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Surgical Management of Benign Tumors of the Heart: A Rare but Important Pathology

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Abstract

Objectives: Primary neoplasms of the heart are relatively uncommon, but represent an important cardiovascular pathology since early diagnosis can be curative. The purpose of this study was to share our institutional experience and surgical management of benign cardiac tumors.

Methods: In this retrospective chart review of patients, we evaluate the incidence, demographics, clinical presentation, histopathological findings, surgical management, and outcomes of patients undergoing procedures at a single tertiary care center for treatment of benign cardiac tumors from January 2000 to October 2013.

Results: 16 patients (4 male and 12 female) with a mean age of 47.3 years (range of 23-79 years) were identified. The most common presenting symptoms were dyspnea (53%), constitutional symptoms (32%), chest pain (26%), and neurological symptoms (16%) and endocarditis (11%). All benign tumors were grossly resected on cardiopulmonary bypass. Myxomas were the most common cardiac tumors, most occurring in the left atrium. There was postoperative mortality. No tumors recurred during our follow-up period.

Conclusions: Early clinical suspicion and use of multiple imaging modalities is a key to early diagnosis of benign cardiac tumors. 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.

Keywords: Cardiac tumor; Primary cardiac neoplasms; Primary cardiac neoplasms; Benign cardiac tumor; Myxoma; surgical resection

Abbreviations: EKG: Electrocardiogram; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; CVA: Cerebrovascular Accidents; EF: Ejection Fraction; CABG: Coronary Artery Bypass Graft; ICU: Intensive Care Unit; CHF: Congestive Heart Failure; HTN: Hypertension; CAD: Coronary Artery Disease; LA: Left Atrium; RA: Right Atrium; LV: Left Ventricle; RV: Right Ventricle; MRI: Magnetic Resonance Imaging

Introduction

Cardiac tumors range from benign lesions to high-grade malignancies over a wide range of ages. Primary neoplasms of the heart are relatively uncommon, with an incidence between 0.001% to 0.3% but represent an important cardiovascular pathology since early diagnosis can be curative.

The medical literature for primary cardiac tumors is quite sparse on anything deeper than epidemiology and case studies on these medical paucities. Several recent studies have found that the incidence of benign primary cardiac tumors greatly outweigh the incidence of primary malignant cardiac tumors [1-4]. Mortality in patients with primary cardiac tumors mimics that of the general population [5]. The most common types of benign tumors demonstrate an extremely high 5, 10, and even 15-year survival rate [6-7].

The most important determinant in mortality of this cohort of patients, like all tumors, is the histological morphology of the tumor [5]. The most common cause of death in patients with primary cardiac tumors is cardiac symptoms associated with a space-occupying lesion and dysrhythmias [1].

The main types of benign primary cardiac neoplasms are myxomas, papillary fibroelastomas, rhabdomyoma, lipomas, and hemangiomas. Most of the in-depth epidemiological work in this area regards the myxoma, the most common type of these very rare tumors. There are several large cohort studies on the clinical presentation and management of myxomas. The incidence of these tumors is higher in females than in males, with an average age of diagnosis of 52 years. These patients most often present with dyspnea, heart failure, or stroke. Friable, irregular myxomas increase the risk for systemic embolism. Presentation of cardiac myxomas can also mimic many infectious, immunological, and malignant processes. Rarely, these tumors can present as the source hypertrophic cardiomyopathy [8]. Another uncommon form of presentation of these tumors is seizure activity [9]. This population exhibited an ejection fraction within current normal limits, making diagnosis of these tumors very difficult [10-13]. The gold standard in diagnosing these tumors has been the electrocardiogram (EKG). CT (Computed Tomography) with cardiac MRI (Magnetic Resonance Imaging) is equally efficacious [2-3].

The location of the tumor is an important clinical indicator as to the histology of the various primary cardiac tumors. Myxomas are most common in the left atrium, although they have been found in the right atrium or with multi-site origins. Non-myxomatous benign tumors are common in the ventricles. Malignant primary cardiac tumors often have a multifocal origin. Secondary tumors are generally localized to the right side of the heart.

