A 12-year Experience with Primary Cardiac Tumors

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Abstract

We report our experience with 117 patients with primary cardiac tumors who underwent surgery at our institute (a referral center) between March 1995 and February 2006.

The patients comprised 47 men and 70 women with a mean age of 44.97 years (range: 2.5- 81 years). The predominant symptom was dyspnea on exertion and palpitation. In all the patients, echocardiography was the main diagnostic tool, but magnetic resonance imaging (MRI) and CT scan and coronary angiography were also performed if indicated. Most of the tumors were found in the left atrium (LA) (77.77%), but the other chambers were also involved with lesser prevalence (right atrium: 7.5%, left ventricle: 5.1%, and right ventricle: 2.5%). Involvement of multiple chambers was found in 8 (6.8%) patients.

All the patients survived the surgical procedure and were discharged from hospital. Follow-up ranged from 1-10 years (mean: 2.4 years). The most prevalent tumor was myxoma (104 cases), followed by sarcoma (4 cases) and fibroma (2 cases). Four patients had secondary (metastatic) cardiac tumors (two Hodgkin lymphoma, one renal cell carcinoma, and one osteosarcoma) and were consequently excluded from the study ***(Iranian Heart Journal 2010; 11 (2):49-54).***

Primary cardiac tumors are benign or malignant neoplasms arising primarily from the myocardium or within a cardiac chamber. These are rare tumors, with large autopsy studies reporting an incidence of between 0.0017% and 0.03%. Most tumors are benign (75%), with the first and second most common being myxoma (50% of all cardiac tumors) and rhabdomyoma (20%). [9]

There has been little experience with primary tumors, and our report is one of the largest series of surgical reports on primary cardiac tumors with evaluation of presenting symptoms, most common echocardiographic findings, sites of involvement, and recurrence of these potentially lethal tumors.

Methods

Between March 1995 and February 2006, a total number of 141 patients were diagnosed as having primary cardiac tumors in our center. Apart from 20 patients who could not undergo surgery due to various reasons, the rest of our study population underwent surgery. Four patients were excluded from the study because of the final diagnosis of metastatic tumors (2 lymphomas, 1 metastatic osteosarcoma, and 1 metastatic renal cell carcinoma). The 117 patients with primary cardiac tumors, who were operated on, were comprised of 47 men and 70 women at a mean age of 44.97 + years (range: 2.5-81 years).

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These cases were evaluated for symptoms, diagnostic methods, site of involvement, nature of tumor found intraoperatively, associated procedures with tumor resection, follow-up, recurrence, and results of histological evaluation of the tumors.

Results

Symptoms

The most common presenting symptoms amongst our patients were dyspnea on exertion (80 patients, 93.6%), and palpitation (42 patients, 35.8%). However, as is reported by previous studies in the literature, a wide range of symptoms was found. (Table I)

Table I. prevalence of symptoms in patients with primary cardiac tumor

Symptoms	number of cases	%
D.O.E	80	93.6%
Palpitation	42	35.8%
Chest pain	18	15.38%
Syncope	15	12.8%
C.V.A	13	11.1%
Fever	9	7.6%
Weight loss	8	6.8%
Asymptomatic	7	5.9%
Vertigo	6	5.1%
T.I.A	5	4.2%
Loss of appetite	4	3.4%
Nausea & vomiting	3	2.5%
Fatigue	2	1.7%
Hemoptysis	2	1.7%
D.V.T	2	1.7%
Disorientation	1	0.8%
peripheral embolism	1	0.8%

DOE: dyspnea on exertion, CVA: cerebrovascular accident
TIA: transient ischemic attack, DVT: deep vein thrombosis

Diagnostic Methods

All the tumors in our patients were diagnosed via transthoracic echocardiography. For 37 patients, transesophageal echocardiography was performed because of equivocal results or for more detailed evaluations of the chambers or the cardiac valves.

The most common echocardiographic findings are listed in Table II.

Table II. The most echocardiographic findings

Finding	number of patients	%
Mitral regurgitation	35	29.9%
Mitral stenosis	6	5.1%
Tricuspid regurgitation	36	30.7%
Mobile mass	13	11.1%
Protruding mass	10	8.5%
Pulmonary	15	12.8%
hypertension	11	9.4%
Pericardial effusion		

It is interesting that most of the patients had good cardiac function as evaluated by ejection fraction (mean: 49.9%).

For 57 patients, coronary angiography was performed because of suspicion to coronary artery disease or as a screening method for the patients over 45 years old: 11 of them had significant coronary artery disease (19.9% of patients who evaluated by angiography).

Other diagnostic methods for the evaluation of the details were computed tomography and angio CT scanning, conducted in 5 patients, and MRI, carried out in 2 patients.

