

# Myxoma of the heart (RCD code: VI-1A.1)

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## Abstract

Myxomas are the most common primary benign intracardiac tumors. Typically they locate in the left atrium (LA) or right atrium (RA) and are composed of cells, that originate from multipotent mesenchyme. The clinical manifestations of myxomas depend on the anatomic location of the tumor, its size, as well as its mobility. We present two case reports that concern the management of myxoma. The first diagnostic approach for assessment of the myxomas is two dimensional echocardiography. Computed tomography (CT) and Magnetic Resonance (MR) play especially role in providing diagnostic information useful toward surgical planning. Surgical operation and resection of the myxoma is a definite and curative therapy. The risk for recurrence of the myxoma is about 2–5 %. The annual clinical surveillance and echocardiographic assesment is recommended to all patients who undergo a myxoma operation. JRCD 2013; 1 (4): 163–166

**Key words:** myxoma, benign cardiac tumor

## Background

Myxomas are the most common primary benign intracardiac tumors although in the general population are extremely rare. Usually they occur sporadically, but 7% may be familial, inherited as an autosomal dominant disorder [1]. They most commonly occur between third and six decade.

Typically they locate in the left atrium (LA) or right atrium (RA) and arise from or near the interatrial septum at the border of the fossa ovalis membrane [1, 2]. Myxomas are smooth, pedunculated tumors, that are gelatinous in texture [3]. Usually they are formed as a compact mass. The spontaneous fragmentation is rare and it concerns mainly myxomas that have multiple fragile villous extensions [1]. Their size may vary – from 1 to 15 cm in diameter, and weight between 15 and 180 g [3]. Histologically, myxomas are composed of cells, that originate from multipotent mesenchyme, which is capable of neural and endothelial differentiation [4].

The clinical manifestations of myxomas depend on the anatomic location of the tumor, its size, as well as its mobility. Myxomas in LA may cause symptoms similar to mitral stenosis [2] and frequently are associated with electrocardiographic evidence of LA hypertrophy [3]. Patients with RA myxomas display features of the right

heart failure [2]. There is evidence that myxomas may cause systemic embolization [2,3].

## Case presentation 1: myxoma of the heart in a 49-year-old woman identified by echocardiography

A 49-year-old woman with persistent atrial fibrillation in the course of Graves' disease was admitted electively (November 2004) to the cardiac department to undergo electrical cardioversion. During the previous hospitalization (October 2004) in the same department due to chest pain, echocardiography was performed. It revealed an enlarged left ventricle with impaired contractility and reduced ejection fraction to about 38%. Coronary angiography was performed and no significant changes in the coronary arteries were observed.

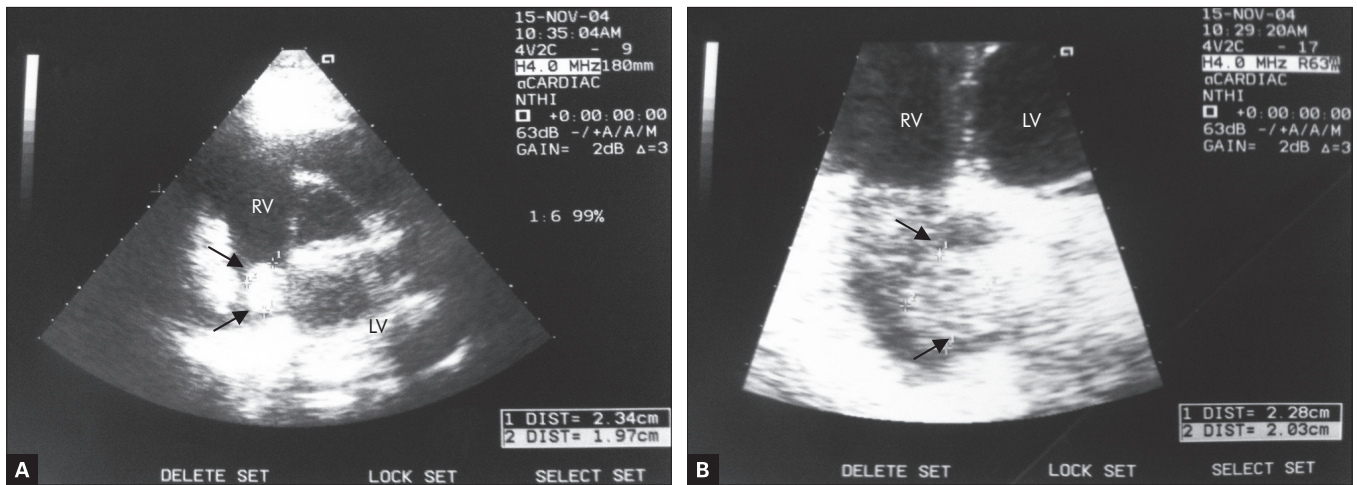
Laboratory tests showed that the level of the international normalized ratio was not in the therapeutic range.

