

# Unotypical cause of heart failure – right atrial myxoma

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**Abstract:** The study presents case of a 62-year-old female patient with symptoms of the heart failure NYHA class III intensifying for the last 2 months. Physical examination at admission showed no significant abnormalities. In two- and three-dimensional echocardiography normal thickness and contractility of the left ventricle wall were found, as well as the normal systolic function of the right ventricle. In enlarged right atrium a structure of the oval shape was found with the features of continuity with atrial septum and bulging of a small part of the structure through the orifice of the tricuspid valve to the right ventricle. In the study no features of blocking the tricuspid flow or the inflow from the main veins and coronary sinus were detected. On the basis of the complete echocardiographic examination and clinical presentation, tentative diagnosis of right atrium myxoma was made. Coronary angiography showed no significant abnormalities in coronary arteries. After cardio-surgical consultation the patient was scheduled for surgical removal of the tumor, which was performed without complications. The histopathological examination confirmed the diagnosis of myxoma. In the follow-up echocardiography carried out after 8 months reduced size of the right atrium and a normal function of the ventricles were demonstrated. There were no features of tumor re-growth.

**Key words:** heart failure, heart tumor, myxoma, transthoracic echocardiography

## INTRODUCTION

Primary heart tumors occur rarely because they are found in about 0.02% of autopsies and 75% of those cases are benign tumors [1,2]. The most common heart tumor is myxoma, which constitutes 50% of benign heart tumors. It occurs more frequently in women. Myxoma is localized in the left atrium in about 75% of patients, while in 18%, it occurs in the right atrium, and the least frequently in a ventricle. In 10% of cases it runs in the family (Carney's syndrome) [3,4]. The diagnostic workup of myxomas is based first of all on the transthoracic (TTE) and transesophageal (TEE) echocardiographic examinations. The transthoracic and transesophageal examinations make it possible for specialists to establish a tentative diagnosis, define the anatomic relations and place of the pedicle attachment, as well as the presence of intravascular clots in the tumor [5]. The final diagnosis of the myxoma is feasible solely on the basis of anatomopathologic examination of the surgically removed mass.

## CASE REPORT

A 62-year-old female patient was admitted to the Cardiology Department of the Medical University due to heart failure NYHA III class, for dyspnea at slight exertion, general weakness, fatigue. These complaints had been gradually becoming more pronounced for two years. On the physical examination on the day of admission no significant abnormalities were found. The heart rate was regular – 90/min., arterial blood pressure of 180/85 mmHg. Above the lung fields regular follicular murmur, no pathological heart murmurs were observed. As far as abnormalities in laboratory tests are concerned, only a slight increase of D-dimers was found – 983 ng/ml. On ECG there were no abnormalities. On the two- and three-dimensional TTE the presence of a moving mass was found. It was oval and had the size of 54 × 34 mm, smooth edges, and intensified echogenicity with internal strongly saturated echoes, and with the features of continuity with atrial septum and projection in the diastole of a slight part of the structure circumference through the orifice of tricuspid valve to the right ventricle (Fig.). No features of blocking the tricuspid flow or inflow from the main veins and coronary sinus were found; the tricuspid insufficiency of the 1st degree was visualised. Enlargement of the right atrium (46 mm) was observed. Moreover, there was a normal thickness and contractility of the left ventricle walls, ejection fraction of 65%, normal systolic function of the right ventricle, without pulmonary hypertension. The transesophageal echocardiographic examination

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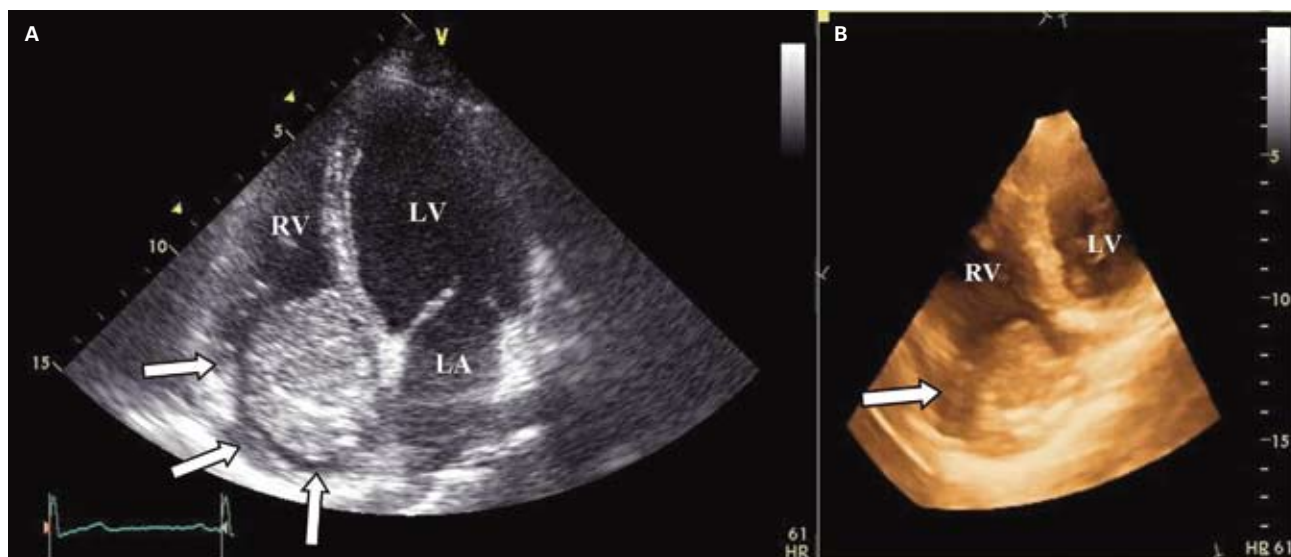
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**Fig.** Apical four-chamber view (phase at the end of diastole), with an oval structure visible (arrows) of intensified echogenic character with the projection in diastole a slight part of the circumference of the structure through the orifice of the tricuspid valve to the right ventricle. **A.** Transthoracic two-dimensional echocardiography. **B.** Transthoracic three-dimensional echocardiography. Abbreviations: LA – left atrium, LV – left ventricle, RV – right ventricle

