My Patient Has a Cardiac Mass!

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Identification of an intracardiac mass on ECHO during the evaluation of a patient with potential symptoms or signs of cardiac disease is an unusual occurrence with profound implications due to the high likelihood of significant pathology. The differential diagnosis includes imaging artifacts, normal anatomic variants, tumour, thrombus, or vegetation. Attention to the characteristics of the intracardiac mass, including the size, shape, location, attachment site, mobility and presence of hemodynamic derangements, in conjunction with knowledge of the clinical setting, are usually sufficient to determine the likely etiology.

Imaging Artifacts and Normal Anatomic Variants

Imaging artifacts are a common occurrence with echocardiography and can simulate an intracardiac mass. These artifacts frequently occur in patients with suboptimal images or a technically difficult study, and often result in the report of an ill-defined, possible or suspected intracardiac mass. Further investigation by transesophageal echocardiography (TEE), which has improved spatial resolution, can usually clarify if a suspected intracardiac mass is real or artifactual.

Variants of normal anatomic structures can be misinterpreted as a pathological intracardiac mass and this possibility must always be considered in the differential diagnosis (Table 1). Thorough knowledge of the anatomy and common variants of anatomic structures in each cardiac chamber by the imaging physician can usually avoid confusion. Additional imaging with TEE, CT scan or MRI scan can usually clarify any uncertainty.

Rachel's case

Rachel is a 70-year-old female who presents with a one-year history of progressive exertional dyspnea and cough. She is a non-smoker and has no history of cardiac disease. She had received several courses of antibiotics for presumed respiratory tract infections without improvement. One week earlier, she suffered a transient loss of vision involving her right eye. A clinical exam is normal with no murmurs or extra heart sounds. A 12-lead ECG demonstrates normal sinus rhythm and a chest X-ray was unremarkable. An ECHO identifies a large mass in the left atrium attached to the atrial septum (Figure 1).

For more on Rachel, see page 17.

Cardiac Tumours

Primary cardiac tumours are rare, with an incidence of 0.001% to 0.030% in autopsy studies. However, identifying that an intracardiac mass is a tumour is essential, due to the necessity for surgical intervention. Approximately 75% to 85% of primary cardiac tumours are benign and only 15% to 25% malignant (Table 2).

Myxomas

Cardiac myxomas are the commonest primary cardiac tumour and account for approximately 50% of all benign cardiac tumours in adults. They are endocardial-based neoplasms that arise from mesenchymal multipotential cells. Myxomas predominantly occur in women between the ages of 40 to 70 years. Most are found in the left atrium (75% to 80%), arise from the atrial septum and protrude

into the left atrial cavity. Less frequently, myxomas are found in the right atrium (15% to 20%) or ventricles (< 5%).

Patients typically present with at least one of a triad of symptoms:

1. obstructive cardiac symptoms due to tumour obstruction of the left atrium or mitral valve (i.e. exertional dyspnea, pulmonary edema, syncope);
2. embolic phenomena (i.e. transient ischemic attack or stroke); or
3. constitutional symptoms (i.e. fever, fatigue, weight loss) due to the release of tumour products or an autoimmune response.

A diastolic murmur from flow obstruction across the mitral valve can be heard in 50% of patients with left atrial myxoma. A tumour plop, or diastolic filling sound with a similar timing to S3, is heard in 15% of patients. ECG is not helpful in making the diagnosis. Transthoracic echocardiography (TTE) is the technique of choice for the initial investigation of a patient with a possible intracardiac mass and has a sensitivity of > 95% for detecting a cardiac myxoma (Figure 1). Demonstration of a left atrial mass attached to the atrial septum is essentially diagnostic for an atrial myxoma. TEE can better visualize the attachment site of an atrial mass if this is unclear on the TTE.

Cardiac myxomas should be treated by surgical resection and the operative mortality is small. Tumour recurrence is observed in 1% of patients with a sporadic myxoma and annual surveillance with TTE is recommended.

There is a familial form of cardiac myxoma (< 7% of patients) associated with pigmented skin lesions and endocrine tumours (Carney's complex). Higher recurrence rates after surgical resection have been reported (12% to 22%). Screening of first-degree relatives by TTE is recommended because of the autosomal dominant transmission.

