

Case report

Giant myxoma of the right sections of the heart obstructing the inferior vena cava: A case report

Khamidulla A. Abdumajidov^{1*}, Khairulla Zh. Buranov²

¹Republican Specialized Scientific and Practical Medical Center of Surgery named after academician V.Vakhidov, Tashkent, Uzbekistan

²Bukhara State Medical Institute named after Abu Ali Ibn Sino, Bukhara, Uzbekistan

Abstract

Objective: We present a rare case of surgical management of giant myxoma of the right heart with obstruction of the orifice of the inferior vena cava (IVC), obstruction of the right atrioventricular orifice, with the development of high pulmonary hypertension, and severe heart failure.

Case presentation: A 39 years old male patients applied to the consultative polyclinic with typical complaints of severe shortness of breath, palpitations, severe weakness, almost bed rest, severe swelling of the lower extremities, with a transition to the anterior abdominal wall (edema). On physical examination signs of severe heart failure NYHA class IV were revealed. Imaging demonstrated giant right atrial myxoma obstructing IVC with base originated in fossa ovalis, obstructing tricuspid valve and protruding in the right ventricle occupying 50% of its cavity. The surgical removal of the giant RA myxoma protruding the right ventricle and obstructing IVC and tricuspid valve was performed under cardiopulmonary bypass and hemodynamic support. The patient was discharged on the 10th day after surgery and only residual minimal tricuspid regurgitation was revealed on follow-up examination.

Conclusion: The described case from our practice proves the possibility of a severe course of giant myxomas of the right heart, with the development of deep hemodynamic complications of systemic and pulmonary circulation. The chosen correct tactics of surgical treatment provided a favorable result, despite the severity of the initial condition and the degree of hemodynamic disturbance. Intraoperative tactics and technique for performing the stages of urgent surgery were chosen based on the situation, the features of the anatomical structure of the tumor, and the severe complications that arose. Our case proves the possibility of adequate implementation of such interventions, when choosing the right tactics of surgical treatment.

Take home message: If clinical signs of circulatory failure, heart failure are detected, it is necessary to perform echocardiography. If mass formations of the heart chambers are detected, immediately refer the patient to specialized cardiac surgery clinics to resolve the issue of surgical correction of pathology.

Key words: myxoma, right atrium, inferior vena cava obstruction, tricuspid valve orifice obstruction, heart failure, cardiac surgery procedures

(Heart Vessels Transplant 2023; 7: doi: 10.24969/hvt.2023.405)

Introduction

The modern development of cardiovascular surgery makes it possible to perform urgent (sometimes for vital indications) interventions in a severe contingent of patients with heart tumors (1-5). As we know, neoplasms of the heart are a rare pathology, only 0.0017-0.02% of cases in the structure of cardiac pathology, and myxomas account for more than 50% of the number of benign heart tumors.

Petrovsky et al. emphasized the difficulties of diagnosis and the complexity of performing surgical interventions in a contingent of patients with myxomas, severe hemodynamic disorders, high pulmonary hypertension and severe circulatory failure (5). In the specialized literature, isolated cases

of rare forms – giant myxomas of the right heart with severe hemodynamic compromise are presented (2, 4, 5-10).

Statistically, myxomas are more often (75%) localized in the left atrium (LA) than in the right atrium (RA) - 15-20%, much less often in the ventricles of the heart and simultaneously on both sides of the interatrial septum (biatrial myxoma). Most myxomas are isolated, but in 7% of cases they are part of hereditary syndromes with autosomal dominant inheritance. In our practice (1, 3), there were cases of cardiac myxomas that were characterized by an extremely severe course. Isolated cases of surgical treatment of giant myxomas of the heart with symptoms of severe heart failure is of interest to cardiac surgeons involved in management of this pathology (2, 5, 6, 8).

Address for Correspondence*: Khamidulla A. Abdumajidov, Republican Specialized Scientific and Practical Medical Center of Surgery named after academician V.Vakhidov, Tashkent, Uzbekistan **Email:** hamidulla06@mail.ru

Received: 18.04.2023 **Revised:** 03.06.2023 **Accepted:** 04.06.2023

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Case report

Patient A., 39 years old, applied to the consultative polyclinic of our Center on May 24, 2019. with typical complaints of severe shortness of breath, palpitations, severe weakness, almost bed rest, severe swelling of the lower extremities, with a transition to the anterior abdominal wall (edema in the lower abdominal quadrants, a protrusion in the navel).

