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Case Series of Intra Cardiac Myxomas: A Single Institutional Experience

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Abstract

Background: Cardiac myxomas are the benign tumors occurring in any of the cardiac chambers but most commonly in the left atrium. Recent advances facilitate appropriate diagnosis and successful surgical management. This study is a retrospective analysis of myxoma cases focused on the clinical features and successful surgical procedures. *Methods:* this study was conducted in madras medical college with a duration of 5 years, which includes 30 patients. Age, sex, site of myxoma, diagnosis and surgical approaches done were recorded. *Results:* A female predominance (73%) with left atrium (70%) being the most common site of myxoma was observed. Trans Thoracic Echocardiography was the choice of diagnosis (83%) and right atrial approach was done in most of the cases (83%). *Conclusion:* According to our experience in the management of 30 cardiac myxomas during a study period of 5 years, we strongly recommend right atrial approach instead of the classical bi-atrial approach.

Keywords: Cardiac Myxoma; Trans Thoracic Echocardiography; Transesophageal Echocardiography; Fossa Ovalis; Right Atrial Technique; Sternotomy; Cardiopulmonary Bypass.

Introduction

Cardiac tumors include benign and malignant neoplasms arising within the cardiac chambers or in the myocardium. Approximately 70% of cardiac tumours are benign and 30% are malignant and potentially capable of invasion or metastasis [1].

Cardiac myxomas are primary cardiac tumours that are generally pedunculated but may have a broad base. They are intracavitary tumours occurring within any of the cardiac chambers, but they have a predilection for the atria, particularly the left. They are usually 5-6 cm in diameter, with a range of 1-15 cm.

Most atrial myxomas, whether left or right, arise from the atrial septum, usually from the region of limbus of the fossa ovalis. About 10% have other sites of origin particularly posterior and anterior atrial walls and the appendage. Most myxomas 80% - 90% are in the left atrium. Right atrial myxomas tend to be more solid and sessile than left atrial myxomas, with a wider attachment to the atrial wall or septum [2]. Myxomas may occasionally be found in right ventricle on the free wall or ventricular septum. Few cases of left ventricular myxomas are also reported. Valvar myxomas arising from mitral, tricuspid and pulmonary valves have also been reported.

Myxomas may produce symptoms of hemodynamic dereangement from obstruction of flow within the cardiac chamber, symptoms associated with emblization and constitutional symptoms.

Hemodynamic Derangement

Myxomas may obstruct pulmonary or systemic venous drainage or may impair flow across the atrioventricular valves. The obstruction is characteristically progressive. When obstruction is

intermittent, syncope, often related to postural change, or sudden death may occur. Impairment of valve closure, either by obstruction of leaflet damage, may cause regurgitation. Although regurgitation is the dominant abnormality in a few patients, as a rule obstruction predominates.

Embolism

A major feature of cardiac myxomas is embolization. Emboli may arise from tumour fragementation or detachment of entire tumour, or from thrombi or infected foci on the neoplasm. Systemic emboli occur in 30%-45% of patients with left atrial myxomas. Embolism from right sided tumours occurs in about 10% of cases and may cause massive fatal pulmonary obstruction. Massive pulmonary embolism complicating left atrial myxoma has been reported [3].

Constitutional Symptoms

In about 30% of patients, the only manifestations of cardiac myxoma are a plethora of constitutional symptoms. Large left atrial myxomas are particularly apt to produce constitutional symptoms. These symptoms include fever, weight loss, clubbing, raynauds phenomenon, myalgia and athralgia. Other unusual manifestations include polycythemia, hemolytic anemia.

Nearly all solitary myxomas are non familial, myxomas have familial occurrence in about 5% of patients. Familial myxomas have autosomal dominant inheritance and are primarily disorders of yound men. Familial myxomas are associated with serotoli cell tumour of testis, cushings syndrome, pituitary tumous, centrofascial and labial lentiginosis, cutaneous myxomas and multiple myxoid mammary fibroadenomas. Familial myxomas have the same histologic appearance as nonfamilial myxomas and produce the same symptoms, however they have a strong tendency to recur.

