Research Article

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Surgical Treatment of Heart Myxomas: the Analysis of 10 Years' Experience

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INTRODUCTION Primary cardiac tumors are rare, and the autopsy detection rate does not exceed 0.003%. Approximately 80% of primary heart tumors are benign, 50% of them are myxomas. Diagnosis of cardiac tumors at present in the presence of transesophageal echocardiography is not difficult in most cases. Surgical treatment of myxomas became possible after the introduction of open-heart surgery under cardiopulmonary bypass into clinical practice. After verification of the tumor, its surgical removal is indicated. In the practice of large cardiac surgery clinics, the experience of radical treatment of heart neoplasms is relatively small; therefore, the analysis of tactical approaches and results of operations in this pathology is of absolute interest.

AIM OF STUDY To present an analysis of 10 years' experience in surgical treatment of cardiac myxomas.

MATERIAL AND METHODS The results of treatment of 58 patients with myxomas who underwent surgical removal of the neoplasm of the heart are presented.

RESULTS Hospital mortality after surgical removal of myxoma was 1.7%, 1 patient died. All patients, we have the long-term period data about, noted a significant improvement in well-being, a decrease or disappearance of dyspnea at rest and/or on exertion, and no recurrence of myxoma was noted.

CONCLUSION The data obtained indicate, on the whole, good immediate results and high quality of life of patients in the long-term period operated on for heart myxomas.

Keywords: myxoma, neoplasm of the heart, extracorporeal circulation, surgical treatment of a heart tumor, pedunculated mass

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BP, blood pressure
CCN, craniocerebral nerves
CHD, coronary heart disease
CPB, cardiopulmonary bypass/artificial blood circulation
CT, computed tomography
CVA, cerebrovascular accident
DUSG, Doppler ultrasonography
ECG, electrocardiography
EchoCG, echocardiography
HR, heart rate
IAS, interatrial septum
ITLN, intra-thoracic lymph nodes
LA, left atrium
LV, left ventricle
MRI, magnetic resonance imaging
MSCT, multispiral computed tomography
MV, mitral valve
RA, right atrium
RV, right ventricle

Primary heart tumors are rare phenomena, the rates of their detection during autopsies does not exceed 0.003% (0.00071–0.0029%) [1]. Approximately 80% of primary heart tumors are benign, 50% of them are myxomas [2]. About 80% of intracardiac myxomas are located in the left atrium (LA), from 7 to 20% are located in the right atrium (RA), and only no more than 10% are found in the left ventricle (LV) or right ventricle (RV) [3-6]. Currently, thanks to transesophageal echocardiography (EchoCG) availability, the diagnosis of heart tumors is not difficult in most cases [7-10].

After the implementation of open-heart surgery using cardiopulmonary bypass (CBP) into clinical practice, the surgical treatment for myxoma became possible. After verification of the tumor, its surgical removal is indicated. Approximately 8% of patients do not survive to surgery, dying from embolism with the fragments of a brain tumor or from atrioventricular valve obturation [11]. The experience of radical treatment for heart neoplasms in the practice of large cardiac surgery clinics is relatively small [12-14], so the analysis of tactical approaches and the results of operations in this pathology are of apparent interest.

The purpose of this report is to present an analysis of the 10-year experience in the surgical treatment for cardiac myxoma.

MATERIAL AND METHODS

For the period from January 2009 to May 2019, surgical interventions for cardiac myxomas were made in 58 patients in the Department of Urgent Cardiac Surgery, Circulatory Support and Heart Transplantation (headed by Professor V. V. Sokolov), of N.V.Sklifosovsky Research Institute for Emergency Medicine, including 14 men and 44 women. The mean age of the patients was 60.7±12.0 years old (from 26 to 82 years). In 27 patients (46.5%), the diagnosis of a heart neoplasm was revealed during a routine examination in absent clinical manifestations.