On physical examination there were cyanosis of the mucous lips, fingertips, severe shortness of breath, sharply aggravated by the slightest movement, palpitations, interruptions in the heart, severe weakness, fatigue. The cervical veins bulge sharply, the pulsation of the latter was visible, an extended apex beat was determined in the 6th intercostal space on the left, along the axillary line. There was an expansion of the borders of the heart to the right. Auscultation revealed a short systolic murmur over the xiphoid process, a pronounced accent of the 2nd tone over the pulmonary artery (PA). We detected weakening of breathing sounds in the lungs on both

sides and anasarca. In the abdominal cavity, pronounced ascites was determined (tense and protruding navel), pronounced edema of the lower extremities with a transition to the scrotum, the lower part of the anterior abdominal wall, where a pronounced expansion of the subcutaneous vessels was determined. His blood pressure was 110/70 mm HG, pulse was irregular - up to 98 beats/ min. The patient noted a noticeable decrease in diuresis; diuresis mainly was achieved with the help of diuretics.

On chest X-ray the pronounced lung ingestion, signs of high pulmonary hypertension (PH), a sharp increase in the RA and right ventricle (RV) were detected.

On echocardiography (Fig. 1) - a giant mass (12.6 x 7.5 cm) in the cavity of the RA was determined, obstructing tricuspid valve (TV) orifice and protruding into the cavity of the RV and occupying almost 50% of RV, it was located close to the mouth of the IVC. The base of the structural formation was the region of the right side of fossa ovalis of the interatrial septum (IAS) - about 3 cm. The condition of the TV was difficult to determine due to poor visualization, but TV regurgitation was severe (3rd degree). Pulmonary artery systolic pressure was determined as 115 mmHG. There was a moderate amount of fluid (up to 650-700 ml) in the pericardial cavity.



Fig.1. Echocardiographic view of giant myxoma of the right atrium obstructing tricuspid valve orifice and protruding into right ventricular cavity

Due to the severity of the condition, the patient was hospitalized in the intensive care unit (ICU), where he underwent emergency intensive care, preparation for surgery for health reasons. Clinical diagnosis: Neoplasm of the right RA, obturating TV and protruding to the RV (most likely - RA myxoma). Severe heart failure (NYHA !V). Severe pulmonary hypertension. Ascites. Anasarca. Bilateral exudative pleurisy (more on the right).

The decision on urgent surgery - removal of the tumor in the conditions of cardiopulmonary bypass was made by a council on the day of the patient's hospitalization, in the ICU with the participation of the cardiologists, cardiac surgeons, reanimatologists and anesthesiologists.

According to vital indications, the patient was operated on. The next morning the patient was taken to the operating room. Introductory anesthesia was performed with technical difficulties. General balanced combined anesthesia. Median sternotomy . Pericardium was opened lengthwise. There was moderate exudative pericarditis. Against the background of drug therapy, changes in hemodynamics (rhythm disturbances, hypotension, acid-base state) were corrected. We performed the traditional cannulation of the ascending aorta and the superior vena cava. Cannulation of the IVC was not possible due to the predominant location of the tumor in this area. A giant neoplasm of the RA was determined by palpation from the outside. Due to labile hemodynamics, it was decided to start artificial circulation without IVC cannulation, the IVC cannula

freely functions as an additional suction in the RA cavity. The anatomical finding – obstruction of the mouth of the IVC by a tumor was revealed intraoperatively. Longitudinal section of RA was done. On the revision: a neoplasm of gigantic size - 13x8cm, dark purple in color, in the form of clusters of grapes, jelly-like structure, with a base of about 2.5-3cm in the region of the fossa ovalis of the IAS were found. A partial removal of a part of the tumor in the area of the IVC was performed by exfoliation. After releasing the orifice of the IVC, it was possible to cannulate the IVC (full extracorporeal circulation). We considered cannulation of the femoral vein to be more time-consuming and dangerous in this situation. It was possible to carry out cardioplegia to the aortic root (antegrade). Asystole. On revision: the presence of a gigantic tumor, 13x8 cm in size, cluster- shaped, dark cherry color, jelly-like structure, mixed density, obstructing TV and protruding in the right ventricular cavity and occupying 45-50% of its cavity was revealed.

