Unexpected Jeopardy: CMR Assist in Diagnosis of Atypical Chest Pain Patient Which Turns Out to be Chronic Aortic Dissection and Cardiac Schwannoma

Runyawan Chotenimitkhun[1]



Runyawan Chotenimitkhun

[1] Cardiac Center, Internal Medicine, Police General Hospital, Bangkok, Thailand

*Address Correspondence to author:
Runyawan Chotenimitkhun, MD
Cardiac Center, Internal Medicine,
Police General Hospital,
Bangkok 10330, Thailand.
email: runyawan_c@hotmail.com

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Abstract

A 60-year-old obese woman who had intermediate probability of coronary artery disease presented with atypical chest pain. Obstructive coronary artery disease was suspected. There were non-diagnostic results from basic cardiac investigation and echocardiogram. She was referred to a stress cardiovascular magnetic resonance (CMR). CMR is a safe comprehensive imaging modality including left ventricular function, stress perfusion study and late gadolinium enhancement to identify myocardial ischemia and myocardial infarction. Moreover, the CMR study can provide information of great vessels e.g. aorta and its complication as in this case. The right-sided structures were easily evaluated without body habitus limitation. She was diagnosed with chronic aortic dissection and right atrial malignant cardiac tumor which pathologically confirmed cardiac schwannoma. Finally, she received an accurate diagnosis which guided prompt surgical management and saved her life.

Keywords: cardiovascular magnetic resonance, cardiac tumor, aortic dissection, chest pain

Nowadays, comprehensive CMR imaging has always been required in clinical practice. It provides the gold standard of left and right ventricular volume and function assessment. The stress CMR perfusion study is used to identify or exclude myocardial ischemia. Late gadolinium enhancement CMR images provide information in tissue characterization of myocardium to prove myocardial infarction. Furthermore, CMR include tissue characterization techniques can help presumptive diagnosis of cardiac mass, discriminate malignant and benign tumor, and differentiate cardiac mass from intracardiac thrombus. The extracardiac structure includes aorta and its complications are also detected at the same period of time in one study. Therefore, CMR study works excellently as a gatekeeper imaging modality for diagnosis in multiple cardiac problems and rare conditions as in this case, which aids proper management.

Case Report

A 60-year-old obese woman with diabetes and hypertension presented with dyspnea on exertion and atypical chest pain for several weeks. The physical examination was unremarkable. There was no evidence of ischemic change on electrocardiography. The high sensitive troponin-T level was less than 50.0 ng/L (0.0-100.0). The result of transthoracic echocardiography was limited due to large body habitus, however, it revealed normal left ventricular systolic function as LVEF 60% by Teicholtz method. The patient was referred to stress cardiovascular magnetic resonance (CMR) to exclude obstructive coronary artery disease (CAD).

CMR findings

The SSFP-cine CMR images showed normal biventricular systolic function with no regional wall motion abnormality (Figure 1). The thickened, immobile, intimal flap was detected in the ascending thoracic aorta, consistent with the classification of Standford type A or Debakey type 2 of the aortic dissection. The ascending thoracic aortic aneurysm was demonstrated. The entry site of the dissection was found above the ostial of the right coronary artery and its termination was before the origin of the right innominate artery (Figure 2). Therefore, adenosine stress CMR was discarded. A moderate aortic regurgitation (regurgitant fraction of 30%) was a sequelae from the ascending aortic aneurysm.

