Clinical presentation and non-invasive diagnosis of right heart masses

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SUMMARY Right-sided cardiac masses do not have a uniform clinical presentation. Successful diagnosis is dependent on a high index of suspicion and appropriate echocardiographic examination. Five cases of right sided intracardiac masses have been detected during the period that echocardiography has been routinely available to us—two of right atrial myxoma, one of right atrial thrombus, and two of right ventricular tumour. M-mode echocardiography identified four of the five cases. Two-dimensional echocardiography was necessary to establish the non-invasive diagnosis in the fifth case. Echocardiography should be used in any patient in whom a right sided mass is suspected, or in those patients presenting with signs of tricuspid regurgitation, tricuspid stenosis, pulmonary stenosis, cyanotic heart disease, progressive right heart failure, constrictive pericarditis, or pulmonary emboli without obvious source.

Although the existence of cardiac tumours has been recognised for over 200 years, until 1950 the diagnosis was believed to be made exclusively at necropsy.1 Prichard wrote that the diagnosis of tumours of the heart “is either impossible or a matter of chance”.1 In 1952, an intracardiac myxoma was first recognised by angiography,2 and in 1955, the first report of excision of an atrial myxoma using cardiopulmonary bypass appeared.3 Since that time, the clinical features of left atrial myxoma have been extensively reported,4-6 and non-invasive methods, including M-mode echocardiography,7 radionuclide angiography,8 9 and phonocardiography,10 have increased the sensitivity of detection of these tumours.

Right sided cardiac masses are less often seen and only scattered reports have suggested the value of M-mode echocardiography in the detection of right atrial11-13 and right ventricular tumours.14 This report describes the non-invasive characteristics of five right sided cardiac masses, and documents that a clinical suspicion may be easily and effectively substantiated.

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Case reports

CASE 1

A 45-year-old woman was admitted for excision of a left parotid tumour. The chest x-ray film showed an incidental calcified intracardiac mass. Though this patient had no cardiac symptoms, she had known of a heart murmur since childhood. She had had a pulmonary embolus nine years before admission after an uneventful childbirth.

Jugular venous engorgement was evident with a prominent systolic regurgitant wave. A grade 3/6 diastolic rumbling murmur with presystolic accentuation was heard best in the second right intercostal space at the sternal border. A systolic murmur was heard intermittently.

After recording the characteristic motion of a normal aortic root with an M-mode echocardiogram, inferior and medial angulation of the transducer disclosed multiple parallel echoes in the region where motion of the tricuspid valve is usually recorded (Fig. 1). In this patient, however, the expected tricuspid leaflet motion pattern was never visualised.

A calcified myxoma was removed from the right atrium. After operation a repeat echocardiogram showed normal tricuspid leaflet motion.

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CASE 2
A 40-year-old woman had a six month history of progressive dyspnoea on exertion, of fatigue, and of weight gain. A two component pericardial friction rub was heard and she was treated with aspirin and then prednisone without much benefit.

Physical examination disclosed a jugular systolic regurgitant wave, bilateral basal râles, a two component friction rub, and a mid-systolic sound.

M-mode echocardiography was at first considered to be normal. Motion of the tricuspid valve could never be recorded, and a pattern of extra echoes from the region of the valve (Fig. 2) was ignored as artefact. Two-dimensional echocardiography with a wide angle phased array ultrasonograph, however, showed a large multilobulated mass in the right atrium which prolapsed into the right ventricle in diastole (Fig. 3). An M-mode echocardiogram obtained from the two-dimensional machine by manually positioning the interrogating ray through the right-sided chambers and tumour mass was identical to that initially recorded by the M-mode machine (Fig. 2).

A 4×10 cm myxoma was removed from the right side of the interatrial septum. Postoperatively, the friction rub was no longer present, and echocardiographic examination disclosed a normal tricuspid valve.

CASE 3
A 69-year-old man with polycythaemia vera was admitted to hospital for increasing abdominal girth and a Budd-Chiari syndrome. After an initially successful side-to-side portocaval shunt had thrombosed, a LeVeen shunt was inserted, and the ascites decreased rapidly. After two months, however, the patient again noted increasing girth.

Physical examination disclosed jugular venous distension with a systolic regurgitant wave. A presystolic gallop, mid-diastolic sound, and grade 1/6 systolic murmur were heard along the left sternal border. Massive ascites and significant ankle oedema were also present.