During current hospitalization, transthoracic and transesophageal echocardiography revealed the presence of a round, movable tumor of about 20×21 mm in size in the right atrium (Figure 1).

Conflict of interest: none declared.

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**Figure 1.** Transthoracic echocardiography. A. Parasternal short-axis view. B. Apical view. The tumor in the right atrium (black arrows). RV – right ventricle, LV – left ventricle

The tumor was evenly saturated; the saturation was similar to the structures of the heart. In addition, transesophageal echocardiography showed no thrombus within the left atrial appendage.

Cardiac magnetic resonance confirmed the presence of an abnormal oval structure in the right atrial cavity, approximately 23 × 19 mm in size. Its middle part was connected to the intra-atrial septum. The structure was movable and showed an intermediate signal on T1 images. Moreover, it was hyperintense on STIR sequences and enhanced after contrast administration. The image corresponded to a myxoma but did not exclude blood clots (Figure 2).

The patient received heparin but there was no change in the tumor's size after 10 days of therapy. Therefore, she was scheduled for urgent cardiac surgery.

The surgery was performed with the use of cardiopulmonary bypass, normothermia, and cold crystalloid cardioplegia. During the procedure, a tumor (myxoma) was shown (30 × 30 mm in size) after the opening of the right atrium, located at the base of the atrial septum. The entire lesion was removed, including the fragments of the intra-atrial septum. After the procedure, low-output syndrome developed, which was treated medically. There were no complications during the postoperative course and wound healing process. The patient was discharged from the cardiac surgery department in good general condition.

## Case presentation 2: myxoma of the heart in a 57-year-old woman identified by echocardiography and magnetic resonance imaging

A 57-year-old woman was admitted to the cardiac department to undergo diagnostic procedures of the tumor in the right atrium.

The tumor was recognized on transthoracic echocardiography in an outpatient clinic. For 2 months before admission, she complained of a nonspecific stabbing chest pain and high blood pressure.

In the cardiac department, the transesophageal echocardiography was performed. It revealed the presence of a lesion in the right atrium that was attached from one side to the atrial septum. The size of the lesion was 23 × 27 mm. Its distal portion was irregular and had polycyclic contours. The image was not typical for a myxoma.

Cardiac magnetic resonance imaging confirmed the presence of an abnormal tissue in the right atrial cavity, approximately 32 × 23 × 31 mm in size. Its middle part was connected to the intra-atrial septum. The structure was movable, hyperintense on STIR sequences, hypointense on T1 and heterogeneous on T2 scans, and enhanced after contrast administration. There were no features of malignancy or infiltration of the surrounding parts of the heart. The image corresponded to a myxoma and, less likely, to a clot (Figure 3, 4, 5).

An abdominal ultrasound examination and a chest X-ray image did not show significant changes. Coronarography did not reveal coronary stenosis.