Diagnosis

Cardiac myxomas were diagnosed only during autopsy before the introduction of angiocardiography [4]. Echocardiogram had come to practice in 1959 through which the diagnosis of myxoma was made easy and noninvasive [5]. Currently, the investigations to diagnose cardiac myxomas are largely dependent on Transthoracic echo (TTE) supplemented by Transesophageal echocardiography (TEE). Cardiac catheterization and angiography no longer constitute the investigation of choice unless other types of cardiac or coronary artery disease require assessment.

Surgical Management

Once the diagnosis of cardiac myxoma has been made, surgery should be done immediately to prevent mortality. An 8% mortality has been reported in patients awaiting operation [6]. complete cure is expected especially if the involved atrial septum is excised and repaired by patch replacement to avoid recurrence [7].

Myxoma excision can be done by a median sternotomy, cardiopulmonary bypass through uniatrial; left atrial/right atrial or biatrial approaches [8], among which biatrial approach has been highly recommended [9]. Although rhabdomyoma is by far the most common primary heart tumour in children and infancy, cardiac myxomas are the most commonly encountered primary heart tumour in adults. In this retrospective study we analyzed the incidence, presentation and operative management of cardiac myxomas.

Materials and Methods

30 consecutive cardiac surgical patients diagnosed with cardiac myxoma admitted in the Department Of Cardiovascular and Thoracic surgery, Madras Medical College, Chennai between April 1, 2003 to March 30, 2008 comprised the sample for this study. Patients operated before August 31, 2005 were analyzed from the case sheets obtained from Medical Record Department. All the patients under the study are classified according to their age, sex, mode of presentation, method of diagnosis, site of the tumor and surgical approach.

Results

Age and Sex

73% (22) of cardiac myxoma occurred in females and 27% (8) of the disease were found to occur in males and amongst the two sex groups, 77% and 100% of the disease occurred between 20-49 years in females and males respectively (Figure 1).

Symptoms

All subjects (100%) included in the study

presented with cardiac symptoms, only 7% (2) of the patients had neurological symptoms (one patient had left hemiparesis and the other had Freidrich's ataxia), 27% (8) of the patients had constitutional symptoms in the form of fever, anemia and raised erythrocyte sedimentation rate (Table 1).

Mode of Diagnosis

Trans Thoracic Echocardiography was diagnostic in 25 cases (83%), Trans Esophageal Echocardiography was required in diagnosing myxoma in 5 cases (17%). 2 cases underwent coronary angiogram for assessing the status of coronary arteries preoperatively and were found to have normal coronaries (Table 2).

Site of Myxoma

Myxomas were found in the left atrium in 21cases (70%), right atrial myxomas were found in 4 (13%) of cases. 2 cases (7%) had right ventricular myxomas. One case was found to have isolated mitral valve myxoma involving the posterior mitral leaflet (PML) and another 2 cases (7%) involving the left atrium and mitral valve (Table 3).

Site of Pedicle Attachment to a Trial Wall

Most commonly the attachment 60% (18) was to the fossa ovalis and with one exception (attachment to the posterior mitral leaflet), the other neoplasms also involved the interatrial septum. Diameters of the atrial myxomas measured laterally and discounting pedicles, ranged from 2cm to 8cm. There were two right ventricular myxomas each involving the trebacular septum and right ventricular free wall (Table 4).

Surgical Approach and Techniques

Majority of the cases 25(83%) were removed by right atrial approach. Bi-atrial technique was used for tumour excision in 3 cases, left atriotomy was done in 1 case (removal of tumour arising from PML of MV). Right venticulotomy was done for tumour retrieval in a case of RV myxoma (Figure 2).

In all 30 cases, the atrial myxomas were excised through a median sternotomy, cardiopulmonary bypass. In all cases, cardiac arrest was achieved with antegrade cold crystalloid cardioplegia with topical hypothermia. Typically a right atrial incision was made parallel to right atrioventricular groove in 25 cases. The superior portion of the inter-atrial septum was incised to visualize the left atrial mass. Once the tumour was visualized from the right atrial aspect, the rest of the atrial septum was incised carefully avoiding cutting through the left atrial mass. Before resecting the other chambers were explored for the occasional multicentric tumour. The pedicle was completely excised along with a cuff of tissue to which it was attached. To minimize the risk of embolization, great care was taken to handle the pedicle gently, and to remove any residual tumour debris from the surgical field.

