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Right ventricular mass detected by cardiovascular magnetic resonance imaging

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- Purpose** : *Right ventricular (RV) mass is very uncommon. We retrospectively reviewed characteristics of RV mass detected by cardiovascular magnetic resonance imaging (CMR).*
- Objective** : *To describe types of RV mass, magnetic resonance (MR) signal intensity, and enhancement of RV mass in CMR.*
- Setting** : *King Chulalongkorn Memorial Hospital, a large medical school, tertiary care and cardiac center for treatment of heart diseases in Thailand.*
- Research design** : *Retrospective review.*
- Methods** : *CMR studies during Jan 1st, 2007 to July 31st, 2009 were reviewed. All patients who had RV mass were recruited. CMR findings were correlated with operative notes, pathology reports and/or clinical diagnosis.*
- Result** : *In total, 243 CMR studies were performed. There were 4 patients (1.65% of total CMR examination) with RV mass: 2 cases were primary cardiac tumor, i.e., malignant fibrosarcoma of right ventricle and benign RV myxoma; and the other 2 cases were intracardiac thrombus. The tumors showed heterogeneous enhancement whereas no enhancement in the thrombus.*

Conclusion : *CMR is useful to characterize tumor and thrombus by using contrast enhancement.*

Keywords : *Right ventricular mass, Cardiovascular magnetic resonance imaging (CMR), Fibrosarcoma, Myxoma, Thrombus.*

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**มนตร์รวี ทুমโสมิต. การตรวจก้อนในหัวใจห้องล่างขวาด้วยเครื่องสะท้อนในสนามแม่เหล็ก
ของหัวใจและหลอดเลือด. จุฬาลงกรณ์เวชสาร 2553 ก.ค. - ส.ค.; 54(4): 313 - 21**

- เหตุผลการทำวิจัย** : ก้อนในหัวใจห้องล่างขวา (*right ventricle*) พบได้น้อยในประชากรทั่วไป เครื่องสะท้อนในสนามแม่เหล็กของหัวใจและหลอดเลือด (*cardiovascular magnetic resonance imaging*) เป็นเครื่องมือหนึ่งที่ช่วยจำแนกชนิดของก้อนได้
- วัตถุประสงค์** : การวิจัยนี้ทำการศึกษาย้อนหลังเพื่อคุณลักษณะของก้อนในหัวใจห้องล่างขวา ที่ตรวจพบด้วยเครื่องสะท้อนในสนามแม่เหล็กของหัวใจและหลอดเลือด
- ประเภทโรงพยาบาล** : โรงพยาบาลจุฬาลงกรณ์เป็นโรงเรียนแพทย์ ศูนย์ตติยภูมิ และมีศูนย์หัวใจ ที่ให้การรักษาผู้ป่วยโรคหัวใจในประเทศไทย
- รูปแบบการวิจัย** : การวิจัยย้อนหลัง
- วิธีการศึกษา** : ผู้ป่วยที่ได้รับการตรวจด้วยเครื่องสะท้อนในสนามแม่เหล็กของหัวใจและหลอดเลือดระหว่างเดือนมกราคม 2550 ถึง กรกฎาคม 2552 ถูกนำมาวิเคราะห์ข้อมูลผ่านจอกอมพิวเตอร์โดยระบบการเก็บข้อมูลของฝ่ายรังสีวิทยา ผู้ป่วยที่มีก้อนในหัวใจห้องล่างขวาถูกคัดเลือกมาเข้าร่วมการวิจัย ข้อมูลที่ตรวจพบจากเครื่องสะท้อนในสนามแม่เหล็กถูกเปรียบเทียบกับผลการผ่าตัด ผลทางพยาธิวิทยา และอาการของผู้ป่วยระหว่างการติดตามผล
- ผลการศึกษา** : ผู้ป่วยทั้งหมด 243 คนได้รับการตรวจด้วยเครื่องสะท้อนในสนามแม่เหล็กของหัวใจและหลอดเลือดในระยะเวลาที่ทำการเก็บข้อมูล มีผู้ป่วย 4 รายพบก้อนในหัวใจห้องล่างขวา โดยผู้ป่วย 2 รายเป็นก้อนเนื้องอกของหัวใจชนิด *malignant fibrosarcoma* และ *RV myxoma* ผู้ป่วย 2 รายเป็นก้อนเลือด (*thrombus*) ก้อนเนื้องอกมี *enhancement* หลังจากฉีดสารสารเพิ่มความทึบของภาพ ในขณะที่ก้อนเลือดไม่มี *enhancement*
- สรุป** : การตรวจด้วยเครื่องสะท้อนในสนามแม่เหล็กของหัวใจและหลอดเลือดสามารถแยกก้อนเนื้องอกและก้อนเลือดโดยใช้สารเพิ่มความทึบของภาพ
- คำสำคัญ** : ก้อนในหัวใจห้องล่างขวา, เครื่องสะท้อนในสนามแม่เหล็กของหัวใจและหลอดเลือด, *Fibrosarcoma*, *Myxoma*, ก้อนเลือด.

