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CME Article

Clinics in diagnostic imaging (126)

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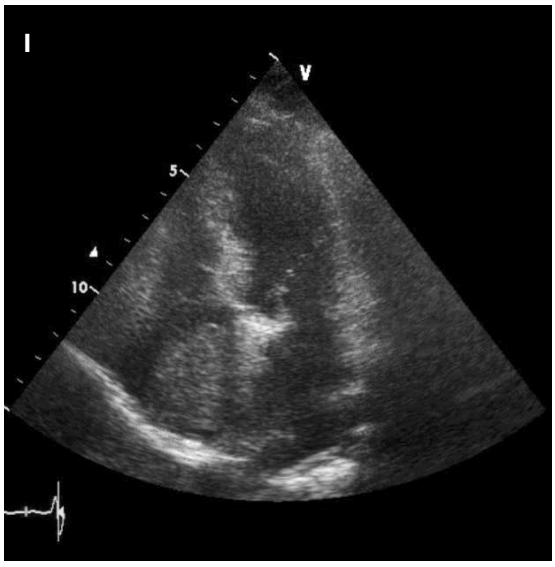

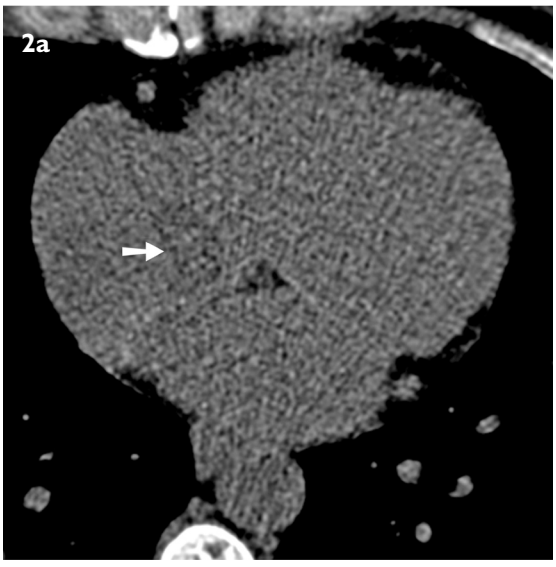

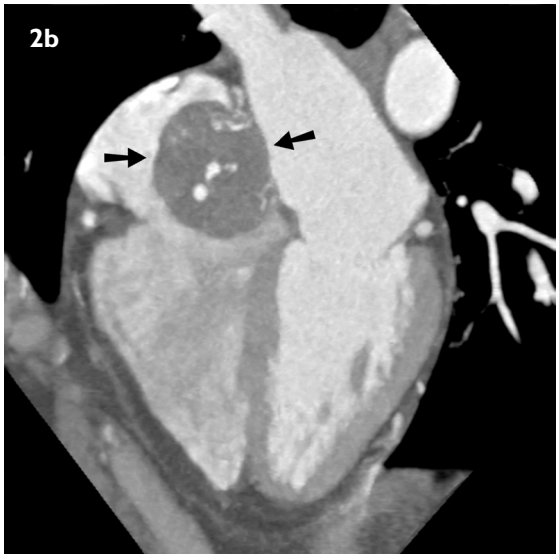

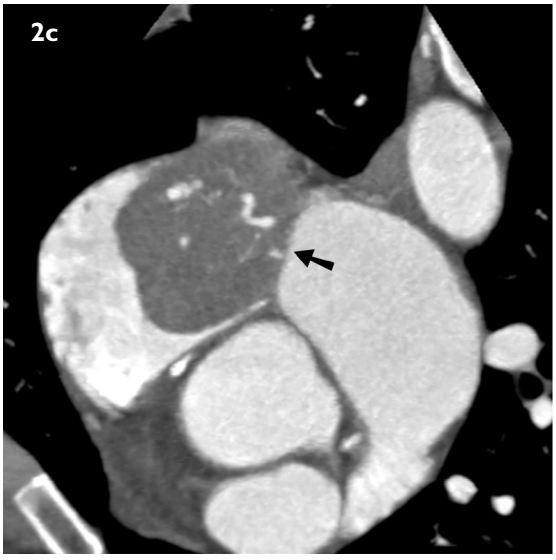

Fig. 1 2D echocardiogram shows the four-chamber view of the heart.

Fig. 2 (a) Non-contrast enhanced multidetector computed tomography (MDCT) image of the heart. (b) Post-IV contrast-enhanced four-chamber view maximum intensity projection (MIP) MDCT image. (c) Short axis MIP MDCT image through the atria.

