Turk Gogus Kalp Dama 2014;22(3):526-533
doi: 10.5606/tgkdc.dergisi.2014.9700

Original Article / Özgün Makale

Cardiac myxomas: a 27-year surgical experience

Kardiyak miksomalar: 27 yıllık cerrahi deneyim

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Background: In this study, we reported the outcomes of surgical resection of cardiac myxomas performed in our center.

Methods: A retrospective review of 99 patients (67 females, 32 males, mean age 49.8±16 years; range 5 to 76 years) who were operated for myxoma between January 1985 and December 2012 was conducted. Preoperative diagnosis was established by transthoracic echocardiography. The mean time to the last follow-up visit was 123.4±85.7 months.

Results: All patients were operated through median sternotomy. The site of origin of the tumor was left atrium in 92 patients and the most common implantion site was interatrial septum in 85 patients. A complete resection of the tumor was performed in all patients except one. Forty five patients (45.5%) were diagnosed with a papillary myxoma, while 54 patients (54.5%) were diagnosed with a solid myxoma. The risk of embolic events in papillary type of myxoma was significantly higher than the solid type (44.4% vs. 9.3%, p<0.01). Early mortality was 5%. Univariant analysis showed advanced age (p=0.04), preoperative presence of atrial fibrillation (p=0.02), peripheral embolism (p=0.03), and need for postoperative inotropic support (p=0.008) as the risk factors for early mortality. Actuarial survival rate was 98%, 94% and 89% at five, 10 and 20 years, respectively. The rate of recurrence was 1.2% in survivors.

Conclusion: Surgical treatment of cardiac myxoma carries a low operative risk and is associated with excellent short and long-term results. However, despite low risk of recurrence in the long-term, the patients should be followed with annual echocardiography.

Keywords: Cardiac tumor; myxoma; surgery; survival.

Amaç: Bu çalışmada merkezimizde yapılan cerrahi kardiyak miksoma rezeksiyonunun sonuçları bildirildi.

Çalışma planı: Ocak 1985 - Aralık 2012 tarihleri arasında miksoma tanısıyla ameliyat edilen 99 hasta (67 kadın, 32 erkek, ort. yaş 49.8±16 yıl; dağılım 5-76 yıl) retrospektif olarak incelendi. Ameliyat öncesi tanı transtorasik ekokardiyografi ile konuldu. Son takip vizitine kadar geçen ortalama süre 123.4±85.7 ay idi.

Bulgular: Tüm hastalar median sternotomi ile ameliyat edildi. Tümörün kökeni 92 hastada sol atriyum olup, en sık yerleşim yeri 85 hastada interatriyal septumdu. Bir hasta hariç tüm hastalarda total rezeksiyon yapıldı. Kırk beş hastaya (%45.5) papiller miksoma, 54 hastaya (%54.5) solid miksoma tanısı konuldu. Embolik olay riski papiller tip miksomalarda, solid tipe kıyasla, anlamlı oranda yüksek bulundu (%44.4'e kıyasla %9.3, p<0.01). Erken mortalite %5 idi. Tek değişkenli analiz sonuçlarına göre ileri yaş (p=0.04), ameliyat öncesi atriyal fibrilasyon varlığı (p=0.02), periferik emboli (p=0.03) ve ameliyat sonrası inotropik destek ihtiyacı (p=0.008) erken mortalite için risk faktörleri olarak belirlendi. Beş, 10 ve 20 yıllık aktüeryal sağkalım oranları sırası ile %98, %94 ve %89 idi. Sağkalanlarda nüks oranı %1.2 olarak tespit edildi.

Sonuç: Kardiyak miksoma cerrahisi, düşük ameliyat riski taşımakla birlikte, mükemmel kısa ve uzun dönem sonuçlar ile ilişkilendirilir. Ancak, uzun dönemde düşük nüks riskine rağmen, hastalar yıllık ekokardiyografi ile takip edilmelidir.

Anahtar sözcükler: Kardiyak tümör; miksoma; cerrahi; sağkalım.

Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2014.9700
QR (Quick Response) Code

Received: December 20, 2013 *Accepted:* February 15, 2014

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Primary cardiac tumors are rare entities with an incidence rate of between 0.0017 and 0.03% in autopsy series when both the benign and malignant histological types are included.[1,2] Approximately 80% of these tumors are benign, and of these, more than half are myxomas.[2]

Myxomas occur in all age groups but are more likely to present between the third and sixth decades of life.[3] Although benign, the tumor has the risk of systemic embolization with subsequent cerebral or peripheral infarctions, intracardiac obstructions, syncope, and sudden death.[4]

In this study, we reviewed our 27 years of experience in the surgical treatment of a consecutive series of 99 patients who were operated on for cardiac myxomas.

PATIENTS AND METHODS

Between January 1985 and December 2012, a total of 112 consecutive patients were operated on for cardiac tumors at our institution, and this study focus on the 99 (88.3%) (67 females, 32 males, mean age 49.8±16 years; range 5 to 76 years) who had myxomas.

The hospital records were reviewed to obtain information regarding the patients' demographics, medical histories, surgical procedures, and pre- and postoperative data. The demographics, preoperative clinical status, and age distribution are summarized in Tables 1 and 2. The majority of the patients were female (67.6%), and all but three were adults with a mean age of 49.8±16 years (range 5 to 76 years). The most common symptoms at admission were dyspnea (74.7%) and palpitation (45.5%). Twenty-four patients (24.2%) also showed symptoms of systemic embolization either to the peripheral or central nervous systems. Futhermore, thromboembolic episodes in the central nervous system presented with stroke in 16 patients, transient ischemic attacks in three others, and epilepsy attacks in one patient.

Table 1. Age distribution of the 99 patients with intracardiac myxomas

Age (years)	n	%
1-10	1	1
11-20	5	5
21-30	8	8.1
31-40	11	11.1
41-50	16	16.2
51-60	29	29.3
61-70	22	22.2
71-80	7	7.1
Total	99	100

Coronary angiography was performed on 64 patients (64.6%) who were over 50 years old or those who had a history of chest pain. In addition, six patients (6.1%) required concomittant coronary artery bypass grafting (CABG) due to coronary artery disease (CAD).

A preoperative diagnosis was established in all of the patients via transthoracic echocardiography (TTE) until 2005 (Figure 1), with transesophageal

Table 2. Clinical profiles of the patients

Variables	n	%
Sex		
Male	32	32.3
Female	67	67,7
Rhythm		
Normal sinus rhythm	83	83.8
Atrial fibrillation	16	16.2
Location		
Left atrial septum	79	79
Left atrial wall	6	6
Inferior pulmonary vein	2	2
Mitral valve		
Anterior	4	4
Posterior	2	2
Right atrial septum	3	3
Right atrial eustachian valve	1	1
Right atrial coronary sinus	1	1
Tricuspid valve	1	1
Right ventricle	1	1
Symptoms		
New York Heart Association		
Class I	11	11.1
Class II	51	51.5
Class III	31	31.3
Class IV	6	6.1
Dyspnea	74	74.7
Palpitations	45	45.5
Angina	13	13.1
Embolism		
Central nervous system	18	18.1
Peripheral	6	6.06
Central nervous system + peripheral	1	0.01
Constitutional		
Fever	22	22.2
Weight loss	6	6.1
Fatigue	15	15.1
Comorbidities		
Coronary artery disease	10	10.1
Aortic stenosis	2	2
Mitral insufficiency	11	11.1
Tricuspid insufficiency	10	10.1
Tricuspid stenosis	1	1

echocardiography (TEE) being performed only when the diagnosis was not clear on TTE. After 2005, TEE was routinely performed preoperatively to inspect all four cardiac chambers. In some of these patients, myxomas were demonstrated by real-time three-dimensional (3D) TEE (Figure 2).

All of the patients were followed up on an outpatient basis at regular intervals, and TTE was performed routinely prior to discharge and subsequently every year afterwards. Four of the patients in this study were lost to follow-up after being discharged.

Statistical analysis

The statistical analysis was performed using the SPSS version 15.0 for Windows (SPSS Inc., Chicago, IL, USA) software program, and the data was expressed as mean ± standard deviation (SD) for continuous variables and as numbers with percentages for categorical variables. Furthermore, a univariant analysis was used to assess the categorical variables as well as the predictors for early and late mortality. The survival analysis was performed using the Kaplan-Meier method, and a p value of <0.05 was considered to be statistically significant.

