Surgical experience with cardiac tumours at the General Hospital, Kuala Lumpur.

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Summary

Fifteen patients underwent surgery for cardiac tumours in General Hospital Kuala Lumpur between October 1984 and June 1989. Twelve of the patients had cardiac myxomas and underwent excision under cardiopulmonary bypass. Two patients had sarcoma, of which one was excised. The other was inoperable. Another patient had a metastalic malignant melanoma which was inoperable.

Of the patients 10 were female and five male. Their ages ranged from 16 to 60 years. All were symptomatic and the commonest mode of presentation was exertional dyspnoea and palpitations. Two presented with cerebral embolisation. The three patients with malignant tumours had constitutional symptoms at the time of surgery.

All patients had echocardiography pre-operatively to confirm the diagnosis of cardiac tumour. Only one patient underwent preoperative cardiac catheterisation and angiography. The surgical approach in all patients was through a median sternotomy and all except one were operated under cardiopulmonary bypass. There was no intraoperative embolisation. There was one perioperative death.

Fourteen patients were followed up for periods ranging from one to 44 months. Three patients with malignant cardiac tumours died. One had recurrence of myxoma 21 months after the initial surgery. We conclude that excision of cardiac myxomas carry a very small risk following which patients have good prognosis. Malignant tumours carry a bad prognosis. From our experience, we conclude that echocardiography is an extremely accurate tool in the diagnosis of cardiac tumours.

Key words: Cardiac tumours, myxoma, atrial myxoma.

Introduction

Primary cardiac tumours are uncommon. Benjamin,[1] found primary tumours in 0.03% of 40,000 autopsy cases. Prior to the introduction of echocardiography and angiography, many of these rare neoplasms were diagnosed by the pathologist in the autopsy room. Currently, with the recent

advances in diagnostic imaging modalities and cardiosurgical techniques, cardiac tumours are often diagnosed with accuracy pre-operatively and surgical excision can be performed with low mortality and provide excellent and sustained symptomatic relief.

In this paper we review retrospectively the case records of consecutive patients who had undergone surgery for cardiac tumours over a four year period between October 1984 to June 1989. The purpose of this review is to discuss the varied clinical presentations of cardiac tumours and analyse our surgical experiences with cardiac tumours.

Patients and Method

The analysis covers 15 consecutive patients who had undergone surgery for cardiac tumours between 1984 to 1989. All available clinical records, roentgenograms, electrocardiograms, cardiac catherisation data, echocardiograms, operation reports and pathological specimens were reviewed.

The ages of the patients (five men and 10 women) were between 16 to 60 years, with a mean of 38.3 years.

Fourteen patients had primary cardiac tumours and one patient had a malignant metastatic melanoma. Of the 14 patients with primary cardiac tumours, 12 had cardiac myxomas and two had primary cardiac sarcoma. The patient population and site of tumour are shown in Table 1.

Table 1
Patient Population

	Myxomas	Malignant Cardiac Tumours	Total
Number of patients	12	3	15
Sex breakdown (M:F)	3 : 9	2 : 1	5 : 10
Ages (years)			
Range	16–60	28–44	16–60
Mean	39.1	35.3	38.3
Site, Left heart : Right heart	10 : 2 +	1 : 2*	11 : 4

\+ Ratio of left atrial to right atrial myxomas. One patient had two left atrial myxomas. None had bi-atrial myxomas.

* One patient had left atrial and left ventricular fibrosarcomas, one had undifferentiated right atrial sarcoma and one patient had metastatic malignant melanoma of the right atrium.

Results

Symptoms: The commonest presenting symptom was exertional dyspnoea – (13 patients). Six patients presented with palpitations as well and a single patient had palpitations only as a presenting complaint. Two patients presented with hemiplegia attributable to cerebral embolisation. Only one patient presented with syncope as well as other symptoms. The distribution of these symptoms is depicted in Table 2.

All patients with malignant cardiac tumours and two with cardiac myxomas presented also with constitutional disturbances inclusive of fever, arthralgia, easy fatiguability and weight loss (710 kg).

Table 2
Symptoms at presentation

			As a single complaint
Dyspnoea	13	(43.3%)	2
Palpitations	6	(20.0%)	1
Chest pain	3	(10.0%)	–
Hemiplegia	2	(6.7%)	1
Syncope	1	(3.3%)	–
Oedema	3	(10.0%)	–
Orthopnoea	2	(6.7%)	–
Total	30	(100.0%)	

Physical signs: Auscultatory findings were variable. The first heart sound was normal in all patients whereas the pulmonary component of the second heart sound was elevated in two patients with myxomas and one with malignant cardiac tumour. Diastolic murmurs were heard in only six patients and all of them had left atrial myxoma. The murmurs were interpreted as that of mitral stenosis in all six instances. Three of the patients had early diastolic murmurs interpreted as opening snaps which in actuality could have been the characteristic tumour "plop". Apical systolic murmurs were heard in six patients with myxoma and one with malignant cardiac tumour. Two patients had a pansystolic murmur at the left lower sternal edge which varied with respiration. This was interpreted as tricuspid regurgitation. One of these patients had a right atrial myxoma and the other a left atrial myxoma.

