SURGICAL EXPERIENCE WITH CARDIAC MYXOMAS

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Background: Cardiac myxomas are the most common benign intracardiac tumours. We studied the clinical presentation of cardiac myxomas and the morbidity, mortality and recurrence rate following surgery at our institution over a 6 year period. **Methods:** This historical longitudinal study was performed at department of Cardiac Surgery, Armed forces Institute of Cardiology and National Institute of Heart Diseases Rawalpindi, Pakistan between January 2002 and March 2008 a total number of 8506 cardiac operations were performed. Of these 34 patients (19 males, 15 females) underwent complete excision of primary or recurrent intracardiac myxomas. Pre-operative diagnosis was established by echocardiography. All patients underwent operation soon after the diagnosis of a myxoma was made. Complete tumour excision followed by close inspection and copious saline irrigation of the cardiac chambers was done in each case. Of the 32 patients who survived the surgery, 29 patients were followed up at regular intervals for recurrence. The mean follow-up period was 34 months. **Results:** Cardiac myxomas constituted 0.40% of the total cardiac operations at our institution. They most commonly occurred in the fourth decade. The commonest location was the left atrium (LA) (79%) followed by the right atrium (RA) (14%). Only one patient had myxoma in the right ventricle (RV). Patients with LA myxoma simulated mitral stenosis clinically whereas patients with RA and RV myxomas presented with features of right heart failure. A smaller percentage presented with embolic and constitutional symptoms. There were two early deaths. One recurrence was noted at 27 months after surgery. No late deaths were observed in the study. **Conclusion**: Cardiac myxomas form a very small percentage of the cardiac cases. A high index of suspicion is essential for diagnosis. Echocardiography is the ideal diagnostic tool as also for follow-up. Immediate surgical treatment is indicated in all patients. Cardiac myxomas can be excised with a low rate of mortality and morbidity.

Keywords: Cardiac myxoma, Tumour, Atrium

INTRODUCTION

Cardiac myxomas are rare benign tumours, which account for nearly 50% of all adult primary cardiac tumours.[1] Approximately 75% to 80% of myxomas are located in the left atrium, 10% to 20% are in the right atrium, and 5% to 10% are in both atria or either ventricle.[2] Typically solitary, pedunculated, and arising in the vicinity of the fossa ovalis, they may on occasion be multicentric, sessile, or attached to other areas of endocardium. Clinically they present with symptoms of haemodynamic obstruction, embolization, or constitutional changes. Diagnosis at present is established most appropriately with two dimensional echocardiography.[3]

Prompt excision using cardiopulmonary bypass, first carried out by Crafoord in 1954[4], has been established as the only acceptable mode of treatment for these tumours.[5] The surgeon must try to prevent fragmentation and intraoperative embolization of the tumour, postoperative recurrence, and the missing of an occasional multicentric lesion. During the past five decades multiple centres have shown excellent surgical therapy results with a decreasing mortality.

This paper reviews the clinical experience and surgical management of cardiac myxomas at a teaching institution over a 6-year period.

MATERIALS AND METHODS

This study was conducted at the Armed Forces Institute of Cardiology and National Institute of Heart Diseases, Rawalpindi, from January 2002 to March 2008. All patients who underwent excision of primary or recurrent intracardiac myxomas were reviewed. A historical longitudinal study was conducted to study the clinical presentation, surgical findings, surgical considerations and associated mortality, morbidity and recurrence of these tumours.

Pre-operative diagnosis was established in all patients by echocardiography. Wherever transthoracic echocardiography (TTE) was equivocal, transesophageal echocardiography (TEE) was performed to confirm the diagnosis. Cardiac catheterization was not done for diagnosis. Coronary angiography was carried out in patients with history of chest pain or those older than 40 years.

Operation was undertaken in all patients soon after the diagnosis of cardiac myxoma was made. One patient needed an emergency procedure because of the severity of symptoms. The standard surgical approach was through a median sternotomy. Cardiopulmonary bypass (CPB) with aortic and bicaval cannulation and moderate hypothermia was used. Myocardial protection was achieved by cold antegrade blood cardioplegia. Heart was not manipulated until the aorta had been cross clamped to avoid tumour fragmentation and systemic embolization. The surgical approach for LA myxomas was left atrial, biatrial or right atrial trans-septal. The approach for RA and RV myxomas was right atrial. An additional right ventriculotomy was also needed for RV myxoma. The objectives of resection were complete tumour resection with full thickness removal of the attachment base and a cuff of interatrial septum to

prevent recurrence. Intramural (subendocardial) resections were used when a full thickness resection would have led to disruption of structural or functional integrity. All four cardiac chambers were thoroughly explored for additional myxomas. The surgically created defect was repaired directly or with pericardial or Dacron patch. Copious irrigation of the atria and ventricles with cold saline was done to eliminate any loose tumour fragments that might have been dislodged during removal of the tumour. All the resected myxomas were subjected to routine histopathological examination.