RESULTS

All patients underwent surgical removal via a median sternotomy soon after the diagnosis of atrial myxoma to avoid embolic complications or sudden death, and aortic arterial, bicaval venous cannulation, and intermittent antegrade cardioplegia were utilized under moderate hypothermia. The mean cardiopulmonary bypass (CPB) time was 69.8±32.1 minutes, and the mean ischemic time was 43.1±25.8 minutes.

Furthermore, care was taken not to manipulate the heart before the aorta was cross-clamped in order to avoid embolization. The tumors were then excised along with a wide margin of tissue surrounding the area of attachment.

The tumor was located in the left atrium in 92 patients (93%), the right atrium in six others (6%), and the right ventricle in an additional patient (1%), and the most common implantation site was the interatrial septum (IAS) for the tumors originating in the left atrium (85%) (Table 2). Only one patient had multifocal tumors in the left atrium, and another had a Carney complex which was previously described by Akbaş et al.[5] Moreover, this patient had surgery for tetralogy of Fallot (TOF) when she was three years old.

The surgical approach was biatrial in 32 patients (32.3%) and uniatrial in the other 67 (67.7%). Furthermore, a left atriotomy was performed on 48 patients (48.5%) and 19 others (19.2%) underwent a right atriotomy, 12 of whom had left atrial myxomas that were accessed via the IAS. In addition, there were six tumors located in the right atrium and one in the right ventricle.

The mass was resected entirely in all of the patients except for one who had multifocal myxomas in the left atria. One of them, which arose from the left inferior pulmonary vein, was pedinculated, and it was resected successfully. However, the posterior wall component was sessile and had invaded almost all of the branches of the pulmonary veins; thus, the mass was only partially resected in this patient. In 55 cases, the tumors were excised along with a cuff of full thickness at the atrial wall. In 11 of these patients, the defect was repaired with a pericardial or Dacron patch (C.R. Bard Inc., Murray Hill, NJ, USA), We also copiously

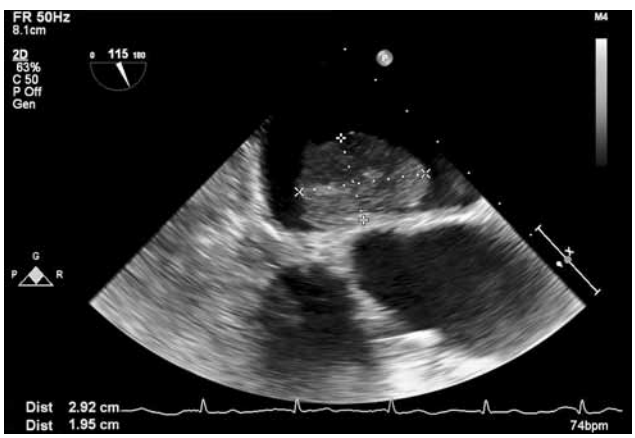

Figure 1. Echocardiographic image of myxoma.

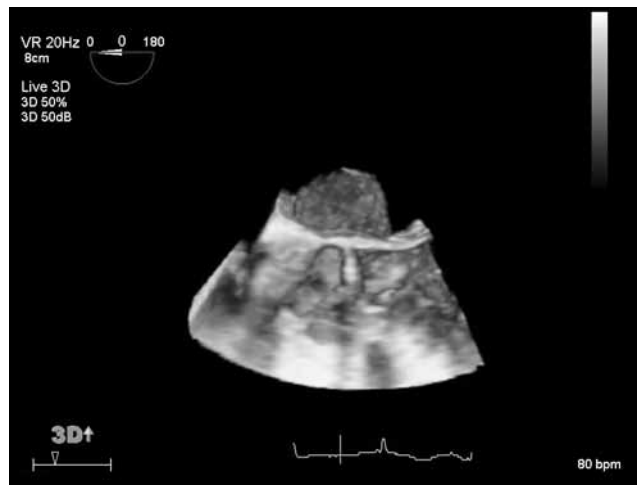

Figure 2. RT-3D transesophageal echocardiography image of myxoma.

irrigated the atria and ventricles with a cold saline solution after the mass resection was performed to avert tumor seeding and intraoperative embolization.

