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Cardiac Myxoma, a Rare but Most Common Encountered Cardiac Tumor: A Single Center Experience

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ABSTRACT

Introduction: Cardiac myxoma is a benign and rare tumor, which can present with a grim phenomenon if the presentation is late or the diagnosis and surgery are delayed. The purpose of this study was to share our institutional experience of cardiac myxoma.

Material and Methods: This retrospective study was conducted to evaluate patients undergoing procedures at a single tertiary care centre for the treatment of cardiac myxoma during January, 2007 to December, 2017. Preoperative diagnosis was made by assessing clinical presentation and doing echocardiography. Complete tumor excision was performed, and all the patients were followed up for recurrence and complications.

Results: A total of 45 cases of cardiac myxoma (13 males and 32 females) with the mean age of 37.5 years old (ranged between 16 and 60 years old) were operated over the period of 10 years. Cardiac myxoma constituted about 0.69% of all cardiac cases operated at our institute. Out of all the subjects, 41, 3, and 1 cases had left atrial, right atrial, and right ventricular involvements, respectively. Additionally, 43 patients (95%) survived the surgery, one recurrence was observed during the follow-up period.

Conclusion: Cardiac myxoma is the most common cardiac tumor account for very small percentage of patients with heart disease. Early clinical suspicion and the use of imaging modalities are key to early diagnosis of this condition. Although these tumors have a risk for severe cardiac and systemic symptoms, referral to experienced centers for prompt surgical resection under cardiopulmonary bypass provides excellent early and long-term results.

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Introduction

Generally, cardiac tumors are rare compared to other heart diseases and tumors of the other organs. The results of autopsy studies demonstrated the overall prevalence of myxoma ranges from 0.002% to 0.33% (1). Approximately 75% of tumors originated from the heart are benign, and cardiac myxoma is the most common cardiac tumor comprising about 30-50% of the cases (2). In addition, about 75%, 23%, and 2% of these tumors are located in the left and right atrium and ventricle, respectively (3, 4).

Although it is extremely rare, the tumors might be found in multiple cavities (4). Myxomas are yellow-brown or green globular-shaped tumors with soft and gelatinous consistency. These tumors often contain the areas of hemorrhage and necrosis. They typically have short broad-based attachment; however, they can be sessile. Papillary forms may have a frond-like surface that is friable and susceptible for

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emolisation (2).

Although cardiac tumors do not significantly contribute to the overall tumor burden, they may cause a variety of cardiac and systemic symptoms. Cardiac tumors can be asymptomatic or have nonspecific symptoms, and they usually present insidiously. The presentation of cardiac myxoma depends on the location of tumor; therefore, the early diagnosis is a real challenge.

Clinically, the symptoms are presented through three mechanisms including intracardiac obstruction, systemic embolism of tumor fragments, or constitutional symptoms via unclear mechanisms (5).

Nowadays, the diagnosis is established most appropriately with two-dimensional echocardiography (6). In addition, cardiac computed tomography (CT) and magnetic resonance imaging (MRI) play an important role in the diagnosis of the tumor. Crafoord carried out prompt excision using cardiopulmonary bypass (CPB) in 1954 for the first time (7). Then, this method was determined as the only acceptable technique of treatment for these tumors.

Although cardiac myxoma has various presentations requiring elective or emergency intervention, the surgeon must try to prevent fragmentation and intraoperative embolization of the tumor, as well as the missing of an occasional multicentric lesion. This study was conducted to share our institutional experience and review the clinical experience, surgical management, and outcome of cardiac myxomas.

Materials and Methods

This retrospective study was conducted at the Department of Cardiovascular and Thoracic Surgery of Super-speciality Hospital, Government Medical College, Nagpur, India. All the patients, who were diagnosed as cardiac myxoma undergoing surgery in our institute during January, 2007 to December, 2017, were enrolled in this study. The subjects were the members of consecutive series of patients, who underwent elective or emergency cardiac surgeries to treat cardiac myxoma with or without valve replacement and other simultaneous cardiac surgery.

The demographic characteristics and clinical outcomes were retrospectively collected from the patient medical records. The follow-up of patients was conducted by contacting patients by making phone calls or by the help of outpatient clinics. Preoperative diagnosis was done in all patients by echocardiography. The pathological diagnosis of cardiac myxoma was made by the Department of Pathology of our centre. All the patients who presented with chest pain or were older than 45 years old underwent coronary catheterization to rule out coronary heart disease (CHD).

Statistical analysis

Data analysis was performed using SPSS, version 10. Continuous variables were presented as mean±standard deviation and in the case of normal distribution, compared using the independent samples t-test. In all the measurements, P-value less than 0.05 was considered statistically significant.