There were no additional heart sounds heard in any of the patients. In two patients (one with a right, the other a left atrial myxoma) murmurs were absent and the heart sounds were entirely normal. In the single patient with metastatic malignant melanoma, gross pericardial rubs were detectable. Two patients considered to have cerebral embolisation had permanent neurological deficits. Both had left atrial myxomas. One patient presented solely with left hemiplegia, the other had right hemiparesis and was in cardiac failure as well.

On clinical assessment alone the diagnosis most often reached was chronic rheumatic valvular heart disease. Nine out of 10 patients who had left atrial myxoma were thought to have rheumatic mitral valve disease prior to echocardiographic analysis.

Electrocardiogram and chest roentgenogram: Electrocardiogram showed that all the patients were in sinus rhythm. P mitrale was present in only one patient. Many had non-specific ST segment and T wave abnormalities but none showed any evidence of ventricular hypertrophy. The patient with metastatic malignant melanoma developed complete heart block prior to surgery.

Chest X-ray revealed cardiomegaly in all patients except one. Left atrial enlargement was evident in seven patients and upper lobe diversion in five. In two patients with right atrial myxoma, the lung fields were normal. No tumour calcification was diagnosed.

Laboratory investigation: Anemia (haemoglobin concentration less than 10gm/dl in females and 12 gm/dl in males) occurred in two patients. Erythrocyte sedimentation rate was elevated (more than 20 mm/hour in males, more than 40mm/hour in females) in eight patients. One patient had polycythaemia (haemoglobin concentration more than 15gm/dl and ESR less than 10mm/hour). There was no incident of leucopaenia while two patients had leucocytosis. Thrombocytopaenia occurred in one patient and thrombocytosis in another. Plasma globulin levels were elevated in three patients (more than 40mg/l).

Diagnostic techniques: Echocardiography was utilised as a diagnostic procedure in all cases. Two dimensional echocardiography was the most accurate technique in diagnosing cardiac myxomas. All patients with myxomas were correctly diagnosed by this technique.

All three patients with malignant cardiac tumours were suspected to have myxomas as well on echocardiography. A significant finding however is that all three had pericardial effusions while none of the true myxomas had any.

Cardiac catheterisation was performed in only one patient. She was the first patient to have undergone excision of a left atrial myxoma at the General Hospital, Kuala Lumpur.

Surgical techniques and findings: The surgical approach in all 15 patients was through a mid-sternotomy incision. Cardiopulmonary bypass was then instituted in 14 patients using moderate hyporthermia and cold potassium cardioplegia. In one patient with an undifferentiated right atrial sarcoma, the tumour had invaded through the right atrial wall and part of the pericardium. A biopsy was taken and no further surgical procedure was undertaken.

In patients with left atrial myxomas the most common approach was through bi-atrial incisions (eight out of 10 patients). One was excised via a left atrial incision posterior to the interatrial groove and one through a right atriotomy only. In two patients with right atrial myxomas the approach was through right atriotomies.

One patient had two left atrial myxomas rendering the total number of myxomas excised to 13. Eleven were in the left atrium with seven attached to the interatrial septum and four to the free left atrial wall. Two were in the right atrium and both were attached to the septum (Table 3). The septal defects created following excisions of those myxomas with septal attachments were closed with a dacron patch in seven patients, and directly sutured in two.

In the patients with fibrosarcoma of the left atrium and ventricle, the approach was through a left atriotomy and left ventriculotomy respectively under cardiopulmonary bypass. The tumour in the left ventricle was obstructing the outflow tract and was excised totally. The left atrial tumour had invaded the mitral valve annulus posteriorly and was left intact.

In the patient with metastatic malignant melanoma of the heart the approach was bi-atrial as it was preoperatively diagnosed as an atrial myxoma. It was found to be a darkly pigmented tumour which extensively involved the right atrium with extension into the right ventricle, left atrium and pericardium. It was also inoperable and only a biopsy was taken.

