well as along the left ventricular short axis from base to cardiac apex. Additional 2-chamber, 4-chamber and left ventricular outflow tract plane SSFP cines are also acquired to provide optimal assessment of the valves. We then acquire a rest perfusion sequence which is planned through the centre of any mass detected in at least two planes followed by a short stack of LGE images in the same planes.

**Benign primary tumors**

The approximate frequency of benign primary cardiac tumours presenting in adulthood is presented in Table 2.
Most benign tumours are intra-cavitary with a narrow attachment point. Those that arise within the peri- or myocardium are usually well defined, with a homogeneous appearance and narrow zone of transition (23).

### Myxoma

Myxoma is the most common primary cardiac neoplasm and is believed to originate from multipotent mesenchymal cell residues within the endocardium (24). Most arise from an area immediately adjacent to the fossa ovalis with a narrow pedicle and villous surface extensions which are friable and at risk of embolization (25). The vast majority develop within the left atrium and are solitary (26-28). Multiple myxomas are rare and usually seen in association with Carney complex which is an autosomal dominant syndrome characterized by multiple myxomas (including extracardiac myxomas), schwannomas and various endocrine tumours (29,30). Mean age at presentation is 50 years with a slight female predominance (31). Clinical symptoms are wide ranging and may relate to intracardiac obstruction (often mimicking mitral stenosis), systemic embolisation (stroke) or a systemic inflammatory response (arthralgia, fevers) (32-34). Prompt surgical resection of myxoma is advocated for definite diagnosis and to reduce the risk of complications (35-39). The risk of recurrence or development of new lesions is low (estimated at 2-5%) and annual TTE follow-up is recommended for a minimum of 4-year post resection (11).

Typical MRI features are a well-defined spherical or ovoid mobile lobulated mass within either atria, sometimes prolapsing through the atrioventricular valve orifice in diastole. They usually appear hypointense to the blood pool on SSFP sequences (2). T1 and T2-weighted images usually show a heterogeneous appearance because of the combination of tumour tissue, necrotic elements and foci of haemorrhage and calcification (40-42) (Figure 1). First pass perfusion is usually absent and LGE is often multi-focal and patchy although may sometimes be homogeneous.

The main imaging differential for myxoma is thrombus. Key discriminating features which are usually reliable for distinguishing thrombus from myxoma are an absence of LGE and lack of atrioventricular valve prolapse (43).

### Papillary fibroelastoma

Fibroelastoma is the most common primary tumour of the cardiac valves although may arise from any endocardial surface (44). They are usually small lesions (<1.5 cm) and are composed of collagen and elastic tissue fibres lined by endothelium with a short pedicle (45). Commonest sites are the atrial side of the mitral valve and aortic surface of the aortic valve leaflets. The majority are asymptomatic and observed serendipitously at TTE performed for another reason but on occasion they may cause symptoms as a result of embolization of tumour fragments or surface thrombus into the systemic or pulmonary circulations (45). Aortic valve fibroelastomas can rarely be a cause of sudden death due to presumed acute obstruction of the coronary artery ostia (46). Prompt surgical resection is the treatment of choice, especially for left sided lesions (40).

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**Table 2**: Approximate frequency of both benign and malignant primary cardiac tumors

<table>
<thead>
<tr>
<th>Primary Cardiac Tumors</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign</td>
<td></td>
</tr>
<tr>
<td>Myxoma</td>
<td>45</td>
</tr>
<tr>
<td>Fibroelastoma</td>
<td>15</td>
</tr>
<tr>
<td>Lipoma</td>
<td>10</td>
</tr>
<tr>
<td>Fibroma</td>
<td>3</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>2</td>
</tr>
<tr>
<td>Cystic tumour of atrioventricular nodal region</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Paraganglioma</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Other</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Total</td>
<td>75</td>
</tr>
<tr>
<td>Malignant</td>
<td></td>
</tr>
<tr>
<td>Angiosarcoma</td>
<td>10</td>
</tr>
<tr>
<td>Sarcomas with myofibroblastic differentiation</td>
<td></td>
</tr>
<tr>
<td>Undifferentiated pleomorphic sarcoma</td>
<td>5</td>
</tr>
<tr>
<td>Osteosarcoma</td>
<td>1</td>
</tr>
<tr>
<td>Leiomyosarcoma</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Fibrosarcoma</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Liposarcoma</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Myofibroblastic tumour</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
<td>2</td>
</tr>
<tr>
<td>Primary lymphoma</td>
<td>2</td>
</tr>
<tr>
<td>Pericardial tumours</td>
<td></td>
</tr>
<tr>
<td>Mesothelioma</td>
<td>2</td>
</tr>
<tr>
<td>Synovial sarcoma</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Total</td>
<td>25</td>
</tr>
</tbody>
</table>
TOE is the modality of choice to detect these small highly mobile lesions and MRI is rarely required. When imaged with MRI features are those of a low signal well circumscribed mobile valve nodule on SSFP sequences, often with peri-lesional flow artifact (47-49) (Figure 2). T1 and T2 weighted images reflect their fibroelastic composition with uniform intermediate signal intensity similar to myocardium. Homogeneous LGE has been reported but in our experience LGE is commonly absent in keeping with a poorly vascularised lesion (50). The main imaging differential is a vegetation of infective endocarditis but distinction is generally straightforward on clinical grounds. On imaging vegetations are invariably associated with valve destruction/incompetence unlike a fibroelastoma with which valve function is usually preserved (51).