In 13 patients (22.4%), the clinical presentation of a voluminous mass in the LA cavity was manifested by the signs of circulation insufficiency (shortness of breath, suffocation, one patient was admitted with orthopnea). Also, 13 patients (22.4%) had complaints of recurrent dizziness and short-term episodes of the loss of consciousness. One patient was admitted unconscious with general cerebral neurological symptoms due to initially suspected exogenous intoxication. Two patients (3.4%) had embolisms in the greater circulation: a female patient had suffered an acute ischemic cerebrovascular accident (CVI) 3 months before the detection of a voluminous mass in the LA cavity; the second patient was admitted with terminal aorta embolism for which an urgent embolectomy from the terminal aorta was performed, and later, the cause of embolism was identified being a voluminous neoplasm in the LA cavity. Two other patients (3.4%) sought

medical advice for a new-onset paroxysm of atrial fibrillation; during the examination, the diagnosis of a voluminous neoplasm in the LA cavity was made. In a 76-year-old female patient (1.7%), myxoma in LA was combined with severe aortic stenosis, complaints were dominated by shortness of breath and pain behind the sternum on minimal physical exertion.

In the vast majority of patients, the diagnosis was based on echocardiography findings (transthoracic or transesophageal echoCG) (Fig. 1). In one case, magnetic resonance imaging (MRI) was performed to verify the location of the tumor, and in one case, computed tomography (CT) was performed (Fig. 2).

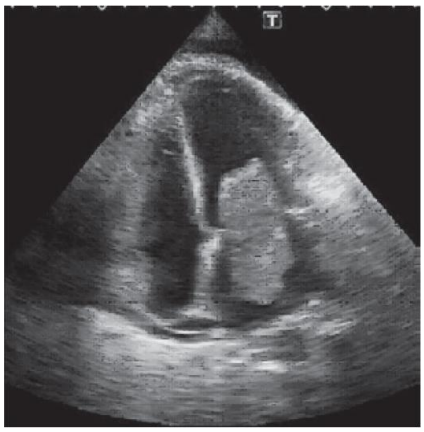

Fig. 1. Echocardiographic signs of a large mass neoplasm of the left atrium, prolapsing into the cavity of the left ventricle and obstructing the mitral valve

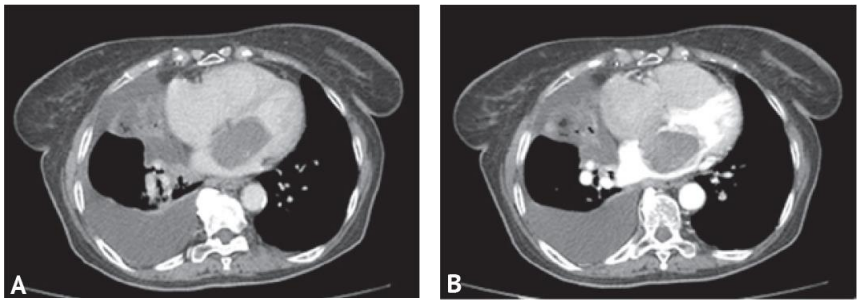

Fig. 2. Computed tomography (A — without contrast; B — with contrast) of the chest. A large myxoma of the left atrium obstructs the left atrioventricular opening; there is a right-sided hydrothorax

On admission, the patients with heart neoplasms were considered as patients in emergency condition. Eight patients (13.8%) with frequent syncopal states were admitted to the intensive care unit and they underwent surgery on the next day after admission. The mean period from the patient admission till surgery was 2.8±1.7 days (from 1 to 7 days).

In all 58 patients, myxomas were located in LA; in 52 patients (89.7%), the tumor originated from the interatrial septum (IAS), usually from the area of the oval fossa; and in 54 (93.1%) it had a clearly defined pedicle.

The technique of surgery included removing the tumor itself and excising its pedicle within the margins of intact tissues. The volume of excised heart tissue depended on the nature and extent of the process.

Access to the tumor located in the LA cavity was obtained either through the anterior wall of the LA behind the atrial sulcus, or through the IAS. In recent years, IAS access has been considered the method of choice; and then it was used in 37 patients (63.8%). Access through the LA was used to remove the LA neoplasm in 21 patients (36.2%).