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CLINICAL PRESENTATION

A 47-year-old woman presented with dyspnoea on exertion for several weeks. She was a non-smoker with a history of hypertension. She underwent laparoscopic cholecystectomy one month prior to presentation. An echocardiogram (Fig. 1) was performed and she was referred for surgical consultation and work-up. An electrocardiogram (ECG) gated, intravenous (IV) contrast-enhanced dual source multidetector computed tomography (MDCT) (Definition™, Siemens Medical Systems, Erlangen, Germany) of the heart and thorax was performed (Figs. 2 a–c). What is the diagnosis?

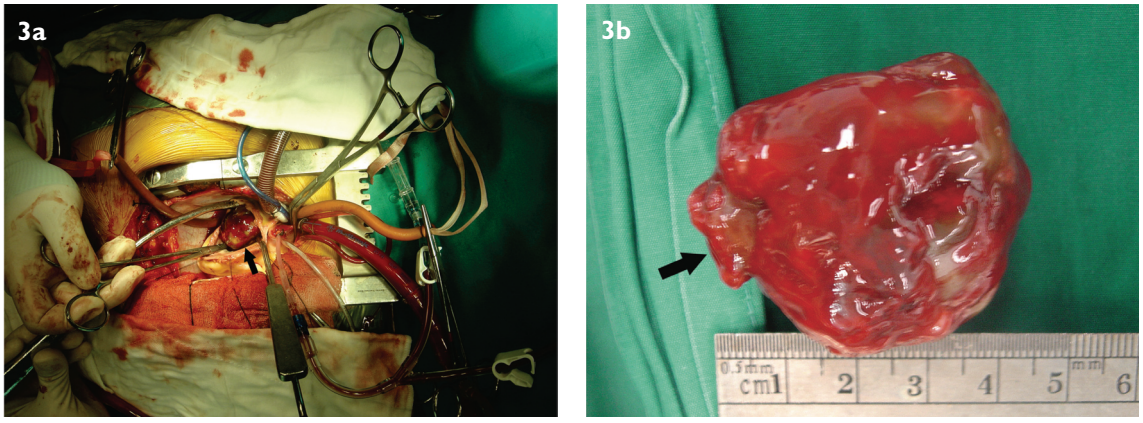

Fig. 3 (a) Operative photograph shows the tumour (black arrow) in the right atrial cavity. (b) Close-up photograph of the resected atrial myxoma shows the stalk where the tumour was attached to the atrial septum (black arrow).

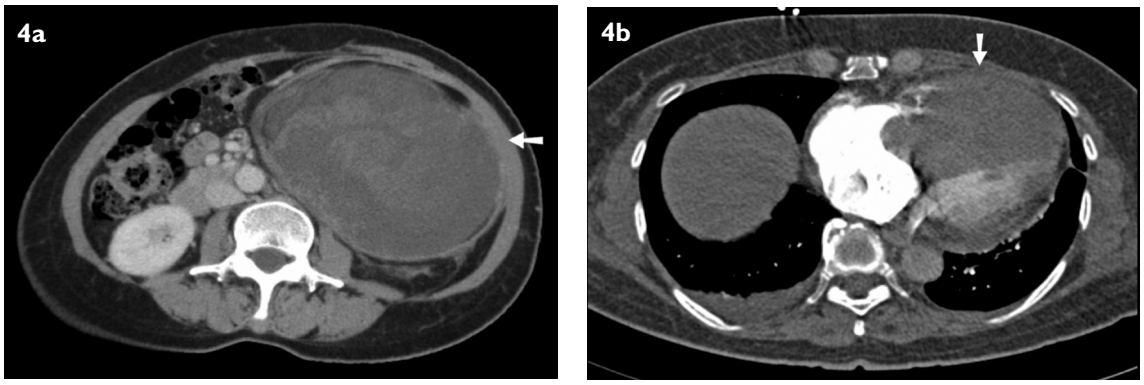

Fig. 4 (a) Contrast-enhanced axial CT image of a 59-year-old woman with a known primary retroperitoneal liposarcoma (white arrow) who presented two years after initial diagnosis of her primary malignancy with acute breathlessness. (b) Contrast-enhanced axial CT image shows non-opacification of the right ventricle due to a soft tissue mass that completely fills and expands the right ventricle.