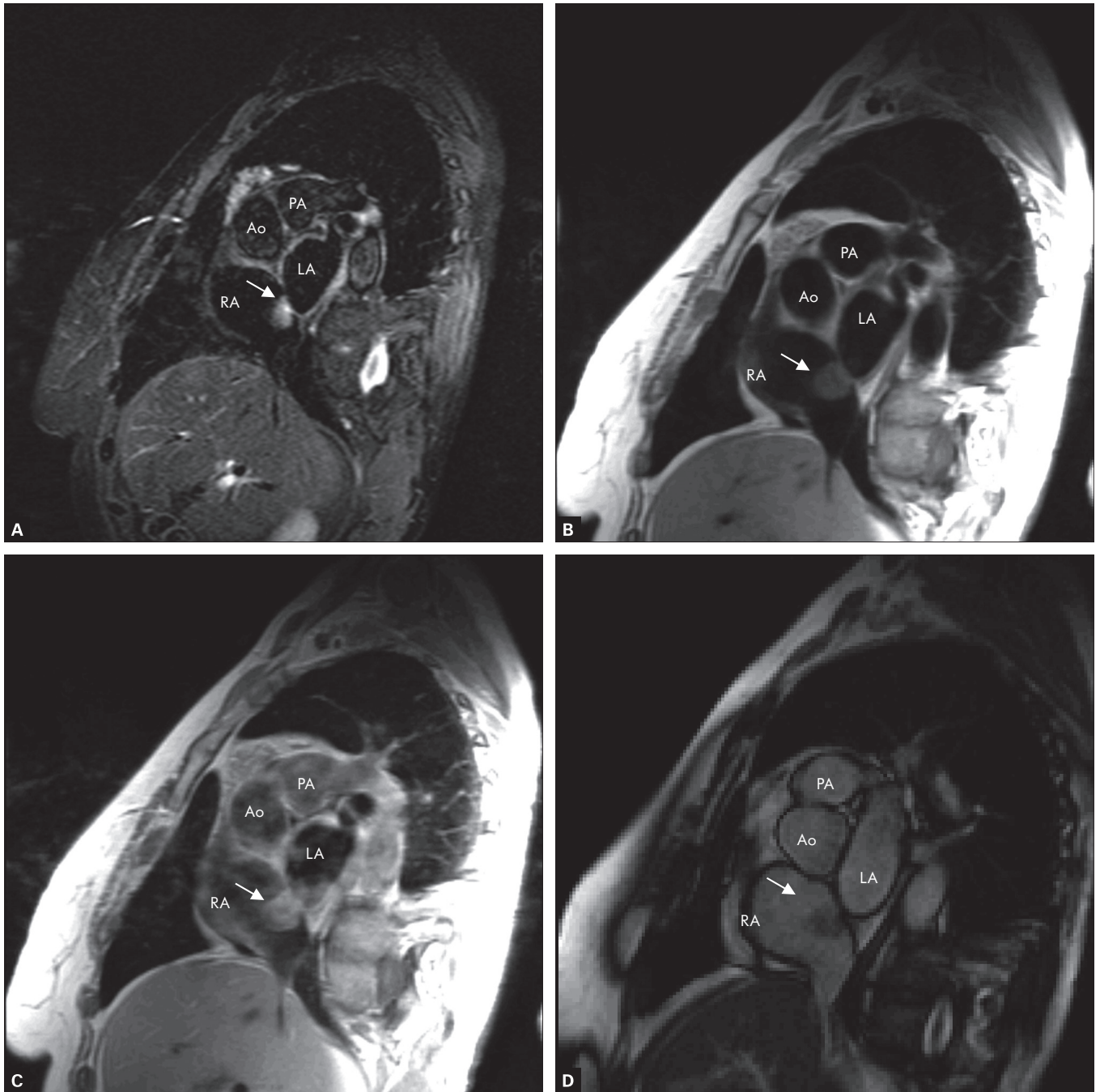
The patient underwent surgery in 2011 with the use of cardiopulmonary bypass, normothermia, and crystalline cardioplegia. During the procedure, after the opening of the right atrium, a myxoma was shown, 20 × 15 mm in size, located at the base of the atrial septum. The tumor was completely removed.

There were no complications during the postoperative course and wound healing process. The patient was discharged from the cardiac surgery department in good general condition.

## Management strategy

Two dimensional echocardiography is the first diagnostic approach and the most valuable tool for assessment of the myxomas [2]. It easily shows the location, size, shape, attachment, mobility and haemodynamic consequence of the tumor [1, 5].

Computed tomography (CT) and Magnetic Resonance (MR) provide a complete multiplanar and non-invasive evaluation of myxomas. In CT they have heterogeneous low attenuation due to their gelatinous consistency whereas in MR they are characterized

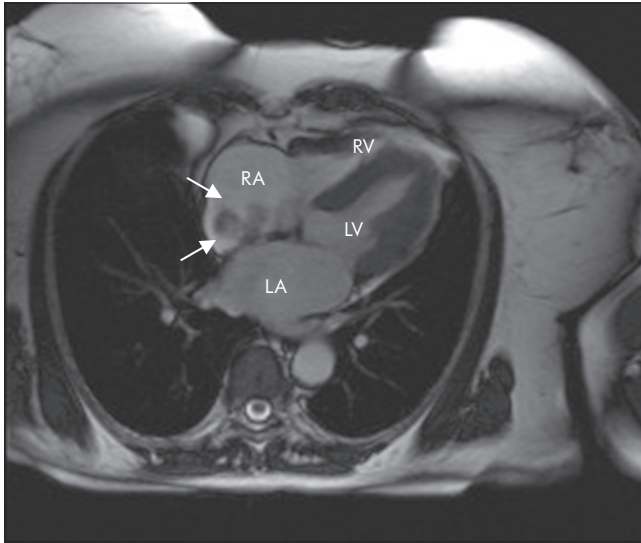


**Figure 2.** Cardiac magnetic resonance. A. STIR sequence. T1-weighted images. B. Spin echo sequence. T1-weighted images. Precontrast. C. Spin echo sequence. T1-weighted images. Postcontrast. D. Sine gradient echo. Short axis view. Atrial projection. The tumor in the right atrium (white arrows). Ao – aorta, PA – pulmonary artery, RA – right atrium, LA – left atrium