Associated procedures consisted of six CABGs, one radiofrequency ablation, two aortic valve replacements, three mitral valve replacements, nine mitral valve reconstructions, five tricuspid valve repairs (three De Vega annuloplasties, one bicuspidization and one septal leaflet repair with a pericardial patch after excising the pedunculated tumor).

All of the resected tumors were sent for a histological examination, and the myxoma diagnosis was confirmed by the presence of disseminated fibrin deposits, the proliferation of capilleries, and blood extravasation. The intraoperatively collected samples were fixed with formalin and routinely processed for light microscopy. Staining was done with hematoxylene and eosin (H-E). The mean tumor dimension at the largest diameter was 5.4±2 cm (range 0.5-9.8 cm). Tumors with a smooth, regular border and a tough consistency were classified as solid. These were stiff and could usually be removed in one piece. Papillary myxomas were characterized by an irregular and gelatinous exterior with a friable, soft consistency. Moreover, they often had a cauliflower appearance which necessitated piecemeal removal. In the current series, papillary myxomas were diagnosed in 45 patients (45.5%) while solid myxomas were detected in 54 others (54.5%). Papillary tumors tended to present with neurological symptoms and more frequent emboli, and the risk of an embolic event with this type of myxoma was significantly higher than for the solid type (44.4% vs 9.3%; $p<0.01$).

Early mortality was seen in five patients (5%), and two patients underwent emergency surgery. One of them was operated on for pulmonary edema and acute heart failure and died from adult respiratory distress syndrome (ARDS) postoperatively. This patient also had chronic renal disease. The other underwent early postoperative surgery due to a femoral embolism when she was an inpatient and died from multiorgan failure even after receiving high-dose inotropic support and inserting an intraaortic balloon pump (IABP). This patient probably also had mesenteric and renal embolization before surgery. In addition, two patients died of progressive acute renal failure that necessitated dialysis, and one died of low cardiac output syndrome (LCOS). Table 3 summarizes the characteristics of the non-survivors. A univariant analysis indicated that older age (63.8±12.07 in non-survivors vs. 49.1±15.8 in survivors; $p=0.04$), the preoperative presence of atrial fibrillation (AF) ($p=0.02$) and peripheral embolisms ($p=0.03$), and the need for the use of inotropes postoperatively ($p=0.008$) were risk factors for early mortality. However, the histological type of tumor ($p=1.0$), gender ($p=0.5$), presence of cerebral embolisms ($p=0.5$), and other preoperative symptoms such as angina ($p=0.1$), dyspnea ($p=0.3$) or palpitation ($p=0.2$), and concomitant surgical procedures ($p=0.1$) had no impact on early mortality.

Table 3. Postoperative mortalities

Variables	Case 1	Case 2	Case 3	Case 4	Case 5
Age (years)	65	74	69	68	43
Sex	Male	Female	Female	Female	Female
Preoperative rhythm	AF	AF	NSR	AF	NSR
Ejection fraction (%)	40	65	65	45	60
New York Heart Association class	IV	II	III	IV	III
Embolism	–	+	–	–	–
Emergency	+	+	–	–	–
Concomitant disease	CRF	–	CRF, MI 3	–	TI 3
Coronary artery disease	–	–	–	(LMC +)	–
Associated procedure	–	Femoral embolectomy	Mitral reconstruction	CABG	Tric reconstruction
Cross clamp time/total perfusion time	50/75	45/70	73/93	87/126	65/80
Inotropic agent	+	+	+	+	+
Intraaortic balloon pump	–	+	–	+	–
Cause of death	ARDS, renal failure	MOF	Renal failure	LCOS	Acut renal failure

AF: Atrial fibrillation; NSR: Normal sinus rhythm; CRF: Chronic renal failure; MI: Mitral insufficiency; TI: Tricuspid insufficiency; LMC: Left main coronary; CABG: Coronary artery bypass grafting; ARDS: Acute respiratory distress syndrome; MOF: Multiorgan failure; LCOS: Low cardiac outflow syndrome.

