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CLINICAL STUDY

Surgery for cardiac myxomas: 12-year experience

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ABSTRACT

INTRODUCTION: Cardiac myxoma is a rare benign cardiac tumor observed in approximately 0.5–1 case per 1 million people per year. The aim of this study is to review our 12-year experience in the surgical treatment of cardiac myxoma with an emphasis on the clinical, pathologic, diagnostic, and surgical features.

METHODS AND PATIENTS: From January 2010 to December 2022, 90 patients (0.67 %) with cardiac myxomas were surgically treated in our institute. Patients´ demographics, cardiac and surgical medical history, surgical procedures, and pre- and post-operative data were analyzed. The median follow-up time was 76 (1–216) months.

RESULTS: The mean age of the patients was 59.4 ± 13.5 years, with a higher prevalence of women. The most common preoperative symptoms were arterial embolism and dyspnea, and 35.6 % of patients were asymptomatic. Only 8.9 % of the patients had systemic and constitutional manifestations. The most common location of cardiac myxoma was in the left atrium, followed by the right atrium. Recurrent myxoma developed in 3 patients (2.7 %), and the mean time of recurrence was 55 ± 19.7. Hospital and long-term mortality were 2.2 % and 15.6 %, respectively.

CONCLUSION: Cardiac myxoma is the most common heart tumor with a low incidence. Surgical excision yields very good short and long-term outcomes with low recurrence rate after surgery, and remains the treatment of choice *(Tab. 4, Fig. 2, Ref. 13)*. Text in PDF *www.elis.sk*

KEY WORDS: cardiac myxoma, cardiac tumor, recurrence, survival.

Introduction

Cardiac myxoma is a rare benign cardiac tumor observed in approximately 0.5–1 case per 1 million people per year (1). The condition may develop in all age groups and is most often found in patients in age range of 30–69 years, more frequently in women than in men (2). Most of the myxomas, up to 80 %, are localized in the left atrium, while 7–20 % are found in the right atrium, and the rest of them are either biatrial, or localized in the right or left ventricle (3).

In this retrospective study, we review our 12-year experience in surgical treatment of cardiac myxoma with an emphasis on the clinical, pathologic, diagnostic and surgical features.

Patients and methods

From January 2010 to December 2022, of the total of 13,402 on-pump open-heart procedures, 90 patients with cardiac myxomas (0.67%) were surgically treated in our institute. Medical records and the registry of cardiac procedures *(https://www.nczisk.sk*) were reviewed for patients´ demographics, cardiac and surgical medical history, surgical procedures, and pre- and post-operative data. Mortality data were obtained upon request from The Healthcare Surveillance Authority registry *(https://www.udzs-sk.sk/en).*

The preoperative signs and symptoms that were analyzed included systemic symptoms, arterial embolism, chest pain, dyspnea, syncope, and arrhythmia. The diagnosis in all patients was established by transthoracic (TTE) and transesophageal (TEE) echocardiography.

Surgical resection was performed through medial sternotomy in most of the patients, except for 9 patients in whom the procedure was performed through video-assisted thoracoscopy (VATS). Standard cardiopulmonary (peripheral in VATS procedures) bypass with bicaval cannulation, mild hypothermia, and cold-blood cardioplegia was used. The operation was performed with the right atrial approach, and in one patient with a transaortic approach. A complete resection of tumors was performed in all cases, and two patients had a repair of iatrogenic atrial septal defect. All resected masses were sent for gross examination and histological characterization. Histological confirmation was obtained by tissue analysis, whereas tumor anatomic characteristics were assessed by the operating surgeon.

The referring cardiologist examined the patients at annual intervals while the median follow-up time was 76 (1–216) months.

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Statistical analysis was carried out using StatsDirect statistical software version 3.2.10 (http://www.statsdirect.com) and JASP statistical softwarel: JASP Team (2021). JASP (0.14.1) (Computer software). Continuous variables were expressed as mean ± standard deviation or mean value and categorical variables were expressed as percentages. Survival data and freedom from recurrence were analyzed with standard Kaplan-Meier actuarial techniques.

Patients´ informed consent was obtained to present this study.

Results

Mean age and EuroScore II values at the time of surgery were 59.4 ± 13.5 years and 2 ± 2, respectively. Sixty (66.6 %) patients were female, and 30 (33.3 %) patients were male, with a mean NYHA (New York Heart Association) class of 1.5 ± 1.2. The preoperative and demographic characteristics are shown in Table 1.