IMAGE INTERPRETATION

The echocardiogram showed a well-defined hyperechoic mass in the right atrium (Fig. 1). The non-contrast enhanced MDCT showed a subtle 4 cm non-calcified mass in the right atrium (Fig. 2a). Contrast-enhanced MDCT showed the mass to have well-defined margins (Figs. 2b & c) and demonstrated attachment of the mass to the fossa ovalis region of the interatrial septum (Fig. 2b; thin black arrow). The mass was predominantly hypodense but contained several serpiginous, enhancing tumour vessels. There was no tumour extension into the superior or inferior vena cava, or any evidence of pulmonary embolism.

DIAGNOSIS

Right atrial myxoma

CLINICAL COURSE

Surgical resection of the mass was uncomplicated (Fig. 3a). Both the gross appearance (Fig. 3b) and histological analysis were in keeping with an atrial myxoma. The patient was well when discharged five days after surgery. No follow-up imaging was required.

DISCUSSION

The commonest causes of a right heart mass in adults in descending order of likelihood are metastasis, myxoma, malignant primary cardiac tumour and thrombus. The physical appearance and location of the mass are important distinguishing factors and MDCT is an ideal modality to demonstrate this. The site of attachment of the mass is a particularly important distinguishing feature.

Atrial myxoma is the commonest primary tumour of the heart and the most frequent intracavitary primary neoplasm. It is commonly diagnosed as an incidental finding during an echocardiogram. In symptomatic patients, the classical presentation has been described as a triad, which includes symptoms due to the obstruction of cardiac blood flow, embolic phenomena

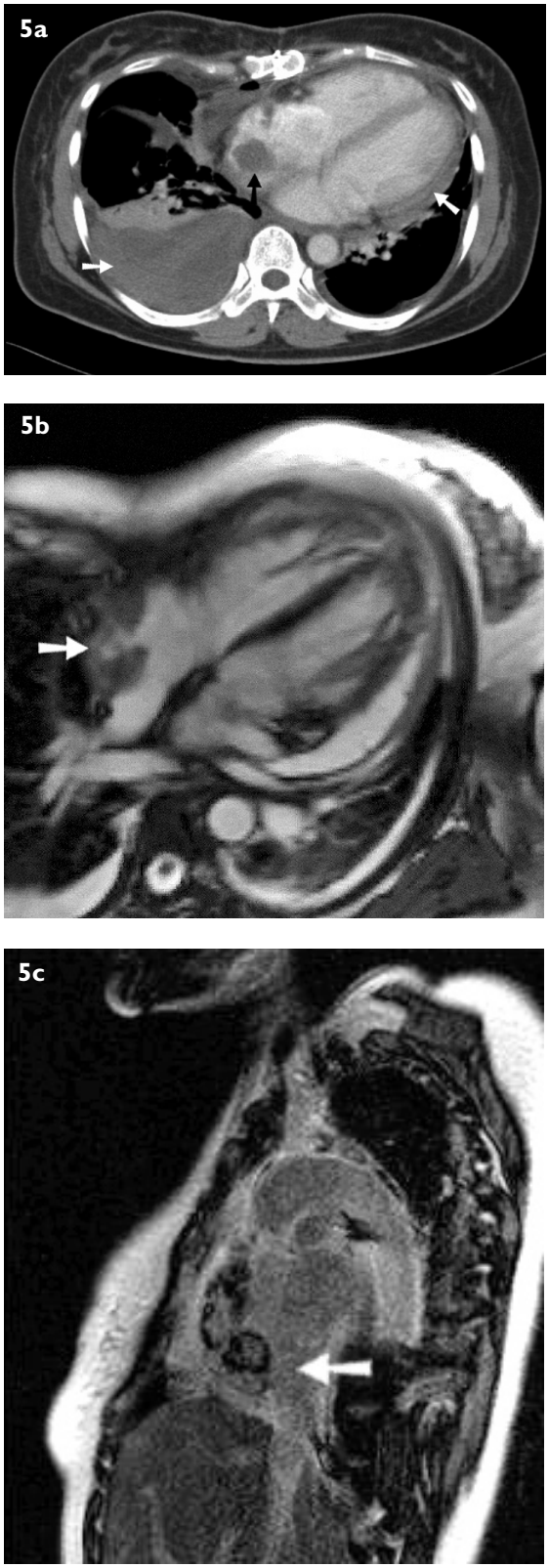

Fig. 5 A 42-year-old Malay woman who presented with chest pain. (a) Contrast-enhanced axial CT shows a hypodense mass in the right atrium (black arrow). A pericardial effusion and right pleural effusion are present (white arrows). (b) T2-W four-chamber view MR image of the same patient shows a broad-based mass with an irregular surface (arrow) along the lateral and posterior walls of the right atrium. (c) Short-axis post-gadolinium inversion recovery gradient echo sequence through the right atrium shows the enhancement of the mass (arrow). Histology was right atrial angiosarcoma.