Twenty-eight patients (28%) had early complications following surgery, including transient AF in 10 (10%), temporary nodal rhythm in nine (9%), and atrioventricular block in four (4%) patients, two of whom (2.02%) underwent the implantation of a permanent pacemaker. In addition, one of these patients had concomitant tricuspid and mitral valve reconstruction and the other had a septal mass resected via the transseptal approach. Furthermore, one patient required reexploration due to postoperative bleeding, one experienced transient postoperative acute renal failure, and two others suffered from pneumonia and mediastinal wound infections, but these patients experienced a full recovery.

Complete follow-up information was available for 90 of the operative survivors, but four patients were lost to follow-up. Of the survivors, six (6.6%) died at a mean follow-up time of 123.4±85.7 months (median 116 months; range 3-322 months). Four of those died from noncardiac causes (i.e., gastrointestinal bleeding, lung cancer, hemorrhagic stroke, and pneumonia), and one patient, who underwent CABG along with myxoma resection, died from myocardial infarction. The cause of death of the other patient was intracardiac flow obstruction with an acute collapse of atrioventricular communication. This patient had a multilocular tumor in the left atrium, and was the one mentioned previously with the pedunculated left inferior pulmonary vein component that was resected successfully who had the sessile posterior wall component that had invaded nearly all of pulmonary vein branches. Since we were only able to partially resect this mass, it is likely that the residual tumor had interfered with the pulmonary venous return to the left atrium thereby leading to the congestive heart failure. The patient underwent emergency surgery but died in the operating room. The only risk factor for late mortality in our study was older age at the time of operation (62.4±13.3 vs. 48.8±15.8; p=0.02).

Upon follow-up, all of the surviving patients were in functional New York Heart Association (NYHA) class I or II. Two patients were reoperated on for mitral valve replacement following myxoma resection at the sixth and 12th postoperative years respectively. One was a patient whose tumor, as noted in the original surgery, was attached to the anterior mitral valve leaflet and showed moderate mitral valve insufficiency on early postoperative echocardiography. The mitral regurgitation had progressed to severe sufficiency after six years. The other patient underwent mitral valve replacement along with myxoma resection at the age of five and developed a mismatch at the mitral mechanical valve. Both patients were classified as NYHA class I on follow-up. No further thromboembolic events or strokes were observed in any of the other patients.

The actuarial survival rates were 98%, 94% and 89% at five, 10 and 20 years respectively (Figure 3), and the rate of recurrence was 1.2% for these patients. One patient showed a recurrence at 81 months after the initial surgery and had undergone a direct closure of the residual defect after the resection and suffered a recurrence of the myxoma on the same site of the original tumor. The recurrent tumor was diagnosed during a routine echocardiographic follow-up at another center. Afterwards, we confirmed the re-occurrence by TEE, and a myxoma measuring 0.8x0.5 cm was identified. The patient chose not to undergo a reoperation and has been followed up via echocardiography every six months since that time.

DISCUSSION

Myxomas are the most common benign intracavitary neoplasms. Their etiology is not fully understood, but they are thought to originate from primitive stromal cells that have the capacity to differentiate along endothelial lines. Studies with neuroendocrine markers using immunohistochemical techniques have stressed another origin for myxomas, and identification of these

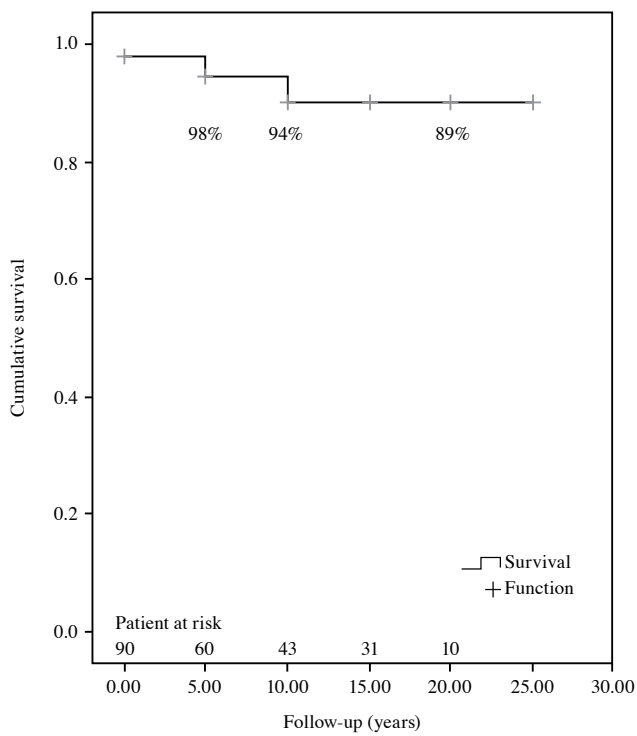

Figure 3. Kaplan-Meier cumulative survival table.