Tab. 1. Preoperative and demographic characteristics.

Variable n= 90	n (%)
Age, years, mean ± SD	59.4±13.5
Gender, male	30 (33.3%)
Euroscore II, mean ± SD	2±2
NYHA class, mean ± SD	1.5±1.2
Smoking	14 (15.6)
Diabetes mellitus	12 (13.3)
Hypercholesterolemia	33 (36.7)
Chronic renal disease	2 (2.2)
Atrial fibrillation	10 (11.1)
Coronary artery disease	9 (10)

NYHA = Ńew York Heart Association, SD = standard deviation

Tab. 2. Perioperative and postoperative characteristics.

Variable n=90	n (%)
CPB, min, mean ± SD	58.2±48.4
Aortic cross clamp, min, mean ± SD	36.7±30
Bleeding, re-exploration	5 (5.6)
Stroke	1 (1.1)
Wound infection	2 (2.2)
Lung infection	1 (1.1)
Dialysis	0 (0)
Postoperative AF	22 (24,4)
Hospitalization, days, mean ± SD	13±7.3
Hospital mortality	2 (2.2)
All-cause mortality, long-term	14 (15.6)

CPB = cardiopulmonary bypass, AF = atrial fibrillation, SD =s tandard deviation

Tab. 3. Associated procedures.

Procedure	n
Atrial septal defect repair	3
Tricuspid valve repair	10
MAZE procedure	9
Coronary artery bypass graftto LAD (LIMA graft 2, SVG graft 3) to RCA (SVG graft 2)	7
Mitral valve repair	6
Mitral valve replacement (mechanical valve)	1
VA-ECMO implantation	1
Septal myectomy	1
Replacement of the ascending aorta due to IMH	1
Aortic valve replacement (mechanical valve)	1

LAD = left anterior descending artery, LIMA = left internal mammary artery, RCA = right coronary artery, SVG = saphenous vein graft, VA-ECMO = veno-arterial-extracorporeal membrane oxygenation, IMH = intramural hematoma

Tab. 4. Location of cardiac myxoma.

Location	n	%
Left atrium	82	88.2
Septum	68	73.1
Superior wall	3	3.2
Lateral wall	2	2.2
Posterior wall	1	1.1
Right superior pulmonary vein	2	2.2
Left superior pulmonary vein	1	1.1
Mitral valve annulus	5	5.4
Right atrium	8	8.6
Septum	5	5.4
Tricuspid valve (2 septal leaflets, 1 papillary muscle)	3	3.2
Right and left atria	1	1.1
Septum	1	1.1
Left ventricle	1	1.1
Septum	1	1.1
Right ventricle	1	1.1
Apex	1	1.1

The most common preoperative symptoms were arterial emboli, dyspnea, chest pain, syncope, arrhythmia supraventricular tachycardia, and paroxysmal atrial fibrillation which were present in 11 (12.2 %), 19 (21.1 %), 8 (8.9 %), 5 (5.6 %), 7 (7.8 %), 3 (3.3 %), and 4 (4.4 %) patients, respectively.

Systemic symptoms of fatigue and fever were recorded in 6 (6.7 %) and in 2 (2.2 %) patients, respectively while features consistent with Carney´s complex were observed in 1 patient (1.1 %). Finally, 32 patients (35.6 %) were asymptomatic.

The operation was undertaken on an emergent or urgent basis and the associated procedures are detailed in Table 3. The peri-