Table 1

Normal anatomic variants commonly misinterpreted as a pathological intracardiac mass

Right Atrium
Chiari network
Eustachian valve
Crista *terminalis*
Lipomatous atrial septum
Fatty tricuspid valve *annulus*
Left Atrium
Atrial septal aneurysm
Transverse or coronary sinus
Pectinate muscles
Ridge between the left atrial appendage
Left upper pulmonary vein
Thoracic aorta
Left Ventricle
Muscle bundles / trabeculations
Accessory papillary muscles
Right Ventricle
Moderator band
Papillary muscles
Trabeculations
Valves
Nodule of Arantius
Lambl's excrescence

Papillary Fibroelastomas

Papillary fibroelastomas are the second commonest benign cardiac tumour and the commonest valve tumour. They are likely underreported because of their small size (usually < 1cm). Papillary fibroelastomas are avascular papillary structures lined by endothelial cells that often have a short pedicle, which results in mobility. They are usually found in adults (> 30 years) on the surface of the aortic or mitral valves, although 15% occur on non-valvular endocardial surfaces. Papillary fibroelastomas are often diagnosed incidentally at the time of surgery or autopsy, or when an echocardiogram is per-

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Table 2
Common Primary Cardiac Tumours

Benign (75% to 85%)
- Myxoma
- Papillary Fibroelastoma
- Lipoma
- Fibroma
- Rhabdomyoma (commonest pediatric tumour)

Malignant (15% to 25%)
- Sarcoma (angiosarcoma commonest type)
- Lymphoma
- Mesothelioma

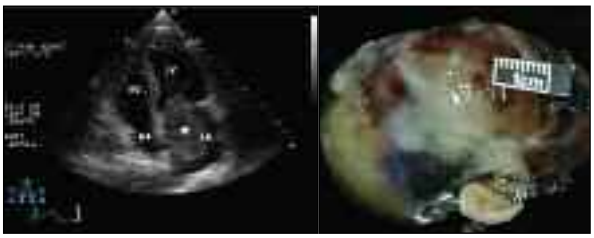

Figure 1. Left atrial myxoma (*) with attachment to the atrial septum visualized on transthoracic ECHO (left) and pathology (right).
(LA = left atrium, LV = left ventricle, RA =right atrium, RV = right ventricle)

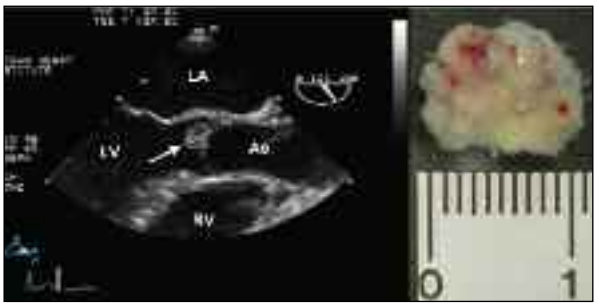

Figure 2. Papillary fibroelastoma (arrow) on the aortic valve visualized by transesophageal echocardiography (TEE) (left) and pathology (right).
(Ao = aorta, LA = left atrium, LV = left ventricle, RV = right ventricle)

formed for an unrelated reason. However, embolization of tumour or thrombus on the tumour can cause symptoms and is reported in > 25% of patients. Rarely, occlusion of the coronary ostium by the tumour can result in cardiac ischemia or sudden death. ECHO is the imaging modality of choice for detecting papillary fibroelastomas (Figure 2). TEE has a better sensitivity than TTE because of the small tumour size.

Papillary fibroelastomas should be surgically removed if a patient has had an embolic event. The native valve can usually be spared. Management of an asymptomatic papillary fibroelastoma is controversial. Left-sided papillary fibroelastomas should likely be removed if they are large (> 1cm) or mobile due to the potential risk of embolization.

Angiosarcomas

Only 15% to 25% of primary cardiac tumours are malignant and 95% of these are sarcomas. Angiosarcomas are the most common primary malignant cardiac tumour in adults. They have a male predominance, usually occurring in individuals between the 3rd and 5th decade. The right heart is preferentially involved (75% of patients) with a predilection for the right atrium. These aggressive intramural tumours grow rapidly, usually extending into the cavity and pericardium. The clinical manifestations depend on the specific tumour site, and may result from blood flow obstruction, local invasion (pericardial effusion/tamponade, arrhythmias), embolization, or may be non-specific (i.e. chest pain, dyspnea, fever or weight loss). Angiosarcomas often appear as an echogenic, irregular, broad based infiltrating right atrial mass on TTE (Figure 3). CT scan or MRI scan can usually better demonstrate tumour infiltration and potential extracardiac extension because of their wider field of view.

Patients with angiosarcoma have a poor prognosis as most have metastases at the time of diagnosis. Average survival is only 6 months to 12 months. Complete surgical resection is the treatment of choice; however, this is rarely possible, and most patients will develop recurrent disease despite resection.

Metastatic Tumours

Cardiac involvement from metastatic disease is 20 times to 40 times more common than a primary

cardiac tumour. Lung, breast, melanoma, leukemia, lymphoma, or kidney tumours are the most common malignancies associated with cardiac metastases. Tumour can spread to the heart by direct local extension, lymphatic channels, hematogenous spread, or cavoatrial spread. The vast majority of cardiac metastases are epicardial and clinically silent, rather than presenting as an intracardiac mass. When symptoms manifest, they usually relate to pericardial involvement. The notable exception is renal cell carcinoma, which can extend up the inferior vena cava into the right atrium. Carcinoma of the lung can also invade the pulmonary veins and extend into the left atrium.