markers suggests the endocardial sensory nerve tissue as the origin.[5] Clinical presentation usually occurs in adulthood, most often between the fourth and sixth decades, and there is a female gender prevalence. In this series, 67 patients (67.7%) were female with a mean age of 49.8 years at the time of surgery.

Cardiac myxomas usually develop in the atria, with approximately 75% originating in the left atrium (93% in our series), 15-20% in the right atrium (6% in our series), and only 3-4% in the ventricles (1% in our series).[2] Most myxomas arise from the IAS at the border of the fossa ovalis, but Yuan.[6] reported that the free atrial or ventricular wall and valve leaflets (7% in our series) are also possible sites of origin.

Although myxomas are benign tumors, they can cause severe physiological disruptions. The structure of the tumor, localization in the heart, and diameter of the mass determine the symptoms and outcome. Most patients present with one or more of the triad of intracardiac obstruction, cerebral or peripheral embolisms, and constitutional symptoms such as fatigue, weight loss, fever, myalgia, arthralgia, and anemia.

In our patients, dyspnea and palpitation were the predominant symptoms. These were caused by intracardiac obstruction and were present in 74.7% and 45.5% of the patients respectively. Moreover, AF was noted preoperatively in 16 patients (16%), and Swartz et al.[7] reported similar findings in approximately 14% of their patients. The arrhythmias may be due in part to increased tumor bulk, which secondarily enlarges the left atrial diameter and alters conduction. However, the presence of the myxoma itself or tumor growth may decrease the left atrial volume, causing the hemodynamic alteration of blood flow from the atria to the ventricle, which can lead to rhythm disturbances.

Embolization can be the initial symptom of cardiac myxomas, and in our series, systemic embolization was present in 25 patients (25.3%). Furthermore, 76% of the recognized emboli in our study involved the central nervous system, and the incidence of systemic embolization from left atrial myxomas ranged from 25-50% according to a study by Goodwin.[8] Most embolic events related to cardiac myxomas involve the cerebral, lower extremity, visceral, and coronary arteries. In addition, a total occlusion of the abdominal aorta may rarely be seen.[9] Moreover, right-sided myxomas can uncommonly develop a pulmonary embolism, and although this is extremely rare in left-sided myxomas, it should still be kept in mind that in patients with atrial septal defect (ASD) or ventricular septal defect (VSD), the mass can protrude into the right side through these abnormalities.[10] The histopathological type of tumor seems to play an important role in the formation of embolic events. Papillary tumors generally have a friable or gelatinous texture and may give rise to emboli by the erosion and fragmentation of the neoplasm (cellular, stromal, or both) or by the erosion of the thrombus covering the tumor surface. In two studies comparing papillary and solid tumors, embolization was typically associated with the papillary variety, with the rate in favor of this type of tumor being 54% to 16% in one series and 58% to 0% in the other.[11,12] Emboli from solid myxomas, although not common, may result from the cracking of the surface as it is subjected to turbulent flow. In our series, the embolic events were primarily associated with the papillary type of myxoma (44.4% vs. 9.3%). On the other hand, the presence of AF has been reportedly related to the increased risk of embolization with no importance attached to the histological type of myxoma.[13] In our population, embolization occurred in half of the patients with AF. Moreover, AF and peripheral embolisms were associated with early mortality. Hence, it is crucial to properly diagnose the patient and choose the appropriate surgical treatment before complications related to embolization develop. In addition, it is prudent to emphasize that patients with papillary myxomas and those with preoperative AF should undergo surgery immediately after diagnosis because of the high risk of embolization.

The location of the tumor may influence the patient's clinical situation. A septal tumor location has a strong association with congestive heart failure while an extraseptal location is correlated with embolic events.[7] However, only four patients out of 25 who presented with embolisms demonstrated extraseptal tumor localization in our study.

