

A CASE OF BI-ATRIAL MYXOMA

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CASE REPORT

A previously fit and well teenager was admitted following a sudden onset of right-sided limb weakness. Physical examination revealed dysarthria, flaccid tone in the right arm and right leg and power estimations of 1/5 and 3/5 respectively on that side. Fundi were normal. Cardiovascular examination revealed fixed splitting of a loud, first heart sound and a systolic murmur at the apex. The patient was apyrexial.

A computerised tomography (CT) brain scan was performed urgently and this revealed several low density areas associated with subtle mass effect in the left internal and external capsule. These appearances were consistent with cerebral infarction.

In view of the murmur, a transthoracic echocardiogram was performed. This revealed a huge mobile and friable intracardiac mass (figures 1 and 2), in both the left and right atrium. It appeared to prolapse across the mitral valve into the left ventricle during the cardiac cycle. The masses were attached to the intra-atrial septum. Appearances suggested a bi-atrial myxoma.

The patient is now making a gradual recovery from these hemiplegic symptoms with much improved strength on the right side, although there is still a residual weakness in both the right arm and right leg.

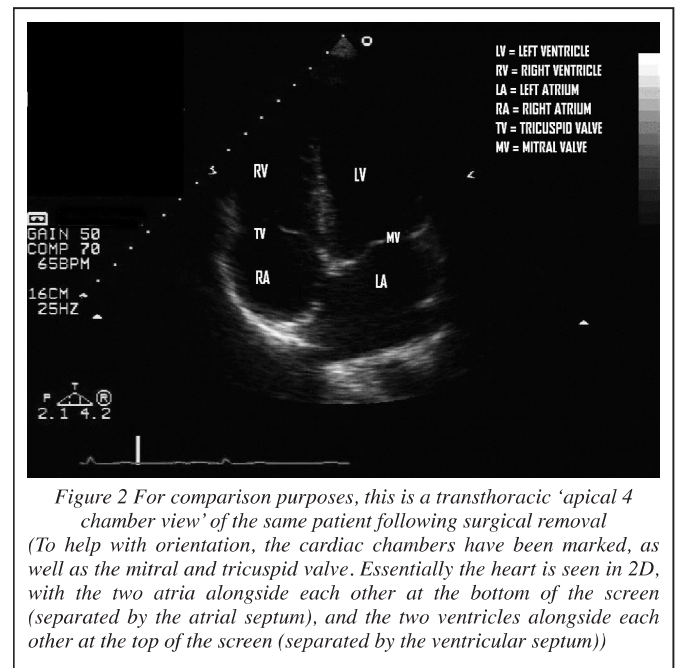


Figure 2 For comparison purposes, this is a transthoracic 'apical 4 chamber view' of the same patient following surgical removal (To help with orientation, the cardiac chambers have been marked, as well as the mitral and tricuspid valve. Essentially the heart is seen in 2D, with the two atria alongside each other at the bottom of the screen (separated by the atrial septum), and the two ventricles alongside each other at the top of the screen (separated by the ventricular septum))

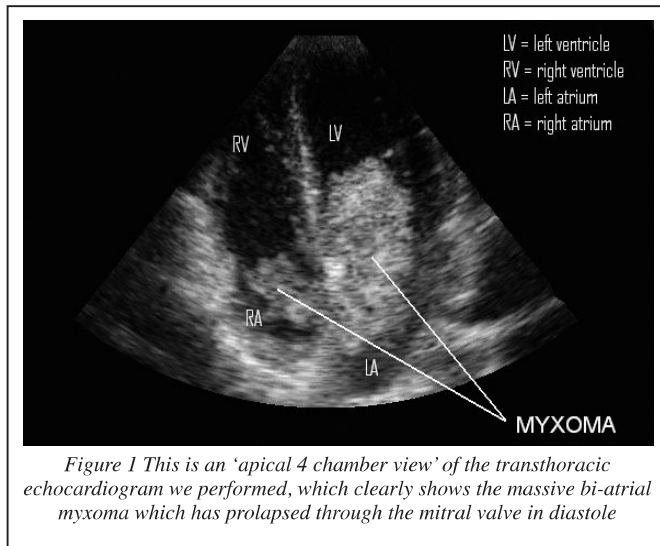


Figure 1 This is an 'apical 4 chamber view' of the transthoracic echocardiogram we performed, which clearly shows the massive bi-atrial myxoma which has prolapsed through the mitral valve in diastole

The patient was urgently transferred to a cardiothoracic surgical centre and the masses were removed under cardiopulmonary bypass. The septum was repaired with a patch of autologous pericardium. Later histology confirmed the typical appearances of an atrial myxoma.

