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Original Article

Operative management of intracardiac myxomas: A single center experience

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ABSTRACT

Background: Cardiac myxomas are the most common benign intracardiac tumors. We studied the clinical presentation of cardiac myxomas, their morbidity, mortality and recurrence rate following surgery at our institution over a period of four years.

Methods: During August 2008 to November 2012, a total of 12023 cardiac surgeries were performed. Amongst these, 50 patients (12 males, 38 females) underwent complete removal of primary or recurrent intracardiac myxomas. Complete tumor excision with a cuff of interatrial septum followed copious saline irrigation of the cardiac chambers was performed in each case.

Results: Forty six patients survived the surgery of which 43 are being followed up at regular intervals for development of recurrence. Myxomas constituted 0.41% of the total cardiac surgeries at our institute. Most of them were noted in the fourth decade. The commonest location was left atrium (74%) followed by right atrium (22%). Only one patient had a myxoma in the right ventricle. Forty six patients (92%) survived the surgery.

Conclusion: Cardiac myxoma excision account for a very small percentage of cardiac procedures. Immediate surgical treatment is indicated in all patients. Cardiac myxomas can be excised with a low rate of mortality and morbidity.

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Introduction

Cardiac myxomas are rare benign tumors, which comprises of 50% of all adult primary cardiac tumors.¹ Approximately 75–80% of myxomas are present in the left atrium, 10–20% in the right atrium, and 5–10% are reported in both atria or either ventricle.² They are typically solitary, pedunculate and arise in

the surrounding area of the fossa ovalis; on occasion they may be multicentric, sessile or attached to other areas of endocardium. Clinically, they present with symptoms of hemodynamic obstruction, or embolization. Diagnosis is most correctly established with two dimensional echocardiography.³ Removal using cardiopulmonary bypass was first carried out by Crawford in 1954⁴; has been documented as the only acceptable mode of

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treatment for these tumors.⁵ The surgeon must try to prevent disintegration and intraoperative embolization of the tumor, postoperative reappearance and missing out of an irregular multicentric lesion. During the past five decades, multiple centers have shown excellent surgical results with a decreasing mortality.

This paper reviews the clinical experience and surgical management of cardiac myxomas at our institute over a four year period.

Materials and methods

This retrospective review was conducted for patients operated for cardiac myxomas during August 2008 to November 2012 at our institute. This study was approved by ethics committee of our institute. Retrospective review of patient's records was done for all the patients who underwent excision of primary or recurrent intracardiac myxomas. Data was collected for clinical and anatomic findings, surgical considerations, associated morbidity, mortality and recurrence of these cardiac myxomas.

Preoperative diagnosis was done in all patients by echocardiography. Cardiac catheterization was not performed for diagnosis. Coronary angiography was carried out in patients with history of chest pain or those older than 40 years.

All patients were subjected to surgery immediately after they were diagnosed with cardiac myxoma. Two patients needed an emergency surgery because of the severity of symptoms. The standard surgical approach was through a median sternotomy. Cardiopulmonary bypass (CPB) with aortic and bicaval cannulation with moderate hypothermia was employed. Myocardial protection was achieved by cold antegrade blood cardioplegia. Heart was not manipulated until the aorta had been cross clamped to avoid tumor 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. The aortic cross clamp in the cases of right sided myxomas also included the main pulmonary artery so as to prevent myxoma fragment migration into the pulmonary arteries. An additional right ventriculotomy was also performed for RV myxoma. The objectives of resection were complete tumor resection with full thickness removal of the attachment base and a cuff of interatrial septum to avoid recurrence. Intramural (subendocardial) resections were used when a full thickness resection was suspected to result into disruption of structural or functional integrity. All four cardiac chambers were thoroughly explored for additional masses. The surgically created defect was repaired directly or with pericardial or polyester patch. Copious irrigation of the atria and ventricles with cold saline was performed to eliminate any loose tumor fragments that might have been dislodged during tumor removal. All the excised myxomas were subjected to routine histopathological examination. The statistical analysis was performed with IBM SPSS version 20.

Result

During a four year period, 50 patients with intracavitary cardiac myxomas underwent surgery at this institution.

Demographic data and presentations are detailed in Table 1 there were 12 (24%) males and 38 (76%) females. The age was 36.05 ± 13.97 (mean $\pm$ SD) years (range: 5.5–60 years). There were 37 (74%) left atrial (LA) myxomas, 11 (22%) right atrial (RA) myxomas, 1 (2%) right ventricular (RV) myxomas and 1 (2%) recurrent myxoma. One patient who had recurrence of LA myxomas was operated previously at our center 6 years back. He had a recurrence at site just adjacent to previous site of origin. None of the patient had familial myxoma. The duration of symptoms ranged from 2 to 6 months. Eighteen patients (36%) were in New York Heart Association (NYHA) Class II, 26 (52%) in NYHA III and 1 (2%) presented in NYHA class IV. Six (12%) patients presented with embolic episode, out of which 3 (6%) patients had cerebrovascular accident (CVA), 2 (4%) patients had femoral artery embolism and 1 (2%) patient had left central retinal artery embolism. Only 3 (6%) patients presented with isolated constitutional symptoms. Most patients with LA myxoma clinically imitated mitral stenosis while most patients with RA myxomas presented with features of right heart failure.

