

# SURGICAL TREATMENT OF HEART MYXOMAS

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## HIRURŠKI TRETMAN MIKSOMA SRCA

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### ABSTRACT

During a period from 1961 until May 2006, there were 50 patients - 32 female and 18 male - with myxoma of the heart who were treated at the Clinic of Cardiac Surgery of the Military Medical Academy in Belgrade. In 43 patients myxoma was situated within the left atrium, in 5 patients within the right atrium, and in one case within the left and the right ventricle each. The classic triad of myxoma clinical presentation is AV valvular obstruction in over 80% of patients, embolization as well as systemic and constitutional symptoms. Diagnostics has been performed mainly by noninvasive methods, mostly echocardiography. All patients were operated using extracorporeal circulation and complete removal of tumor was done, followed by pathohistological verification. Valvular damage was solved by either implantation of the artificial valve or reconstruction of the natural one. Operative mortality rate was 6%. There was no recurrence.

**Key words:** heart neoplasms, myxoma, cardiac surgical procedures

### INTRODUCTION

Primary heart tumors are rare. The incidence of primary heart neoplasms is between 0.17–0.19% (1, 4). Seventy-five percent of primary heart neoplasms are benign, and 25% malignant (2, 3). Approximately 50% of benign heart tumors in adults are myxomas (2, 5). Although according to histological criteria benign, their localization and possible complications can lead to „malignant“ behavior.

In 1559 Realdo Colombo was the first who described heart neoplasm (6, 7). In 1809 Alden Allen Burns from Edinburgh described valvular obstruction caused by atrial tumor (8). In 1845 King reported serial of six atrial tumors, nowadays recognized as myxomas (9). The first successful surgical removal of myxoma using cardiopulmonary by-pass was performed by Crafoord in Sweden in 1954 (10). Kay was the first who removed myxoma of the left ventricle in Los Angeles in 1959 (11).

### PATIENTS AND METHODS

At the Clinic of Cardiac Surgery at the Military Medical Academy in Belgrade during a period from 1961 until May 2006 there were 50 patients with heart myxomas who had been diagnosed and operated.

In 64% of cases patients were female (32), and in 36% male (18). They were all between 15–66 years, average age 46.7 years.

Classic triad of clinical presentation of myxoma consists of intracardial obstruction with possible congestive heart failure, signs of embolism and systemic – immune-constitutional symptoms: fever, body mass loss, fatigue, myalgia and arthralgia. Associated anomalies were atrial septal defect - ostium secundum type, together with mitral and tricuspidal valve disease.

### SAŽETAK

Tokom perioda od 1961. godine do maja 2006. godine bilo je 50 bolesnika, 32 žene i 18 muškaraca, sa miksomom srca koji su lečeni na Klinici za kardiohirurgiju Vojnomedicinske akademije u Beogradu. Kod 43 bolesnika miksom je bio lokalizovan u levoj pretkomori, kod 5 u desnoj pretkomori a kod jednog bolesnika u svakoj od obe komore. Od klasične trijade kliničkog ispoljavanja miksoma AV valvularna stenoza je bila u preko 80% bolesnika, a takođe i embolizacija te sistemski i konstitucionalni simptomi. Dijagnoza je učinjena pretežno putem noninvasivnih metoda, većinom ultrasonografijom. Svi bolesnici su operisani u ekstrakorporalnoj cirkulaciji kada je rađeno kompletno uklanjanje tumora, sa potonjom patohistološkom verifikacijom. Valvularno oštećenje je rešavano implantacijom veštačke valvule ili nativnog zaliska. Operativni mortalitet je bio 6%. Nije bilo recidiva.

**KLjučne reči:** tumori srca, miksom, kardiohirurške procedure

The diagnosis of myxoma itself at the same time represents the indication for surgical procedure. Surgical resection is the only efficient therapeutic option for patients with heart myxoma and it should not be delayed because of possible consequences due to obstruction or embolization.

All operations were performed using extracorporeal circulation and medial sternotomy approach, with cannulation of ascendent aorta and both cava veins, in moderate hypothermia with local heart cooling. Manipulation with the heart before the cardiopulmonary by-pass establishment has been extremely delicate because of fragility and embolic tendency of myxoma.

In case of myxoma of the left atrium, venous cannula is usually placed through the right atrial wall and the right appendage, while in a case of myxoma of the right atrium - it is usually placed through the superior vena cava. Myxoma in the oval fossa region used to be removed via suitable atriatomic approach along with excision of its attachment at septum, which is commonly sutured directly, exceptionally by means of pericardial patch.