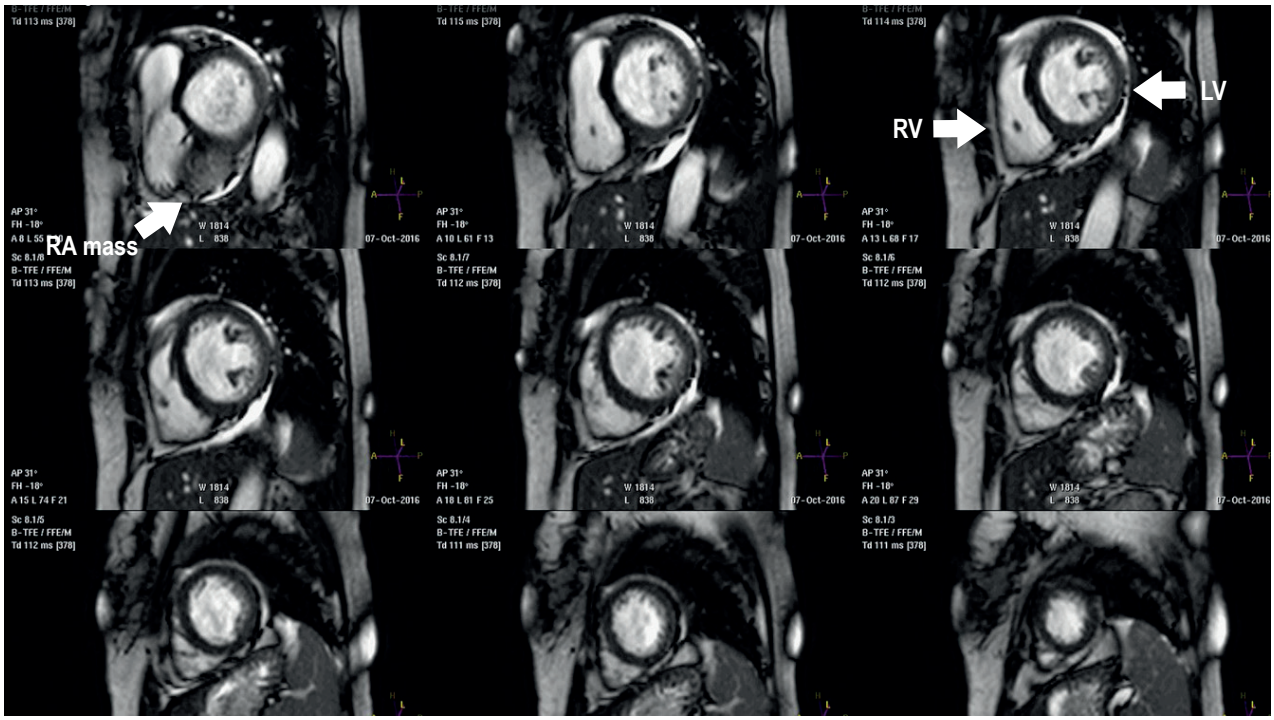


Figure 1: The SSFP-cine CMR images showed normal biventricular size and systolic function with no regional wall motion abnormality. A large, heterogenous signal intensity, right atrial mass was demonstrated. (LV: Left ventricle ; RV: Right ventricle ; RA: Right atrium)

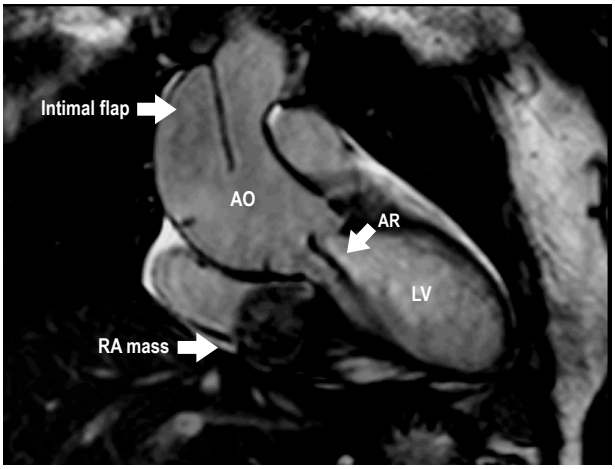


Figure 2: Aneurysmal change and dissection (intimal flap) of the ascending thoracic aorta. A large, heterogeneous signal intensity of right atrial mass attached to the interatrial septum. (LV: Left ventricle ; RA: Right atrium ; AO: Aorta, AR: Aortic Regurgitation)

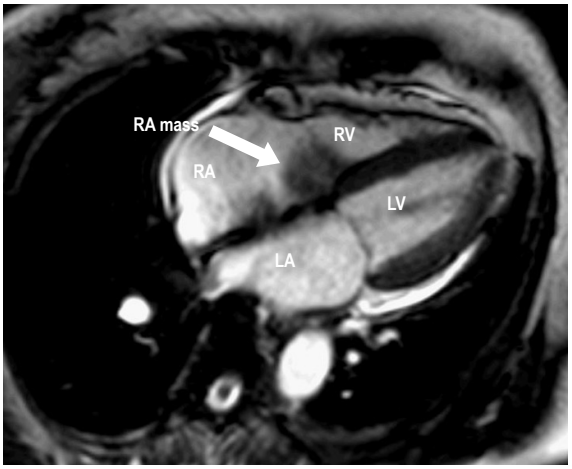


Figure 3: A large right atrial mass attached to the interatrial septum, moving across tricuspid valve without hemodynamic obstruction. (LV: Left ventricle ; RV: Right ventricle ; LA: Left atrium ; RA: Right atrium)