Procedures to remove benign neoplasms of the heart are curative and reduce the risk of pulmonary embolism [14] and ischemic stroke [15]. The Kuroczyński study described the several key facets to the operative management of patients in this category [16]. First, the procedures have low

operative risks and excellent benefits for the patient. The time between the diagnosis and operative treatment of these patients is a critical value; the sooner the operation can be performed, the better outcome of the patient. Regular follow-up is needed for the appropriate continued management of these patients. The gold standard in monitoring these patients in follow-up is the EKG [2]. The time between diagnosis and treatment is crucial because these tumors have a documented risk of causing an initial ischemic stroke in patients, especially those younger than 50.

In this retrospective chart review of patients with cardiac tumors, we evaluate the incidence, demographics, clinical presentation, histopathological findings, surgical management, outcomes and factors influencing success of patients undergoing procedures for treatment of cardiac neoplasms of patients from January 2000 to October 2013 with a cardiac neoplasm treated at the University of Kentucky Medical Center. The purpose of this study was to share our institutional experience and outcome of surgical management of benign cardiac tumors.

Materials and Methods

The medical records of all patients with cardiac tumors from January 2000 to October 2013 who underwent surgery at the University of Kentucky Medical Center in Lexington, Kentucky were reviewed. A waiver of consent was obtained from the Institutional Review Board. Subjects for the study were identified by the institutional cardiothoracic surgery database. Patients enrolled in the study were males or females older than 18 years of age who were treated in the cardiothoracic surgery department for surgical management of cardiac tumors. All patients with suspected cardiac mass were evaluated with echocardiogram and/or transesophageal echocardiogram or cardiac magnetic resonance. Diagnosis was confirmed with surgical excision. Data are presented as mean or number and percentage. Statistical calculations were performed using Microsoft Excel software (Microsoft Corporation, Redmond, WA, USA).

Results

A total of 16 patients were diagnosed with a primary benign cardiac tumor. We retrospectively reviewed demographics, critical presentation, diagnostic data, location of lesions, operative reports, surgical outcome through medical records and clinical follow-up, pathology reports, and echocardiogram results. Pathological data collected included tumor type, location, and tumor diameter.

Demographics

The mean age of diagnosis was 47.3 years (age range of 23-79 years), with 25.0% (n=4) being male and 75.0% (n=12) being female. Table 1 summarizes the patient characteristics at the time of presentation. Over 40% of our population reported past or active tobacco use and hypertension. Over 30% of our cohort had diabetes and were obese.

Diagnosis

There was no one defining clinical presentation for the patients within our cohort. The most common presenting symptoms were dyspnea (56%), constitutional symptoms (32%), neurological symptoms (25%) and chest pain (19%). Twelve percent of our population presented with thromboembolism or endocarditis. Table 2 summarizes the clinical presentations.

The primary diagnostic and imaging tool used to diagnose patients with a cardiac tumor was transthoracic echocardiography (87.5%, n=14). Two-dimensional intraoperative transthoracic echocardiography of large left atrial myxomas are demonstrated in figure 1. Transesophageal echocardiogram was used intraoperatively as a matter of routine. Several other methods were used as diagnostic tools in our cohort. The use of MRI and CT scans was quite prominent as primary and secondary diagnostic tools (75.0%, n=12). Figure 2 demonstrates a cardiac MRI of a left atrial myxoma. The average pre-operative ejection fraction (EF) for patients was difficult to determine due to missing data in the electronic medical record system. Of the 7 patients whose EF was reported, 4 had normal EF, 1 had a slightly low EF, and 2 had greatly decreased EF.

Operative treatment

All benign tumors were grossly resected, with the exception of one benign hemangioma that was only biopsied due to the risk of resection. One patient with sepsis from an infected patch was treated with an exploratory sternotomy with debridement of the sternal infection and replacement of the patch.

Median sternotomy was utilized in all patients. Cardiopulmonary bypass was conducted with bicaval cannulation, moderate systemic hypothermia, deep topical cooling, and cardioplegic cardiac arrest. Adequate cardiopulmonary bypass was achieved in 93.8% (n=15) of our patients. One patient also underwent a coronary artery bypass

	Patient Characteristics						
Gender	CHF	Diabetes	HTN	Obesity	CAD	Active Tobacco Use	Past Tobacco Use
Male	50% (2)	0%	50% (2)	25% (1)	25% (1)	75% (3)	75% (3)
Female	0%	33.3% (4)	50% (6)	33.3% (4)	16.7% (2)	31.3% (5)	50% (6)
Total	12.5% (2)	25% (4)	50% (8)	31.3% (5)	18.8% (3)	50% (8)	56.3% (9)