The base of the tumor, with a wide diameter of more than 3 cm, originated from the fossa ovalis of the IAS on the right, from the side of the RA. Using the "Palms" method, the tumor was removed into the wound from the tricuspid foramen and the RV. A gigantic tumor was removed fragmentarily, the base of the tumor determined in the region of the foramen ovale on the right side of the RA, was processed and isolated. The base of the tumor was excised with elements of the IAS (Fig. 2).

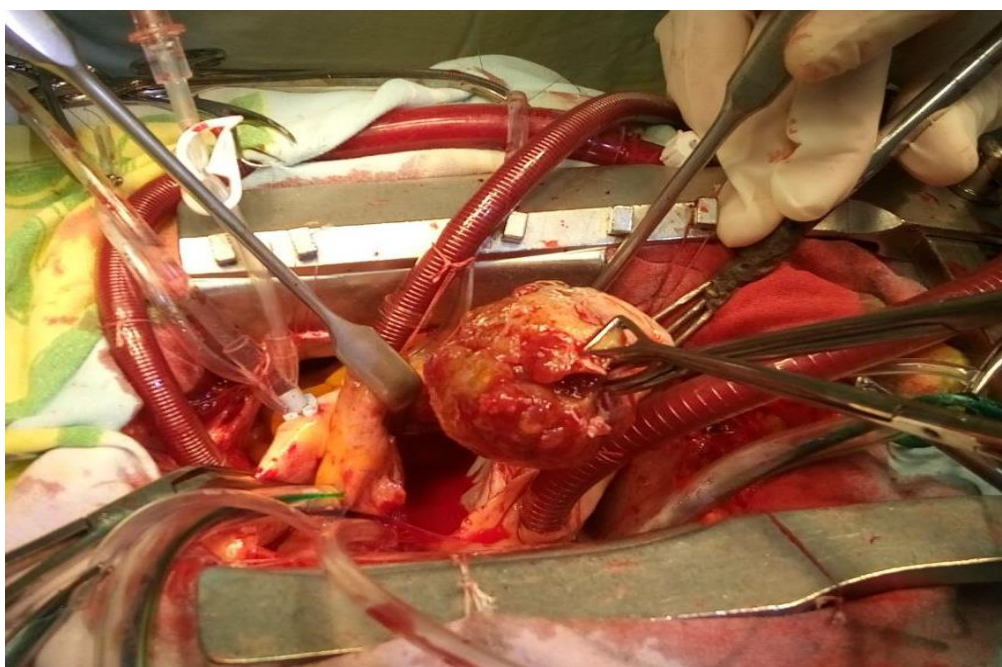


Figure 2. Removal of the bulk of the right atrial myxoma, after fragmentary removal of a part of the tumor in the area of the inferior vena cava.

Heart, Vessels and Transplantation 2023; 7: doi: 10.24969/hvt.2023.405

Surgery for RA myxoma obstructing IVC and AV orifice

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Remote area of myxoma was treated with betadine, alcohol, washed with saline. The removed tumor was 13x8 cm in size, of the above-mentioned consistency, shape and color. During the revision, no structural changes were found in the TV, volumetric dilatation of the fibrous ring of the TV was noted. Taking into account the absence of morphological changes in the TV, it was decided to perform a moderate correction - suture annuloplasty of the fibrous of annulus of the TV according to De Vega (keeping regurgitation up to 1.5-2 degrees). Washing the cavity of the heart and pericardium and restoration of IAS with a two-row stitches were performed.

Revision established exudative pleurisy on both sides. The pleural cavities were opened, about 2 liters of serous fluid were evacuated on the right, and 1.5 liters on the left. Warming up to 37C.

Triple air embolism prophylaxis and removal of the clamp from the aorta were done. Cardiac activity recovered independently, with rhythm disturbances. After medical therapy, hemodynamics was stabilized. Under infusion of cardiotoxic drug (Dobutamine 6-7 mcg/min), it was possible to decannulate the vena cava, cardioplegia and aorta (in stages). Myocardial electrode was left in RV. Hemostasis, sewing of the pericardium with rare interrupted sutures were made; drainages were placed in the cavity of the pericardium and mediastinum; approximation of the edges of the sternum with steel wire and restoration of the wound of the sternum in layers were done.