Mortality and follow up: In the whole group there was one peri-operative death (6.7% of total; 8.3% of myxoma group). The patient died of right heart failure eight hours after surgery. During surgery the patient was found to have a tense and dilated main pulmonary artery and poorly

Table 3
Sites of attachment/origin of 13 atrial myxomas

Atrium	Total number	Attachment	
		Septal*	Free wall
Left	11	7	4
Right	2	2	0

* Septal attachments were in the form of predunculated or sessile stalks most commonly within the area of the fossa ovalis.

contracting and hypertrophied right ventricle. The features suggest a long standing left atrial outflow obstruction by the myxoma with subsequent development of severe pulmonary hypertension. There were no incidents of intraoperative embolisation.

Post operative follow-up for the remaining 14 patients ranged from one month to 44 months (mean 14.8 months). During this time all the patients with malignant cardiac tumours had died. The patients with fibrosarcoma and metastatic melanoma died one month after surgery. The patient with undifferentiated sarcoma survived for 13 months after the biopsy.

In patients with atrial myxomas, recurrence developed in one patient (8.3%) 21 months after excision of a left atrial myxoma. The patient was reoperated and a myxoma in the right atrium and one in the right ventricle were totally excised. The postoperative course was uneventful.

No evidence of embolisation was detected during the follow-up period and no patient required subsequent valve replacement or repair. Good surgical results were achieved in all patients who had excision of cardiac myxomas.

Discussion

Atrial myxomas: In our experience with cardiac tumours we have found that they are seldom diagnosed on history and physical examination alone. Most cardiac myxomas presented with signs and symptoms that mimic mitral valve disease if they are in the left atrium. The right atrial tumours present with signs and symptoms of congestive cardiac failure.

Auscultatory findings vary greatly as were found in other reviews.[2,3] In brief they consisted mainly of a mid-diastolic murmur mimicing mitral stenosis. In additional 25% of the patients presented with an early diastolic murmur which were interpreted as an opening snap which were most likely to be a tumour "plop". Although left atrial myxomas are classically thought to simulate mitral stenosis we have found this to be true in only 50% of our cases.

Atrial myxomas can be diagnosed with accuracy with the aid of an echocardiogram. In our series we found it not necessary to proceed with cardiac catheterisation except in one case. Cardiac catheterisation which had been the diagnosis method of choice up to 1968 is not without hazards in patients with cardiac myxomas.[4,5]

Surgery was recommended on the basis of echocardiographic diagnosis in all our cases. They were all diagnosed as cardiac myxomas. In our experience all three malignant tumours that were referred for surgery had features of a myxoma on echocardiography. Perhaps the salient finding of coincidental pericardial effusion could alert us to the diagnosis of malignancy. In centres with facilities for magnetic resonance imaging it would possibly help in distinguishing these tumours apart.

The surgical excision of atrial myxomas carries a small risk. There was only one perioperative death in our series in a patient with longstanding disease and severe pulmonary hypertension. There was no tumour embolisation perioperatively. This experience is shared with reports from other centres.[5,6]

Long term follow-up studies are rare[9] and the true incidence of tumour recurrence is difficult to ascertain. The reported range is five to 14%. In our series with survivors followed up for a mean of 14.8 months the rate of recurrence was 8.3%.

The cause of recurrence is unclear. The aetiologies implicated are inadequate tumour resection, intracardiac implantation during excision and multicentric growth.[10] In our single case of recurrence the most probable cause is multicentric tumour growth as the recurrent myxomas were in chambers opposite to the original tumour.

Malignant cardiac tumours: Metastatic tumours to the heart are 20 to 30 times more common than primary tumours.[11] During our period of review we had three malignant cardiac tumours of which only one had a metastatic tumour. Our low incidence of metastatic tumour was most likely due to the fact that very few patients with widespread malignancy were subjected to echocardiography. In addition, possible cardiac metastases do not manifest with cardiac signs or symptoms until they are very large or extensive, or if the sole metastatic site is the heart as it was in our case.

Sarcomas are the most commonly encountered primary malignant tumour accounting for approximately 20% of all cardiac tumours.[12] Although arising from the endocardium or the pericardium more often than the myocardium, these highly malignant tumours tend to extend through all layers and invade mediastinal structures.[13] This growth pattern often makes these cardiac tumours irresectable and the patient survival measured only in weeks or months. In one of our patients the tumour had invaded through the right atrial wall and into the pericardium.

When diagnosed during life, palliative resection has been attempted but with limited success. On one of our patients the indication for surgery was the relief of left ventricular outflow tract which was obstructed by the sarcoma.

From our experience we conclude that the excision of cardiac myxomas carries a small risk, following which the patient has a good prognosis with sustained symptomatic relief. Life long follow-up and frequent echocardiography to detect tumour recurrence is mandatory. Malignant cardiac tumours carry a poor prognosis and echocardiography is an extremely accurate tool in the diagnosis of cardiac myxomas.

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