Operations were performed under conditions of CPB and spontaneous hypothermia (33-35°C). The duration of CPB in patients with isolated removal of the neoplasm from the LA cavity was mean 41.7±9.1 minutes (from 26 to 68 minutes), the mean time of aortic compression was 21.5±13.2 minutes (from 12 to 41 minutes). In 6 patients they refused to mobilize the aorta for clamping due to the adhesive process in the pericardial cavity and the need to reduce surgical trauma, and so the operation was performed on a beating heart with normothermia.

In 24 cases, the myxoma removal was combined with other cardiac interventions. Three patients underwent a mitral valve replacement for the pedicle attachment area was located in the immediate vicinity

of the A3 segment of the MV anterior leaflet. Myxoma removal was combined with an aortic valve replacement for a degenerative calcified defect in 2 cases, with making the anastomosis between the internal thoracic artery and the anterior interventricular branch of the left coronary artery and with aortocoronary bypass grafting of the right coronary artery for ischemic heart disease in 6 cases. In 6 patients, after the left ventricle myxoma removal due to severe annulectasia and tricuspid valve insufficiency, the latter was repaired using De Vega technique. Finally, IAS plastic surgery with a xenopericardial patch after the excision of the broad base of the neoplasm was required in 7 patients. The mean duration of CPB in this group of 24 patients was 72.8±17.4 minutes (from 50 to 106 minutes), the mean time of aortic compression was 50.1±16.3 minutes (from 28 to 76 minutes).

The removed neoplasms were evaluated macroscopically. The sizes varied from 3 x 2.5 x 1.5 cm to 10 x 7 x 5 cm. In 45 cases (77.6%), the tumor had a jelly-like consistency (Fig. 3), which, due to possible fragmentation, could lead to embolism in various arterial pools; in the remaining 13 cases (22.4%), the neoplasm had a dense consistency with a clearly defined capsule (Fig. 4) and even a calcification in one patient. Tumors of this type were more common in patients with the clinical evidence of obturation of the left atrioventricular orifice.

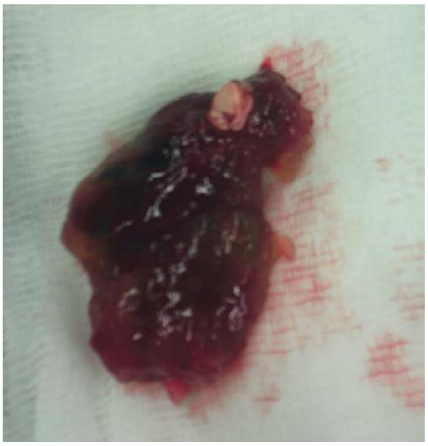

Fig. 3. Removed myxoma (jelly-like consistency)

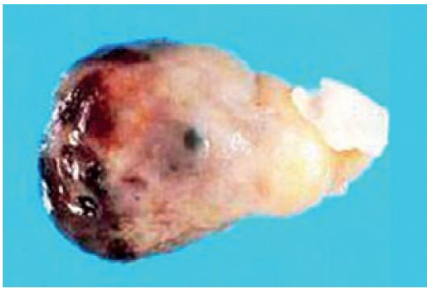

Fig. 4. Removed myxoma (dense consistency)

The diagnosis of myxoma of the heart in all patients was confirmed by morphological studies of the removed tumor (Fig. 5, 6).