Intracavitary mass of the right ventricle is very uncommon. Incidence of tumor originated from right ventricle is estimated 0.13% in all primary cardiac tumors. ⁽¹⁾ Differential diagnosis of right ventricular (RV) mass depends on patient's age, underlying disease and radiological findings. Transthoracic echocardiography (TTE) is widely used for initial evaluation in patients suspected of intracardiac mass. However, TTE has several limitations such as operator dependent, poor acoustic window in patient who has large body habitus, or chronic obstructive airway disease, and poor tissue characterization. Cardiovascular magnetic resonance imaging (CMR) is currently a noninvasive tool which has excellent tissue characterization and image resolution. CMR is usually used to localize lesions and determine tumor extension before surgery. The purpose of our study is to retrospectively review CMR findings in patients diagnosed with RV mass correlated with operative and pathological findings.

Materials and Methods

All patients who were referred to receive CMR at King Chulalongkorn Memorial Hospital from Jan 1, 2007 to July 31, 2009 were retrospectively reviewed on picture archiving and communication system (PACS) monitor. All studies were examined with 1.5 Tesla MR Scanner (MR Signa Excite HD, GE, USA) with eight-channel cardiac coil placed over the anterior chest wall. The standard CMR protocol for cardiac mass included ECG-gated steady state free precession (SSFP) white blood cine images (repetitive time 3.9 msec, echo time 1.7 msec, flip angle 45°, matrix 224 x 224, field of view 360 mm x 288 mm,

section thickness 8 mm without gap) in basic cardiac views including apical long axis (2-chamber, 3-chamber, 4-chamber views) and multislice short axis views. Additional axial or coronal SSFP white blood images of the entire heart were performed. Axial or coronal double inversion recovery (IR) black blood T1-weighted images (T1-WI) (repetitive time 1200 msec, echo time 40 msec, matrix 256 x 256) and double IR T2-WI with fat suppression (repetitive time 1300 msec, echo time 90 msec, matrix 256 x 256) were obtained for evaluation anatomy and tissue characterization. 2D FGRE gadolinium first pass perfusion (repetitive time 8.9 msec, echo time 26 msec, flip angle 25°, receiver bandwidth 125 kHz) were acquired to assess tumor vascularity. Post contrast double IR T1-WI with fat suppression (repetitive time 1250 msec, echo time 40 msec, matrix 256 x 256), and inversion recovery delayed enhancement images (repetitive time 6.9 msec, echo time 3.3 msec, flip angle 20°, receiver bandwidth 31.25 kHz, inversion time 200 -280 msec, matrix 256 x 192) were performed. Clinical information, age, sex, and CMR findings were recorded. Final diagnosis was concluded by operative note, pathology report or clinical diagnosis after follow up.