Table 1: Patient characteristics at presentation
CHF: Congestive Heart Failure; HTN: Hypertension; CAD: Coronary Artery Disease

	Tumor Type				
Presenting Symptoms	**All benign tumors (16)**	**Myxoma (10)**	**Papillary fibroelastoma (4)**	**Fibroma (1)**	**Hemangioma (1)**
Dyspnea	0.56	60% (6)	25% (1)	100% (1)	100% (1)
Chest Pain	0.19	10% (1)	0	100% (1)	100% (1)
Palpitations	0	0	0	0	0
Neurological	0.25	30% (3)	25% (1)	0	0
Constitutional	0.25	40% (4)	0	0	0
Heart Failure (right or left)	0.0625	0	25% (1)	0	0
Atrial fibrillation	0.0625	10% (1)	0	0	0
Ventricular tachycardia	0	0	0	0	0
Thromboembolism	0.125	10% (1)	25% (1)	0	0

Table 2: Clinical Presentation

graft (CABG) procedure in addition to their gross resection with cardiopulmonary bypass.

Pathology

The histologic type and location of the tumors is summarized in table 3. The median size of resected tumors was 4.8 cm for benign tumors. No patients had multiple tumors. Accurate margin information was not available for those undergoing gross resection of their masses. Myxomas were the most common tumor found in our cohort (62.5% of all benign tumors in our study, n=10).

Morbidity and mortality

Morbidity and mortality data are summarized in table 4. The predominant complications were pulmonary, with four patients suffering from respiratory failure and three from pleural effusions. The median length of hospital stay was 9.6 days for our cohort. The length of intensive care unit (ICU) stay as well as the presence of postoperative complications was indicative of a longer hospital course.

There was one in-hospital mortality in our cohort. This patient presented pre-operatively with congestive heart failure and underwent excision for a right ventricular mass, a Maze procedure to correct atrial fibrillation, and a tricuspid valve replacement for tricuspid regurgitation. On post-operative day (POD) #0, the patient had increased chest tube output from a mediastinal hemorrhage that required sternotomy to correct. Following this procedure, the patient was placed on mechanical ventilation and was never extubated. On post-operative day #18, the patient developed respiratory failure, right-sided cardiac failure, atrial fibrillation with rapid ventricular rate and acute renal failure. The patient expired in the ICU on POD#19.

Survival

In our cohort, there was one in-hospital mortality. Thirty-day survival was confirmed in eleven of our patients and one-year survival was confirmed in 8 patients.

Recurrence

In our limited study time and cohort, we experienced no recurrence of disease in our patients.

	Tumor Location			
Tumor Type	LA	RA	LV	RV
Myxoma	80% (8)	10% (1)	10% (1)	0
Papillary Fibroelastoma	75% (3)	0	25% (1)	0
Fibroma	0	0	100% (1)	0
Hemangioma	0	0	100% (1)	0

Table 3: Tumor type and location
LA: left atrium; RA: Right Atrium; LV: Left Ventricle; RV: Right Ventricle

Benign	
Total	16 (100%)
Morbidity	
Cardiac	0
Respiratory Failure	4 (25%)
Pleural Effusion	3 (18.8%)
Early Mortality	1
Late Mortality	0

Table 4: Major clinical outcomes: Morbidity and mortality of operative treatment of cardiac tumors

Discussion

Cardiac tumors may be primary (benign or malignant) or secondary (metastases of the heart). Primary tumors of the heart are abnormal growths that arise from the normal tissues that compose the heart or heart valves. Primary cardiac tumors have an incidence of 5% of all cardiac tumors and secondary tumors are 95% of all cases. Approximately 75% of all primary cardiac tumors are benign.

The most common primary tumor of the heart in the literature and in our study is the myxoma, which makes up approximately 77% of all primary tumors of the heart [17,18]. Atrial myxomas have a predilection for female sex (80% of atrial myxomas in our series were female, n=8). They are most common in the left atrium (80% in our series) and in solitaire. Myxomas with multi-site origins are the only myxomas that have demonstrated an ability to recur after surgical excision [7]. Papillary myxomas have a much higher rate of overall incidence, as well as a higher rate of embolism compared to solid myxomas [11]. The Ikeda case study described a pulmonary embolism secondary to a right atrium myxoma with a tail-shaped projection [14]. This case was treated with urgent surgical resection. A pulmonary embolism is a well-known risk for patients diagnosed with a primary cardiac tumor. Embolism is more likely in myxomas that are gelatinous or lobulated versus solid neoplasms.