A large 50x48x49 mm, heterogeneous signal intensity of right atrial mass attached to the interatrial septum without stalk, and moving across tricuspid valve without hemodynamic obstruction (Figure 3), there was no abnormal flow of tricuspid regurgitation or stenosis. The mass was iso-signal intensity relative to myocardium on T1-weighted image (Figure 4) and hypersignal intensity on T2- weighted image (Figure 5) and no fat tissue on T1-weighted with fat-saturated image (Figure 6). The central part of the tumor showed mixed high and low (heterogeneous) signal intensity on T2-weighted image representing hemorrhagic and necrosis. There was a peripheral or ring enhancement with minimal contrast uptake within the central core of the tumor during the first pass perfusion study, and on late gadolinium enhanced images (Figure 7), indicating central hemorrhage and necrosis. This appearance would exclude a thrombus, but rather resembled a cardiac tumor. All findings are not consistent with typical characteristics of any type of tumors. In addition, there was no myocardial infarction or scar (Figure 8).

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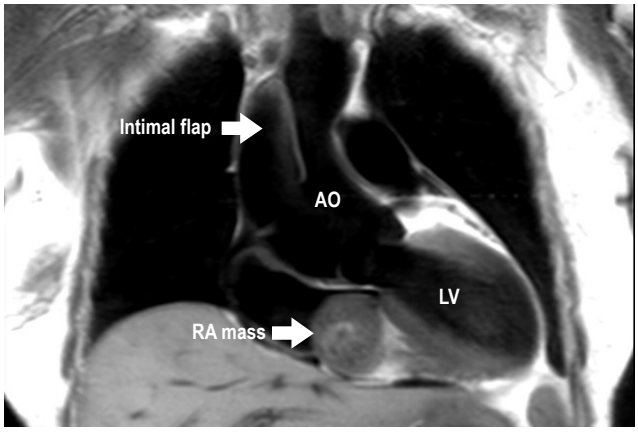


Figure 4: T1-weighted dark blood image, the RA mass showed isosignal intensity relative to myocardium on T1-weighted spin echo image. Aneurysmal change and dissection (intimal flap) of the ascending thoracic aorta.

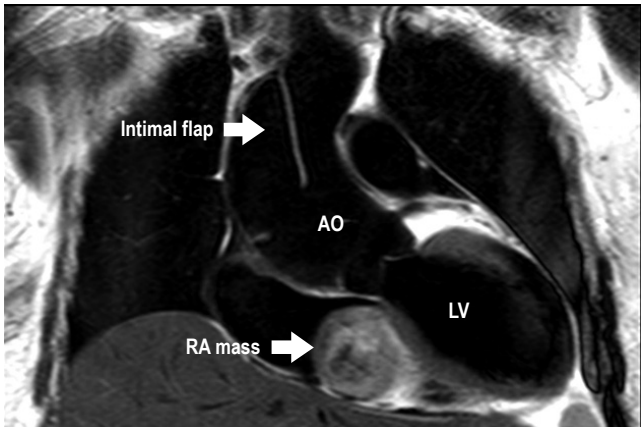


Figure 5: T2-weighted dark blood image, the RA mass showed hypersignal intensity. The central part of tumor showed mixed high and low (heterogenous) signal intensity on T2- weighted image represent hemorrhagic and necrotic tumor.

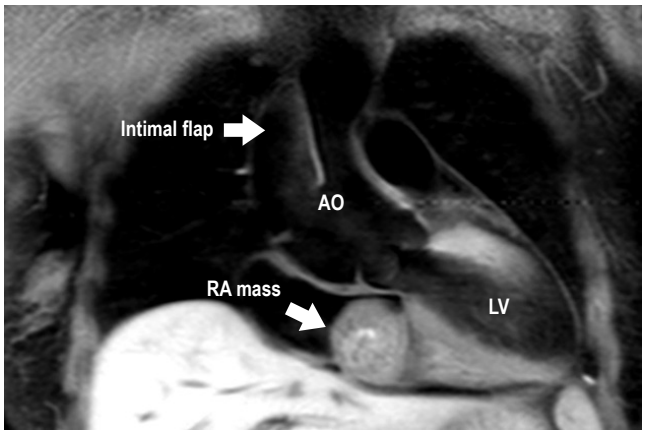


Figure 6: T1-weighted dark blood image with fat suppression showed brightness persisting on fat saturated image, excluding fat-originating tumor. Aneurysmal change and dissection (intimal flap) of the ascending thoracic aorta.

