Journal of BUON **15:** 51-55, 2010

ORIGINAL ARTICLE

Cardiac myxoma: clinical presentation, surgical treatment and outcome

L. Velicki, S. Nicin, B. Mihajlovic, P. Kovacevic, S. Susak, M. Fabri

Institute of Cardiovascular Diseases of Vojvodina, Sremska Kamenica - Clinic for Cardiovascular Surgery, Novi Sad, Serbia

Summary

Purpose: *To analyse the clinical characteristics and patterns of cardiac myxomas and to evaluate the clinical course and surgical treatment results.*

Methods: *In this retrospective study we evaluated the clinical presentation, diagnostic methods and surgical treatment outcome in all patients (n=17) with cardiac myxoma treated at the Institute of Cardiovascular Diseases of Vojvodina between the 1.1.2006-31.1.2009.*

Results: *Patients with cardiac myxoma constituted 0.52% of all patients having had heart operations for different reasons. The average patient age was 56.59 years and 12 (70.88%) patients were female and 5 (29.12%) male. Left atrial myxomas were by far more frequent (16 patients, 94.11%) than right atrial myxomas (1 patient, 5.88%). The average time of symptom presence was around 12 months while 4 patients (23.53%) were asymptomatic. The most common symptom was dyspnoea (53%) and fatigue (41%). Total extirpation of tumor mass was achieved in all cases. The tumor size ranged from 1.7×1 cm to 9.5×3.3 cm (average of 2×2.86) and histological examination confirmed the diagnosis of cardiac myxoma in every instance. The 30-day mortality after surgical extirpation of atrial myxomas was 0%.*

Conclusion: *Cardiac myxoma may mimic a huge variety of other cardiac diseases. Echocardiography plays an important role in diagnosing this disease. Because of relatively low risk, surgical treatment is recommended for most patients with excellent postoperative prognosis.*

Key words: cardiac myxoma, cardiac surgery, heart neoplasm

Introduction

Cardiac neoplasms are uncommon and the vast majority of them are benign and can be successfully treated by surgery with excellent postoperative results. Their clinical course may be deceiving mimicking other cardiac diseases thus imposing precise detection and diagnosis [1]. With the advent of new imaging modalities, above all echocardiography, the diagnosis is made easily and promptly with high degree of accuracy.

Cardiac myxomas are by far the most common primary cardiac tumors in adults with an estimated incidence of 0.5 per million population per year [2]. Around 80% of myxomas are localized in the left atrium, of which 75% involve the interatrial septum, 10-20% are found in right atrium while the rest are either biatrial or in right/left ventricle [3]. Cardiac myxomas are most frequently diagnosed in the 5th decade of life, with mean age of 50 years, and 90% of patients are aged 30-60 years [4]. There is a female predominance with a ratio of 2:1.

Cardiac myxoma is regarded as a benign neoplasm but can be lethal because of embolic complications or atrioventricular obstruction [1]. Most myxomas have broad-base endocavitary connection (Figure 1), while a few have a narrow pedicle which is usually attached to the interatrial septum (Figure 2). The consistence of myxomas varies greatly from soft, gelatinous and very friable to firmer or even calcified mass.

With the advent of noninvasive imaging techniques, diagnosing cardiac tumors has never been easier. Transthoracic echocardiography (TTE), computerized tomography (CT) and magnetic resonance imaging (MRI) can accurately determine tumor size, location, attachment and mobility. If there is an involvement of valvular apparatus, careful anatomical as well as function-

Correspondence to: Lazar Velicki, MD, MSc. Pecka 14, 21000 Novi Sad, Serbia. Tel: +381 63 772 33 99, Fax: +381 21 66 22 059, Email: velar@sbb.rs
Received 19-02-2009; Accepted 13-04-2009