Result

In total, 243 CMR studies were performed. Indications of the studies were shown in Table 1. There were 10 patients referred to exclude intracardiac mass by CMR (table 2). Two cases of RV tumor were found. Another two cases of RV thrombus from myocardial viability study were recruited (total 4 from 243 patients, 1.65% of total CMR examination).

Table 1. Indications of patients referred for cardiovascular magnetic resonance imaging.

| Indication | Number of study |
|--|-----------------|
| Post myocardial infarction | 102 |
| Myocarditis | 3 |
| Intracardiac mass | 10 |
| Coronary artery disease | 90 |
| Congenital heart disease | 27 |
| Arrhythmogenic right ventricular dysplasia | 4 |
| Hypertrophic obstructive cardiomyopathy | 1 |
| pericarditis | 2 |
| Valvular heart disease | 2 |
| Heart transplantation | 1 |
| Total | 243 |

Table 2. Patients referral to excluded intracardiac mass.

| Indication | Age (years) | Sex |
|-------------------------|-------------|-----|
| R/O intracardiac mass | 18 | M |
| RV mass | 14 | F |
| LV mass | 68 | M |
| R/O RA mass | 50 | M |
| RV sarcoma | 63 | M |
| R/O metastatic lymphoma | 68 | F |
| R/O RA mass | 67 | M |
| R/O RA mass | 37 | F |
| R/O intracardiac mass | 61 | M |
| R/O RA mass | 51 | F |

The first patient was a 61-year-old Thai male presented with one-month history of dyspnea and functional class changed from class I to IV. The patient underwent echocardiography from other hospital. Echocardiography demonstrated a large well-defined mass in right ventricle extending into right ventricular outflow tract. CMR revealed a 6.8 x 5.1 x 2.2 cm lobulated mass consisted of iso to hypersignal

intensity on T1WI, hypersignal intensity on T2WI with fat suppression, and heterogeneous enhancement (Figure 1). This mass was originated from the junction of interatrial and interventricular septum near the opening of coronary sinus and protruded through tricuspid valve into right ventricle and right ventricular outflow tract. The operative findings showed a 6 x 7-cm board based tumor originated from the Triangle

of Koch (triangular area between septal wall of right atrium, coronary sinus orifice, and tricuspid valve) extended into right ventricle. Excision biopsy with partial tumor removal was done. Pathological finding was high-grade fibrosarcoma.

The second patient was a 14-year-old Thai female presented with dyspnea and syncope. Echocardiography showed masses in right atrium and right ventricle. CMR demonstrated a 5.6 x 2.0 x 5.2-cm right atrial mass extending into superior vena cava (SVC) (Figure 2). This mass consisted of iso to hypersignal intensity on T1WI, hypersignal intensity on T2WI with fat suppression, and no contrast enhancement. Another mass in right ventricle had isosignal intensity on T1WI, hypersignal intensity on T2WI with fat suppression, and heterogeneous enhancement. This mass occupied almost in entire

right ventricle and right ventricular outflow tract (RVOT). Moderate amount of pericardial effusion was seen. Operative findings revealed a large well-encapsulated right ventricular mass. Mass in right atrium was thrombus. Total tumor removal was done. Pathology showed thrombus in right atrium and myxoma in right ventricle.

Two cases of RV thrombus were referred for assessment of myocardial viability by CMR. Both patients had extensive myocardial scar and severe right and left ventricular dysfunction. Mass was found at RV apex without contrast enhancement. These cases were diagnosed as RV thrombus (Figure 3). The patients received anticoagulant and follow up echocardiography one month later. Those thrombi were of decreased size.