Two (4%) patients with LA myxomas had associated moderate mitral regurgitation (MR) which required mitral ring annuloplasty and 1 (2%) patient had associated rheumatic mitral stenosis which required open mitral commissurotomy. In one patient repair of anterior mitral leaflet (AML) was required as the tumor was attached to it. All the mitral valve procedures also underwent a ring annuloplasty with rigid ring of appropriate size. In another patient with right atrial myxomas, tricuspid valve (TV) repair was performed as tumor was arising close to the anterior tricuspid leaflet and the leaflet was injured during the excision of the tumor. One patient underwent coronary artery bypass grafting with LA myxoma removal. Grafts were put on left anterior descending artery and obtuse marginal artery.

Table 1 – Demographic data.

	N (%)
Age, yrs (mean $\pm$ SD)	5.5–60 (36.05 $\pm$ 13.97)
Sex (male/total)	12/50 (24%)
Location	
Left atrium	37 (74%)
Right atrium	11 (22%)
Right ventricle	1 (2%)
Recurrent myxoma	1 (2%)
NYHA class	
NYHA 2	18 (36%)
NYHA 3	26 (52%)
NYHA 4	1 (2%)
Presentation	
Embolic episode	6 (12%)
a) Cerebrovascular accident	3 (6%)
b) Femoral artery embolism	2 (4%)
c) Central retinal artery embolism	1 (2%)
Constitutional symptoms	3 (6%)
Associated lesions	4 (8%)
Moderate mitral regurgitation	2 (4%)
Mitral stenosis	1 (2%)
Coronary artery disease	1 (2%)

NYHA – New York Heart Association class.

Morbidity and mortality

Forty-Six patients (92%) survived the surgery and were discharged from the hospital. There were four hospital mortalities. One patient with LA myxomas died of CVA due to embolic occlusion of the left middle cerebral artery. One patient with RA myxomas had massive pulmonary embolism at the time of presentation. Patient was operated in emergency. Postoperatively, patient died on postoperative day 5 because of persistent severe right ventricle dysfunction. Two patients died because of massive myocardial infarction in early postoperative period with persistent low cardiac output. Postoperative morbidity is shown in (Table 2). Two patients had femoral artery embolism as presenting complaint requiring embolectomy. Diagnosis of myxomas was confirmed by histopathological examination of the embolus. Out of 46 patients who were discharged from the hospital, three patients were lost to follow-up. Complete follow-up was available for 43 patients. All the patients were alive and in NYHA class I or II at a median follow-up of 28 ± 11.2 months (Table 3). We gave low dose anticoagulation for six weeks in the form of warfarin to maintain the international normalized ratio (INR) between 2.5 and 3.5 in the patients who required mitral valve repair.

Pathologic findings

All the LA myxomas ($N = 37$) were pedunculated except two which were sessile. Among the RA myxomas, 9 were attached to the fossa ovalis and two were attached to the suprahepatic IVC. RV myxoma was attached to the ATL. The recurrent LA myxoma was located in the LA and was pedunculated. The tumor size ranged from 2×3 cm to 6×10 cm and weighed 5–30 g. The external appearance of the tumors was usually that of a gelatinous, sessile or pedunculated mass with either a villous or smooth surface. Microscopic examination confirmed the diagnosis in all the patients. Typically, the histopathological examination revealed proliferation of capillaries, blood extravasations and dispersed fibrin deposits.

Discussion

Cardiac myxomas are neoplasms of the heart with an estimated incidence of 0.5 per million.¹ the exact incidence in

Table 3 – Post operative functional result.

Symptomatic status of patient	No. of patients
Asymptomatic	29
NYHA Class I	8
NYHA Class II	4
Non-specific chest pain	1
Improving neurological deficits	1
Electrocardiogram:	
Atrial fibrillation	3
Abnormal P wave	1
Echocardiography:	
Mild Mitral regurgitation	1

India are not known. In our series, cardiac tumors accounted for 0.41% of all cardiac operations performed at our institution compared to approximately 0.3% reported in western literature.^{1,2} The tumors were predominantly found in female similar to published literature. In our population mean age at presentation was 36 years in contrast to 55 years in other published literature.³

Clinically LA myxomas mimicked mitral stenosis in our series. Also, all cases of RA myxomas were associated with some tricuspid regurgitation probably related to mechanical interference of the TV by the myxoma. Few patients in the current study had constitutional symptoms contrary to other studies.^{3,4} On comparing the tumor consistency in patients with or without embolization, all tumors with embolization had soft gelatinous morphology. Echocardiography revealed polypoidal type of tumor with irregular shape and mobile surface in all these patients.⁵

Echocardiography is the investigation of choice as it is non-invasive and allows preoperative diagnosis with fair degree of accuracy, it can rule out the tumor in other chambers and there is no risk of tumor embolization. Transthoracic echocardiography can determine the location, size, shape, attachment and mobility of a tumor. The transoesophageal approach is particularly helpful to notice the site of attachment and morphological features of left atrial and ventricular myxomas.