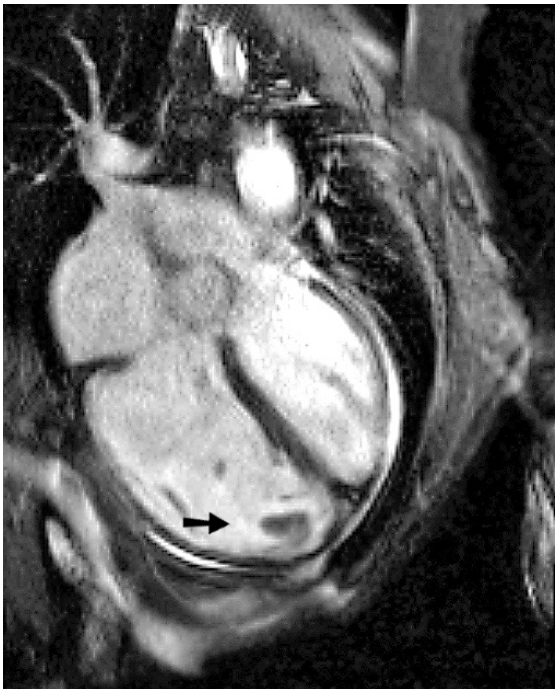

Fig. 6 Four-chamber view of a bright blood MR image cine sequence of a 60-year-old man with arrhythmogenic right ventricular dysplasia (ARVD) shows significant dilatation of the right ventricle (RV) and a hypointense thrombus in the apical part of the RV (arrow).

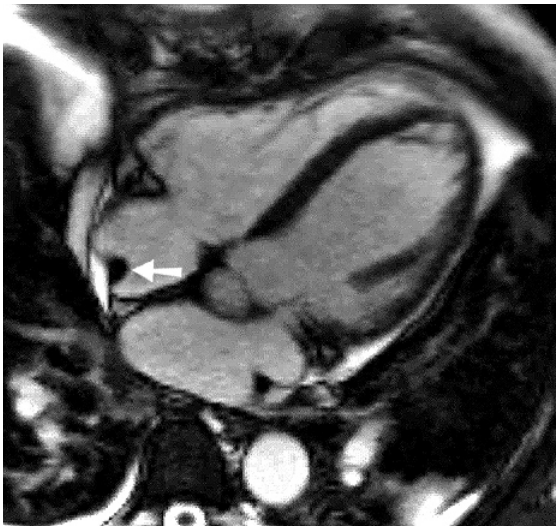

Fig. 7 Axial MR image shows the crista terminalis (arrow) – a normal embryological remnant and mimic of a right atrial mass.

and constitutional symptoms.[1] There is a slight female preponderance and patients are typically in the age range of 30–60 years. The majority of atrial myxomas (75% of cases) are found in the left atrium. The right atrium, as in the case illustrated above, is a less common site (20% of cases).[2] The diagnosis of an atrial myxoma can usually be made on its radiological appearance alone, due to its characteristic location and attachment to the interatrial septum at the border of the fossa ovalis. In

addition, atrial myxomas tend to be spherical or ovoid with smooth lobulated borders, which is typical of benign cardiac tumours. This is in contrast to malignant tumours, which have a more irregular and infiltrative appearance. Although not demonstrated in this case, it has been reported that radiographically-apparent tumoural calcification is more common in right atrial myxomas compared to the left.[1] Thus, a radiologist should include myxoma as a differential diagnosis when faced with a calcified mass on the right side of the heart.