The surgically created septal defect was closed by the following methods: in 22 instances, by pericardial patch closure with running monofilament suture. In 3 instances by direct closure was done. Two patients required mitral valve replacement in addition to myxoma excision. One patient required mitral valve repair in addition to myxoma excision.

The mean aortic cross clamp time and Total cardiopulmonary bypass time recorded for each approach. Short cross clamp time and cardiopulmonary bypass time were noted in right atrial approach.

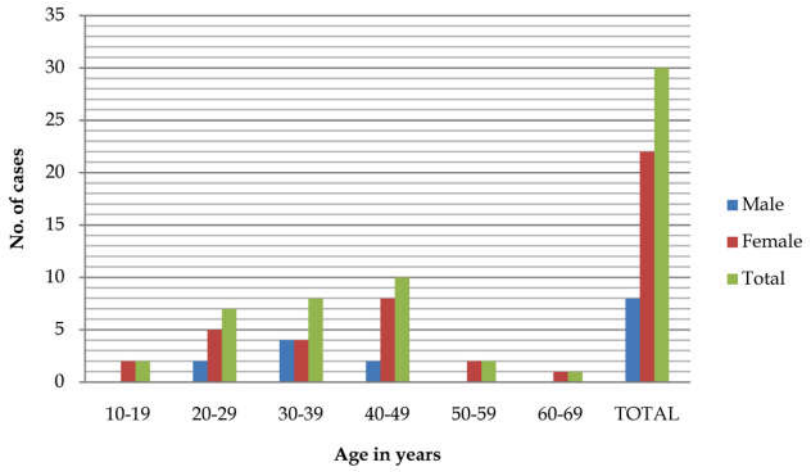

Fig. 1: Age and sex distribution

A female predominance (22/30) with most of the cases falling in the age of 20 to 49

Table 1: Symptoms

Cardiac Symptoms	Cardiac & Neurologic Symptoms	Cardiac & Constitutional Symptoms
30	2(7%)	8(26.6%)

All cases were cardio symptomatic with a few presented with neurological and constitutional symptoms additionally

Table 2: Mode of Diagnosis

TTE	TEE	CAG
25(83%)	5(17%)	2

TTE -Trans Thoracic Echocardiography, TEE -Trans Esophageal Echocardiography, CAG- coronary angiogram. TTE was done in majority of the cases. CAG was required in 2 cases and the coronaries were found normal.

Table 3: Site of myxoma

Left Atrial	21(70%)
Right Atrial	4(13%)
Right ventricular	2(7%)
Left Atrial and MV	2(7%)
Mitral valve	1(3%)
Total	30

Myxoma was observed more commonly in the left atrium followed by the right atrium with a very few in other sites

Table 4: Site of Pedicle Attachment to Atrial Wall

Left Atrial Septum fossa ovalis	18(60%)
Left Atrial Septum not involving FO	05(17%)
Fossa Ovalis in Right. Atrium	04(14%)
Mitral Valve (PMF)	01(3%)
Trebacular Septum in RV	01(3%)
Right Ventricular Wall	01(3%)
Total	30(100%)

Fossa ovalis (FO) was the most common site for pedicle attachment and a few other sites were also observed during the study
PMF- posterior mitral leaflet

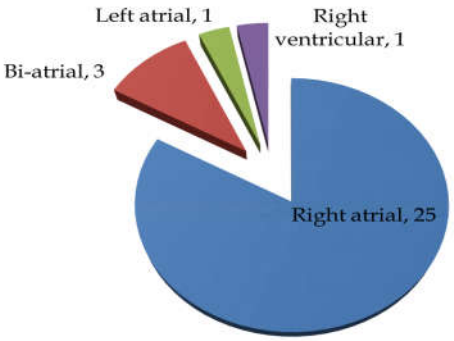

Fig. 2: Surgical Approach and Techniques

Right atrial approach was used in most of the cases instead of the conventional bi – atrial method.