Results

Demographic profile

During 10 years, 45 patients with intracavitary cardiac myxomas underwent surgery at the institution, 13 and 32 of whom were male (29%) and female (71%), respectively. The patients' demographic characteristics are detailed in Table 1. Overall, the mean age of the patients at the point of tumor diagnosis was 37.5 years old (ranged from 16 to 60 years old), and females were disproportionately affected (71%). The cardiovascular risk factors included hypertension, diabetes mellitus, and positive history of tobacco use in 29%, 11%, and 40% of the subjects, respectively.

Clinical presentation and diagnosis

The most common manifestations of disease were dyspnoea (n=21, 46%), acute embolism (n=14, 31%), and chest pain (n=9, 20%). Approximately 55% (n=25) of the patients presented with the functional classes of III or IV based on the New York Heart Association classification. In addition, five patients (11.1%) presented with the constitutional symptoms of fever, palpitations, and weight loss.

Transthoracic echocardiography was the

Table 1. Demographic characteristics of the patients

	N (%)
Age, yrs. (mean _ SD)	16-60(37.5±15.6)
Sex (female/total)	32/45(71)
Risk factors	
Hypertension	13(29)
Diabetes mellitus	5(11)
Tobacco	18(40)
NYHA Class	
II	16(35)
III	24(53)
IV	1 (2)
Presenting symptoms	
Dyspnea	21(46)
Chest pain	9(20)
Palpitations	0(0)
Constitutional symptoms	5(11)
Thromboembolism	14(31)
Location	
Left atrium	41 (91)
Right atrium	3 (6)
Right ventricle	1 (2%)

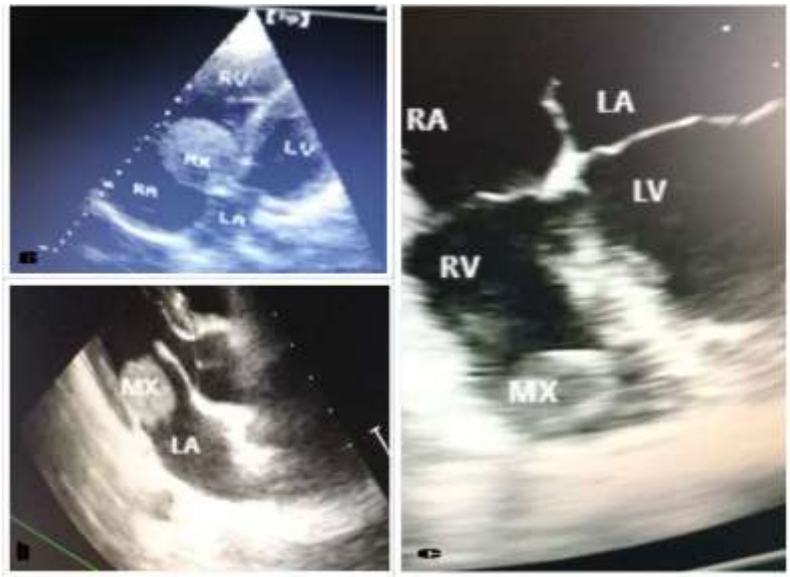

Figure 1. Two-dimensional transthoracic echocardiogram of right atrial myxoma (a), left atrial myxoma (b), and right ventricular myxoma (c)

primary instrument used for the diagnosis (100%, n=45). Additionally, two-dimensional intraoperative transthoracic echocardiography demonstrated right and left atrial and right ventricular myxomas (Figure 1). Intraoperative transesophageal echocardiography is utilized as a routine diagnostic tool in our institute. In this study, the use of MRI and CT scans was not quite prominent as primary diagnostic tools.

The cardiac CT and CT pulmonary angiography were executed for one patient with right atrial myxoma to rule out pulmonary embolism. Moreover, coronary catheterization was done in all patients older than 40 years old (33%, n=15), and none of them had significant CHD. Most of the patients with left atrial myxomas clinically mimicked mitral stenosis, while most of the patients with right atrial myxoma presented with the symptoms of right heart failure.

Surgical management

All the patients were subjected to surgery immediately after they were diagnosed as cardiac myxoma. The standard surgical approach was through a median sternotomy. Furthermore, CPB with aortic and bicaval cannulation with moderate hypothermia were employed.