Intracardiac Thrombus

Intracardiac thrombus is most likely to develop in the left ventricle or left atrium. It can be detected by ECHO and can occasionally be confused for a cardiac tumour when located in the left atrium (Figure 4). Left atrial thrombus is generally located in the appendage or on the posterolateral wall and has no stalk. In contrast, an atrial myxoma is usually attached to the atrial septum and has a stalk. The clinical setting is invaluable in differentiating these two entities. Intracardiac thrombus is almost always associated with atrial fibrillation, mitral valve disease, left atrial enlargement or left ventricular dysfunction. Intracardiac thrombus should be treated with anticoagulation to target an international normalized ratio of 2 to 3, or 2.5 to 3.5 if a mechanical mitral valve is present.

Vegetations

The hallmark lesion of infective endocarditis is the vegetation, a mass of microorganisms, fibrin, and inflammatory infiltrates on the endocardial surface. Echocardiographically, vegetations appear as mobile irregular echogenic masses that are usually attached to a valve. Distinguishing whether a valve mass represents a vegetation or papillary fibroelastomas can be difficult. Vegetations usually occur on the upstream (low pressure) side of the valve, attach along the line of leaflet coaptation, may be multiple and are often associated with valve destruction or perivalvular extension. In con-

More on Rachel's case

Echocardiography demonstrated a large left atrial mass attached to the atrial septum, which was diagnostic for left atrial myxoma (Figure 1, left). The patient underwent urgent surgery with removal of the left atrial mass and pathology confirmed the diagnosis of atrial myxoma (Figure 1, right). Her post-operative course was uncomplicated. She has an annual transthoracic echocardiogram to evaluate for recurrent tumour.

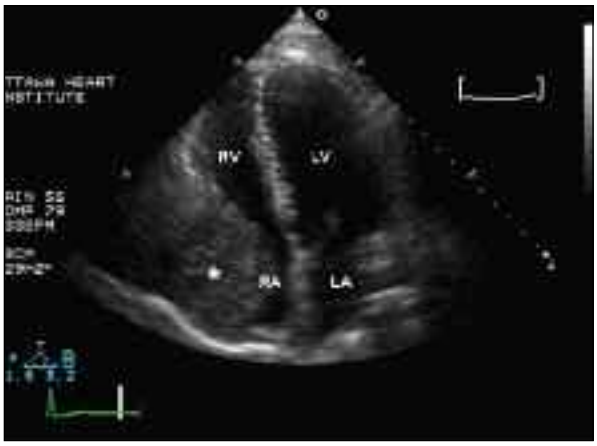

Figure 3. Right atrial angiosarcoma (*) visualized by transthoracic echocardiography (TTE).
(LA = left atrium, LV = left ventricle,
RA =right atrium, RV = right ventricle)

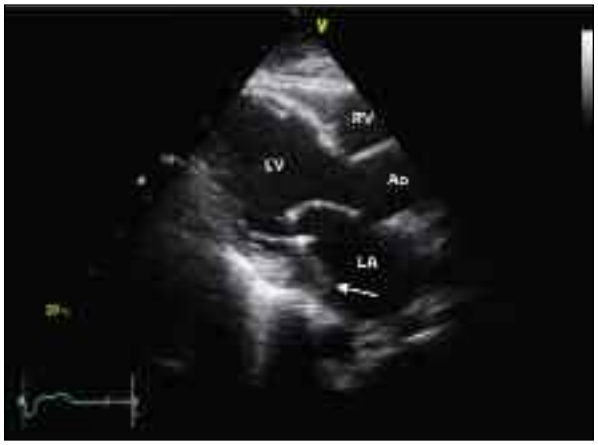

Figure 4. Left atrial thrombus (arrow) in a patient with previous mitral valve repair visualized on TTE.
(Ao = aorta , LA = left atrium, LV = left ventricle,
RV = right ventricle)

trast, papillary fibroelastomas often occur on the downstream side of the valve, attach to the mid leaflet, are usually solitary and are uncommonly associated with valve dysfunction. However, the clinical context is the most important distinguishing feature. Vegetations usually occur in the setting of infection, while papillary fibroelastomas are often identified in patients who are asymptomatic or who have had a potential embolic event. Other non-infective valve masses, such as non-bacterial thrombotic endocarditis or Libman Sacks endocarditis associated with systemic lupus erythematosus, can usually be distinguished from infective endocarditis by the clinical setting.

Patients with a suspected vegetation of infective endocarditis should have blood cultures drawn and receive intravenous antibiotic therapy directed at the inciting organism.

Resources
1. Burke A, Jeudy J, Virmani R. Cardiac tumours: an update. Heart 2008;94:117-123.
2. Butany J, Nair V, Naseemuddin A, et al. Cardiac Tumours: Diagnosis and Management. Lancet Oncol 2005;6:219-228.
3. Peters PJ, Reinhardt S. The Echocardiographic Evaluation of Intracardiac Masses: a Review. J Am Soc Echocardiogr 2006;19:230-240