Case Report

The Atrial Myxoma, the Learning Model for Cardiac Tumor Assessment Using Magnetic Resonance Imaging: A Case Report

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Abstract
Cardiac tumor is still considered as the one of the most difficult conditions in cardiology in terms of diagnosis and treatment. The atrial myxoma is a common primary benign cardiac tumor presenting in adults.[1] The diagnosis of cardiac tumor always relies on the clinical examination and results obtained from the imaging tools used. This article selects the atrial myxoma as a learning model to demonstrate the assessment of the cardiac tumor when using Magnetic Resonance Imaging (MRI) with the different techniques including the tissue characterization method which has already been established as a key diagnostic method for the diagnosis of cardiac tumor and mass.

Keywords: cardiac tumor, atrial myxoma, tissue characterization, MRI

Cardiac tumor is considered as the one of the most difficult conditions in cardiology in terms of diagnosis and treatment. All tumors can be classified into two simple categories, benign and malignant tumor. A benign tumor is unable to invade an adjacent organ and to metastasize to a remote organ. Conversely the malignant tumor is determined by its ability to aggressively invade a neighboring organ and to metastasize to a remote organ.[2] On the other hand, a tumor is also differentiated into the primary and the secondary tumor.[2] A primary tumor is defined as the first tumor in the body that originates in the specific organ and it is named after the origin location. The secondary tumor comes from spreading or metastasis of the primary tumor and it will form a new tumor mass in another part of the body.

Atrial myxoma is the most common primary benign cardiac tumor that forms the first tumor cell and starts its growing in the heart without aggressive invasion and metastasis. Myxoma comes from the Latin and means a myxiod tumor of primitive connective tissue that is composed of mucoid (mucus-like) substance in the background.[3] The atrial myxoma is the most common tumor in adults especially among females. The mean age at tumor presentation is around 50 years old.[4] The typical location of the atrial myxoma is the left atrium (75-85%).[4,5] The second most common location is the right atrium.[4] The clinical features of atrial myxoma depends on the size and location of the tumor. The tumor in the left heart may produce symptoms and signs of valvular obstruction or an embolic event.[6]

The atrial myxoma is described in a macroscopic pathology character as a polypoid (resembling a polyp) or lobulate, with a round or oval contour. The atrial myxoma is the tumor with a stalk that arises from the interatrial septum at or near the fossa ovalis. With this appearance the atrial myxoma can be confidently diagnosed. If the tumor stalk does not appear, then atrial thrombus should be considered in the differential diagnosis.[7] The microscopic characters of the atrial myxoma are heterogeneous with a cystic area of polysaccharide rich myxoid substance, hemorrhage or

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Received: August, 3, 2015.
Revision received: August, 3, 2015.
Accepted after revision: August, 5, 2015.
Bangkok Med J 2015;10:42-49.
E-journal: http://www.bangkokmedjournal.com

hemosiderin, fibrosis and calcification in about half of all cases that cause heterogeneity of the tumor tissue. Although the atrial myxoma typically attaches to the endocardial surface near the fossa ovalis by its stalk, it may also have other attachment sites within a given chamber such as at the posterior wall, anterior wall or atrial appendage.[8,9]

Assessing atrial myxoma by MRI

Transthoracic echocardiography (TTE) is always the first line tool for assessing cardiac mass. Although the transesophageal echocardiography (TEE) is a semi-invasive tool it is still superior to the transthoracic echocardiography (TTE) in defining the characteristic of the tumor.[10] The obtained findings of the tumor from an echocardiogram include the location, the gross appearance and the extension of the tumor which may be sufficient clues to diagnose some cardiac tumors and mass.[11] That said, however, echocardiography may not be able to differentiate the type of tumors in a more specific manner (by using the tissue characterization method) because echocardiography may not provide the signal intensity that is different enough to distinguish each principle composition of the assessed tumor. Nowadays, MRI is considered a good alternative diagnostic tool for cardiac tumor assessment because it acquires a diagnostic image with high resolution and high soft tissue contrast that allows for an improved tumor classification than the tissue characterization method.[12]

Case presentation

This is the case of a Thai female patient aged 54 years with a history of a recent ischemic stroke who was sent to perform a cardiac MRI to define the left atrial mass that was documented by echocardiogram. Echocardiogram showed a 4.6x2.8 cm, mobile, lobulated mass attached to the left site of the inter-atrial septum with stalk. A hyperechoic density at the central area of mass was observed. The left atrial mass was protruding into the left ventricle in diastole. Mild to moderate MR was also visualized on echocardiogram. The provisional diagnosis of the cardiac mass is left atrial myxoma.