Myocardial protection was achieved by intermittent antegrade cold blood cardioplegia. The heart was not manipulated until the aorta cross clamped to avoid tumor fragmentation and systemic embolization. The surgical approach for LA myxomas was left atrial, biatrial, or right atriotomy trans-septal. The approach for right atrial and ventricle myxomas was right atriotomy. The myxomas arising from atrial septum were resected with the excision of uninvolved atrial septum and rim of approximately 5 mm. The atrial septal defect was closed with autologous pericardial patches. Concomitant mitral valve replacement (MVR) and tricuspid annuloplasty (De Vega) were performed in one patient (2%) with large left atrial myxoma involving mitral annulus and in one patient with right atrial myxoma (2%), respectively. Copious irrigation of the atria and ventricles with cold saline was done to eliminate any loose tumor fragments that might have been dislodged during tumor resection. All the resected myxomas were sent to laboratory for routine histopathological examination (Figures 2 and 3).

Pathological findings

In this study, 41 (91.1%), 3 (6.6%), and 1 (2.2%) patients had left and right atrial and right

Figure 2. Resected pedunculated myxoma

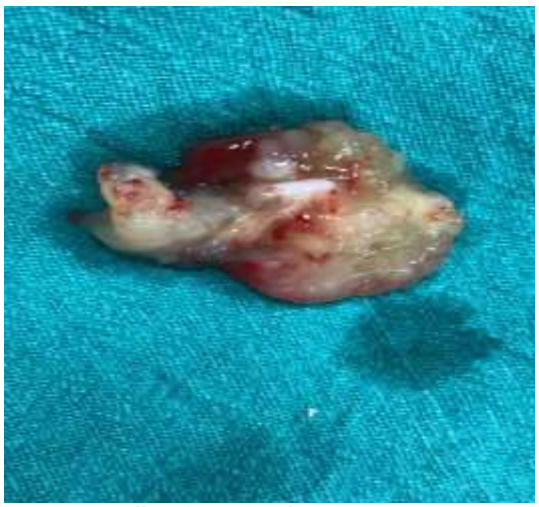

Figure 3. Papillary form of cardiac myxoma

ventricular myxomas, respectively. The mean size of the resected tumors was 3.9 cm for benign tumors. All the left atrial myxomas (n=41) were pedunculated except for one, which was sessile. The tumors were yellow-brown or green globular-shaped masses with soft and gelatinous consistency (Figure 2) and often contain the areas of haemorrhage and necrosis. The papillary forms of myxoma were present in 10 patients (22%), all of whom had embolism (Figure 3). Microscopic examination confirmed the diagnosis in all the patients. Typically, the histopathological examination revealed capillary proliferation, blood extravasation, and fibrin depositions.

Outcomes (morbidity, mortality, and follow-up)

In this study, the mean length of intensive care unit stay and total hospital stay were 3.6 and 8.6 days, respectively. Postoperative complications included bleeding requiring reoperation, renal failure requiring dialysis, pneumonia, pleural effusion, and respiratory failure, which occurred in 6.6% (n=3) of the subjects. Moreover, perioperative mortality was 4.4% (n=2) due to postoperative complication and cerebrovascular accident as a result of cerebral embolism.

The mean follow-up period was 8.5 years ranged from 4 months to 9.2 years. Recurrence was observed in one patient (2%), who was previously operated for left atrial myxoma in our institute three year before the presentation.

Discussion

Cardiac myxoma is a neoplasm of the heart with an estimated incidence of 0.5 per million (8). The exact incidence in India was not described before. In our study, cardiac tumor, which was predominantly cardiac myxoma, accounted for 0.69% of all cardiac operations (6500) during the study period performed at our institution compared to approximately 0.3% reported in western literature.

We never encountered with any other primary cardiac tumor pathology during the study period. This figure (0.69%) was slightly higher than that reported in the literature (approximately 0.3%) (9). In congruence with the literature, the tumor was more prevalent in females (71%). In our population, the mean age at the point of presentation was 37.5 years in contrast to the previous studies (55 years) (10).

The clinical presentations observed in our study population, such as dyspnoea on exertion, palpitations, congestive heart failure, syncope, haemoptysis, embolic events, atypical chest pain, paroxysmal nocturnal dyspnoea, fever, fatigue, weight loss, and night sweats were reported in previous studies (11-14). Left atrial myxoma with preoperatively systemic embolism is usually a fatal complication that was reported in 25-50% of patient (15-18).

In our study, 31% of the patients had preoperative embolic event, whose tumors were extremely friable with soft gelatinous morphology i.e. the papillary form of myxoma. To the best of our knowledge, this type of cardiac myxoma was not described in detail in previous studies. Two-dimensional echocardiography is the gold standard method for the diagnosis of myxoma with a sensitivity of 100% (19, 20).