M-mode echocardiography showed a mass behind the tricuspid valve with an initial clear space in diastole (Fig. 4). A two-dimensional echocardiogram confirmed the presence of a mass, 3 to 4 cm, in the right atrium attached to the end of the LeVeen shunt (Fig. 5). A superior vena cavagram and injection through the LeVeen shunt disclosed what was presumed to be a large thrombus at the end of the catheter which had migrated into the right atrium.

During treatment with continuous heparin infusion serial two-dimensional echocardiograms showed diminution in the size of the mass.

CASE 4
A 53-year-old man with progressive dyspnoea on exertion had been aware of a heart murmur for four years. She was admitted to hospital because of praecordial pain unrelated to exertion.

Physical examination disclosed a thrill along the left sternal border where there was a grade 4/6 harsh
Fig. 2  M-mode echocardiogram obtained from a 40-year-old woman with a right atrial myxoma showing numerous echoes practically filling the right atrial cavity. (Reproduced with permission from Cohen MV. Correlative Atlas of Adult Cardiac Disorders: Noninvasive Diagnostic Techniques. Mt. Kisco: Futura Publishing Company, 1980.)

Fig. 3  Systolic and diastolic frames from an apical, four chamber real time two-dimensional echocardiogram recorded in the same patient with a right atrial (RA) myxoma described in Fig. 2. The tumour prolapses into the right ventricle (RV) in diastole. (Reproduced with permission from Cohen MV. Correlative Atlas of Adult Cardiac Disorders: Noninvasive Diagnostic Techniques. Mt. Kisco: Futura Publishing Company, 1980.) IV, interventricular; LA, left atrium; LV, left ventricle; MV, mitral valve; Pul, pulmonarv.
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Fig. 4 M-mode echocardiogram from a 69-year-old man with a thrombus at the end of a LeVeen shunt which had migrated into the right atrium, showing the anterior tricuspid valve leaflet (ATL) and the thrombus behind it. This echocardiogram with the initial clear space in diastole before the mass prolapses through the valve annulus is indistinguishable from that of a right atrial myxoma. This clear space is caused by the delay of movement related to the inertia of the mass, and is independent of whether the mass is thrombus or tumour.

The M-mode echocardiogram disclosed hypertrophy of the interventricular septum (17 mm). There was a mass of echoes in the right ventricular outflow tract (Fig. 6) which could not be detected in the body of the right ventricle. The abnormal mass appeared to extend further into the outflow tract in late systole. Examination of the pulmonary valve showed a normal “a” wave and high frequency systolic fluttering (Fig. 7).

The right ventricular angiogram showed a large, pedunculated, lobulated mass in the outflow tract of the right ventricle. There was an infundibular outflow tract gradient of 70 mmHg.

Removal of a leiomyoma abolished the echoes from the right ventricular outflow tract.

Fig. 5 Frame from the apical, four chamber two-dimensional echocardiogram from the same patient described in Fig. 4. The thrombus prolapses from the right atrium (RA) into the right ventricle (RV) in diastole. IV, interventricular; LA, left atrium; LV, left ventricle; MV, mitral valve.

Case 5
A 5-day-old infant was noted to have cyanotic attacks on the second day of life which progressed to generalised cyanosis on the third day.

The baby was slightly cyanotic with a respiratory rate of 60 per minute. A lower left sternal border heave and grade 2/6 systolic ejection murmur loudest in the pulmonary area and absent at the apex were found.

There were multiple abnormal echoes in the right ventricular outflow tract that could not be detected in the body of the right ventricle (Fig. 8). The tricuspid valve itself was noted to be normal.

A rhabdomyoma arising from the region of the crista supraventricularis and partially obstructing the right ventricular outflow tract was resected.
Discussion

Before the widespread availability of non-invasive methods, the diagnosis of a right-sided tumour could be made only during cardiac catheterisation and usually the finding was unsuspected. As the introduction of catheters into right-sided chambers containing a mass can be hazardous because of dislodgement of portions of the tumour or adherent thrombus, a non-invasive technique which could establish the diagnosis and possibly obviate the necessity for cardiac catheterisation is safer. In the nine years before the introduction of echocardiography at Montefiore Hospital only two right-sided tumours were detected, whereas in the past two years echocardiography has enabled us to detect five, an experience similar to that reported elsewhere. The present five cases indicate that the non-invasive diagnosis of right-sided masses is feasible. Because of the ease with which the diagnosis can be made with M-mode and two-dimensional echocardiography, one's index of suspicion should be higher and more patients should be screened for the presence of masses. Thus, in addition to obstruction at either the tricuspid or pulmonary valve, right heart failure especially when unaccompanied by left-sided disease,
progressive ascites, pulmonary emboli without evidence of peripheral venous disease, "pericardial rubs" without other evidence of pericarditis, fever without evident source, and fatigue and constitutional symptoms without explanation justify consideration of right-sided masses as a possible cause.