Two-dimensional (2D) echocardiography is a safe and accurate method for diagnosing cardiac tumors because it is noninvasive and does not pose the risk of tumor embolization. Furthermore, 2D echocardiography can quantify the tumor size, shape, attachment, and mobility and can screen other cardiac chambers accurately for additional tumors. We obtained excellent results with TTE, but in the last six years, we have routinely used TEE before performing these operations. Moreover, TEE is particularly helpful for evaluating the posterior left atrial wall, atrial septum, and right atrium, which often are not well displayed on TTE, to exclude the possibility of biatrial multiple tumors.[14] In some cases, we used 3D TEE since it has the ability to visualize desired structures in multiple planes and evaluate tissue characteristics. However, in 2D TEE, the attachment site can be located, but

complete delineation and measurement of the area is not possible.[15] Recently at our center, we have increasingly been using 3D TEE in general clinical practice at our center.

Coronary embolization is very rare, but it should be kept in mind, and cardiac catheterization should be carried out on all patients with angina pectoris, regardless of their age and the presence or absence of cardiac risk factors. However, none of the patients in this study had coronary embolization, and atherosclerotic CAD was diagnosed in only four patients.

The aim of the surgery must be the complete resection of the tumor as soon as possible after the diagnosis is established because of the high risk of sudden death from a thromboembolism or valvular obstruction. Moreover, special care must be taken to avoid intraoperative embolization of the myxoma. Therefore, the surgical approach should allow for only minimal manipulation of the tumor, provide adequate exposure for a complete resection, and allow for the inspection of all four heart chambers in order to minimize the chance of recurrence.[16] The surgical access to the myxoma may vary depending on the tumor location. For a left atrial myxoma resection, there are generally three different approaches: left atrial (48.5% in our series), transeptal (12% in our series), and biatrial (32.3% in our series). In our clinic, the transseptal approach via the right atrium is rarely used, but some investigators advocate this method because it facilitates wide excision (by giving direct access to the fossa ovalis where most tumors attach) without mandating direct handling. It also affords adequate mitral exposure and allows for the visualization of cardiac chambers for concomitant tumors.[17] In cases when the transseptal approach via the right atrium does not provide adequate exposure, the superior transseptal approach (Guiraudon technique) can easily be performed. Furthermore, although the biatrial approach affords superb exposure, it has been criticized as being responsible for a high incidence of arrhythmias and conduction disturbances after the resection of left atrial myxomas.[18] In our experience, the incidence of rhythm disturbances were encountered in 31% of the patients who underwent a biatriotomy, whereas this rate was 21% when an isolated left atriotomy was performed.

Cardiac myxomas may recur postoperatively. Possible explanations for this include incomplete resections, intraoperative displacement, embolization, and multifocal genesis. Garatti et al.[11] recommended the use of a wide excision and patch repair, and they had a recurrence rate of 1% for this procedure, but Centofanti et al.[19] found that a simple excision was satisfactory, and they observed no recurrence in their patients during the follow-up period. In addition, according to McCarthy et al.,[20] the risk of tumor recurrence is much higher with the familial variety of myxomas (10%). The rate climbs even hire with the patients have the Carney complex syndrome (21%) or when multiple myxomas are present (33%). We performed a wide excision and patch repair on only 11 patients while 55 patients had the mass excised with a cuff of full thickness at the atrial wall. Furthermore, the tumors were resected without an associated endocardial part in 44 others. Our recurrence rate at our center is quite low with these surgical techniques (1.2%), which is comparable with the literature.[11,19] Our experience has led us to believe that total extirpation of the tumor with a cuff of surrounding normal tissue and copious irrigation of the cardiac chambers may help prevent recurrence. Postoperative echocardiography is also recommended annually for detecting recurrent myxomas.

Conclusion

Myxomas are the most frequent type of cardiac tumors and may present with a wide range of symptoms ranging from being completely asymptomatic to life-threatening catastrophic consequences. After a diagnosis has been established, prompt surgical excision must be performed. Excision of cardiac myxomas carries a low operative risk and gives excellent short- and long-term results. Annual follow-up visits that include echocardiographic surveillance are recommended since there is a risk of recurrence, especially in familial cases.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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