Cardiac MRI was performed. With the gradient echo CINE MRI pulse sequence on the four-chamber view and short axis view showed the left atrial mass with a size of 7.84x2.26 cm, mobile with plunging to and fro from the left atrium to the left ventricle. The left atrial mass had an irregular surface with a multi-lobular contour, and was attached to the inter-atrial septum by its stalk and the mass occupied nearly all of the left atrium. The mass had two broad-base stalks and two attachment sites at the left side of the inter-atrial septum at the proximal and distal to the fossa ovalis. The left atrial mass produced an inhomogeneous hypointensity compared to the myocardium on the gradient echo CINE MRI images, and a heterogeneous hypointensity among isointensity to myocardium in the background on a T1W bb spin echo image and also produced the heterogeneous hyper-, hypo-, iso intensity of some areas in tumor on T2W bb spin echo image. This may be some of the area of chronic hemorrhage that produced the hypo-intensity on T1W and T2W bb images. The T2W bb with fat saturation images revealed a bit hyperintensity of mass at the area that was compatible with the T2W bb that was confirmed for the area of fluid. Normal pericardial thickness is observed on T1W bb image. By the first pass perfusion study with gadolinium contrast injection revealed no significant contrast perfusion into the mass body that indicates no hypervascularization. By delayed contrast enhancement study revealed heterogeneous contrast enhancement of fibrotic or necrotic area of the mass with the prominent small area hypoenhancement of the probable calcium. By the location, the gross appearance of tumor with stalk attached to the inter-atrial septum and the characteristics of the heterogeneous signal intensity of the mass that were shown on MRI images, the left atrial myxoma is highly suggestive (see Figure A-G).

The patient was treated by the surgical resection of the left atrial mass with inter-atrial septum closure by Dacron patch. The resection mass was sent for pathology study, and this reported the gross description of mass as a lobulated yellowish white mixoid mass with the size of 2.0x4.0x9.0 cm. The microscopic examination of the mass composition is described as distant scattered stellate cells with round to oval nuclei among the mixoid stroma with focal calcification and surrounded by similar flattened cells. By the pathological examination results, the left atrial mass was definitively diagnosed as a left atrial myxoma.

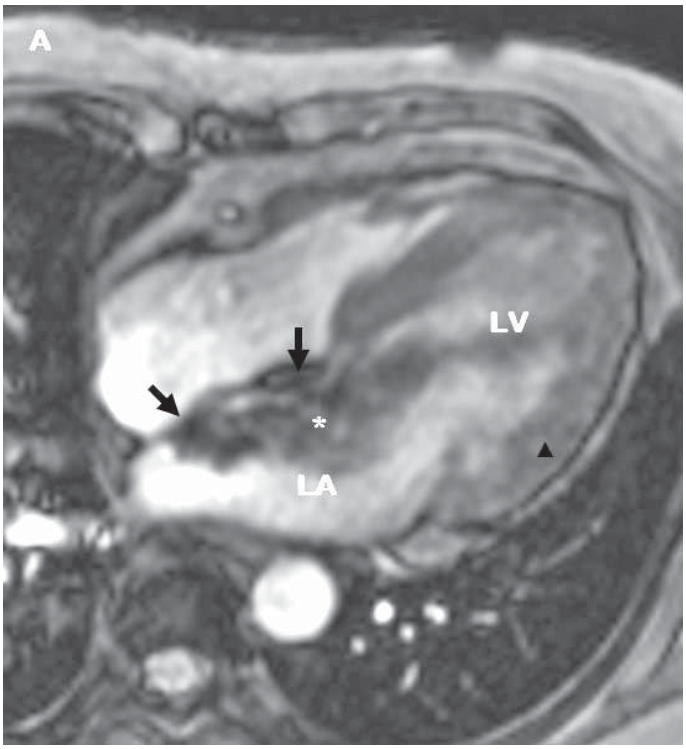


Figure A: The gradient echo CINE MRI image demonstrates the left atrial mass (*) atrial myxoma with two stalks that attaches to the left side of the inter-atrial septum (black arrow). The myxoma produces heterogeneous hypointensity among the iso intensity (to myocardium) background (▲).