Table 5: Mean ACC Time and CPB Time

S. No.	Approach	ACC Time (Mean) (Minutes)	CPB Time (Mean) (Minutes)
1	Rt. A trial	49.96	86.76
2	Bi-a trial	100.6	153.0
3	Left a trial	108.0	148.0
4	Right ventricular	66	124

Mean ACC and CPB time was shorter in right atrial approach compared to bi - atrial method.
ACC- aortic cross clamp time, CPB- cardiopulmonary bypass time

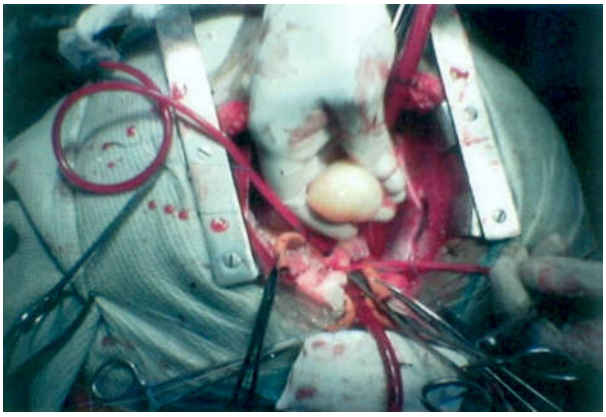

Fig. 3: Intraoperative tumour removal through Right Atrial approach

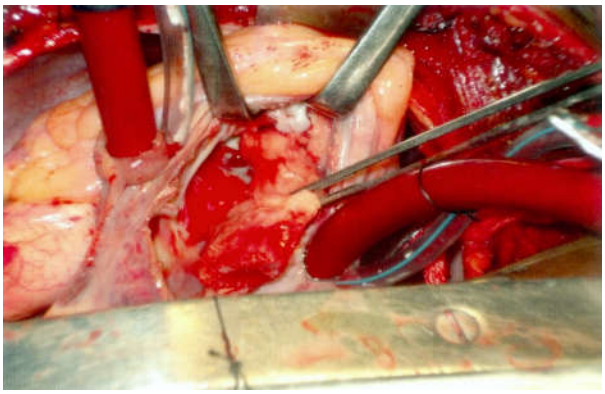

Fig. 4: Left Atrial Myxoma excised through Right Atrium

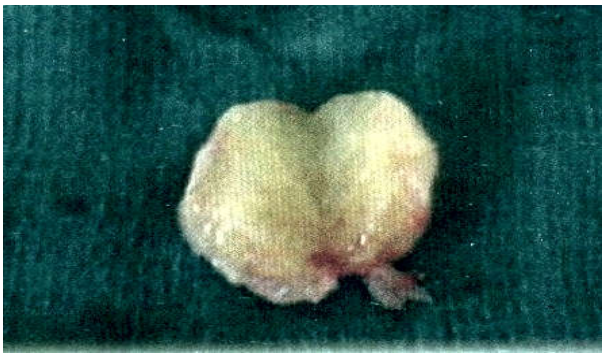

Fig. 5: Left Atrial Myxoma mass of a patient

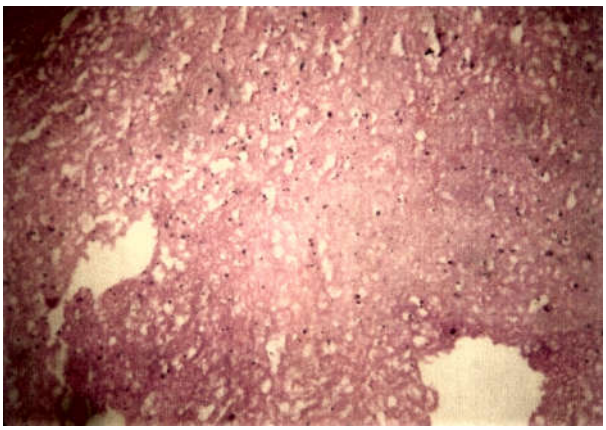

Fig. 6: Histopathological photograph of Cardiac Myxoma

Discussion

Prevalence

Primary cardiac tumours are rare, and 80% of them are benign. They account for 5% to 10% of all cardiac neoplasms, the balance of which are secondary tumours (therefore metastatic and malignant). Myxomas account for approximately 50% of all primary cardiac neoplasms, and have been reported in all cardiac chambers. Although about 75% of myxomas occur in the left atrium and 25% in the right [10]. Multiple myxomas have been reported in 5% of patients with left atrial of left ventricular myxomas. Myxomas predominantly occur in woment in the 3rd to 6th decades of life, and familial tendency have been reported [11]. In our experience myxomas were found to occur predominantly between 20-49 years age group (83%). the tumour was found to occur predominantly in females in 73% and males in 27%, however in both sex groups the tumour occurred mostly between 20-49.

Clinical Manifestations

Although small tumours may be asymptomatic, the clinical manifestations of atrial myxomas, when they occur are well recognized. The most common findings are symptoms of valvular obstruction and signs of peripheral embolization [3]. In our study group all patients 30 (100%) presented with any one of the cardiac symptoms in the form of dyspnea on exertion, palpitation, paroxysmal nocturnal dyspnea, chest pain and discomfort. This is attributed to the fact that most cases seek medical attention only when they become symptomatic and screening for such cases are far from their reach. We encountered 1 case of atrial myxoma with systemic embolization and one case was associated with Freidrich's Ataxia. Constitutional symptoms such as fever, weight loss, arthralgia and anemia were noted in 27% of the cases (8) in our study.

Diagnosis

Historically cardiac tumours have been diagnosed by angiocardiography and M-mode echocardiography [12].