Transesophageal echocardiography is superior to transthoracic echocardiography for identifying the size, shape, point of attachment, and motion characteristics of the atrial myxoma (21, 22). We routinely use two-dimensional transthoracic echocardiography for preoperative diagnosis and coronary catheterization is reserved only for special situations like suspected CHD or patients older than 40 years old.

In several cases, newer imaging techniques such as cardiac MRI and ultrafast-CT might be required. These techniques provide non-invasive, high-resolution cross-sectional views of cardiac structures (23). Once the diagnosis of cardiac myxoma is made, the emergent surgery is performed. Surgery has indication in all patients diagnosed as myxoma and is considered urgent since the risk of hemodynamic decompensation and embolism is present.

The mortality rate was between 8% and 10% due to embolic complications during the interval between diagnosis and surgery (24, 25). In our study, there was no preoperative death.

The gold standard of treatment of myxoma is curative surgical resection, and complete resection is best performed through a median sternotomy with total CPB and cardiac arrest (26, 27). Myxoma can be excised by less invasive or minimally invasive cardiac surgery using right

thoracotomy approach. There is a controversy surrounding the ideal surgical approach to achieve complete excision.

Jones et al. revealed that a surgical approach via biatrial approach allows minimal manipulation of the tumor, provide adequate exposure for complete resection, provide the inspection of the four heart chambers, minimize recurrence, and is safe and efficient (28). Other researchers considered the exposure of the left atriotomy approach to be adequate and demonstrated the low recurrence rates and the safety of the technique, although not to its attachment (29).

The trans-septal approach through right atriotomy, suggested by Chitwood, gives good access to the myxoma with minimum manipulation, which provides the inspection of all cardiac chambers. The right atrial and ventricular myxomas are approached through right atrium. Right atrial myxomas demand more care during cannulation. In this study, all the left atrial myxomas were successfully resected using biatrial approach except for two cases. In these two cases, the myxomas were pedunculated; therefore, they were excised through left atrial approach.

There is a general agreement regarding the necessity of full thickness resection with clear margins to minimize the risk of recurrence. In our institution, the myxomas arising from atrial septum were excised with the excision of uninvolved atrial septum with the rim of approximately 5 mm. Additionally, the atrial septal defect was closed with autologous pericardial patches.

Valvular damages should be corrected with annuloplasty, repair, or replacement (30, 31). In the present study, concomitant MVR and tricuspid annuloplasty (De Vega) were performed in one patient (2%) with large left atrial myxoma involving mitral annulus and one patient (2%) with right atrial myxoma, respectively.

The recurrence of a sporadic myxoma is rare occurring for 1-3% of the cases (32). The risk of postoperative recurrence was correlated with young age, positive family history of myxoma, inadequate resection, intraoperative implantation, or multicentric growth (33, 34). In this study, only one recurrence was observed (2.2%) in a previously operated patient with left atrial myxoma three years after first surgery. Therefore, a regular follow-up of all patients by non-invasive method is mandatory. The major complications of the surgery included tumor embolism, supraventricular arrhythmias, and the requirement of permanent cardiac pacing due to conduction disturbances. In our series, five patients had the episodes of supraventricular arrhythmias, which were controlled medically.

The results of surgical resection are generally very good with most series reporting operative mortality rates of less than 5% (30, 31). In the current study, perioperative morbidity and mortality (4.4%) were low and comparable with other reports.

Conclusion

Cardiac myxoma is rare and form a very small percentage of the cardiac tumors, which can present with a grim phenomenon. Symptomatology and clinical features are not specific, diagnosis is made by transesophageal echocardiography, in case of doubt, other modalities of investigation should be used, once a diagnosis of myxoma is made, surgery should not be delayed. The prognosis for patients after surgical resection is excellent. A close follow-up of all the patients is mandatory because the reappearance of myxomas has been documented at various intervals.

Limitation of the Study

The limitations of our study include its retrospective and descriptive nature and small cohort size. Retrospective studies are susceptible to selection and recall bias. However, considering the rarity of the disease, a prospective study is impractical.

Disclosure

Authors report no conflict of interest. The study was approved by the Ethics Committee of our institution SSH & GMC, Nagpur. Because of its retrospective nature, patient consent was waived. The cardiac procedures performed were in accordance with the routine standard of care being followed at the institution during that period. Consent for surgery was obtained routinely from all individual participants included in the study. This research received no specific grant from any funding agency in the public, commercial, or non-profit sectors.

Conflict of Interest

The authors declare no conflict of interest.

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