Echocardiography seems to be ideally suited for making the non-invasive diagnosis of a right-sided cardiac mass. In the present series, four of the five masses were correctly identified by M-mode techniques while the fifth was also detected, though the M-mode echocardiogram was initially misinterpreted. Of the three right atrial masses, only the echocardiographic pattern of the thrombus (Fig. 4) resembled that previously described for left atrial myxomas with an echo-free interval following initial diastolic separation of the leaflets and then appearance of multiple linear echoes behind the anterior valve leaflet. This echo appearance is not specific for myxoma, but may be seen with any lesion prolapsing through the atrioventricular valve.16

In the other two patients with right atrial masses, the M-mode echo patterns were less characteristic. The mass of echoes in the vicinity of the tricuspid valve in case 1 was correctly identified because its pattern of motion during the cardiac cycle resembled that of a tricuspid valve. In case 2, however, the echo pattern was initially dismissed as artefact or that of an extracardiac structure because of the absence of identifiable landmarks and the lack of characteristic motion of the tumour mass. This underlines the difficulty there may be using M-mode echocardiography to show right atrial lesions.

The two right ventricular tumours produced abnormal echo patterns in the right ventricular outflow tract, a region that is easily approached by standard M-mode views.14 Thus, the possibility of a mass should be entertained if abnormal echoes in this normally echo-free space are detected.

Because M-mode echocardiography is dependent on echo pattern recognition, this technique is neither completely sensitive nor specific. The M-mode echo has the greatest chance of detecting a right atrial mass if it passes through the tricuspid valve since the valve becomes a valuable landmark in identification of the abnormal echoes. If the mass is sessile or does not prolapse through the valve, it may be overlooked. Because the tricuspid valve is the only identifiable landmark of the right atrium, attempts to scan the remaining parts of the chamber are usually unsuccessful. Furthermore, echoes from a very large mass which obscures normal tricuspid valve movement and other landmarks may be misinterpreted.

Two-dimensional echocardiography, however, can study the right atrium with ease and thus can visualise masses within this chamber in locations other than just behind the tricuspid valve. It can detect masses even if they are not pedunculated and separated from

Fig. 8 M-mode echocardiogram of a 5-day-old boy with a rhabdomyoma showing the mass of echoes in the right ventricular (RV) outflow tract. AML, anterior mitral valve leaflet.
the surrounding myocardium, or do not pass through the tricuspid valve. A two-dimensional echo study is also more likely to be able to distinguish a mass from a possible artefact or other structure. Pacemaker and Swan-Ganz catheters, Eustachian valves, and transducer artefacts identified as masses by M-mode techniques can be correctly distinguished by two-dimensional echocardiography.

Extra echoes recorded within the right ventricle by an M-mode echocardiograph may also be reflected from structures other than tumours, such as hypertrophied muscle bands, papillary muscles or Swan-Ganz catheters, thus confusing the identification of right ventricular masses. On the other hand, two-dimensional echocardiography provides a wider field of view and enables the observer to visualise movement of actual structures, therefore avoiding many of the potential difficulties of M-mode echocardiography.

Two-dimensional echocardiography, however, has its limitations. It cannot readily distinguish thrombus from tumour. Furthermore, the technique's success is still dependent on technical factors such as chest wall configuration and degree of lung hyperinflation, though the ability to visualise cardiac structures from the subxiphoid area and apex is a distinct advantage over M-mode echocardiography. Despite these limitations, the availability of real time, two-dimensional echocardiography is a major advance in our ability to diagnose right-sided cardiac masses in a safe, non-invasive manner, with a high sensitivity and specificity.

The appropriate treatment of right-sided cardiac masses is dependent upon accurate diagnosis. An appreciation of the diverse presentations of these masses and the use of echocardiography for diagnostic screening of these patients will assist in proper identification and in the institution of definitive treatment. It cannot be stressed enough that both the general practitioner and cardiologist must have a high index of suspicion, or these potentially curable disorders will be overlooked.

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