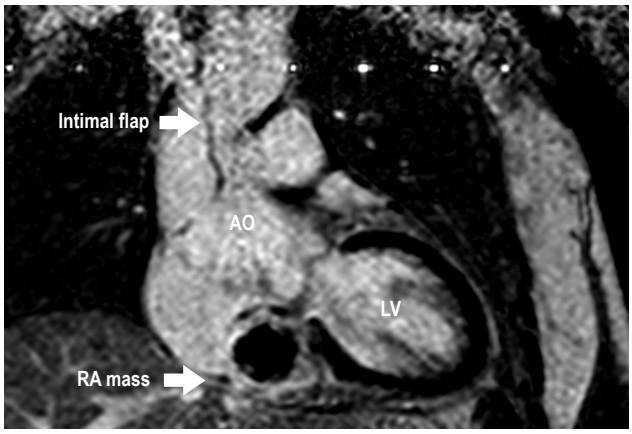


Figure 7: Late gadolinium enhanced image showed peripheral enhancement with minimal contrast uptake within central core of tumor, representing central hemorrhage and necrosis. This appearance excluded a thrombus.

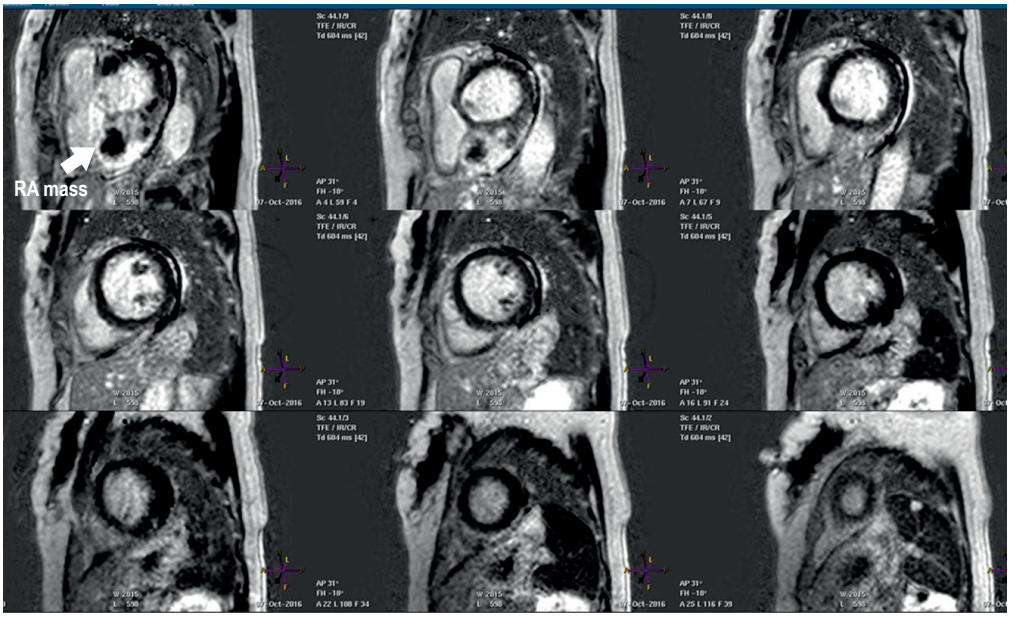


Figure 8: Late gadolinium enhanced image showed no myocardial infarction or scar.

The patient was diagnosed with chronic aortic dissection with right atrial cardiac tumor. The patient underwent urgent aortic surgery, whereby her right atrial tumor was removed without complication and she was discharged 2 weeks later. The gross pathologic report and microscopic evidence of the right atrial mass revealed cardiac schwannoma (Figure 9 and Figure 10).

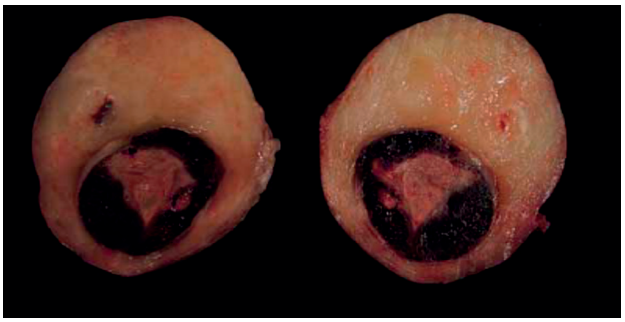


Figure 9: Grossly, the tumor is a round encapsulated light brown rubbery mass. The cut surfaces of mass show a well-circumscribed area of hemorrhage and necrosis with calcific nodule in the mass. The remaining area of mass show homogeneous myxoid white and light yellow surface.

