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Case of Dilated Cardiomyopathy in Combination with Left Atrial Myxoma and Left Atriomegaly (Rare Clinical Case)

Abstract

Case description. A 41-year-old male patient F. was admitted to the Department of Surgical Treatment of Acquired Heart Diseases of the National Amosov Institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine with a diagnosis: Dilated cardiomyopathy. Mitral-tricuspid insufficiency. Left atrial myxoma. Left atriomegaly. Pulmonary hypertension. The patient was operated with heart failure symptoms of NYHA functional class IV.

After a week of medical preparation and additional examination, the patient was successfully operated on (removal of the left atrial myxoma, imposition of support rings on the left and right atrioventricular openings, paraanular plicacy of the left atrium). The postoperative period proceeded without significant complications. On the 12th day after the operation, the patient was discharged in satisfactory condition. In the remote period the patient died suddenly 3 years later.

Conclusion. Taking into account the initial serious condition of a patient with advanced heart disease with a reduced left ventricular ejection fraction, left atriomegaly (7.8 cm), pulmonary hypertension (50 mm Hg) and comorbidities, complex reconstruction of the left heart in atriomegaly and ventriculomegaly leads to an improvement in the functional state of the myocardium and morphometric indicators of left atrium and left ventricular ejection fraction.

Keywords*: mitral-tricuspid insufficiency, left atrial plication, cardiopulmonary bypass, low contractility of left ventricle, pulmonary hypertension.*

Introduction. Dilated cardiomyopathy (DCM) is a clinically significant risk factor for surgical treatment of patients with mitral-tricuspid insufficiency, especially with low left ventricular (LV) ejection fraction. Left ventriculomegaly in combination with reduced left ventricular contractility is an additional risk factor for plastic interventions in mitral-tricuspid insufficiency [1, 2, 3, 4]. Concomitant left atriomegaly leads to compression of LV, bronchi, resulting in significant respiratory disorders and heart failure. Dilation of left atrium (LA) is a factor that prevents the restoration of sinus rhythm and increases the risk of thromboembolic complications [2, 5, 6, 7, 8, 9]. Joining of giant LA myxoma to this complex pathology further increases the risk of surgery [10, 11, 12, 13, 14]. Combination of all the factors determines the feasibility of a comprehensive reconstruction of the left parts of the heart during surgical correction.

Case description. A 41-year-old male patient F. was admitted to the Department of Surgical Treatment of Acquired Heart Diseases of the National Amosov Institute of Cardiovas-

cular Surgery of the National Academy of Medical Sciences of Ukraine on August 2, 2012 with complaints about shortness of breath, swelling of the lower extremities, interruptions in the work of the heart. Diagnosis: DCM. Functional mitral-tricuspid insufficiency, giant LA myxoma, left atriomegaly, pulmonary hypertension. The patient was operated with heart failure symptoms of NYHA functional class IV. Advanced heart disease was noted since 2000. Myxoma was diagnosed three months ago. The patient had atrial fibrillation (AF) since 2006 (for 6 years). In 2011, there was an episode of thromboembolism in the brachial artery, the thrombus was removed.

According to the electocardiography, AF with a heart rate of 86 bpm and incomplete left bundle branch block were noted.

According to the echocardiographic examination before the operation (Fig. 1, Table 1), a volumetric formation with clear rounded edges (6.2x8.2 cm) with petrification sites was found in the LA cavity. The myxoma was tightly connected with the surface of endocardium of the left atrium especially in the zone of the oval fossa. In the LA appendage, a blood clot was detected (transesophageal examination). Functional mitral-tricuspid insufficiency. Dilatation mainly of the left parts of the heart. Pulmonary hypertension (pulmonary artery systolic pressure was 50 mm Hg), sharply reduced LV contractility (left ventricular ejection fraction = 17%).