Excision of the attachment or the base of myxoma localized close to atrioventricular valves has to be performed much more delicately, avoiding valvular and conduction system damage.

Ventricular myxoma is to be approached through tricuspidal valve, and myxoma of the left ventricular outflows through the aortic valve. After myxoma removal, in case of valvular damage annuloplasty or implantation of the artificial valve is to be done.

All removed myxomas were pathohistologically analyzed and diagnosis was verified.

## RESULTS

Eighty-six percent of myxomas were found at the left atrium, 10% at the right atrium, 2% at the left ventricle and 2% at the right ventricle. There were neither multiple, nor biatrial tumors.

In majority of our patients (over 80%) cardiac symptoms prevailed, predominantly with symptoms of malfunction of AV valve. Embolism appeared in 5 (10%) patients. Three youngest patients (6%) were asymptomatic. Functionally, the patients belonged into NYHA I – NYHA IV categories.

Routine diagnostic methods combined with echocardiography and heart catheterization during last three decades were used in myxoma diagnostics (table 1).

**Table 1.** Diagnostic methods.

DIAGNOSTIC METHOD	LA	RA	LV	RV
invasive diagnostic only	4	2	0	0
invasive diagnostic + echocardiography	13	1	1	1
echocardiography only	23	1	0	0
other - explorative atriotomy	3	1	0	0

The operations performed on our patients are shown in table 2.

**Table 2.** Type of operation.

OPERATION	LA	RA	LV	RV
excision of myxoma	21	3	3	
excision + valve reconstruction	6	1		1
excision + valve implantation	16			
excision + suture of the asd		1		

The emergency operation was performed in three patients; while in the rest 47 patients the operation was an elective procedure. Forty seven of fifty operated patients went home without complications. Three patients (6%) died. During the last 26 years mortality rate was zero. There were no relapses.

## DISCUSSION

Myxomas could be localized within all heart cavities, but they show special predilection for the left atrium in 75% of cases (1-4, 12, 13). Atrial myxomas generally arise from the interatrial septum at the border of the fossa ovalis, but they can originate from any other location within the atrium, including appendage (4). In 10–20% cases they can be found into the right atrium (2, 12, 13), and the rest of the percentage is equally divided between both ventricles (2, 12), which is in accordance to our results. Myxomas of the right atrium are more likely to have broad-based attachments than the left atrial tumors, they are also more likely to be calcified (5), and thus visible on chest radiographs. Ventricular myxomas occur more often in women (2) and children, which is also proved by our series. Localization within ventricles is rather rare (2, 4, 12-14). Of all our 50 patients, there was just one with myxoma of the right ventricle and one with myxoma of outflow tract of the left ventricle, which were both successfully removed (14).

Myxomas occur with peak incidence between the third and sixth decade of life and tend to be more common in women (4, 13-15), as was also our experience.

About two thirds of myxomas are oval or round, polypoid, pedunculated, mobile, tumors with smooth or slightly lobulated surface, yellowish-green to brownish color, frequently covered with thrombus (2, 4, 13, 15). Mobility depends on the length of the stalk and the width of attachment to the heart, as well as the amount of collagen in the tumor (4). Less common villous or papillary myxomas are gelatinous and fragile and prone to fragmentation and embolization, occurring about one third of the time (15). The average size of myxoma is about 5 cm (4), but much larger myxomas have been also reported. Their weights range is from 50–60 g (16), they could be even bigger. In our series the largest one of 152 g was the myxoma of the left atrium (13). Most myxomas appear to grow rapidly, but growth rates vary and occasionally tumor growth arrests spontaneously (4). There are reports of slowly growing myxomas in surgically high-risk patients during 79 months with growth rate of 0.2 cm per year (17).

About 5% of myxoma patients show familial pattern of tumor development; approximately 20% of them have „complex myxomas“ or Carney’s complex: associated conditions such as adrenocortical nodule hyperplasia, Sertoli cell tumors of the testicle, pituitary tumors, multiple myxoid breastfibroadenoma, cutaneous myxomas, and facial and labial spots (18). In spite of careful investigation, we have never determined any of these conditions in our patients.

Histologically, myxomas are composed of polygonal cells and capillary channels within an acid mucopolysaccharide matrix (4). The base of the myxoma contains a large artery and veins that connect them with subendocardium and in most cases they tend to grow into the overlying cardiac cavity rather than into surrounding myocardium (18). All our removed myxomas have been histologically verified, and dilemma arose only in one case.

The classical triad of myxoma clinical presentation is intracardiac obstruction with congestive heart failure (67%), embolization (29%), systemic or constitutional symptoms of fever (19%) and weight loss and fatigue (17%) (13-15, 19, 20). The nature of these symptoms depends on size, localization and way of tumor attachment.