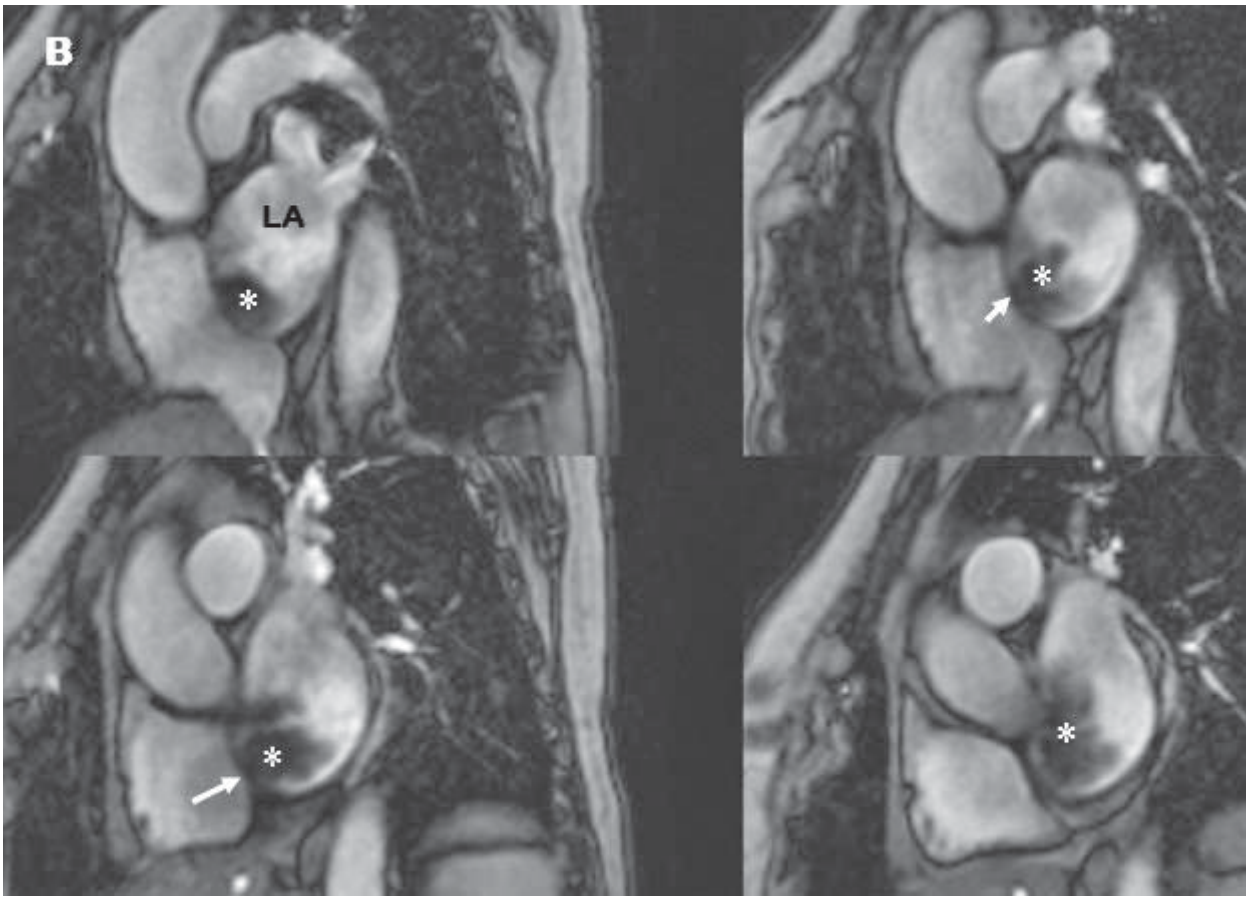
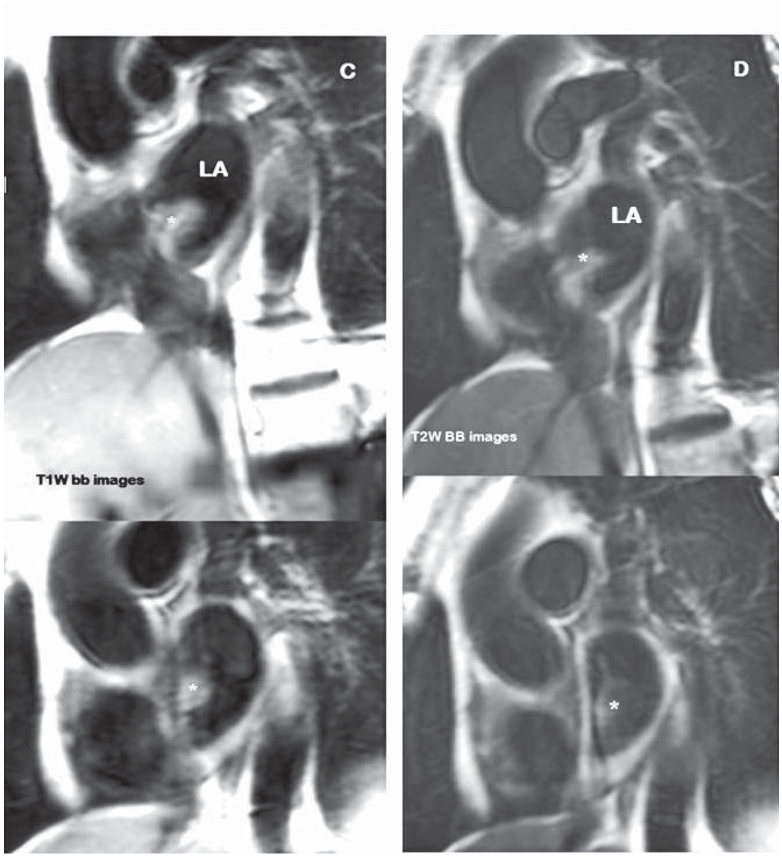