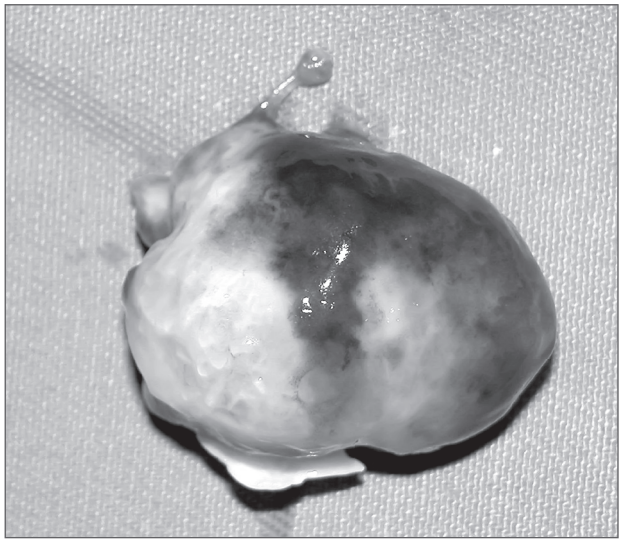

Figure 1. Myxoma (with a wide pedicle) with low embolisation potential extirpated *in toto* from the left atrium.

Figure 2. Myxoma (with a narrow pedicle) with high embolisation potential extirpated *in toto* from the left atrium.

al examination must be performed prior the operative treatment. Preoperative transesophageal echocardiography (TEE) can reconfirm type, location and structure of tumor mass, quantify valvular stenosis or regurgitation and guide cannulation.

The purpose of this study was to analyse the clinical characteristics and patterns of appearance of cardiac myxomas and also to evaluate clinical course and surgical treatment results in a series of patients diagnosed and treated at our department.

Methods

This retrospective study included all patients operated for cardiac myxoma at the Institute of Cardiovascular Diseases of Vojvodina between 1.1.2006 and 31.1.2009.

All patients underwent standard clinical examination, chest X-ray and laboratory exams. Diagnosis of cardiac neoplasm was initially established using TTE and later confirmed by TEE.

The data were collected from the patients' records and included main symptoms, date of surgical treatment, type of surgical treatment, histological verification of tumor type and information on postoperative outcome.

Results

Seventeen patients with cardiac myxoma were operated at our Clinic. These patients represented 0.52% of all patients having had heart operations for different reasons. The average patient age was 56.59 years (range 40-73). There were 12 (70.88%) females and 5 (29.12%) males (female to male ratio 2.4:1). The vast majority of tumors were left atrial myxomas (16 patients, 94.11%); only one (5.88%) was located in the right atrium. The average time of symptom presence was around 12 months (range 4-40) while 4 patients (23.53%) were asymptomatic. The most common symptom was dyspnoea (53%) and fatigue (41%) (Table 1). Total extirpation of tumor mass was achieved in all cases, followed by cauterisation of the tumor area.

Twelve (70.59%) patients had sinus rhythm, and 5 (29.41%) atrial fibrillation (Table 2). The discovery of tumor mass in the heart was achieved by echocardiography (TTE and TEE) in all cases. The average size of the left atrium was slightly enlarged (4.15±0.8 cm). All patients but one had good ejection fraction (Table 3). Only one (5.88%) patient had significant mitral regurgitation (+3), while 16 patients had mitral regurgitation ≤ +2. Ten

Table 1. Clinical findings of patients with cardiac myxoma

Preoperative symptoms	*No. of patients*	*%*
Asymptomatic	4	23.53
Dyspnoea	9	52.94
Palpitation	6	35.29
Systemic emboli	4	23.53
Syncopal episodes	2	11.76
Angina	2	11.76
Fatigue	7	41.18

Table 2. Preoperative electrocardiographic findings in patients with cardiac myxomas

Preoperative rhythm	*No. of patients*	*%*
Sinus rhythm	9	52.94
Sinus tachycardia	2	11.76
Sinus bradycardia	1	5.88
Atrial fibrillation	5	29.41

Table 3. Preoperative echocardiographic findings of ejection fraction

Preoperative EF	*No. of patients*	*%*
Normal (> 50)	16	94.12
Reduced (30-50)	1	5.88
Poor (<30)	-	-

EF: ejection fraction

patients with left atrial myxoma had tumor mass prolapsing into the left ventricle during diastole (Figure 3).