**Lipoma**

Lipomas are an encapsulated conglomerate of slow growing mature adipose tissue. They can occur anywhere...
in the heart but are most frequently seen within the atrial septum and epicardium where they grow into the pericardial space. Subendocardial lipomas are broad based lesions which protrude into the cardiac chambers and may cause obstructive and compressive effects but most probably remain asymptomatic and are discovered incidentally (52-54). There is a reported association with atrial arrhythmias and conduction abnormalities. Surgical resection is rarely needed except with very large and symptomatic lesions (40).

Lipomatous hypertrophy of the inter-atrial septum is hyperplasia of normal septal fat and also reflects a benign entity. The key distinguishing feature from lipoma is sparing of the mid-septal region (fossa ovalis) giving rise to their classical “dumbbell” appearance.

Lipomas and lipomatous hypertrophy have a highly variable appearance on echocardiography making confident distinction from other cardiac tumours like myxoma challenging (55). MRI appearances are typical owing to definitive characterization of fat signal which appears of uniformly high signal on both T1 and T2-weighted imaging and fully suppresses with fat saturation (Figure 3). Due to their avascular nature there is no first pass or LGE with these lesions (56).

Haemangioma

These are slow flow vascular malformations composed of endothelial lined venous channels with an intervening connective tissue stroma (56). There is no chamber predilection and they have been described in many locations including with the pericardial space. Most probably remain asymptomatic but large lesions are associated with exertional dyspnoea and may be considered for surgical resection.

On MRI they display isointense signal to myocardium on T1-weighted images due to slow flow and of diffuse high signal intensity on T2-weighted images. First pass and delayed enhancement is intense and prolonged and may appear heterogeneous depending upon the amount of fibrous tissue and any calcific foci (57).

Fibroma

Fibromas are the most common paediatric cardiac tumour but they are occasionally seen in adults (58-60). Typically, they have an intramural location within the interventricular septum or left ventricular free wall (61). Fibromas have been associated with Polyposis syndromes (familial adenomatous polyposis) and Gorlin-Goltz syndrome (62,63). Histologically they are well-defined lesions composed of collagen and fibroblasts with some calcific foci which help differentiate them from rhabdomyomas. Heart failure is the most common clinical presentation due to obstruction of blood flow or interference with valvular function. Ventricular arrhythmias and sudden death have also been reported (63).

On MRI fibromas are usually sharply demarcated and

![Figure 3 Atrial septal lipoma. (A) 4-Chamber T2-weighted black blood image showing a well-defined uniformly high signal lesion within the interatrial septum (curved arrow); (B) 4-Chamber T1-weighted black blood image again showing the lesion as having uniformly high signal in keeping with a fat composition; (C) 4-Chamber T2-weighted black blood image with fat suppression showing complete and uniform signal suppression (curved arrow). RA, right atrium; LA, left atrium; LV, left ventricle.](image-url)
of uniformly low signal intensity compared with adjacent
myocardium on both T1 and T2-weighted imaging due
to their dense fibrous composition (64). A lack of first pass
enhancement is typical and LGE is commonly intense
and uniform although peripheral and heterogenous LGE
patterns have also been described (65,66) (Figure 4). The
main imaging differential of a fibroma with little or no
delayed enhancement is focal hypertrophic cardiomyopathy
and distinction can be challenging. Careful review of
the cine images is the best means of discrimination with
a fibroma showing no regions of contractility which
can usually be appreciated in most cases of “mass-like”
hypertrophic cardiomyopathy (65).

Cardiac paraganglioma

Paraganglioma is a very rare tumour which originates
from neuroendocrine ganglia cells which tend to lie
within the atrioventricular grooves and at the root of the
great vessel origins (67). Up to 50% of these tumours
secrete catecholamines which explains the typical clinical
presentation of palpitations, flushing and headaches.
Patients may also be hypertensive. Although histologically
benign curative resection is often difficult due to extensive
vascularity and often complex relationship with the
adjacent coronary arteries.

On MRI these lesions usually range in size from 4-8 cm
appearing well circumscribed. They are isointense to
myocardium on T1- and of intensely high signal on T2-
weighted images (“light bulb bright”) (Figure 5). First pass
and LGE is usually intense and uniform (23).

Cystic tumour of the atrioventricular node

This is one of the rarest benign cardiac tumours with
only a few cases reported in the literature. It is postulated
to be an inclusion cyst of endodermal origin containing
keratinous debris. Most reported lesions are small
(<2 cm) and located at the base of the interatrial septum
in the atrioventricular nodal region where they may cause
interference with cardiac conduction leading to heart
block and potentially lethal arrhythmias. Surgical resection
is therefore advocated (68,69).

Described MRI features are those of a well-defined high
T1 and high T2-signal nodule in the AV nodal region of
the lower septum (Figure 6). Homogeneous LGE has been
reported (70).

Conclusions

A comprehensive cardiac tumour imaging protocol has
been described and the spectrum of benign primary tumour
MRI appearances has been reviewed. Part 2 of this review
will focus on primary malignant cardiac tumours, cardiac
metastases as well as the MRI features of some potential
tumour mimics.

Disclosure: The authors declare no conflict of interest.
Figure 5 Cardiac paraganglioma. (A) Coronal T1-weighted black blood image showing a well-defined intermediate signal lesion at the root of the great vessels and indenting the roof of the right ventricle (arrows); (B) coronal T2-weighted black blood image showing the lesion to be of intensely high signal (arrows). RV, right ventricle.

Figure 6 Cystic atrioventricular nodal tumour. Axial T1-weighted black blood image showing a well-defined uniformly high signal lesion within the lower margin of the interatrial septum (arrows). RV, right ventricle; LV, left ventricle.

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44. Bruce CJ. Cardiac tumours: diagnosis and management. Heart 2011;97:151-60.