After additional drug therapy, with stable hemodynamics, blood gases, the patient was transferred to the ICU. During 2 days In the ICU the patient was stabilized, extubated, his hemodynamic parameters were corrected and the patient was subsequently transferred to the department.

The postoperative period was relatively stable. The phenomena of circulatory decompensation were gradually eliminated by medications.

A follow-up examination was performed, there were no additional shadows in the cavity of the right and left heart chambers, the valves were functioning normally, there was a residual minimal tricuspid regurgitation. The patient was discharged in a relatively stable condition on the 10th day after the operation.

Discussion

This case of myxoma of the heart is rare in clinical practice; a special course of a complicated tumor with severe disorders of intracardiac hemodynamics in cardiac surgery was described by a few authors (6).

The reason for such complications is the late detection of the disease, the lack of early visualization of the heart, the ignorance of specialists about the possibility of such complications in heart neoplasms, and much more. But, the fact is that detectability of cardiac myxomas has improved significantly with the introduction of two-dimensional ultrasound examinations into practice. Therefore, it is understandable how difficult it was to diagnose an intracardiac neoplasm in the absence of echocardiography. Indeed, they were mostly diagnosed by chance, or after the catastrophe, at autopsies (4, 5). In this regard, our case also confirms the need to improve knowledge in this area, especially for first-line doctors, where our patients apply at first. Our patient also went to the local polyclinic, where a heart disease was suspected with the above complaints, he was referred to a cardiologist for echocardiography. At the place of residence, echocardiography confirmed the suspicion - an intracardiac structural formation, in the right sections, with obstruction of the right atrioventricular orifice. The patient was referred to our center. In the conditions of a consultative polyclinic, due to a serious condition, only repeated echocardiography with Doppler echocardiography was performed, where the diagnosis was confirmed: a structural neoplasm of the heart; tumor of the RA with obturation of the TV, possibly myxoma; high pulmonary hypertension; ascites; bilateral exudative pleurisy.

Only 2 cases of giant myxomas of RA similar to our were reported by Nivargi et al. (8) and Sato et al. (10), they described similar variants of RA myxoma, but did not describe in detail the obstruction of the mouth of the IVC and how this was managed surgically.

In such cases, the most important thing is to determine the correct treatment tactics. The patient was urgently hospitalized in the ICU, where he underwent intensive therapy for heart failure, as a preoperative stage of treatment. There are similar cases in the literature, when the outcome of treatment was decided by the correct tactics - urgent preparation for an emergency operation to remove the tumor for health reasons. As described above, all the manipulations were carried out, strictly according to the protocol, correctly and in a timely manner.

As for the operation itself: significant difficulties have already arisen at the stage of induction of anesthesia, intubation and stabilization of hemodynamics. It was especially difficult to perform cannulation due to the significant size of the tumor, and blocking the IVC

We think that the right decision was made - to start artificial circulation on "one vein" - the superior vena cava and additional suction (using a cannula for the IVC - as a suction) in the RA. Further removal of a part of the tumor by exfoliation also justified itself, since it became possible to quickly cannulate the IVC and achieve complete cardiopulmonary bypass.

The further course of the operation was also performed adequately, which was confirmed by a relatively stable weaning from cardiopulmonary bypass and cardioplegia. The subsequent evacuation of about 2 liters of exudative fluid from the pleural cavities made it possible to further stabilize hemodynamics. The operation was successfully completed and the patient was transferred to the ICU, and infusion of dobutamine 7 mcg - as a prevention of possible pulmonary hypertension crises. The relatively stable course of the early postoperative period is the result of an adequately performed operation and all stages of providing this intervention.

Conclusion

The described case from our practice proves the possibility of a severe course of giant myxomas of the right heart, with the development of deep hemodynamic complications of systemic and pulmonary circulation. The chosen correct tactics of surgical treatment provided a favorable result, despite the severity of the initial condition and the degree of hemodynamic disturbance. Intraoperative tactics and technique for performing the stages of urgent surgery were chosen based on the situation, the features of the anatomical structure of the tumor, and the severe complications that arose. Our case proves the possibility of adequate implementation of such interventions, when choosing the right tactics of surgical treatment.

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Ethics: Informed consent was obtained from patient before all procedures.

Peer-review: Internal and external.

Conflict of interest: None to declare

Authorship: Kh.A.A. and Kh.Zh.B. equally contributed to case management and preparation of manuscript

Acknowledgement and funding: None to declare

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