Surgical excision of myxomas was performed under extracorporeal circulation and moderate hypothermia. In all cases of left atrial myxomas, surgical approach was through the right atrium and interatrial septum. Total extirpation of tumor mass together with an adjacent cuff of endocardium was achieved in all cases followed by cauterisation of the tumor area where applicable. Interatrial septum was the most frequent (82.35%) place of origin of atrial myxoma (Table 4). Five (29.41%) patients needed valvular surgery (reconstruction of mitral annulus) and 1 patient with right atrial myxoma needed atrial septum defect closure.

The tumor size ranged from 1.7×1 to 9.5×3.3 cm (average 2×2.86) and histological examination confirmed the diagnosis of cardiac myxoma in every instance. The 30-day mortality after surgical extirpation of atrial myxomas was 0%. The average time spent in hospital after surgical treatment was 8.29 days.

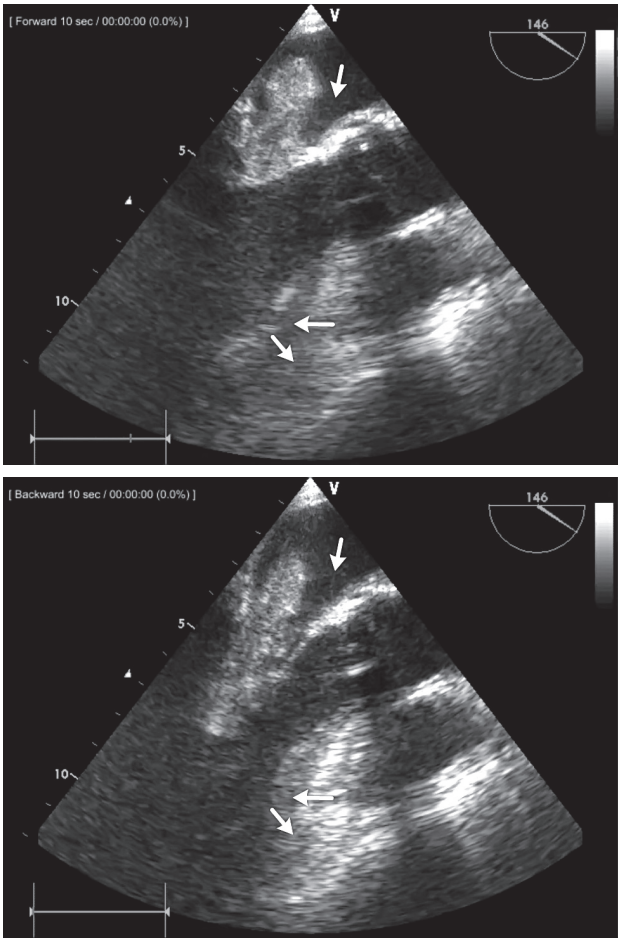

Figure 3. Preoperative transesophageal echocardiography showing left atrial myxoma from the Figure 1 prolapsing into the left ventricle during diastole (arrows).

Table 4. Place of origin of myxoma (intraoperative findings)

Origin of myxoma	*No. of patients*	*%*
Left atrial myxoma		
Interatrial septum	13	76.47
Mitral ring	1	5.88
Posterior atrial wall	1	5.88
Right atrial myxoma		
Interatrial septum	1	5.88

Discussion

Cardiac myxomas are the most frequent primary cardiac tumors in adults (30-50% in most pathologic studies) [5]. Realdo Colombo was the first to describe a case of primary cardiac tumor in 1559 [6]. Advances in imaging technology and the rapid progress in the field of cardiovascular surgery brought a larger awareness of those tumors and facilitated surgical treatment. Crafoord was the first to successfully remove a left atrial myxoma in 1954 using cardiopulmonary bypass [7].

Their embryological origin is currently thought to be entrapped embryonic foregut, and hence they are derived from multipotent mesenchymal cells capable of both neural and epithelial differentiation [8]. Recently, it has been established that cardiomyocyte-specific transcription factors in these tumors favor their derivation from mesenchymal cardiomyocyte progenitor cells [9].

The average patient age in our series was 56.59 years. This is in concordance with other surgical series

of different geographical areas [10,11]. On the other hand, female to male ratio was slightly higher than the average [2.4:1], and this overestimation may be attributed to the relatively small sample size. The 30-day mortality was 0% which could also be attributed to the small sample size. Reported 30-day mortality rates are up to 3-5% [12]. Higher mortality rate is attributed to concomitant diseases of the patients, prolonged cardiopulmonary bypass (CPB) time (usually due to combined valvular and revascularization surgery**)** and poor preoperative ejection fraction (EF).