Other types of benign primary tumors include papillary fibroelastomas, rhabdomyomas, fibromas, hemangiomas, teratomas, lipomas, paragangliomas, and pericardial cysts. Papillary fibroelastomas were the second most common in our series, which is consistent with the epidemiology of cardiac tumors. The benign neoplasms with the highest hazard ratios are lipomas and papillary fibroelastomas. Correcting for variables such as diabetes mellitus, age at surgery, presence of coronary artery disease, and hypertension is a requirement for understanding the survival likelihood of patients with these types of tumors, as both fibromas and lipomas have a significantly decreased apparent hazard ratio if the factors are left uncorrected.

Although our review did not include pediatric patients, this population has a very similar experience to the adult population [17]. The presence of benign tumors is relatively common in children, with the rhabdomyomas being the most common primary tumor. Unlike adults, cardiac tumors are much more common in males than in female pediatric patients. They present with symptoms of heart congestion; symptoms such as heart failure, arrhythmia, cerebrovascular accidents (CVAs), and paresthesias. These tumors can also be asymptomatic in children. Treatment for these cases follows the same pattern as adults, with surgery being the definitive option. The post-surgical prognostic indicators of poor recovery include ventricular outflow obstruction and distant thromboembolism [19]. In pediatric patients, the presence of rhabdomyomas has a strong correlation with the child also have tuberous sclerosis complex.

Clinical manifestations of cardiac tumors depend on the tumor type, location, size, and friability, and can include symptoms related to valvular or inflow-outflow tract obstruction, arrhythmias, thromboembolism, or constitutional symptoms [20]. Therefore, symptoms may be nonspecific and can mimic other cardiac diseases, making diagnosis difficult. Obstructive symptoms may cause chest pain, shortness of breath, or syncope. Constitutional symptoms may include cachexia, fever, or arthralgia. The majority of the tumors of the heart has a benign course and is not directly fatal; however, even benign tumors can be lethal due to direct extension into the electrical conduction system of the heart (resulting in dysrhythmias or complete heart blocks). Emboli are more common in smaller tumors with irregular surfaces. In our series, none of our patients with a cardiac tumor had a history of embolic stroke.

Diagnosis is often delayed since symptoms can mimic those of more common disorders. Diagnosis is confirmed by imaging, which is useful

to collect data on the size, mobility, myocardial invasion, relationship with adjacent structures and chamber location of the mass and other characteristics that help to determine diagnosis and prognosis. Echocardiography is the preferred modality for diagnosis. Transesophageal echocardiography is better for visualizing atrial tumors and transthoracic echocardiography is better for visualizing ventricular tumors. Cardiac MRI and cardiac CT can be useful complementary diagnostic tools. Cardiac MRI is useful in characterizing the extent of a mass and its relationship with adjacent structures. Often, tissue characterization with MRI can aid in generating differentiation considerations and can aid in treatment planning for surgical cases. Figure 1 and Figure 2 demonstrate the utility of cardiac MRI for the characterization of cardiac masses. Biopsy is not necessary since imaging studies can often distinguish benign from malignant tumors.

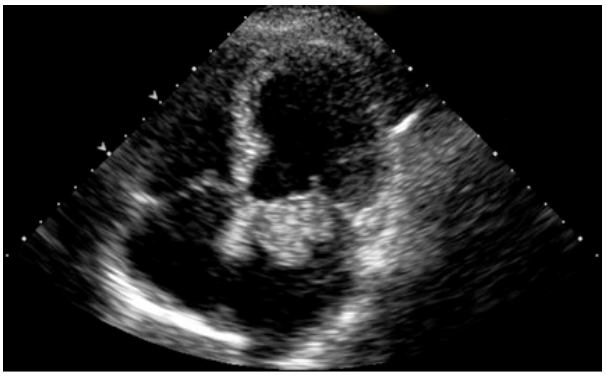

Figure 1: Two-dimensional intraoperative transthoracic echocardiography of a large left atrial myxoma

The patient initially presented with severe intermittent dyspnea. The patient subsequently underwent resection of the gelatinous left atrial myxoma via sternotomy with cardiopulmonary bypass. At the time of resection, there was a very small stalk attached to the anterior leaflet of the mitral valve near the anterior leaflets attachment to the annulus, which was repaired after removal of the myxoma. The patient had resolution of presenting symptoms following the procedure.