With respect to the sites of involvement and method of resection, the most common chamber involved by a tumor was the left atrium (91 patients, 77.77%). The other chambers were involved at lesser frequencies (Table III).

Table III. Sites of involvement by primary cardiac tumors

Site of involvement	number of patients	%
Left atrium	91	77.77%
Right atrium	9	7.6%
Left ventricle	6	5.1%
Right ventricle	3	2.5%
Multiple chambers	8	6.8%
Involvement of valve	12	10.2%

Multiple-chamber involvement was found in 8 (6.8%) patients; in one case, the tumor involved 3 chambers (LA, RA, and LV).

Involvement of the valve was found in 12 (10.2%) patients, and it necessitated mitral valve replacement in 3 patients and mitral valve repair in 1 patient. In the other cases, no procedure was performed on the valve.

In all the cases, surgical excision was performed via a mid sternotomy with the aid of cardiopulmonary bypass, which was established using the ascending aorta and dual venous cannulation.

The approach of the tumors was trans-septal or with the opening of both atria. We believe that the biatrial approach confers a good evaluation of the four chambers of the heart for additional tumors.

In the patients with the involvement of the interatrial septum, the involved part was resected and the defect was repaired with primary closure (55 cases) or with pericardial patch (46 cases).

Additional procedure was pulmonary artery embolectomy in one case, which was complicated by the embolization of the tumor particles to the pulmonary artery.

In regard to recurrence and familial history, the patients were followed for 1-10 years (mean: 2.4 years). During this time, 10 cases had recurrence (one patient had two episodes of recurrence). The time of recurrence was between 9 months and 18 years (the latest case was involved in our study at her first episode of recurrence). In all the cases, the chamber of recurrence was the same that was involved in the first instance except for one patient who had recurrence with multiple tumors in the LA and LV chambers.

The main symptoms at recurrence were dyspnea on exertion (6 cases), arrhythmia (one case), cerebrovascular accident (one case), and 2 patients were asymptomatic at the time of recurrence and were diagnosed in routine follow-up.

Amongst our patients, one case had a familial history of cardiac myxoma (in her mother and brother) (0.8%). She was involved by the tumor when she was 17 years old. She had two episodes of recurrence at 33 and 35 years old.

Histological evaluation

Amongst the 117 patients with a diagnosis of primary cardiac tumor, the most prevalent type was myxoma (104 cases, 88.88%), followed by sarcoma and angiosarcoma (4 cases, 3.4%), fibroma (2 cases, 1.7%), and rhabdomyoma (one case, 0.8%).

Discussion

Primary cardiac tumors are rare with large autopsy studies reporting an incidence of between 0.0017% and 0.03%. Females are involved more than males,[1,2] with a mean age of 40 years old.[2,3] In this study, the mean age of the patients was 44.97 years with the involvement of females being more than that of the males (60% vs. 40%). Most of the tumors in our patients were benign, and myxoma was as the most prevalent pathological diagnosis (104 patients, 88.88%), followed by fibroma and hemangioma (each 2%), as benign tumors and 4 (3.4%) patients were involved by sarcoma and angiosarcoma as malignant tumors. Other series have reported 16-25% of all primary tumors as malignant tumors,[1,3,4.5,6,7,8,9] which is much higher than that found in this study.

Although myxomas are considered benign for a lack of aggressive mitotic activity and metastasis, these tumors can cause cardiac insufficiency by interfering with atrioventricular or valvular function. In his study, the most common symptom was dyspnea due to heart failure (93,6%) and echocardiographic evidence of valvular dysfunction was found in 35% (M.R: 29.9% and M.S: 5.1%). Additionally, in 10 patients, a protruding mass through the valve was found. Fifteen patients had echocardiographic evidences of pulmonary artery hypertension (12.8%), too.

Cardiac tumors may be a source of cerebral or peripheral emboli (embolism up to 40% in

some series).[10,11] Embolic events may occur in the pulmonary vascular tree.[12] In our study, the prevalence of ischemic cerebrovascular accident, TI.V.A, T, I.A, and peripheral arterial embolism was 11.1%, 4.2%, and 0.8%, respectively. One patient was involved by the pulmonary embolization of tumor fragments of a myxoma, located in the right ventricle. Total embolic events occurred in our study was 16.1% of our patients, which is comparable with the results of a study performed by Grande.[1]

Cardiac tumors, especially myxomas, have a wide variety of symptoms at presentation as noted in the literature (Table IV) but may be asymptomatic at all and are discovered during routine evaluations (up to 13% in some series).[1,11] In our series of 104 patients involved by myxomas, we discovered 7 (6.7%) asymptomatic patients. The most useful method for the diagnosis of cardiac tumors is echocardiography.[1] In our study, echocardiography was performed for all suspected patients (transthoracic or transesophageal or both) with invaluable data on atrioventricular valve involvement (35%), protruding mass through the valve (8.5%), and presence of a mobile mass in a chamber (11.51%). In addition to these findings, 11 (9.4%) patients had at least moderate to severe pericardial effusion (Table II).Other diagnostic methods of diagnosis (magnetic resonance imaging-MRI- and CT scan) were performed in 7 patients, if indicated. Coronary angiography was carried out in 57 (48.7%) patients, and we found concomitant coronary artery disease in 11 (19.2% of those studied by this method) patients. As a result, we recommend coronary angiography in all patients who represent symptoms of ischemic heart disease or those who are over 45 years old.