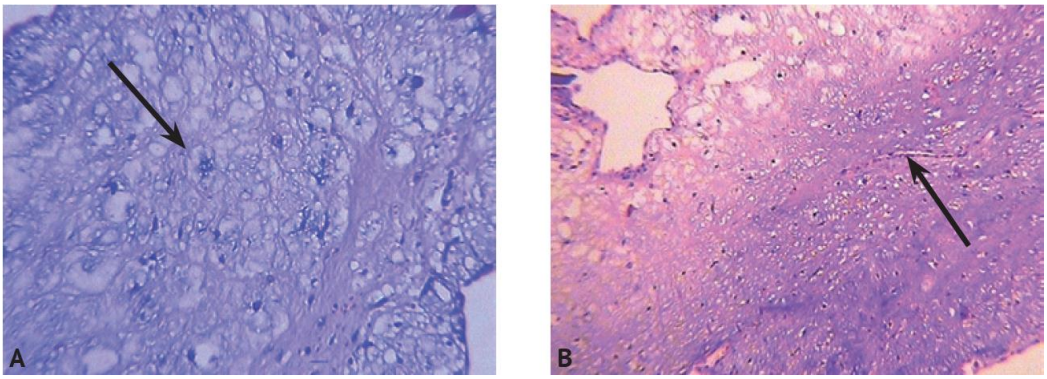

Fig. 5. Morphological features of myxoma of the heart. A — the main morphological feature — star-shaped (arrows) and elongated cells in the myxomatous stroma with reticulin and collagen fibers. Periodic acid-Schiff (PAS) stain, magnification x400; B — fine-grained stromal matrix, delicate collagen fibers, thin-walled vessels (arrows) and numerous mucus-forming cells. Hematoxylin and eosin staining, x100 magnification

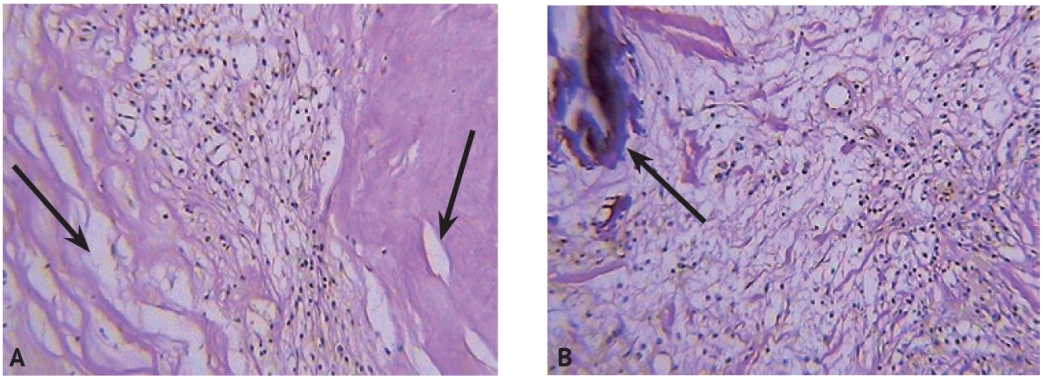

Fig. 6. Morphological features of fibromyxoma (false myxomas according to Virchow). Van Gieson stain. Magnification x200. A — predominance of collagen fibers in the capsule and tumor tissue (arrows) and fibroblasts in the edematous matrix; B — focal calcification (arrows), multiple collagen fibers in the myxomatous stroma

RESULTS AND DISCUSSION

Of the 58 operated patients, one died during the in-hospital period, and the overall hospital mortality rate was 1.7%.

A multiple organ failure syndrome was the cause of death of a 76-year-old patient with LA myxoma, calcified aortic valve defect and significant concomitant pathology (cerebrovascular disease, chronic brain ischemia, post-CVA condition, moderate type 2 diabetes mellitus at decompensation stage, chronic obstructive pulmonary disease), who underwent a combined operation to replace the aortic valve and remove myxoma from LA.

All patients for whom the long-term follow-up data were available had a significant health improvement, the relief or disappearance of the shortness of breath at rest and/or during exercise. In 6 patients, the initial atrial fibrillation persisted; and in 8 others, the episodes of sinus rhythm impairments were reported. Signs of circulation insufficiency existing before surgery disappeared or significantly decreased in the long-term period. Nine patients had to take diuretics periodically, and 7 took cardiac glycosides, which was due to a decreased LV contractility to some extent in the presence of coronary heart disease.

Myxoma is the most common benign intracavitary heart tumor. Harken B. G. et al. [15] described a "75" rule for myxomas. According to this rule, 75% of all heart neoplasms are myxomas, 75% of all myxomas are localized in the LA, 75% of myxomas originate from IAS, and 75% of these tumors have a pedicle at the attachment site.

Data from other authors confirm Harken's "rule" or give figures close to it. Most of the myxomas (75%) are localized in the LA [16-19], of which 90% are attached to the IAS near the oval fossa [20, 21]; myxomas in the RA are 3-4 times less common than in the LA [16, 18-20], myxoma of the heart ventricles account for 2.5–4% [19, 20].

In our case series, 100% of myxomas were localized in the LA; in 87.7% of patients, myxoma growth came from the IAS; and in 92.7% of cases, there was a clearly defined pedicle.