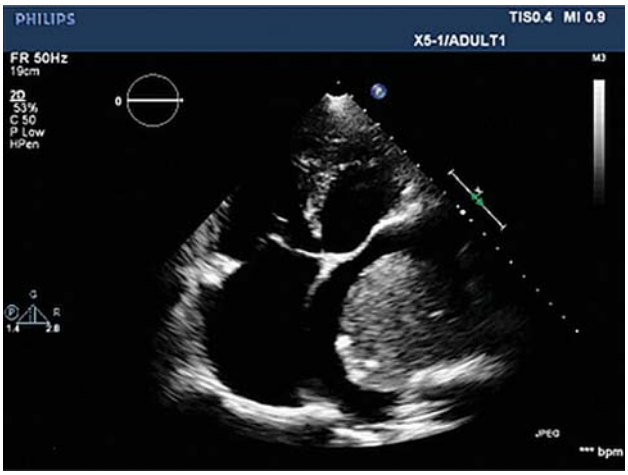

Fig. 1. *Echocardiogram of giant left atrial myxoma (through esophageal examination)*

The patient also underwent speckle-tracking echocardiography (Fig. 2), which made it possible to identify weak zones of contractility of LV.

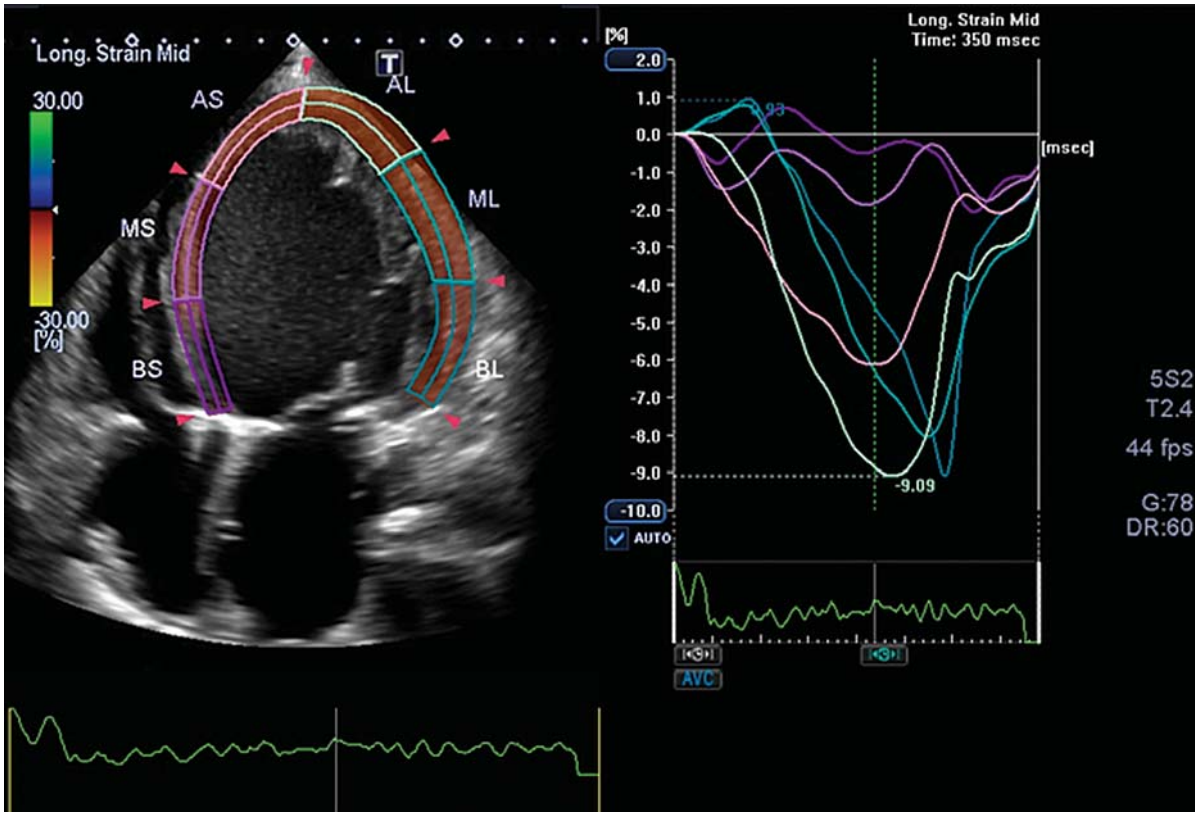

Fig. 2. *Speckle-tracking echocardiography of the left ventricle*

According to the radiography of the chest cavity, cardiomegaly was detected (Fig. 3).