Figure B: The left atrial tumor mass (atrial myxoma) with stalk is demonstrated on the gradient echo CINE MRI (*) on the short axis view of the atrium. The mass attaches to the left side of the inter-atrial septum (white arrow).

ISSN 2287-0237 (online)/ 2287-9674 (print)



Figures C, D: The left atrial myxoma (*) with stalk is demonstrated on T1W and T2W bb spin echo images on the short axis view of the atrium. Figure C: The left atrial myxoma produces heterogeneous hypo- and iso intensity in the T1W bb and produces heterogeneous hypo - and iso intensity that and produces heterogeneous hypo- hyper and iso intensity on the T2W bb images (Figure D). Probable some chronic hemorrhage in the tumor is suggested by the evidence of hypo-intensity that is produced by the same area in the tumor and is visualized on both T1W and T2W bb spin echo images.

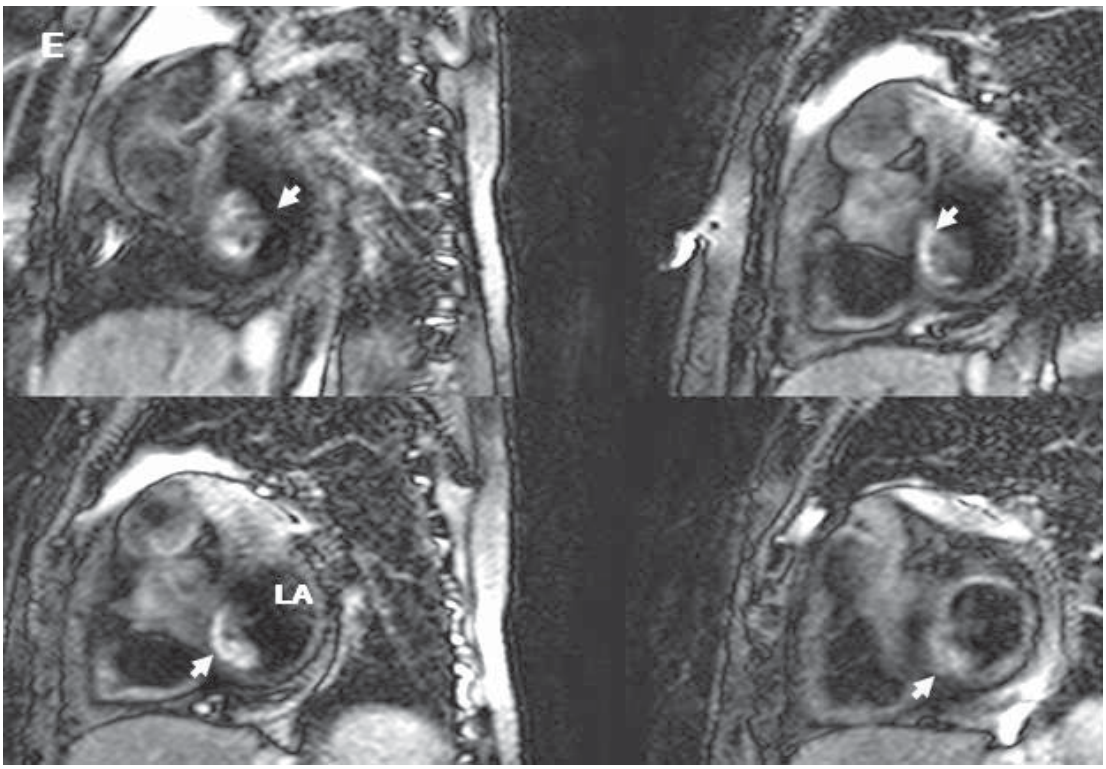


Figure E: The T2W bb with fat saturation demonstrates the hyperintensity of the mass (arrow) on the isointense background that is compatible with the hyperintense area on T2W bb that indicates an area of fluid.

ISSN 2287-0237 (online)/ 2287-9674 (print)

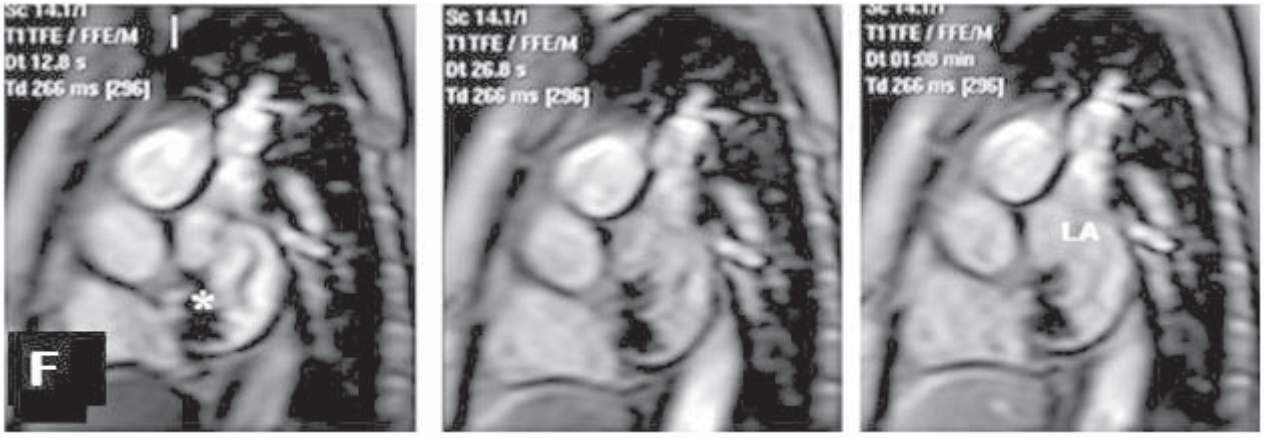


Figure F: The first pass contrast injection study reveals no contrast perfusion into the tumor mass (*) and this indicates no hypervasularization of tumor mass.