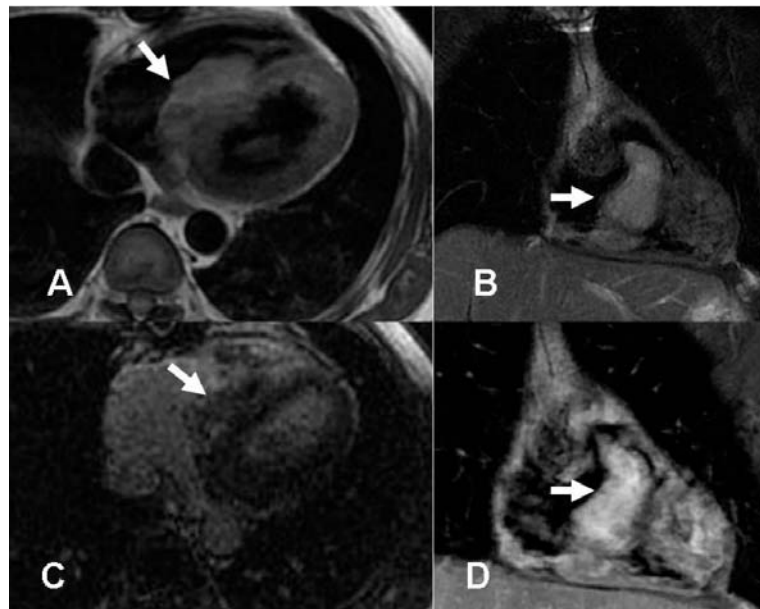


Figure 1. A case of fibrosarcoma of right ventricle (arrowed) showed right ventricular (RV) mass with iso to hypersignal intensity on T1WI (A), hypersignal intensity on T2WI with fat suppression (B), and heterogeneous enhancement on delayed enhancement imaging (C) and post contrast T1WI with fat suppression (D).

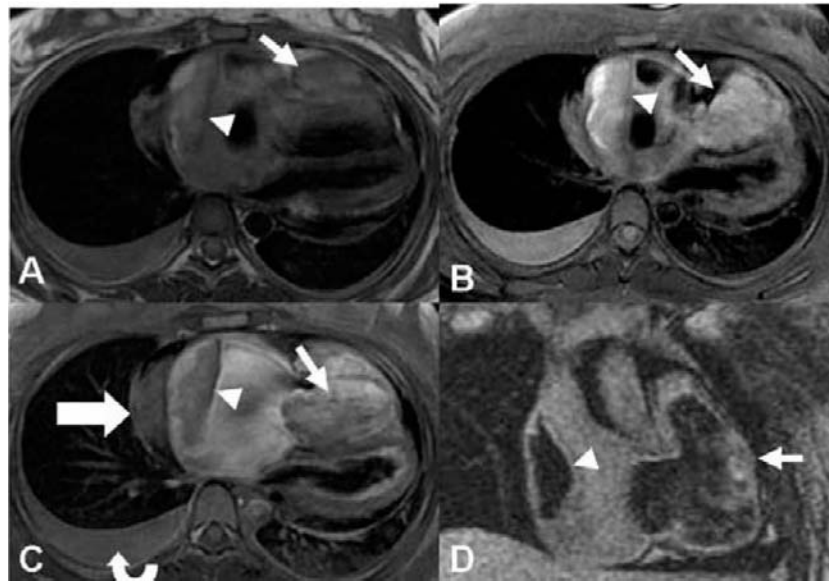


Figure 2. A case of RV myxoma (arrowed) shows isosignal intensity on T1WI (A), hypersignal intensity on T2WI with fat suppression (B), and heterogeneous enhancement on post contrast T1WI with fat suppression (C), and delayed enhancement imaging (D). Thrombus in right atrium (arrow head) consisted of iso to hypersignal intensity on T1WI, hypersignal intensity on T2WI with fat suppression (B), and no enhancement (C and D). Patient had pericardial effusion (thick arrow) and right pleural effusion (curved arrow).