DISCUSSION

Atrial myxomas are benign tumours of the heart. They are uncommon, with an incidence in the United Kingdom (UK) of approximately 1:1,000,000/year⁽¹⁾. Bi-atrial myxomas are extremely rare and according to most reports only account for 2–4% of atrial myxoma cases^(2,3,4). Left atrial myxomas in isolation are much more common, accounting for between 60–89% of cases^(3,4,5). They usually arise from the intra-atrial septum, although they have been reported to arise from other areas of the atrium on occasions.

Clinical presentation

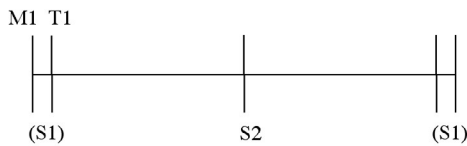
Symptoms often depend on which cardiac chamber is affected as commonly they include effects related to valve/chamber obstruction. With left-sided (mitral) obstruction this includes dyspnoea on exertion, dizziness or syncope (when significant obstruction occurs). Right-sided (tricuspid) obstruction tends to result in peripheral oedema, fatigue and occasionally ascites. Constitutional symptoms including weight loss, fever and arthralgia are very common. The tumours also tend to be quite friable and it is estimated that embolisation accounts for 26% of presenting symptoms with the majority of these affecting the cerebral circulation⁽⁶⁾.

Differential diagnoses to be considered in young patients presenting with a murmur and stroke

- Infective endocarditis
- Atrial septal defect/PFO* or ventricular septal defect (with paradoxical embolus)
- Atrial myxoma

*patent foramen ovale

The splitting of heart sounds



Fixed splitting of first heart sounds

Pacemaker, mitral stenosis
atrial myxoma

S1: first heart sound

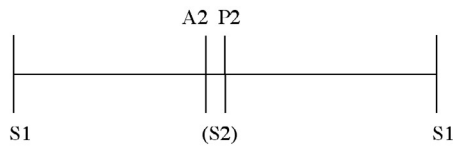
S2: second heart sound

M: mitral valve

T: tricuspid valve

A2: the aortic component of S2, made
by the shutting of the aortic valve

P2: the pulmonary component of S2,
made by the shutting of the
pulmonary valve



Fixed splitting of second heart sounds

Atrial septal defect

In our patient, there was fixed splitting of S1 on auscultation. This is an uncommon finding and is caused when M shuts at a different time to T. The commonest cause in most patients is a pacemaker, where activation of the right ventricle occurs before the left. Conditions of high left atrial pressure (such as mitral stenosis) or atrial myxomas (as in this case) are also recognised causes.

Splitting of S2 is a more common finding in the auscultation of patients. In a normal subject, we can usually appreciate a split of S2 during the respiratory cycle. S2 is produced by the combination of aortic and pulmonary valve closure. During inspiration, the negative intrathoracic pressure causes more blood to enter the right side of the heart. This forces the pulmonary valve to stay open longer, and thus shut later than the aortic valve. During expiration, the valves shut together. *Fixed* splitting of S2 occurs when equalisation of the two atrial pressures can occur, such as in a large atrial septal defect.

Physical examination findings

On auscultation of the heart, the findings of a left atrial myxoma may be similar to those of mitral stenosis due to valve obstruction. Common findings are:

- a loud first heart sound due to delay in mitral valve closure (It may be split)
- an early low-pitched diastolic sound – ‘a tumour plop’ may be heard
- a systolic murmur may be heard if the tumour causes mitral regurgitation

Clubbing is occasionally seen. Jugular venous pressure and right-sided heart failure signs may be present if the tumour is situated in the right atrium.

Management

Blood tests are not helpful in the diagnosis of myxoma, but are certainly required in the overall evaluation of patients. Normocytic, normochromic anaemias are common and sometimes haemolysis may occur⁽⁷⁾.

Echocardiography is the gold standard test in making a diagnosis. Transoesophageal echo provides the highest sensitivity and specificity in making a diagnosis, but usually a transthoracic approach is adequate. CT/magnetic resonance imaging scans contribute little to the diagnosis.

Treatment is within a surgical realm. Resection of the tumour is usually successful with low recurrence rates⁽⁸⁾ (4.7%). There are no specific medical treatments and in particular there is no evidence that anticoagulation confers any benefit.

SUMMARY

The most important lesson to learn from this case is the

importance of transthoracic echocardiography in the investigation of stroke in the young. Although myxomas are rare entities, (and in this case a bi-atrial myxoma, which is an extremely rare entity), our cardiology department here at the Royal Lancaster Infirmary has seen two cases within a six-month period.

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