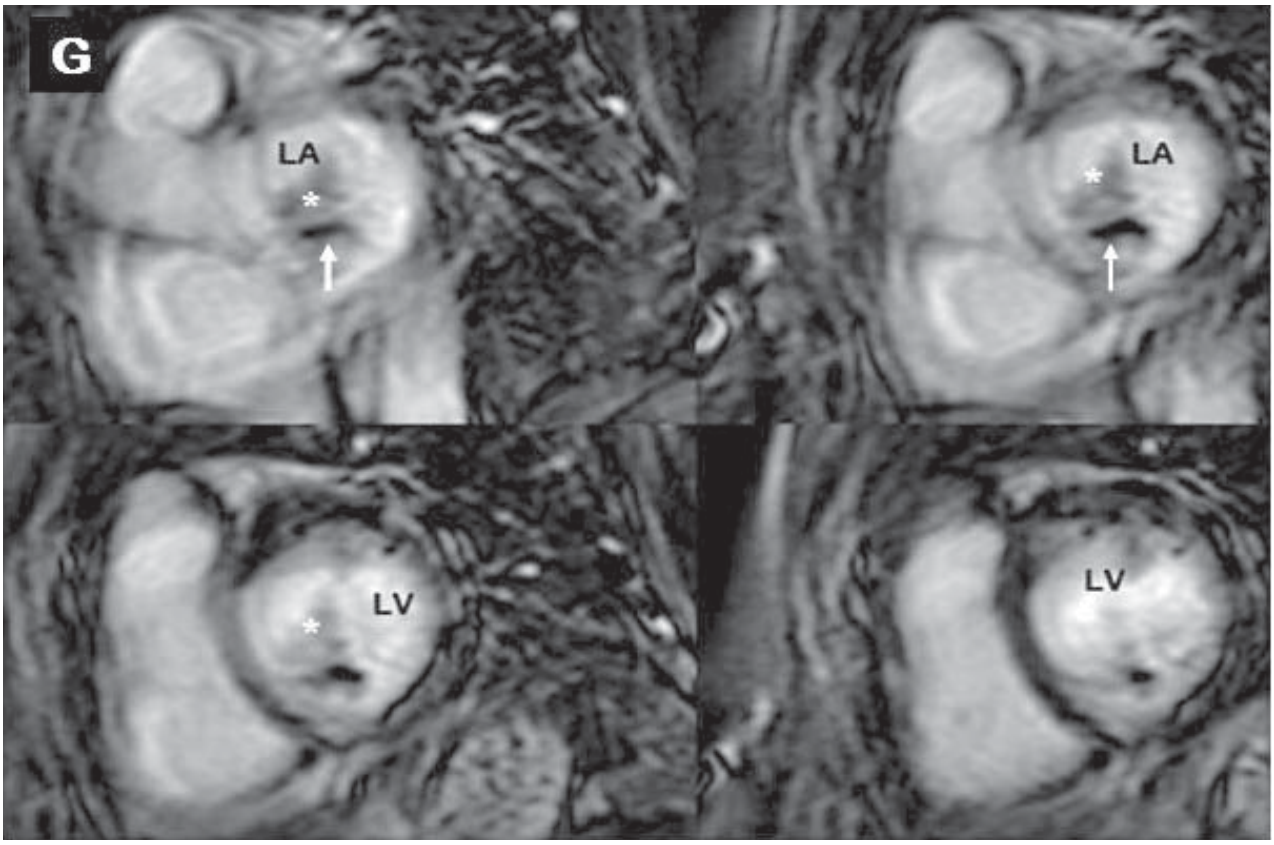


Figure G: The delayed contrast enhancement study reveals contrast enhancement of the left atrial myxoma at the central area (*) and this indicates tissue necrosis of tumor mass and the prominent minimal hypo-enhancement area may indicate a hemosiderin or calcium (white arrow).

ISSN 2287-0237 (online)/ 2287-9674 (print)