confirmed the diagnosis of the right atrial myxoma. Coronary angiography showed no significant changes in the coronary arteries. After the consultation with a cardio-surgeon the patient was scheduled for surgical treatment. At the Department of Cardiac Surgery of the Medical University uncomplicated surgical removal of the myxoma was performed. Histopathologic examination confirmed the diagnosis of myxoma. On the follow-up echocardiography eight months later, reduced size of the right atrium cavity (44 mm), moderate tricuspid insufficiency and efficient function of the ventricles (ejection fraction of 63%) were detected. No features of the tumor re-growth were observed.

## DISCUSSION

Finding echogenic mass in the heart requires the differentiation which takes into consideration first of all: a thrombus, myxoma or another tumor, malignant tumor (especially metastases from the breast or lung cancers and/or sarcoma, which is often localized in the right atrium), lipomatous overgrowth of inter-atrial partition [5]. Echocardiography showed the myxoma as an oval or polypous irregular mass. Most frequently it is connected to atrial septum, or more rarely a free atrial wall, with a pedicle. This mass was non-homogenous with areas of necrosis, degeneration, bleeding or the presence of cysts. Three-dimensional echocardiography allows a more precise evaluation of the sizes and mobility of pathological masses than the two-dimensional echocardiography does.

Sometimes the differentiation diagnosis based on the echocardiogram is difficult; attempts are made at digital anal-

ysis of the texture of tumors in order to make their evaluation easier [6].

Myxoma is most frequently detected in patients in the third or fourth decade of their lives. To establish the diagnosis of myxoma in a younger person requires the exclusion of a dominant autosomal Carney's syndrome. In such patients myxomas are multi-focal, and after removing them surgically their recurrence is more frequent [5,7].

The clinical presentation of heart tumors depends on the localization, size and presence of the pedicle which determines the mobility of the tumor, thus its penetration in the phase of diastole into the atrial-ventricular orifice and the ventricular cavity. The symptoms of myxoma are not typical and include general ones (e.g. weight loss, raised body temperature, dyspnea after exertion, fatigue, pains and dizziness), supraventricular arrhythmias (atrial fibrillation, supraventricular tachycardia, supraventricular beats). Torn off fragments of the tumor may cause embolism of peripheral blood vessels [8]. Physical examination may reveal systolic or diastolic heart murmurs; the myxoma tone is present in about 15% of patients. Some myxomas can induce inflammatory state, confirmed in laboratory tests (increased ESR, leucocytosis), as well as anemia, thrombocytopenia or thrombocytosis. It is believed that the occurrence of general symptoms is associated with the production of cytokines by the tumor, especially of interleukin 6 [5]. No such features were detected in our patient.

Right atrium myxoma can produce symptoms of right-ventricular cardiac insufficiency, stenosis or insufficiency of the tricuspid valve, symptoms of pulmonary embolism or pulmonary hypertension, and general symptoms, e.g. weakness, fever or weight loss [5]. In the case of the closure of the venous orifice that can occur, sudden cardiac death may result.

Translocation of the tumor from the atrium to the ventricle can cause the damage to the valve apparatus or a tendinous chord. Clinical symptoms in the case of right atrium occur at a two or three times bigger mass. In the described patient despite normal tricuspid flow or inflow from main veins and coronary sinus, the filling of the right ventricle was hampered, which caused venous stasis and the development of cardiac insufficiency.

Detection of myxoma is an indication for urgent cardio-surgical intervention due to the speedy growth of the tumor, considerable risk of embolism, and blocking the route of the outflow. Cardio-surgical treatment consists in the removal of the tumor with the pedicle. After the operation a regular echocardiographic follow-up is necessary in order to detect tumor recurrence, which is observed most frequently approximately four years after surgery. In patients with a high risk of recurrence (myxoma that runs in the family, genetic forms) echocardiography should be carried out every year or two for 15 years after the operation [5,7]. In the case of lower risk patients at our centre, we perform follow-up visits up to five years after the intervention. To sum up, the described case illustrates the necessity of taking into consideration in differential diagnosis untypical causes of heart insufficiency that might be cured by means of surgical treatment.

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