Majority of the patients (> 80% in our series) have dominant symptoms of obstruction of the mitral and tricuspidal valve with signs of mitral stenosis, right heart failure all the way to pulmonary edema, which appeared in three of our patients. Large ventricular myxomas may mimic ventricular outflow obstruction. The left ventricular myxoma is able to produce the same effect as subaortic or aortic stenosis (14), whereas right ventricular myxomas can simulate right ventricular outflow track, pulmonary valve obstruction or pulmonary thromboembolism (12).

Embolization due to myxoma fragility represents the second most common mode of presentation in 30–40%

patients (2, 4, 5). In our series this percentage was even lower – 10%, and it includes three cerebral, one femoral and one coronary embolism. We have never noticed pulmonary thromboembolism as a consequence of right-sided myxomas (4, 12). Coronary embolism with acute myocardial infarction with left atrial myxoma is rarely described (21), and we have faced it in one case.

The majority of patients with heart myxomas, including ours, meticulous investigation reveals constitutional symptoms: weight loss, fatigue and fever, followed by laboratory disturbances such as leukocytosis, elevated sedimentation rate, hemolytic anemia, elevated C-reactive protein with disturbed immunoglobulin levels with increased circulating IgG (22). The recent discovery of elevated levels of interleukin-6 in patients with myxoma has been linked to variety of associated constitutional symptoms (23).

Infection arising in the myxoma is rare complication presenting with picture of infective endocarditis (19).

The diagnosis of heart myxoma is, together with classic clinical examination, positively established with echocardiography, where the sensitivity of 2-D mode is 100%, thus replacing angiocardiography (24, 25). Transesophageal echocardiography (TEE) provides the best information concerning size, location, mobility and attachment and it can detect even the smallest myxomas of 1–3 mm in diameter (26). In patients older than 40 years planned for a surgery, coronarography is necessary to exclude occlusive coronary disease. Magnetic resonance of the heart is still not a routine diagnostic mean, unless in cases of ventricular located myxomas (12), myxomas of the right atrium extending into superior vena cava, as well as in unclear cases.

In a case of completely diagnosed myxoma, there is an absolute indication for surgical treatment: half-urgent because of possible embolization complications or even urgent because of possible development of pulmonary embolism (13), what had happened three times in our series during last 45 years (27).

Surgical resection is the only efficient therapeutic option for patients with heart myxoma. Medial sternotomy is common approach with ascending aortic and bicaval cannulation and extracorporeal circulation is usually employed. Manipulation of the heart before aortic cross-clamping and initiation of cardiopulmonary by-pass is minimized in deference to the known friability and embolic tendency

of myxomas, especially in cases of right atrial myxomas, where both vena cava may be cannulated directly, as we used to do. Atrial myxomas should be approached atrially and biatrially - transeptally (28), what is also our approach of choice. Ventricular myxomas should be approached through the AV valve. We found a rare localized myxoma of the left ventricular outflow that had simulated aortic stenosis, thus it had been removed approaching through aortic valve (14). Regardless of surgical approach, an ideal resection encompasses myxoma as well as a portion of the ventricular wall or interatrial septum to which it is attached.

There is still a dilemma whether the excision of full thickness wall is necessary or the excision of only an endocardial attachment is sufficient to prevent recurrence of myxoma. Our policy is to resect full thickness whenever possible. However, only partial thickness resection of the area of tumor attachment has been performed when anatomically necessary without a noted increase in recurrence rate (29). Some authors favored excision of the full thickness of the wall at the attachment (25). We usually excise radically the part of septum with base of tumor, and subsequent defect is reconstructed by direct suture or by means of pericardial or synthetic „patch“ (13).

Hospital mortality rate was about 5% (15). Excision of ventricular myxomas has operative risk up to 10%, but the experience is still insufficient. In some larger series mortality of about 8% was reported (25). Mortality rate in our group of 50 patients was 6%.

Atrial-ventricular valve damages arising due to mobility and friction of myxoma through the valve nowadays are solved by anuloplasty and valvular reconstruction; years ago by implantation of artificial valves.

The most common complication during surgery is heart rate disorder, up to development of complete A-V block, but this has never appeared in our series.

Recurrence was rare; approximately 1–4%, and could be explained by incomplete resection or – extremely rare – by unrecognized multicentric myxoma growth (4, 29). We have had no relapses.

Heart myxomas, benign tumors which could be cause of serious complications due to their clinical features and strategic position, are rare and relatively simple diagnosed by echosonography. The only effective treatment is surgical removal, that is connected with low operative mortality.

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