The commonest cause of a right heart mass is a metastatic mass (Figs. 4a & b). The commonest route is via transvenous extension through the inferior vena cava, such as in hepatocellular carcinoma and renal cell carcinoma. Spread may also occur via the haematogenous route, lymphatic channels or via direct extension from a mediastinal neoplasm. The presence of a primary tumour is normally apparent by the time it metastasises to the heart, making the diagnosis obvious. Primary malignant tumours in the right atrium are mainly soft tissue sarcomas, with angiosarcoma being the most common subtype in adults (Figs. 5a–c). The characteristics of malignant primary tumours are heterogeneous enhancement, multichamber involvement, intramural spread, extension into pulmonary veins or arteries, broad attachment, pericardial effusion (often haemorrhagic) and thickening, features of mediastinal invasion such as lymphadenopathy, and lastly, pulmonary metastases. Thrombi, although overall the most frequent cardiac masses, are uncommon on the right side of the heart and are usually iatrogenic due to central, venous lines or dilated cardiomyopathy (Fig. 6). The physical appearance of a thrombus varies greatly depending on its haemosiderin content, which is age-related. Nevertheless, thrombi tend to demonstrate "layering" against the myocardium, close attachment to the catheter tip, rarely enhance and are usually adjacent to areas of abnormal wall motion seen on cine images. They can be confirmed on MR imaging using a delayed contrast-enhanced sequence with a long inversion time, which will demonstrate the thrombus as being hypointense relative to the surrounding myocardium and slow flow or stagnant blood.[3]

The crista terminalis is an example of a pseudomass that can be mistaken for a right atrial lesion (Fig. 7). The crista terminalis is a fibromuscular ridge formed by the junction of the sinus venosus and primitive right atrium. It is found on the posterolateral wall of the right atrium extending between the openings of the superior and inferior vena cavae in a craniocaudal direction.

In summary, atrial myxoma is the second most common cause of a right heart mass after a metastasis. Its characteristic radiological appearance often allows for diagnosis based on imaging alone. Thrombi are uncommon on the right side of the heart and are usually associated with either a central venous line or right-sided cardiomyopathy.

ABSTRACT

A 47-year-old woman presented with dyspnoea on exertion for several weeks. Echocardiogram and multidetector computed tomography of the heart showed a right atrial mass typical of an atrial myxoma. This was confirmed on histology. The imaging features of atrial myxoma and other conditions presenting as a right-sided cardiac mass are discussed.

Keywords: atrial myxoma, cardiac tumour, heart neoplasms, myxoma, right atrial mass

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SINGAPORE MEDICAL COUNCIL CATEGORY 3B CME PROGRAMME
Multiple Choice Questions (Code SMJ 200905B)

	True	False
Question 1. The following statements regarding atrial myxoma are true:		
(a) It is more common in the right atrium.	☐	☐
(b) Right-sided lesions are more often calcified compared to left-sided lesions.	☐	☐
(c) It is commonly asymptomatic.	☐	☐
(d) It is the most common intracavitory primary cardiac tumour.	☐	☐
Question 2. Regarding right heart masses:		
(a) Cardiac thrombus is more common in the right side of the heart compared to the left.	☐	☐
(b) Primary malignant tumours are the most common cause of a right heart mass.	☐	☐
(c) A primary malignant cardiac mass is more common than a metastatic cardiac mass.	☐	☐
(d) A crista terminalis is an example of a pseudomass that can be mistaken for a right atrial lesion.	☐	☐
Question 3. The following are imaging features of a primary malignant cardiac tumour:		
(a) Pericardial effusion.	☐	☐
(b) Homogeneous enhancement.	☐	☐
(c) Single-chamber involvement.	☐	☐
(d) Broad attachment.	☐	☐
Question 4. Regarding a cardiac thrombus:		
(a) It is overall the most frequent cardiac mass.	☐	☐
(b) It can be associated with central venous lines.	☐	☐
(c) It usually enhances.	☐	☐
(d) MR imaging is a useful modality for diagnosis.	☐	☐
Question 5. The following statements regarding cardiac metastasis are true:		
(a) Diagnosis is usually obvious due to the presence of a primary tumour.	☐	☐
(b) The haematogeneous route is the most common route of metastasis.	☐	☐
(c) It is an uncommon cause of a right heart mass.	☐	☐
(d) It can be found in renal cell carcinoma and hepatocellular carcinoma.	☐	☐

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SUBMISSION INSTRUCTIONS:
(1) Log on at the SMJ website: http://www.sma.org.sg/cme/smj and select the appropriate set of questions. (2) Select your answers and provide your name, email address and MCR number. Click on "Submit answers" to submit.

RESULTS:
(1) Answers will be published in the SMJ July 2009 issue. (2) The MCR numbers of successful candidates will be posted online at www.sma.org.sg/cme/smj by 15 July 2009. (3) All online submissions will receive an automatic email acknowledgment. (4) Passing mark is 60%. No mark will be deducted for incorrect answers. (5) The SMJ editorial office will submit the list of successful candidates to the Singapore Medical Council.

Deadline for submission: (May 2009 SMJ 3B CME programme): 12 noon, 7 July 2009.