According to the results of coronarography (August 6, 2012), atherosclerotic lesions of the coronary arteries were not detected. The risk of surgery according to the EuroSCORE II scale was 23.34%. The operation was performed on August 10, 2012, the volume of the operation was as follows: removal of LA myxoma + imposition of support rings on the left and right atrioventricular openings + paraanular plication of LA + thrombectomy from the LA appendage. Duration of the operation was 175 minutes. The operation was performed under conditions of cardiopulmonary bypass (CPB) (81 min) and general hypothermia (33.0 °C). Aortic cross-clamping time was 94 minutes. Intraoperative blood loss was 250 mL.

The operation was performed as follows (surgeon: prof. Volodymyr V. Popov). Median sternotomy. Aorta and vena cava cannulation. Start of CPB. Hypothermia to 33.0 °C. Antegrade cardioplegia through the aortic root with Custodiol solution 500 mL + retrograde cardioplegia through the coronary sinus with Custodiol solution 2000 mL and external cooling of the heart with ice solution. LA was dissected: myxoma in the capsule, spherical (8 cm in diameter), attached on a wide base 3x2 cm at the upper edge of the antennae of the left pulmonary veins. The myxoma was removed fragmentarily due to its large size (Fig. 4), the basement was removed, coagulated and stitched. The LA appendage was bandaged from the outside, resected. Paraanular plasty of the back wall of LA was made (Fig. 5). A 32 mm St. Jude Medical supporting ring was superimposed on the left atrio-

Table 1

Echocardiographic parameters of the heart before operation

Parameters (units)	Values
Body surface area (m^2)	1.72
LV EDVI (mL/m^2)	196.5
LV EDV (mL)	338
LV ESVI (mL/m^2)	163.4
LV ESV (mL)	281
LV SVI (mL/m^2)	33.1
LV SV (mL)	57
LVEF	0.17
LAD (cm)	7.8
PASP (mm Hg)	50.0
Mitral valve	Moderate regurgitation
Tricuspid valve	Moderate regurgitation
Aortic valve	ΔAV 7 mm Hg, no regurgitation

EDV, end diastolic volume; EDVI, end diastolic volume index; ESV, end systolic volume; ESVI, end systolic volume index; LAD, left atrial diameter; LVEF, left ventricular ejection fraction; PASP, pulmonary artery systolic pressure; SV, systolic volume; SVI, systolic volume index.