Transseptal angiocardiography has been used in numerous centers, but tumour embolism has been reported with use of this method[13]. Pulmonary levophase angiocrdiography has proved a reasonably accurate method of visualizing a left atrial myxoma. Yet in our series, cardiac catheterization was not used as the sole radiological

method of diagnosing a myxoma, however 2 cases underwent catheterization for assessing their coronary artery status preoperatively.

In 25 of our patients, the cardiac mass was diagnosed preoperatively by Trans Thoracic Echocardiography. 5 patients with left atrial myxoma required Trans Esophageal Echocardiography for the confirmation. In our study 2-dimensional echocardiography was diagnostic in 83% of cases and TEE was needed for confirmation in rest of the cases surgical findings corroborated with the echocardiographic findings in all but one case where myxoma was found to be adherent extensively to roof of left atrium, mitral valve and inter-atrial septum.

Surgical Observations and Recommendations

When a diagnosis of left atrial myxoma is made, surgery should be done without delay. Current surgical techniques for treating atrial myxomas include median sternotomy with total cardiopulmonary bypass, using moderate hypothermia with cold crystalloid cardioplegia with minimal touch technique of the heart to prevent tumour embolization, which can be a serious intraoperative complication of this procedure [14].

Standard surgical approach for the management of left atrial myxoma is Bi-atrial [15,16], but in our experience in the management of 30 cases, we used right atrial approach alone for the management of 25 cases of cardiac myxoma without involving mitral valve, 20 cases were approached through right atrium and one case was approached through left atrium. All the four cases of right atrial myxoma were operated through right atrium. Of the two right ventricular myxomas, one was removed through right atrium and the other required a right ventriculotomy. Bi-atrial approach was used in 3 cases were tumour was hugely enlarged and adherent to mitral valve. The mean aortic cross clamp time and Cardiopulmonary bypass time was shorter in patients operated through right atrium compared to patients who were operated through Bi-atrial and left atrial approach. All cases included in the study were discharged and no deaths have occurred. From our study right atrial approach as an alternative to Bi-atrial approach in the management of cardiac myxomas arising from left atrium is worth mentioning.

Conclusion

All the cases included in the study (100%) presented with cardiac symptoms which warranted them for medical attention. However, a systematic screening is essentially required to diagnose and treat cases at asymptomatic level with a view to prevent morbidity.