In the presence of myxoma in the left heart, especially if the tumor is mobile and has a jelly-like consistency, there is a risk of embolism of various arterial pools with the tumor fragments, which may result in myocardial infarction, impaired cerebral circulation, kidney and spleen infarction, acute ischemia of the upper or lower extremities. Mobile large size myxomas, especially of dense consistency, can cause a short-term or prolonged obturation of the left atrioventricular orifice, which can be manifested by chronic heart failure, syncopal states, and even sudden death [16, 20, 22].

Myxoma of the heart is considered a benign tumor. The main morphological feature of such a tumor is star-shaped "spider-like" or elongated cells with cytoplasmic processes (Fig. 5A). They accumulate mucus and express non-specific markers: lysocyl, alpha-1-antichymotrypsin and alpha-1-antitrypsin, often located around thin-walled vessels or in the myxoid matrix containing mucopolysaccharides. The myxoid stroma consists of delicate reticulin and collagen fibers with fibrin deposits, capillary-type vessels (Fig. 5CB).

Histogenesis of myxomas has been associated with mesenchymal tissue, which determines their frequent location in the area of the atrial septum.

Myxomas must be differentiated from soft-tissue tumors (fibroids, lipomas, etc.) that have undergone mucosal degeneration — the so-called false myxomas by Virchow. Such tumors are characterized by the presence of a capsule, the predominance of the original tissue structures, a higher density, a lower severity of myxomatosis, and the presence of focal calcification (Fig. 6). Hyperchromia and mitoses sometimes appear

in the cell nuclei. Such soft-tissue tumors with myxomatous degeneration acquire the properties of mesenchyme and sometimes have locally destructive (invasive) growth.

Due to the fact that myxoma is more often localized in the LA and can cause the development of life-threatening complications, the essence of the ideology of managing patients diagnosed with neoplasms in the LA cavity is to treat them as patients with urgent condition, which implies emergency hospitalization, shortening the preoperative examination time and preparation for surgical treatment. In our study, the mean time from hospital admission to surgical treatment was 3.1 days, but in recent years we have been increasingly striving to reduce the period of hospital stay before surgery to 1-2 days. Meanwhile, if there is no suspicion of pathology caused by embolic syndrome, the examination mandatorily contains only laboratory tests, ECG, echocardiography, chest X-ray and coronary angiography in patients older than 50 years or with an ischemic history.

The most common complications are the heart failure, syncopal conditions, or sudden death due to an atrioventricular valve obstruction. Myxoma is also a source of embolism of the cerebral vessels and peripheral vasculature. Clinically, this is manifested by stroke in young patients. Such embolic symptoms are not as frequent as minor embolic manifestations, such as temporary visual disturbances and loss of consciousness [23].

Alterations in the leaflets of the heart valves in myxomas are less common than in other neoplasms of the heart, and may be associated with mechanical trauma to the tumor, especially in its calcification. In some cases, chord separations occur, which is caused by their mechanical tension when the tumor wedges into the atrioventricular orifice [24]. Mitral valve replacement was performed in 3 of our patients.

According to various authors, currently, with modern surgical techniques, the hospital mortality rates after removal of the heart myxoma make 2.3-5.0% depending on the volume and complexity of surgery operation. The survival rate of operated patients in the long-term period is satisfactory, and most patients experience a significant improvement in their health status or a complete recovery [13, 25-27].

Relapses of myxoma occur in 4-7% of patients [28]. According to F. Gerbode et al., R.C. Read [29, 30], relapses of myxoma are extremely rare and occur, as a rule, in cases where the tumor attachment site is not resected. The use of radical surgical techniques helps to avoid this complication [31]. Meanwhile, G. M. Soloviev and L. V. Popov [13] believe that the resection of the tumor attachment site is not necessary; it is sufficient to remove the endocardium at this site. Whenever possible, we tried to remove the base of the tumor, but technically this was not always possible, however, no myxoma recurrences were noted in the analyzed group of patients.

Thus, in general, the obtained data have shown good immediate results and a high quality of life in the long-term among the patients who were operated on for myxoma of the heart.

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