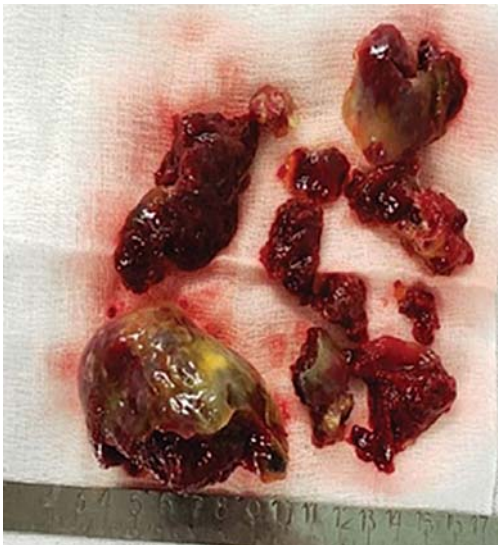

Fig. 4. *Resected myxoma*

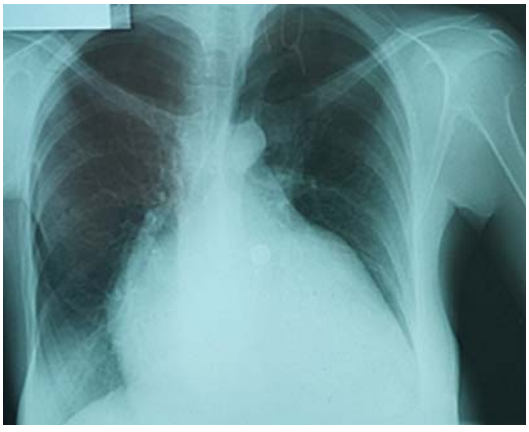

Fig. 3. *X-ray of the chest before surgery*

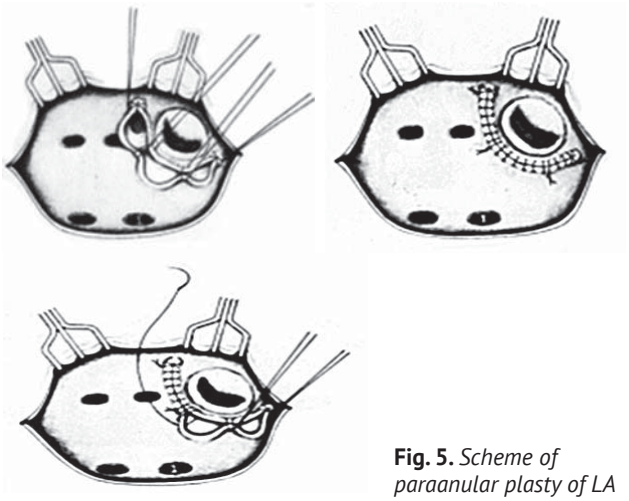

Fig. 5. *Scheme of paraanular plasty of LA*

ventricular opening and fixed with 17 separate sutures. Right atrium was dissected. A 32 mm St. Jude Medical supporting ring was superimposed on the right atrioventricular opening with 14 separate seams. The heart resumed its activity through depolarization. After stopping the CPB, the pressure was 220 mm H_2O in LA, central venous pressure was 80 mm H_2O, arterial pressure was 120/80 mm Hg.

Inotropic support after stopping the CPB: dobutamine 6.2 μg/kg/min. The postoperative period in the intensive care unit lasted 168 hours. Inotropic dobutamine support was 6.2 μg/kg/min on the first day after surgery, 3.6 μg/kg/min on the second day, 4.4 μg/kg/min on the third day, 4.4 μg/kg/min on the fourth day, 2 μg/kg/min on the fifth day, and on the sixth day it was already suspended. Creatine kinase-MB values: 31 u/L (on August 11, 2012, i.e. the second day after surgery). While the patient was in the intensive care unit, moderate liver failure (total bilirubin up to 65 μmol/L) was observed. On the seventh day, the patient was transferred to the surgical department. Drug therapy was successfully continued (anticoagulant therapy, amiodarone, antibiotic therapy, etc.). The wound healed by primary intention. Table 2 compares echocardiographic indicators (before surgery and at discharge).

Table 2 provides information on morphometric and hemodynamic features in early postoperative period. In the first period, the values were expected and unremarkable. The wound healed by primary intention. Electrocardiography at discharge: AF with a frequency of 77 beats/min. Blood test at discharge: hemoglobin 131 g/L; RBC $4.6x10^{12}$/L; ESR 23 mm/h; WBC $9.4x10^{9}$/L. Total bilirubin decreased to 24 μmol/L. Radiography of the chest cavity showed a decrease of heart shadow (Fig. 6).

Echocardiogram at discharge is shown in Fig. 7.