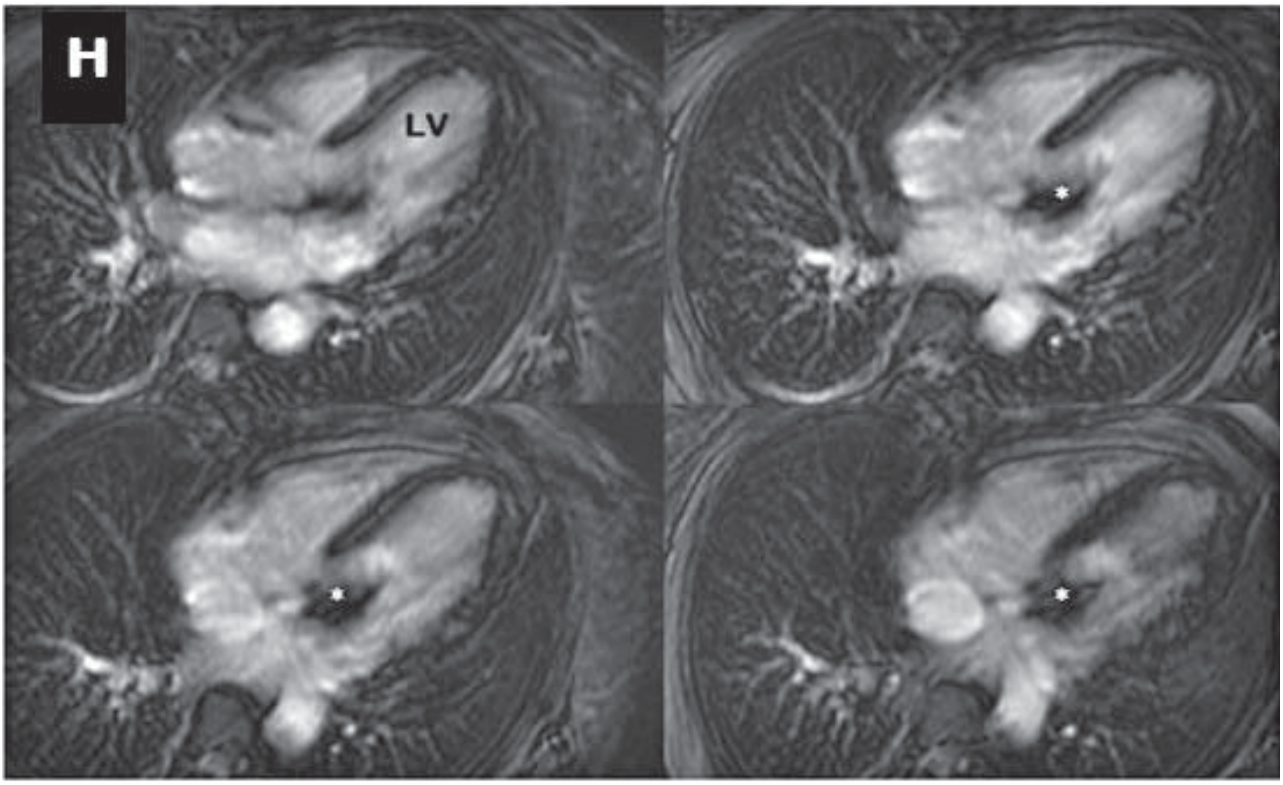


Figure H: The delayed contrast enhancement study reveals an heterogeneous contrast enhancement of the tumor (*) and this indicates some necrotic tissue of the tumor and the inhomogenous hypo-enhancement of the tumor that indicates a hemosiderin or calcium.

Discussion

The initial step for ease of differential and definitive diagnosis of any cardiac tumor is accomplished by the tumor's location. The most common location of an atrial myxoma is in the left atrium. The left atrial mass that must be differentiated from the atrial myxoma is thrombus and may be include tumors such as lipoma, rhabdomyoma, malignant histocytoma which may also originate in the left atrium. MRI has been a good alternative tool to provide both gross appearance and the composition characteristics of the tumor. To demonstrate the mass contour in all four cardiac chambers on MRI, images use the gradient echo CINE pulse sequence on a horizontal axis (4-chamber) view. By the CINE MRI pulse sequence, the structural contour, the mobility and the functional effect of the tumor, including the invasion of the tumor into neighboring areas of the tumor origin are visualized simultaneously. In assessing the cardiac tumor to include the structural contour, the mobility and the functional effect using the gradient echo CINE MRI pulse sequence, it is most necessary to perform multi-slice CINE imaging with whole heart coverage, otherwise the tumor attachment and invasion site may be missed. The typical gross pathology of the atrial myxoma is described as a benign, lobulate mass with stalk that arises from the inter-atrial septum at or near the fossa ovalis which is a remnant of a thin fibrous sheet that covers the foramen ovale during fetal development.[13] The tumor with a stalk appearance at the atrial myxoma and is attached at or near the fossa ovalis makes the atrial myxoma very unique which is an important diagnostic clue for the atrial myxoma. However, the other left atrial mass that mimics the atrial myxoma, especially the thrombus, must be taken into account. Thrombus is most likely to occupy the posterior wall of the left atrium and the left atrial appendage and is often correlated with atrial fibrillation and which is the clue to consider a thrombus. To discriminate among tumors that are classified in the same group, such as originating in the same place, tissue characterization method will play a role. The atrial myxoma is a heterogeneous mass as mentioned above but a thrombus is a homogeneous mass and it also produces a different signal intensity on MRI images that varies directly to the thrombus age. As we know that the atrial myxoma is a benign tumor, the characteristic of a benign tumor must be visualized on the images. This includes slow growth, a resemblance to the tissue of origin (well differentiated), well circumscribed, lack of invasion and absence of metastases. The sign of invasion of the tumor can be observed on gradient echo CINE images by the evidence of the thickening of the area as being an invasion during the heart movement. A well circumscribed