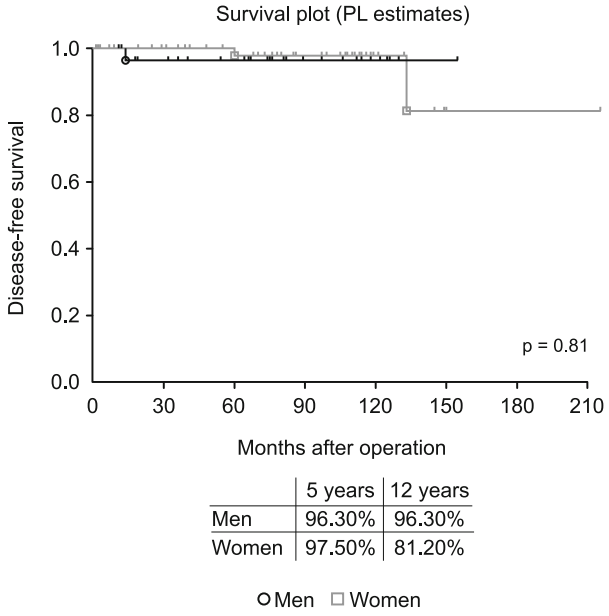
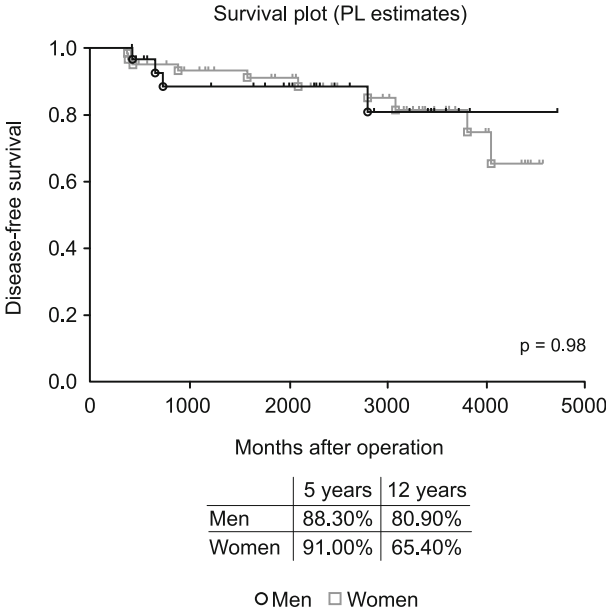


Fig. 1. Kaplan-Meier survival free of recurrence curve.



	5 years	12 years
Men	88.30%	80.90%
Women	91.00%	65.40%

Fig. 2. Kaplan-Meier survival curve.

operative and postoperative characteristics are shown in Table 2. Hospital mortality was 2.2 % (2 patients). The first patient who had re-exploration due to severe postoperative bleeding later developed a sternal wound infection which was treated with vacuum therapy. The second patient was treated postoperatively by implanting a veno-arterial extracorporeal membrane oxygenator (VA-ECMO). As to associated procedures, MAZE and repair of the mitral and tricuspid valves were performed, and the postoperative course was complicated by a posterior wall hematoma.

In this series, the location of myxomas was mainly in the left atrium (88.8 %), followed by that in the right atrium (8.6 %). The patient with Carney complex had biatrial cardiac myxomas at index operation. The locations of cardiac myxomas are shown in Table 4.

Recurrent myxoma developed in 3 patients (2.7 %), and the mean time of recurrence was 55 ± 19.7 months. The patient with the Carney complex had two reoperations for recurrent myxomas. The second patient with recurrent myxoma had a sporadic form of the disease. The third patient had a history of neck and shoulder angiofibromas and resection of an angiofibroma in the scrotum with no history of familial or hereditary form of the disease. The 12-year survival free of myxoma recurrence in men and women were 96.3 % and 81.3 %, respectively (Fig. 1) (p = 0.81).

The all-cause long-term mortality was 15.6 % (14 patients) (Tab. 2). The 12-year survival in men and women were 81.6 % and 65.5 %, respectively (p = 0.98) (Fig. 2).

Discussion

Intracardiac myxoma is the most common tumor of the heart with an estimated incidence of 0.5–1 case per 1 million people per year (1). Among operations with the use of cardiopulmonary bypass, 0.3 % are resections of cardiac myxoma (4). In our study, 0.67 % of the total number of operations done with the use of cardiopulmonary bypass were resections of cardiac myxomas, and this rate is consistent with the data published previously.

Moreover, recent studies of patients with cardiac myxomas showed a higher prevalence of the disease in women (range 53–77.4 %) (5,6). Similarly, in our cohort, the incidence of cardiac myxomas was more prevalent in women (66.6 %).

Cardiac myxoma can be observed in all age groups, and the mean age reported in our study (59.4 years) is comparable with the mean age range of 42 to 66 years reported in previous studies (7, 8). It is rarely found in younger patients and only as hereditary and familial forms of the disease. In our study, cardiac myxoma was diagnosed in two younger patients aged 35 and 22 years with a history of Carnex , and skin and scrotal angiofibromas, respectively.

