Abnormal diastolic movement of the interventricular septum caused by a prolapsing right atrial myxoma

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SUMMARY A 40 year old woman with cough and exertional dyspnoea was found to have a large right atrial myxoma by M mode and cross sectional echocardiography. Prolapse of the tumour into the right ventricle occurred during diastole with sufficient force to cause mechanical distortion of the interventricular septum. Septal motion became normal after surgical resection of the myxoma.

Right atrial myxomas rarely occur. The clinical findings are diverse and echocardiography is the single most useful investigation in confirming the diagnosis. In the present case the characteristic echocardiographic signs were accompanied by abnormal diastolic distortion of the interventricular septum caused by the impact of a large and highly mobile tumour.

Case report

A 40 year old woman presented with a five month history of non-productive cough and progressive exertional dyspnoea. She had been treated for anaemia three months before presentation. On examination the jugular venous pressure was raised 7 cm with a prominent "a" wave and positive Kussmaul's sign. The pulse was 80 beats/minute, sinus rhythm; and blood pressure was 130/90 mmHg. A loud third heart sound (tumour plop) was followed by a long low pitched mid-diastolic murmur with presystolic accentuation and both were increased in intensity by inspiration.

The following laboratory results were noted: haemoglobin 12.5 g/dl, white blood cell count 9.1 × 10⁹/l, erythrocyte sedimentation rate 40 mm in first hour, total protein 80 g/l (normal 57-77), albumin 44 g/l (35-47), C-reactive protein 37.7 mg/l (normal <18.0), IgM 2.8 g/l (normal 0.7-2.5), total bilirubin 21 µmol/l (normal 0-18), alkaline phosphatase 121 U/l (normal 10-110), aspartate transaminase 234 U/l (normal 10-30), lactate dehydrogenase was normal.

The electrocardiogram, chest radiograph, and pulmonary function tests were normal. Phonocardiography confirmed the clinical findings. M mode echocardiography demonstrated a mass of echoes behind the tricuspid valve; these filled the right ventricular chamber during diastole and produced paradoxical posterior bulging of the interventricular septum. The abnormal echoes disappeared during ventricular systole (fig 1). Cross sectional echocardiography demonstrated a large mobile pedunculated tumour mass in the right atrium which originated from the interatrial septum and prolapsed into the right ventricle in diastole distorting the interventricular septum (fig 2). The right atrium and ventricle were dilated and the left atrium and ventricle were normal. The right atrial pressure trace showed striking "a" waves reaching 16 mm Hg with a mean pressure of 12 mm Hg. Contrast injection into the superior vena cava confirmed the echocardiographic diagnosis of prolapsing right atrial myxoma and showed patent right and left pulmonary arteries. A perfusion lung scan was normal.

At operation a large gelatinous globular mass (8 × 6 × 5 cm) with an haemorrhagic surface filled the right atrium. Its broad base originated from the fossa ovalis and the tumour prolapsed into the right ventricle. It was successfully removed. Histology confirmed the tumour to be a benign atrial myxoma.
Fig 1  M mode echocardiogram showing a cloud of echoes from a prolapsing right atrial myxoma (Myx). The echoes fill the right ventricular chamber (RV) during diastole and disappear during ventricular systole. The interventricular septum (IVS) is displaced posteriorly (arrows) in diastole by the large prolapsing tumour mass. LV, left ventricle; MV, mitral valve.

Fig 2  Cross sectional echocardiogram of apical four chamber view showing diastolic distortion (arrows) of the interventricular septum (s) by a large prolapsing right atrial myxoma (Myx). RA, right atrium; RV, right ventricle; TV, tricuspid valve; LA, left atrium; LV, left ventricle.

The patient made an uneventful recovery and subsequent echocardiography demonstrated normal interventricular septal motion.

Discussion

About a third to a half of primary cardiac tumours are myxomas. Ninety per cent occur in the atria but only about 20% are found in the right atrium.1–3 The symptoms and physical signs of right atrial myxoma are diverse.4–6 Echocardiography is the most sensitive non-invasive method for detecting right atrial myxomas and for excluding a concomitant left atrial myxoma.5,7 The tumours typically appear as a dense cloud of echoes behind the tricuspid or mitral valves. Cross sectional echocardiography allows additional assessment of tumour size and mobility, including the detection of myxomas too small or not sufficiently mobile to prolapse through the tricuspid valve, differentiation of myxoma from infective vegetations on the tricuspid valve,8 and the presence of haemodynamically important tumour emboli in the right and left pulmonary arteries.9

An additional and striking echocardiographic sign was noted during cross sectional examination of our patient that was consistent with a large and highly mobile tumour. Rapid diastolic excursion of the mass through the tricuspid valve and into the right ventricle was arrested by its sudden impact against the upper interventricular septum; this caused the septum to bulge posteriorly towards the left ventricle. The M mode appearance of this event thus simulated reverse or "paradoxical" motion of the interventricular septum. The association of "paradoxical" septal motion and right atrial myxoma has been noted in two previous case reports. Goldschlager et al described right atrial myxoma presenting as congenital heart disease with a right to left shunt; paradoxical septal motion during systole with a normal right ventricular dimension was present but its cause was unexplained.10 Right ventricular enlargement and paradoxical septal motion in systole were attributed by Yuste et al to the right ventricular volume overload of tricuspid regurgitation due to the presence of a large right atrial myxoma, although the abnormal septal motion persisted after its removal.11 In our case the primary abnormality of septal motion occurred in diastole and septal motion became normal after removal of the tumour. This case demonstrates that the impact force of a large mobile right atrial myxoma is sufficient to distort the normal shape of the interventricular septum, a substantial structure. Massumi has described the diagnosis of large mobile right heart myxomas through detection of palpable tumour shocks and audible plops as the tumour is hurled vigorously against intracardiac structures in various phases of the cardiac cycle.6 Even when these physical signs are not detectable mechanical distortion of intracardiac structures may represent their echocardiographic counterpart.
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