All the patients were followed up on an outpatient basis at regular intervals. They underwent clinical examination, chest X-ray, electrocardiography and echocardiography.

The statistical analysis was made by using SPSS version-12. All continuous variables were presented as Mean±SD and categoric variables were expressed as percentages.

RESULTS

Thirty-four patients of cardiac myxomas underwent operation at this institution during this 6-year period. Nineteen (55.88%) patients were male and 15 (44.11%) were female. The mean age was 33±15 years (range 5 years to 65 years). There were 27 left atrial (LA) myxomas (79.41%), 5 right atrial (RA) myxomas (14.70%), 1 right ventricular (RV) myxoma (2.94%), and 1 recurrent myxoma (2.94%). The duration of symptoms ranged from 2 to 6 months. Sixteen patients (47.05%) were in New York Heart Association (NYHA) Class II, 13 patients (38.23%) were in NYHA Class III and 1 patient (2.94%) presented in NYHA Class IV. Three patients (8.82%) presented with embolic episode, 2 patients of LA myxoma with cerebrovascular accident (CVA), and 1 patient of RV myxoma with multiple bilateral pulmonary embolism. Only one patient (2.94%) presented with constitutional symptoms of fever, palpitations and weight loss. Most patients with LA myxomas mimicked mitral stenosis clinically while most patients with RA and RV myxomas presented with features of right heart failure. A total of 12 patients (35.29%) required coronary angiography. None of them had any evidence of coronary artery disease.

Of the 27 LA myxomas, 23 (85.18%) arose from the interatrial septum, 2 (7.40%) from the posterior LA wall, 1 (3.70%) from the lateral LA wall, and 1 (3.70%) near the annulus of the posterior leaflet of mitral valve. Of the 5 RA myxomas, 4 (80%) arose from the interatrial septum and 1 (20%) from the lateral RA wall. The RV myxoma arose from the RV free anterior wall and anterior tricuspid leaflet. The recurrent myxoma was situated in the LA and arose from the interatrial septum.

Twenty-four LA and five RA myxomas were resected with a cuff of full thickness normal atrial tissue (interatrial septum 27 cases, atrial wall 2 cases). The surgically created defect was closed primarily in 11 patients, with autologous pericardial patch in 16 patients, and with Dacron patch in 2 patients. In 3 patients of LA myxomas intramural (subendocardial) resection was done. The RV myxoma was resected intramurally from the RV free wall and papillary muscles of tricuspid valve through a right atriotomy and right ventriculotomy. The tricuspid valve was excised along with the tumour and was replaced with a 30mm Carpentier Edwards bioprosthetic valve. The recurrent LA myxoma was resected with full thickness of interatrial septum and Dacron patch closure of the defect.

Associated procedures included mitral valve repairs in 2 patients of LA myxomas, tricuspid valve repair in 1 patient of RA myxoma and tricuspid valve replacement in the patient of RV myxoma.

The tumours ranged in size from 2×3 cm to 6×10 cm. Of the 27 patients of LA myxomas, all had a pedunculated tumour except 2 which were sessile. Of the 5 RA myxomas, 2 were pedunculated and 3 were sessile. RV and Recurrent LA myxomas were sessile. The gross appearance of the tumour was usually that of a soft, gelatinous, sessile or pedunculated mass with either a villous or smooth surface. Microscopic examination confirmed the diagnosis of myxoma in every patient. Histologically the tumours were composed of polygonal and stellate cells in a vascular, acid mucopolysaccharide stroma. Intramural haemorrhage and calcification were occasionally present.

Among 34 patients who underwent operations at our institution for cardiac myxomas, there were 2 (5.88%) early deaths (death occurring within 30 days of operation). One patient had preoperative cerebral infarct due to embolic occlusion of the right middle cerebral artery. The second patient died of myocardial infarction and low cardiac output.

Postoperative complications occurred in 7 patients (Table-1). Three patients (8.82%) had episodes of supraventricular arrhythmias which were controlled medically, and 2 patients (5.88%) had a transitory atrioventricular block requiring temporary pacing.

Complete follow up is available for 29 of the operative survivors. Three patients were lost to follow up. Mean follow up was 34±22 months. There were no late deaths (death occurring after 30 days of operation). Most patients were asymptomatic or in NYHA class I at follow up (Table-2).