As reported in the literature (1), the most common location of cardiac myxomas was in the left atrium (72–92 %), followed by the right atrium (0.7–7.5 %), right (0.7–2.5 %) and left (0.7–3.6 %) ventricles, and in heart valve in fewer than 1 % of cases. *Fossa ovalis* in the interatrial septum was the most common site the cardiac myxomas were attached to. In our cohort, the most common location was in the left atrium (88.2 %), with the interatrial septum being the most common site of attachment (73.1 %). One patient with Carney complex had a biatrial cardiac myxoma.

The clinical presentation and the signs observed in patients with cardiac myxoma depend on the location and mobility of the tumor and are divided into 3 groups: hemodynamic (dyspnea, chest pain, arrhythmia, syncope, and sudden death), systemic embolization, and systemic and constitutional symptoms (anemia, fever, weight loss, fatigue, arthralgia, myalgia and Raynaud phenomenon) (1). However approximately 3.2 % to 46.4 % of the patients with cardiac myxoma are asymptomatic. In our cohort, 35.6 % of the patients were asymptomatic, and the hemodynamic manifestation was the most common clinical presentation (as also reported in the literature), and only 8.9% of the patients had systemic and constitutional symptoms (in contrast with a previously reported incidence range of 16.9 % to 32.4 % (1)).

The diagnosis in all patients was established by TTE and TEE. Transthoracic echocardiography and TEE show a 90 % to 96 % accuracy in diagnosing cardiac myxoma. Transthoracic echocardiography is the simplest and most useful examination, but does not provide additional information on the nature, structure, and mobility of cardiac myxoma, On the other hand TEE may accurately assess tumor characteristics (1).

A genetic analysis may provide valuable information on a patient with familial and Carney complex forms of the disease. Familial myxomas and syndrome myxomas (Carney complex) are often multicentric and account for most recurrences. In our study, there was only one patient with the Carney complex who had a biatrial myxoma at initial presentation and two recurrences of the tumor. Familial myxomas and Carney complex are associated with a mutation of the PRKAR1A gene (9, 10). Carney complex is a syndrome consisting of cardiac myxomas, spotty pigmentation (lentiginosis), and endocrine overactivity, while the condition is inherited in an autosomal dominant pattern and presents a variable phenotype (11).

Median sternotomy with the use of cardiopulmonary bypass is the most common approach used during cardiac myxoma resection. In our study, 90 % of the procedures were performed through median sternotomy. Only 10 % were performed through VATS with very good results. Bianchi G et al (7) also reported successful outcomes with VATS.

According to our experience, and in agreement with the literature (12), the approach via right atriotomy is the ideal one. The complete removal of the tumor mass and its attachment site was the basic principle of excision. In cases where tumors were attached with a wide base to the interatrial septum, the affected areas were excised along with the tumors. These iatrogenic defects were closed with a pericardial patch. Moreover, other approaches, such as left atrial or biatrial approaches, are also reported by other authors (13).

According to our experience and in agreement with the data from the literature, the surgery for cardiac myxoma resection is associated with a low rate of postoperative complications and uneventful postoperative period with minor complications, while atrial fibrillation is the most common complication reported (1), including the incidence of 24.4 % in our cohort.

The recurrence rate reported in our study was 2.7 % and was observed in 3 patients, namely in a patient with Carney complex, in a patient with a history of neurofibroma tumors (non-hereditary non-familial form) and in a patient with a sporadic form of the disease. Recurrence is commonly observed in patients with familial cardiac myxoma or Carney complex form, and very rarely in patients with a sporadic form (1).

The hospital mortality of 2.2 % observed in our study is within the range of mortality of 0 % to 10 % reported in previous publications (1).

The long-term survival reported in the literature is high and ranges between 84 % and 96 % (1). Similar results with a long-term survival of 84.4 % are also reported in our study.

Due to the retrospective characteristics of the study, several limitations can play a role, especially the long-term follow-up. Despite its limitations, this study provides an insight into the natural history of cardiac myxomas and shows a good long-term outcome after surgical resection.

In conclusion, cardiac myxoma is the most common tumor among heart tumors and has a low incidence. Surgical excision produces very good short and long-term outcomes and remains the treatment of choice. The recurrence rate is low and may be observed months or years after surgery, mainly in patients with Carney complex and familial forms, and rarely in those with a sporadic form.

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