Benign tumors of the heart grow relatively slowly, which enables them to be undiscovered for a long period of time till the moment they do start causing some significant clinical manifestations. They can have many clinical presentations mimicking a broad range of cardiac and non cardiac diseases. Even though myxomas are considered benign tumors, their location is what that makes them "malignant". The clinical presentation is mainly consisted of improper hemodynamic status related to blood flow obstruction, rhythmic disturbances, embolic potential and systemic (constitutional) symptoms. Intracardiac location of these masses are related to obstruction of blood flow whether they are located in the atria or ventricles. Even the atrial myxoma can, sometimes, obstruct the pulmonary or systemic venous drainage and interfere with valve function. Because of their enormous size, they consume most of the cavity room leading to decreased cavitary blood volume thus afflicting the normal hemodynamic status of the heart. These patients can have an increased risk of acute cardiogenic shock or sudden cardiac death [13]. If there is invasion of the underlying tissue, rhythmic disturbances can occur. Cardiac myxomas are known for their high embolisation potential. They can produce recurrent strokes, peripheral or pulmonary embolisation. These events are reported in about 30-40% of the patients [14]. There is a difference between embolisation potential regarding the site of myxoma. Mitral valve myxomas are more likely to embolise due to motion of mitral leaflets and a higher pressure in the left ventricle. It has been noticed that polypoid tumors give rise to embolisation more frequently than solid ones. Systemic symptoms are common and include long-term fever, weight loss, joint pains, anaemia, high erythrocyte sedimentation rate, leucocytosis etc.

The Carney complex includes LAMB (lentigines, atrial myxoma, mucocutaneous myxoma and blue naevi) and NAME (naevi, atrial myxoma, myxoid neurofibromas and ephelides) syndromes. It has been noticed that, in some cases, there is a familial clustering of the above mentioned findings associated with multiple and multifocal myxomas [15]. Carney complex is inherited in an autosomal dominant fashion with variable penetration, and 7% of all cardiac myxomas are considered to occur within this complex.

TTE has been considered the procedure of choice for diagnosing and assessing atrial myxoma. On the other hand, TEE can provide high-quality images of the heart structures, allowing a clear presentation of structures that are hard to visualize by TTE [16]. For these reasons, TTE may be considered as initial diagnostic tool, and once the neoplasm is discovered the patient should undergo further TEE examination to precisely delineate the relation between tumor and the adjacent structures. For more complex tumors or those with greater extent, the combination of echocardiography, CT and MRI generally provides excellent anatomical definition for preoperative planning. CT and MRI may reveal important features of primary cardiac tumors such as infiltrating growth and extracardiac extent, criteria that can help distinguish benign from malignant lesions. Coronary angiography is done in patients older than 40 years of age, those having any risk factors for coronary artery disease, or if there is potential involvement of coronary arteries [15].

Once a myxoma is diagnosed surgery should proceed urgently. Such an aggressive approach is justified by the constant threat of valvular obstruction or pulmonary or systemic embolisation, a likelihood which is extremely difficult to predict. Myxomas can be successfully excised using hypothermic CPB. Intraoperative manipulation of the tumor should be kept to a minimum during preparation for CPB and before clamping the aorta [17].

Myxoma recurrence has been reported to occur at different rates in surgically treated patients (0.4- 14%) [18]. Sadeghi et al., adding together the results of 16 series of cases of atrial myxoma totaling 194 patients, reported a 7% recurrence rate [19]. Nevertheless, because sporadic recurrence is not clearly predictable, continuous postoperative follow-up of all patients by noninvasive methods is mandatory. Annual follow-up including echocardiography is preferred [20].

Conclusion

Cardiac tumors, although rare, represent a significant clinical problem. Unrecognized heart tumors may imitate many other heart diseases. If not timely diagnosed, cardiac neoplasms may lead to the development of heart failure, embolisation, and sudden cardiac death. Surgical extirpation of tumors is a safe and effective way of treatment.

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