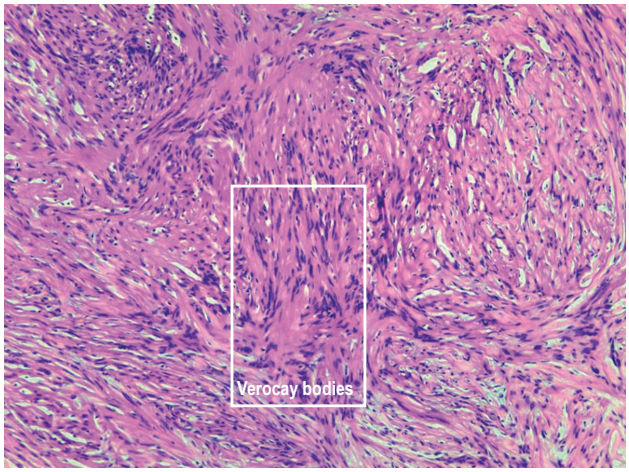


Figure 10: Microscopically, the sections show circumscribed, thick fibrous encapsulated by pseudo-capsule, alternating hypercellular (Antoni A) and hypocellular (Antoni B) areas of low-grade spindle cells neoplasm. Occasional nuclear palisading around fibrillary process (Verocay bodies) are found in Antoni A area (H&E stain, 600x800).

Discussion

The CMR characteristics of cardiac mass such as non-enhancing central tumors following gadolinium administration, raises fibroma (central calcification) or sarcoma (central necrosis) or organized chronic thrombus (peripheral fibrous content) into the relevant differential diagnosis. The specific location (endocardial/ intracavitary, myocardial, or epicardial/ pericardial) of the lesions, the main chamber or structure involved can be further narrowed down into different diagnosis[1-2]. However, a definite diagnosis of cardiac tumor can only be completed after histological examination.

In this case, the most tissue characteristics of this cardiac tumor appeared benign, including single, well-defined border without tissue plane invasion and pericardial or pleural involvement. However, its large size, the right heart involvement, heterogeneous tissue on T1-and T2-weighted images and the contrast enhancement of the cardiac tumor made it impossible to disregard a malignant tumor cannot be excluded.

Primary cardiac schwannoma is an extremely rare disease.[3] So far, only 17 cases of this disease have been reported in the literature .[3-5] It frequently arises from the nervous system of the heart, which resides in epicardial fat and contains both myelinated and unmyelinated axon located on the right side of the heart, particularly in the right atrium and close to cardiac plexus. However, it can also be found in other sites of the heart [4]. Most of them are asymptomatic or symptom related to their location, causing extrinsic compression and pericardial effusion; or they may lead to valvular obstruction, thromboembolism, heart failure, sudden death, etc [3-5]. Some previous cases reported as a malignant tumor with an aggressive presentation. But they can have a benign and asymptomatic or non-specific presentation such as in the patient reported here (for months to years) as well as in other previously reported cases [5]. The early detection and consequently smaller tumor size or location favored a lack of symptoms. Similarly to previous reports [3-5], the distinct characteristics of a cardiac schwannoma on CMR is the ring enhancement with minimal contrast uptake within central core during perfusion study or post-gadolinium enhancement images. This finding corresponds to gross pathology of an encapsulated light brown rubbery mass.

Mostly, a patient diagnosed with chronic aortic dissection (onset > 90 days)[6] has asymptomatic or non-specific symptoms, similar to this case. The typical findings in CMR of chronic aortic dissection including thickened, immobile, intimal flap, demonstrated in this case except thrombus in the false lumen. However, the aortic or intimal flap calcification which represents chronicity may not visible by CMR [7]. CMR in the aortic dissection provides good diagnostic accuracy of the extent of the disease, localization of entry and re-entry identification of the intimal flap, presence of pericardial effusion, aortic regurgitation, and carotid artery dissection or coronary involvement, while it does not require ionizing radiation or iodinated contrast[6,8]. However, despite the abovementioned excellent performance of CMR, several methodological and practical limitations preclude the use of this modality in the majority of cases and also in unstable patients[6-8].

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Conclusions

Comprehensive CMR imaging can provide an accurate diagnosis in unusual clinical presentation of unexpected life-threatening conditions. CMR overcomes a limitation of echocardiogram in diagnosis of intracardiac mass and aortic diseases especially among obese patients. The unique technique of CMR can differentiate tissue characteristics of cardiac mass and exclude thrombus. Beside the diagnosis, CMR aids in excellent surgical planning as in this case that where the patient undergoes underwent lifesaving surgery without complication. and save her life.

Conflict of Interest

None

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