MRI may be of considerable value in differentiating the type of tumor.[21] (sarcoma) and the third patient had RV myxoma with embolization of the tumor into the pulmonary vasculature. Two other patients had multiple tumors.

Table IV. Some presenting symptoms of patients with primary cardiac tumors

Syncope +	weight loss
Tamponade	fever
CVA**+	pleural effusions
DOE **+	pericardial effusion+
Fatigue	acute limb ischemia+
Ankle edema	AF* rhythm+
Palpitation+	WPW*
Diaphoresis	PAT*
CHF**	transitional visual loss
TIA**+	sweating
Angina+	erythematous rash
Arthralgia	myalgia
Cerebral aneurysm	asthma
Psychiatric presentation	

*references:1,2,13,14,15,16,17,18,19,20

** CVA: cerebrovascular accident, DOE:: dyspnea on exertion , WPW: wolf Parkinson white , CHF: congestive heart failure, TIA: transient ischemic attack + symptoms which are present in our series

Scince Crafoord first attempt to remove a left atrial myxoma in 1954, [22] surgical excision has become routine. We excised LA myxomas through a biatrial approach during total cardiopulmonary bypass support with bicaval cannulation and removed the full thickness of the septum. We believe that the biatrial approach offers accurate exploration of all the four cardiac chambers and avoids excessive tumor manipulation, reducing the risk of embolization. After resection, repair of the defect was performed by primary closure in 55 patients and by pericardial patch in 46 patients in the cases of extensive resection of septum.

During surgery, we evaluated the atrioventricular valve. Twelve patients had involvement of the valve but in most of them, preservation of the valve was possible. Three cases needed mitral valve replacement and in one case, preservation of the valve was possible with mitral valve repair.

Another important consideration is the prospect of familial myxoma. Often the tumor is diagnosed in a young patient (<20 years) and may be accompanied by a complex of conditions that can include lentigines, nevi and adrenal and pituitary tumors.[1] When the patient is young, it is critical to extend echocardiographic testing to other members of family. In our study, there was one female who had a family history of myxoma in her

mother and brother. She was involved by atrial myxoma at 17 years old (before our study started). Afterwards, she had 2 episodes of recurrence at 33 and 35 years old.

We had 10 recurrences: two patients had two episodes of recurrence and one of them was the case of a familial tumor (noted above).

The earliest recurrence occurred within 9 months of the first operation and the latest one was 16 years after the first operation. In the literature, the longest period that is reported as the time of recurrence is 10 years after the procedure. Therefore, all myxoma patients (especially in the cases of familial type) should undergo annual echocardiography.

The most prevalent symptom at the time of recurrence was dyspnea on exertion. The other symptoms were arrhythmia and cerebrovascular accident. Two patients had no symptoms, and the tumor was discovered during a routine follow-up.

Arrhythmias and other conduction alterations are frequent, either in the immediate postoperative period or later.[2] In our experience, 5 patients developed arrhythmia in the intermediate period. 2 patients developed atrial fibrillation, and 1 patient developed paroxysmal supra ventricular tachycardia (PSVT), which was controlled with medical therapy. One patient developed recurrent episodes of ventricular tachycardia and was treated with ICD (implantable cardioverter defibrillator).

Bateman and colleagues[23] reported that arrhythmias could be caused by trauma to the specialized conduction tissue during myxoma resection and also by atrial distention during the biatrial approach.[24]

The prognosis in the other non-malignant tumors is highly related to their respectability.[7] In our series, all the benign tumors were grossly respectable, except for one patient who had extensive involvement of three chambers in his second relapse.

Our data, in general, support the case for aggressive surgical intervention. A cure can be achieved in patients with benign tumors, and palliation can be extended both to patients with malignant lesions and to those with unresectable or partially resectable benign tumors. Be that as it may, improvement in adjuvant therapy will be necessary in order to increase survival times. In conclusion, the advancement in diagnostic techniques and the routine practice of echocardiography not only have conferred the detection of cardiac tumors but have also improved the prognosis for many patients.

Conflict of Interest

No conflicts of interest have been claimed by the authors.

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