tumor is observable on gradient echo CINE images. The next step is to confirm the myxoma findings by the tissue characterization method. This can be done with MRI images. The tissue characterization method using MRI is done by examining the different composition substances of the tumor or mass producing different signal intensity on the MRI image. About 50% of the atrial myxoma tumor tissue is composed of polysaccharide, myxoid substance, hemorrhage or hemosiderin, fibrosis and calcification. The T1W and T2 bb spin echo pulse sequence and T1W inversion recovery pulse sequence with contrast injection are used to differentiate the type of tumor by the tissue characterization method. The polysaccharide and the myxoid substance are expressed into isointensity on T1Weighted and T2Weighted black blood spin echo images as myocardium, the acute stage of hemorrhage produces a hypointensity on T2Weighted because of the paramagnetic effect of the deoxyhemoglobin and the sub acute hemorrhage gives a hyperintensity signal on T2W bb image because of the effect of methemoglobin that shortens the T2 relaxation time. Furthermore, chronic hematoma will produce the hypointensity on both T1W bb and T2W bb because of the effect of ferritin and hemosiderin,[14,15] the fibrotic component produces a white band of delayed contrast enhancement on the T1W inversion recovery with the contrast injection images but produces hypointensity on T1Weighted and T2Weighted black blood spin echo images. The fast T1 gradient first pass gadolinium contrast perfusion is used to assess hyper-vascularization of the tumor mass. The malignant tumor is always hypervasculaized and this is evident by the moderate to strong perfusion enhancement. Mild contrast enhancement on the first pass perfusion image may be found in benign tumors in about 40-50% of all cases.[16] The disadvantage of MRI in assessing the cardiac tumor is the inability to detect any calcium component. Calcium component will produce hypointensity on both spin echo black blood and gradient echo MRI including the T1 gradient inversion recovery images that may be confused with thrombus. In the majority of tumors, a heterogeneous signal intensity is often produced on T1W and T2W bb spin echo images therefore it is sometimes a bit difficult to specify the type of tumor exactly. The tissue characterization method, on an MRI, is the easy way, to begin with, when we look for the main background tissue. The key components that can transform into the signal intensity on MRI then reveal the probable origin of the signal intensity on MRI images.

Conclusion

The assessment of cardiac tumor by using imaging tools, especially MRI, needs a comprehensive knowledge and understanding of both macroscopic and microscopic pathologic characteristics of the tumor with clinical sign correlations including the knowledge of imaging techniques. In general, the macroscopic characteristic of a tumor is used to diagnose the cardiac mass in the majority of cases because of the limitation of the imaging tool in using the tissue characterization method. Hence the microscopic information is often ignored. MRI is a diagnostic tool that has been established to perform the tissue characterization that relies on the difference of the signal intensity among the tumor components. This article emphasizes the use of the tissue characterization method in diagnosing the cardiac tumor on MRI by using the atrial myxoma as a learning model.

The atrial myxoma is selected to be a learning model for cardiac tumor assessment on MRI because it has typical characteristics of its own in that it is a tumor with stalk and it attaches to the endocardial wall of the atium at the fossa ovalis. In addition, the atrial myxoma is a very common tumor that is often seen and therefore can be used as a model for further repeating the practice of using MRI assessment. In the past, the majority of tumors have always been diagnosed by virtue of its location and its being malignant but in the present day we have MRI to transform the microscopic component of tumor to the specific intensity on images which provides important and key information to classify the tumor. The comprehensive combination of the knowledge of the pathologic characteristics of tumors and the imaging techniques is the most important key in cardiac tumor diagnosis when using MRI. However, the drawback of the MRI in assessing the cardiac tumor by tissue characterization is that is it not able to detect the calcium component as CT scan does. In case where the signal intensity is not shown with more contrast, such as in small tumors, the differentiation of tumor by location and its unique appearance may be used as the most important clues.

ISSN 2287-0237 (online)/ 2287-9674 (print)

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ISSN 2287-0237 (online)/ 2287-9674 (print)