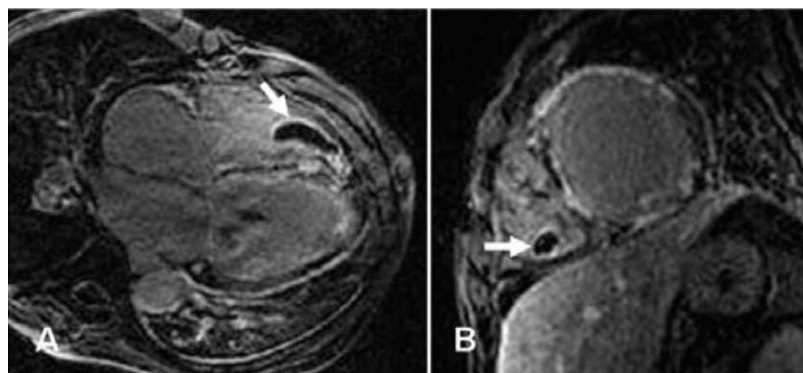


Figure 3. A case of RV thrombus shows no delayed enhancement in 4-chamber and short axis views (arrowed).

Discussion

Right ventricle is a rare location for primary cardiac tumor. The most common mass in right ventricle is thrombus. Common tumor originated in

right ventricle is rhabdomyoma and secondary deposit of angiosarcoma.⁽²⁾ Our study showed two rare cases of RV tumor detected by CMR, fibrosarcoma and RV myxoma.

Fibrosarcoma of the heart is extremely rare. Incidence of primary cardiac fibrosarcoma from autopsies is between 0.0017% and 0.35% in all primary cardiac tumors (benign and malignant) or 3.3% in the group of primary malignant cardiac tumor. ⁽³⁾ Tumor originates from mesenchymal structure of the heart with predominant fibroblastic differentiation. ⁽⁴⁾ CMR features usually show hypointensity on T1WI, hyperintensity on T2WI with fat suppression and enhancement after contrast medium injection. The iso to hypersignal intensity of the tumor on T1WI in our patient is possibly explained by intratumoral hemorrhage.

Myxoma is the most common primary benign cardiac tumor; 75 - 80% of myxoma originate in left atrium; 18% in right atrium; less than 2.8% in biatrial. ⁽¹⁾ Less than 3 - 4% are located in ventricle. ⁽⁵⁾ Unusual location of myxoma associates with familial myxoma syndrome or Carney's syndrome consisted with multiple cardiac myxoma with atypical location, endocrine overactivity, skin hyperpigmentation, and extracardiac tumors. ⁽⁶⁾ Symptom of RV myxoma is nonspecific. If tumor obstructed RV outflow tract, it may lead to syncope, pulmonary embolism, and sudden death. ⁽⁷⁾ Pericardial effusion in our patient of RV myxoma was explained by RVOT obstruction and right sided heart failure. Typical CMR features of myxoma show hypointense on T1WI, hyperintense on T2WI and heterogeneous enhancement due to high extracellular water content. ⁽²⁾

Benefits of CMR in cardiac mass are to differentiate tumor and thrombus, and determine tumor extension especially pericardial or extracardiac invasion. We found that contrast enhancement can accurately differentiate tumor from thrombus.

Our review shows no contrast enhancement in all intracardiac thrombi whereas all tumors have heterogeneous enhancement. Evaluation of tumor extension by CMR is also useful. CMR can assess local extension to adjacent structures such as pericardium or extracardiac metastasis to the lung or liver. We, however, found that differentiation between benign and malignant tumors of right ventricle by using CMR is difficult. Our benign and malignant cases had similar tumor signal intensities in both T1WI, T2WI with fat suppression and post contrast enhancement. Tissue characterization of RV tumor is impossible for specific diagnosis. Patient's age, underlying disease, location and extension of tumor are still important for differential diagnosis primary cardiac tumor.

Conclusion

Primary cardiac tumor in right ventricle is rare. CMR has benefits in evaluation of tumor extension and differential diagnosis of tumor versus thrombus by using contrast enhancement. CMR findings of benign and malignant RV tumors by are unspecific and difficult to characterize by CMR.

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