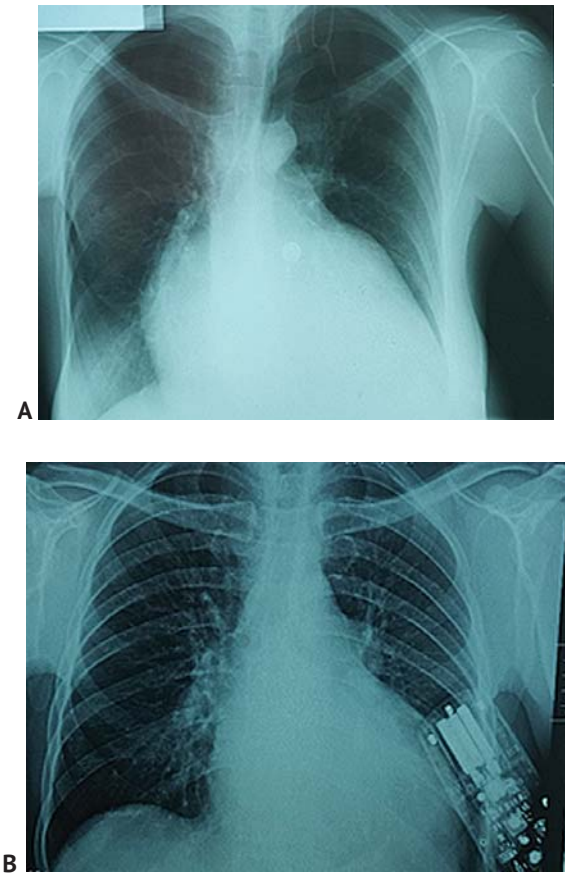

Fig. 6. *Radiography of the chest cavity before surgery (August 2, 2012) (A) and after surgery (August 17, 2012) (B)*

Table 2

Changes in echocardiographic parameters before and after operation

Parameters (units)	Before the operation	At discharge
Mitral valve	Moderate regurgitation	ΔMV 10 mm Hg, no regurgitation
Tricuspid valve	Moderate regurgitation	ΔTV 6 mm Hg, no regurgitation
LV EDVI (mL/m²)	196.5	177.3 (-9.8%)
LV EDV (mL)	338	305 (-9.8%)
LV ESVI (mL/m²)	163.3	137.2 (-16.0%)
LV ESV (mL)	281	236 (-16.0%)
LV SVI (mL/m²)	33.1	43 (+29.9%)
LV SV (mL)	57	69 (+29.9%)
LVEF	0.17	0.23 (+35.3%)
LAD (cm)	7.8	5.0 (-35.9%)
PASP (mm Hg)	50	40 (-20.0%)

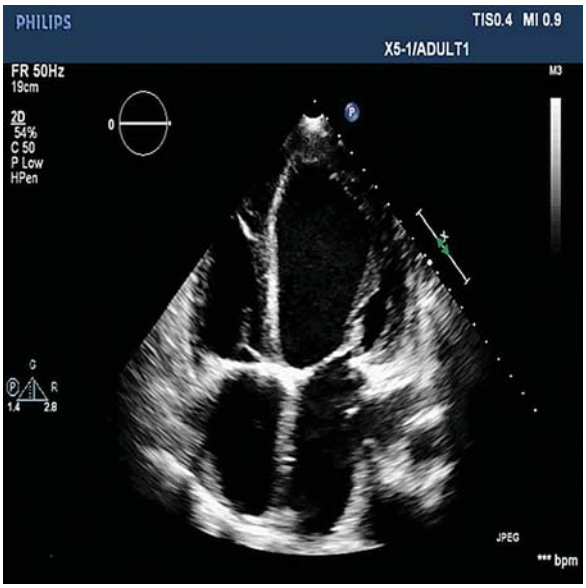

Fig. 7. *Echocardiogram at discharge*

The patient was discharged on August 22, 2012 (12 days after surgery) in satisfactory condition for further treatment in a cardiology hospital at the place of residence. Six months after discharge the patient was examined at the National Amosov Institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine. Echocardiographic examination data remained almost unchanged. The patient noted certain improvement in the condition compared to his preoperative state. The patient did not attend the Institute anymore. According to information provided by his wife, the patient died suddenly 3 years later, in November 2015.

Discussion. In the correction of mitral-tricuspid heart disease due to DCM, the factor of left atriomegaly is clinically significant at the hospital stage, and, to an even greater extent, in the distant period. The level of thromboembolic complications, as well as the level of cardiovascular insufficiency in the group without correction of left atriomegaly in the distant period reaches a critical value, especially in combination with long-existing AF [3, 7, 8]. Left atriomegaly in the presence of a tachyform AF contributes to the formation of blood clots in the cavity of the LA and thromboembolic complications occurring in this case. Extreme dilated LA contributes to the progression of heart and respiratory failure due to compression of the bronchi, trachea and posterior wall of LV [1, 5, 6, 7]. Therefore, we consider it necessary to carry out LA reduction.