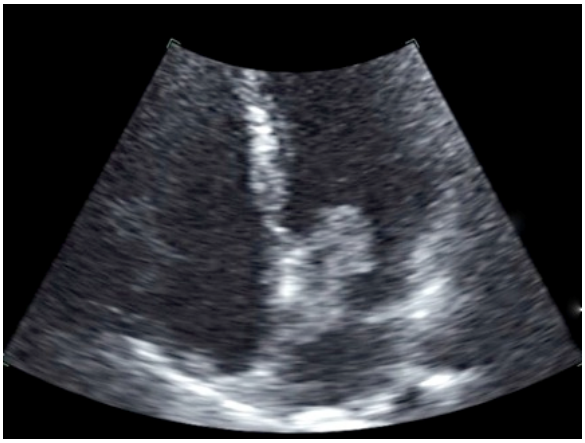

Figure 2: Cardiac Magnetic Resonance Imaging (MRI) of left atrial myxoma

Routine cardiac MRI performed in order to characterize the left atrial mass and to identify additional cardiac masses demonstrated a large 4 × 1.7 cm mass in the left atrium attached to the inter-atrial septum. The mass is minimally enhanced by perfusion and has significantly lower signal on early and late gadolinium enhancement images compared to the myocardium. Myxomas are typically pedunculated masses located in the left atrium. There was no other intracardiac masses identified based on perfusion imaging, early and late gadolinium enhancement imaging.

For benign tumors, treatment is surgical excision followed by serial echocardiography over five years to monitor for recurrence. Prompt surgical excision minimizes the sequelae of obstruction, embolization, and arrhythmias [21]. Surgery is typically curative, with a 95% survival rate at 3 years (with the exception of rhabdomyomas, most of which regress spontaneously and do not require treatment). Excision of intracardiac tumors involve establishment of cardiopulmonary bypass and cardioplegic arrest with sufficient exposure of the tumor to achieve complete resection without embolization. Perioperative tranesophageal echocardiography can facilitate visualization of all four cardiac chambers for concomitant lesions. Lin, et al. examined the long-term effects of using a gross resection via a midline sternotomy using cardiopulmonary bypass [6]. They reported excellent 5 and 10-year survival rate with a very low risk of needing a secondary surgery. Iribarne found that a minimally invasive operation had equal safety to the traditional resection approach, with no compromise in tumor resection margins, and a reduced length of mean hospital day by 2 days [22].

We found that our series closely reflected those represented in the literature in terms of prevalence, presentation and modalities for diagnosis and resection. Myxomas and solid tumors were more common and best excised in one part. Complete surgical excision on cardiopulmonary bypass is the preferred method of treatment.

Conclusions

Primary neoplasms of the heart are relatively uncommon but not rare and represent an important cardiovascular pathology since early diagnosis can be curative. Early clinical suspicion and use of multiple imaging modalities is a key to early diagnosis. Although benign cardiac tumors have a risk for severe cardiac and systemic symptoms, referral to experienced centers for prompt surgical intervention provides excellent early and long-term results. Surgery is typically curative, with a 95% survival rate at 3 years for most tumor types.

Our small cohort of patients verifies the same trends reported in larger, longitudinal studies on these tumors. In our study, we found dyspnea to be the most common presenting symptom and left atrial myxomas to be the most common primary cardiac tumor. The location of the tumor is an important clinical indicator as to the histology of the various primary cardiac tumors. Clinical manifestations of cardiac tumors depend on the tumor type, location, size, and friability, and can include symptoms related to valvular or inflow-outflow tract obstruction, arrhythmias, thromboembolism, or constitutional symptoms.

We also confirmed benign tumors to be a left-sided phenomenon, which parallels trends found in the literature. The clinical outlook for patients receiving this diagnosis is one of optimism due to two factors: very low mortality/morbidity and a very low rate of recurrence after the creation of negative resection margins during surgical management.

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