We at our institute rely exclusively on two dimensional transthoracic echocardiography for preoperative diagnosis and cardiac catheterization is reserved only for special situations like suspected coronary artery disease or patient's age >40 years.⁶ Once the diagnosis of cardiac myxomas is made, the surgery is performed without delay.⁷

There is still argument regarding the most appropriate surgical approach to achieve complete excision of intracardiac myxoma. The LA approach gives a direct and fast access to the myxoma although not to its attachment.⁸ The BA (biatrial) approach not only gives good exposure but also facilitates the easy extraction of the tumor with minimum handling and also permits good examination of all the cardiac chambers⁹ but, the technique is more radical. The trans-septal approach through right atriotomy suggested by Chitwood⁷ is more practical and conservative (Fig. 1). It gives good access to the myxoma with minimum handling and allows inspection of all cardiac chambers (Figs. 2 and 3). The RA and RV myxomas are approached through right atrium. RA myxomas demand more care during cannulation. At our institution, all the LA myxomas were successfully excised by the right atrial, trans-

Table 2 – Surgical findings and results.

Findings and complications	No. of patients
Left atrial myxoma	37
a) Pedunculated	35
b) Sessile	2
Right atrial	11
a) Fossa ovalis	9
b) Inferior vena cava	2
Mortality	4 (8%)
Femoral artery embolus	2

Fig. 1 – Intraoperative view of LA myxoma showing the view from right atrium.

septal approach except in six cases. In three cases, LA myxomas was approached through left atrium and in other three patients, biatrial technique was used in view of the large size of the myxomas (Fig. 4). After excision of the tumor, we routinely explore all the cardiac chambers for additional tumors as it can be missed by the preoperative echocardiography, though not evident in this series.

There is general agreement about the necessity to proceed to a full thickness resection with clear margins to minimize the risk of recurrence.^{2,10} Although 7 (14%) of our patients underwent intramural resection; one presented with recurrence at the resection site. Recurrence of a sporadic myxoma is unusual, occurring for 1–3% of cases.^{11,12} The risk of recurrence after surgery is correlated with young age, family history of myxoma,^{13,14} inadequate resection, intraoperative implantation, or multicentric growth. Recently, interleukin-6¹⁵ and vascular endothelial growth factor¹⁶ have been identified as markers for these tumors. The average recurrence occurs about 30 months after removal of the first myxoma.¹⁷ Only one

Fig. 2 – Intraoperative view of excision of LA myxoma by creating the plane in left atrium taking care not to injure the myxoma.

Fig. 3 – Reconstruction of interatrial septal defect with polyester patch.

recurrence was observed (2.0%) in a previously operated patient six years after first surgery. Though the friable nature of the tumor could have produced 'seeding' at the time of surgery, but growth from a new focus in the septum seems to be the most likely cause of recurrence. Therefore, a regular follow-up of all patients by non-invasive method is mandatory.¹⁸

In this study, we noticed that in all those patients with embolic events, the tumor was extremely friable. Meticulous care during surgery may prevent these unfortunate events and furthermore also reduce the risk of tumor implantation. The grade of MR lessened postoperatively in patients where no additional procedures were performed as this was most likely caused by mechanical interference of the mass with the mitral valve.

A high incidence of arrhythmias and conduction disturbances, both early and late, have been reported by Bateman and colleagues.¹⁹ Although the pathophysiology of this was unclear, they were thought to be related to a possible surgical injury to the transmission pathways (biatrial approach) or extreme retraction of the heart (left atrial approach). In this study, 3 patients had episodes of supraventricular arrhythmias which were controlled medically and 2 patients had

Fig. 4 – Biopsy sample of left atrial myxoma.

transitory atrioventricular block required temporary pacing.^{2,20} Operative morbidity and mortality in this series was low and comparable with other reports.^{2,3,21,22}

Conclusion

Cardiac myxomas form a very small percentage of the cardiac tumors. Instant surgical treatment is essential in all patients. A close follow-up of all the patients is mandatory because the reappearance of myxomas has been documented at various intervals.

Conflicts of interest

All authors have none to declare.

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