Among the 30 cases studied in the institution, Atrial myxoma's incidence was more in females 22(73%) compared to males 8(27%) which correlated with International studies [2,12]. Myxoma's typically occur between 3rd to 5th decade which was observed in our study also [12]. Trans thoracic Echo (TTE) was the investigation of choice which was seen in our study (83%) again correlated with other studies [17,18].

Right atrial approach was the choice of surgical excision in majority of the cases in our study, in which the mean ACC and CBP time was comparably less than the bi-atrial approach. We conclude that cardiac myxomas are a rare group of tumours and cardiovascular surgeons must be familiar with this condition as only a tip of an iceberg present to us, right atrial approach could be used as an alternative to the classical bi-atrial approach.

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References

1. Mcallister HA,Fenoglio JJ,Tumours of the cardiovascular system, atlas of tumor Pathology, series 2. Armed forces Institute of Pathology, 1978.
2. St .john Sutton MG, Mercier LA, Guiliani ER, Lie JT.Atrial myxomas : a review of clinical experience in 40 patients. Mayo clinic proc 1980; 55:371.
3. Kang N, Clifford FH. Massive pulmonary embolism complicating left atrial myxoma; J Thoracic and Cardiovascular surgery 2001; 121:814-5.
4. Goldberg HP, Glenn F, Dotter CT, Steinberg I. Myxoma of the left atrium: Diagnosis made during life with operative and postmortem findings. Circulation 1952; 6:762–7.
5. St John Sutton MG, Mercier LA, Giuliani ER, Lie JT. Atrial myxomas. A review of clinical experience in 40 patients. Mayo Clin Proc 1980; 55:371–6.
6. Comer TP,Arbegast R, Schmalhorst WR. Left atrial

myxoma-diagnostic and surgical aspects. Calif Med 1973 Apr; 118:18-20.

7. Walton JA, Khan DR, willis PW: Recurrence of a left atrial myxoma. Am J cardiol 1972; 29: 872.
8. Piazza N, chughtai, T, Toledano K, sampalis J, Liao C , Morin JF. Primary cardiac tumors: eighteen years of surgical experience of 21 patients. Can aj cardiol. 2004 Dec; 20(14):144-8.
9. Murphy MC, Sweeney MS, Putnam JB Jr, et al. Surgical treatment of cardiac tumors: a 25-year experience. Ann Thorac Surg 1990; 49:612–18.
10. Silerman NA. Primary cardiac tumours.Ann surg 1980; 191:127-38.
11. Powers JC,Falkoff M,Heinle RA,et al. Familial cardiac myxoma: emphasis on unusual clinical manifestations.J Thorac cardiovasc surg 1979; 77; 782-9.
12. Sutsch G,Jenni r,Von Segesser L,Schneider J.Heart tumours : incidence , distribution,diagnosis of cardiac myxomas. 1:schweiz Med Wochenschr. 1991 Apr 27; 121(17):621-9.
13. Pindyck F, Oierce EC, Bron MG, et al. embolization of left atrial myxoma after trans-septal cardiac catheterization.Am J cardiol 1972; 30:569-71.
14. Comer TP, Arbegast R, Schmalhorst WR. Left atrial myxoma-diagnostic and surgical aspects. Calif Med 1973 Apr; 118:18-20.
15. Kabbani SS, Jokhadar M, Meada R, et al. Atrial myxoma: report of 24 operations using the biatrial approach. Ann thorac surg 1997; 63:697-700.
16. Semb BK. Surgical considerations in the treatment of cardiac myxoma. J Thorac cardiovasc Surg 1984; 87:251-9.
17. O Neill MB Jr, Grehl TM, Hurley Ej.Crdiac myxomas: a clinical diagnostic challenge. Am J Surgery 1979; 138:68.
18. Meller J, Teichholz Le, pickard AD, Matta R, Litwak R, Herman MV, et al. Left ventricular myxoma: echocardiographic diagnosis and review of the literature. Am J Med 1977; 63:816.