Table-1: Surgical Morbidity (n=34)

Complications	No. of Patients	%
Supraventricular arrhythmias	3	8.82
AV heart block	2	5.88
Prolonged ventilation	1	2.94
Pericardial effusion	1	2.94
Total	**7**	**20.58**

Table-2: Postoperative Functional results

Symptomatic status of the patient	No.	%
Asymptomatic	18	62.06
NYHA Class I	6	20.68
NYHA Class II	1	3.44
Nonspecific chest pain	2	6.8
Improving neurological deficit	1	3.44
Palpitations	1	3.44

DISCUSSION

Myxomas are the most common primary tumour of the heart[1] with an estimated incidence of 0.5 per million population.[6] Cardiac myxomas constituted 0.39% of the total (8506) cardiac operations during this period at our institution. This figure is the same as that reported in the literature (approximately 0.3%).[6,7] In our study there was a higher incidence in male sex (55.88%) contrary to the female predominance reported in other studies (almost 75% of the myxomas occur in female).[8] Our patients conform to the age distribution and the relative tumour occurrence in the left and right atria evident in other series.[2,9,10]

The location, size, and mobility of cardiac myxomas determine their clinical features. Most patients present with one or more of the triad of embolism, intracardiac obstruction, and constitutional symptoms. Of our patients with myxomas, 30 of 34 (88.23%) had congestive heart failure or palpitations or both, and 3 patients (8.82%) had embolic disease. A higher frequency of embolization up to 30–40% was reported in western series.[11,12]

Echocardiography is non-invasive and allows preoperative diagnosis with fair degree of accuracy. It can rule out tumour in other chambers and there is no risk of tumour embolization. Transthoracic echocardiography can generally be used to determine the location, size, shape, attachment, and mobility of a tumour. The transoesophageal approach is particularly helpful in detecting the site of insertion and morphological features of atrial and ventricular myxomas.[13]

We have come to rely solely on two-dimensional echocardiography for preoperative diagnosis and believe that catheterization, a potentially harmful procedure in myxoma, is not indicated for diagnosis.[11] However, if coronary artery disease is suspected, or patient's age is greater than 40 years, coronary arteriography is advised to evaluate the coronary arteries.

Once the diagnosis of cardiac myxoma is made, the operation should be carried out without delay.[14] Complete resection is best performed through a median sternotomy with total cardiopulmonary bypass and cardiac standstill.

The ideal surgical approach to achieve complete excision of intracardiac myxoma is still controversial. Jones and associates believe that a surgical approach to atrial myxomas should allow minimal manipulation of the tumour, provide adequate exposure for complete resection of tumour, allow inspection of four heart chambers, minimize recurrence, and be safe and efficacious. They use biatrial approach to myxoma.[15]

Others consider the exposure of the left atriotomy approach to be adequate and have demonstrated the low recurrence rates and the safety of the technique.[16] The trans-septal approach through right atriotomy suggested by Chitwood gives good access to the myxoma with minimum handling and allows inspection of all cardiac chambers. [14] The RA and RV myxomas are approached through right atrium. RA myxomas demand more care during cannulation.

At our institution, the approach for LA myxomas was left atrial in 5 cases, biatrial in 9 cases and right atrial trans-septal in 13 cases. However all surgeons described an adequate exposure in their reports.

There is general agreement about the necessity to proceed to a full thickness resection with clear margins to minimize the risk of recurrence.[2,17] Although 14% of our patients underwent intramural resection, none presented recurrences at the resection site. Recurrence of a sporadic myxoma is unusual, occurring in 1–3% of cases.[18,19] The risk of recurrence after surgery is correlated with young age, family history of myxoma,[20,21] inadequate resection, intraoperative implantation, or multicentre growth. Recently, interleukin-6[22] and endothelial growth factor[23] have been identified as markers of these tumours. The average recurrence occurs about 30 months after removal of the first myxoma.[24] Only one recurrence was observed (2.94%) in this series, 27 months after the first operation. In retrospect the friable nature of the tumour could have produce 'seeding' at the time of operation, but growth from a new focus in the septum seems the more likely cause. A regular follow up of all patients by non-invasive method is mandatory because the recurrence of myxoma have been documented at various intervals and are not clearly predictable.[25]

A high incidence of arrythmias and conduction disturbances, both early and late, have been reported by Bateman and colleagues.[26] Although the pathophysiology of this was unclear, they were thought to be related to a possible surgical injury to the conduction pathways (biatrial approach) or excessive retraction of the heart (atrial approach). In this study 3 patients had episodes of supraventricular arrhythmias which were controlled medically and 2 patients had transitory atrioventricular block requiring temporary pacing. Operative mortality and morbidity in this series was low and comparable with that reported by others.[7,8,18]

The present study has several limitations. Retrospective studies are susceptible to selection and

recall bias. However, considering the rarity of the disease, a prospective randomized study is impractical.

CONCLUSION

Cardiac myxomas form a very small percentage of the cardiac cases. Immediate surgical treatment is indicated in all patients. These tumours can be excised with a low rate of morbidity and mortality. The prognosis for patients after surgical resection is excellent.

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