Complex cardiac myxoma

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Introduction

Cardiac myxomas are the most common benign intracardiac tumour, and are more common in women. Since many patients suffer from cerebral or systemic embolism, early diagnosis is vital to plan for surgical intervention. Surgical excision is advocated as soon as possible, particularly in left atrial myxoma, because of the high risk of valvular obstruction and systemic embolisation. Patients with a family history of the disorder are at greater risk of tumour recurrence.[1]

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Case report

We report a case of a man who underwent three separate operations for myxoma resection, two of which were for recurrent tumour. His tumours involved the interatrial septum, the mitral valve annulus and the atrial wall.

A 37-year-old previously healthy male presented to the emergency department with a right-sided hemiparesis. Clinical examination revealed him to be afebrile, normotensive and in sinus rhythm. There was no audible murmur. There was no evidence of skin pigmentation or cutaneous tumour. Absent bilateral femoral pulses were noted.

Urgent vascular ultrasound was performed, which revealed a saddle embolus at the aortic bifurcation, with evidence of emboli seen in the renal arteries.

Urea and electrolytes revealed acute renal failure, with a urea of 39 mmol/L and creatinine of 536 µmol/L. His immunological profile and calcium were normal, and investigations for myeloma and renal vasculitis were negative.

An aortogram confirmed saddle embolus. Therefore he underwent urgent bifemoral embolectomy of what proved to be embolic myxoma. A computerised tomography (CT) scan of the brain confirmed left parieto-occipital infarction.

Subsequent transthoracic echocardiography showed a large left atrial myxoma attached to the interatrial septum and prolapsing through the mitral valve into the left ventricle (figure 1). He was stabilised and sent for urgent cardiac surgery to resect the tumour. Intra-operative transoesophageal echocardiography (TOE) confirmed the myxoma (figure 2). At operation the left atrium was incised vertically. The myxoma was attached via a thin stalk to the mitral valve (MV) annulus, hence the stalk and tumour were resected. Histology confirmed that the tumour was a benign myxoma. This site of myxoma is extremely rare.[2]

He went on to make a good recovery, with normalisation of his renal function and only a residual mild right-sided weakness from his stroke. The patient's family history revealed that his brother had also had a cardiac myxoma excised five years earlier but had had no recurrence at follow-up. In addition, his mother had died suddenly of an unexplained stroke in her mid-thirties.

He presented again three years later with a transient ischaemic attack. Subsequent echocardiography confirmed a recurrent left atrial myxoma. This had developed rapidly, since his transthoracic echo only six months earlier had been normal. Operative findings this time showed the mitral valve annulus to be free of myxoma but recurrent tumour was visible on the anterior atrial wall, close to the incision site. The resected specimen was confirmed as atrial myxoma, with no evidence of sarcomatous change.

On routine echo follow-up one year later, a third left atrial myxoma was discovered, like the first, on the MV annulus. In fact, at the

Figure 1. Transthoracic echo M-Mode picture, showing the orifice of the mitral valve between the anterior and posterior mitral valve leaflets (AMVL and PMVL, respectively) virtually occluded. The interventricular septum (IVS) and posterior wall (PW) of the left ventricle are annotated for reference points. M denotes the large atrial myxoma seen in this patient

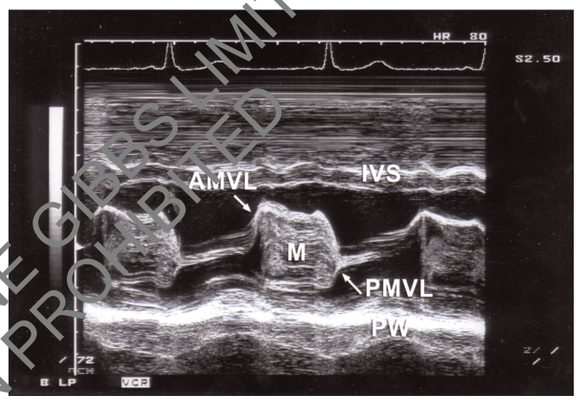

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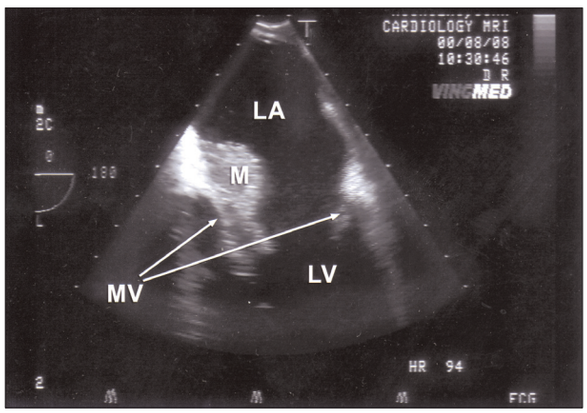
Figure 2. Transoesophageal echo, demonstrating a large myxoma (M), adherent to the mitral valve (MV) annulus and prolapsing into the left ventricle. The left atrium (LA) and left ventricle (LV) are annotated for reference points



time of surgery, two separate myxomas were located at different locations, both adherent to the MV annulus. These were surgically resected, histology again confirming myxoma.

This represents a unique and challenging case, and highlights the need for close follow-up in this group of patients. It also represents a surgical dilemma in that for technical surgical reasons, fourth-time surgery is thought not to be feasible in this patient. Indeed, should recurrence occur again, perhaps his only option would be heart transplantation. There is only one report in the literature of a woman undergoing cardiac transplantation for recurrent disease.[3]

Discussion

Cardiac myxomas are the commonest intracardiac tumour: they are most often located within the left atrium, comprising around 75% of all cardiac myxomas. They are usually attached to the interatrial septum. Although the majority of myxomas are sporadic, a smaller number may be familial or part of the Carney complex.[1] Familial cases tend to be inherited in autosomal dominant fashion or as part of the Carney complex. The latter is associated with multiple cutaneous tumours (myxomas), skin pigmentation and adreno-cortical disease.[4]

The vast majority of myxomas are diagnosed inadvertently. Indeed, a recent series of 70 patients showed that only around 5% were picked up clinically. Usually combinations of either mitral or tricuspid valve disease are suspected initially,[5] with subsequent echocardiography demonstrating the myxoma.

Key messages

- Cardiac myxomas are the commonest intracardiac tumour
- They present most often with dyspnoea, constitutional symptoms and embolic phenomena
- Early surgical resection is advised
- The long-term recurrence rate is low in sporadic cases
- Familial/Carney's complex cases have a much higher recurrence rate after resection
- Concise family history and examination are mandatory
- Follow-up should include sequential echocardiography

Once diagnosed, a concise history and examination are required in an attempt to delineate between familial and sporadic myxoma. We suggest that it is important to distinguish the two groups as familial patients tend to have a much higher recurrence rate after surgery.[6] In contrast, patients with sporadic myxoma can expect to be free of recurrence long term more than 95% of the time. Familial myxoma has been reported to recur as long as 20 years after the initial resection.[7] Therefore, long-term echocardiographic follow-up of these patients is mandatory, as is echocardiographic screening of the family. Initially, echocardiographic screening should be performed every three months as recurrences can occur rapidly.

Conflict of interest

None declared.

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