Conclusion. Adequate plastic correction of functional mitral-tricuspid insufficiency, along with the elimination of myxoma and LA plasty, significantly improved the morphometry of the left heart and the contractility of LV already at the early hospital stage [4, 6, 9]. Elimination of giant LA myxoma, together with the maternal basement, is an important element of radicalism and the absence of relapse in the long term. LA reduction in the surgical treatment of mitral-tricuspid valve diseases is a desirable procedure in patients with left atriomegaly.

This operation leads to a significant improvement in the morphometry of LA, which is accompanied by a positive clinical effect, both in the hospital and in the distant periods after surgery. In the presence of left ventriculomegaly in combination with reduced contractility of LV, mitral valve repair should be performed with the maximum possible preservation of the sub-valve structures of the mitral valve. Thus, careful medical preoperative preparation, complex reconstruction of the left heart during the correction of mitral-tricuspid valve diseases (in DCM), in combination with LA myxoma and left atriomegaly, reduces the risks of complications at the hospital stage even in an extremely difficult patient and makes it possible to transfer him to the waiting list for transplantation.

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Випадок дилатаційної кардіоміопатії в поєднанні з міксомою лівого передсердя та лівою атріомегалією (рідкісний клінічний випадок)

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Резюме. При корекції мітрально-тристулкової вади серця за рахунок дилатаційної кардіоміопатії фактор лівої атріомегалії є клінічно значущим на госпітальному етапі, а ще більшою мірою – у віддаленому періоді. Рівень тромбоемболічних ускладнень, як і рівень серцево-судинної недостатності, в групі без корекції лівої атріомегалії у віддаленому періоді сягає критичної величини, особливо в поєднанні з давно існуючою фібриляцією передсердь. Ліва атріомегалія за наявності тахіформи фібриляції передсердь сприяє виникненню тромбів у порожнині лівого передсердя та тромбоемболічним ускладненням, що мало місце в цьому випадку. Дилатоване ліве передсердя сприяє прогресуванню серцевої та дихальної недостатності за рахунок компресії бронхів, трахеї та задньої стінки лівого шлуночка. Тому ми вважаємо за необхідне проведення редукції лівого передсердя.

Повне збереження клапанних і підклапанних структур під час пластики мітрального клапана дає змогу в більшій мірі зберегти скоротливість лівого шлуночка, зменшити його об'єм, особливо за наявності вентрикуломегалії. Наявність міксоми лівого передсердя в поєднанні з іншою патологією є вкрай рідкісним клінічним випадком, а також суттєвим фактором ризику та обов'язково потребує радикального усунення.

Адекватна пластична корекція відносної мітрально-тристулкової недостатності водночас з усуненням міксоми та пластикою лівого передсердя дозволила значно покращити морфометрію лівих відділів серця та скоротливість лівого шлуночка вже на ранньому госпітальному етапі. Усунення гігантської міксоми лівого передсердя разом із материнською основою є важливим елементом радикальності та відсутності рецидиву у віддалені терміни. Редукція лівого передсердя під час хірургічного лікування мітрально-тристулкової вади – бажана процедура у пацієнтів з лівою атріомегалією. Ця операція призводить до значного покращення морфометрії лівого передсердя, що супроводжується позитивним клінічним ефектом, як на госпітальному, так і у віддаленому періодах після операції.

За наявності лівої вентрикуломегалії в поєднанні зі зниженою скоротливістю лівого шлуночка пластику мітрального клапана потрібно виконувати з максимально можливим збереженням підклапанних структур мітрального клапана.

Отже, ретельна медикаментозна доопераційна підготовка, комплексна реконструкція лівих відділів серця при корекції мітрально-тристулкової вади на тлі дилатаційної кардіоміопатіїї, поєднаної з міксомою лівого передсердя та лівою атріомегалією, допомагає зменшити ризики ускладнень госпітального етапу навіть у вкрай важкого пацієнта та дати можливість перевести його на лист очікування для трансплантації.

Ключові слова: *мітрально-тристулкова недостатність, пластика лівого передсердя, штучний кровообіг, низька скоротливість лівого шлуночка, легенева гіпертензія.*

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