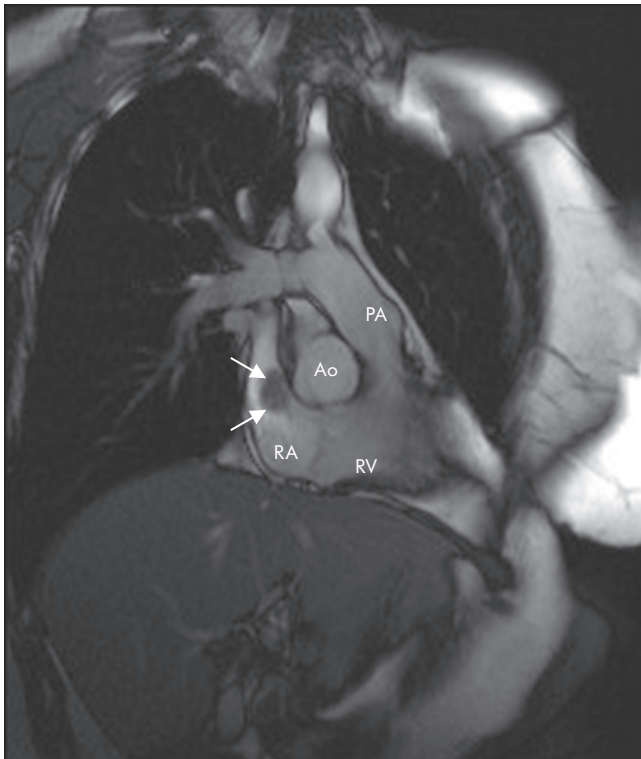
by increased signal intensity on T2 weighted images [1]. These techniques play especially role in providing diagnostic information useful toward surgical planning.

In order to avoid the risk of cardiovascular complications and embolisation, prompt surgical operation and resection of the myxoma are required as soon as a myxoma diagnosis is made [6]. If anatomically feasible the proper surgical intervention is the resection of the tumor with a margin of normal tissue and it is recognized as a definite and curative therapy [7]. Postoperative recovery is rapid.

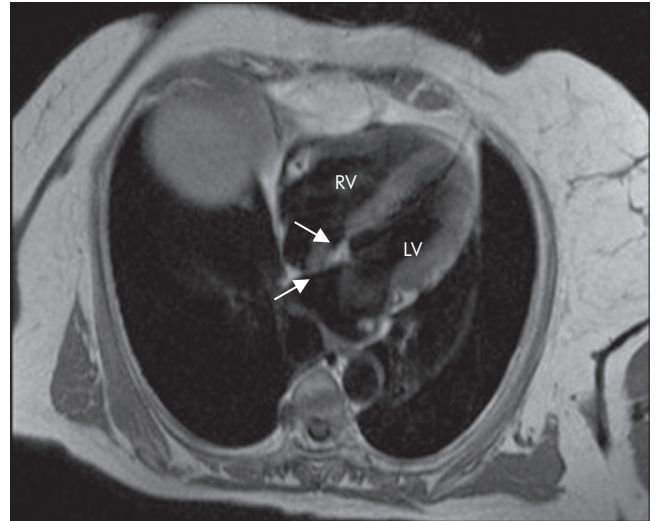
However, atrial arrhythmias or atrioventricular conduction abnormalities may occur [3]. Patients after the surgery are at risk for recurrence of the myxoma (2–5 %) [3], but the risk is much more common with familial myxomas than with sporadic tumours [1]. The annual surveillance, clinical as well as echocardiographic, is recommended to all patients who undergo a myxoma operation [8].



**Figure 3.** Cardiac magnetic resonance. Four-chamber long-axis view. Sine gradient echo. The tumor in the right atrium (arrows). RV – right ventricle, LV – left ventricle, LA – left atrium, PA – pulmonary artery



**Figure 4.** Cardiac magnetic resonance. Two-chamber view. Right ventricular projection. Sine gradient echo. The tumor in the right atrium (arrows). RV – right ventricle, RA – right atrium, Ao – aorta, PA – pulmonary artery



**Figure 5.** Cardiac magnetic resonance. Four-chamber long-axis view. Spin echo T1-weighted images. The tumor in the right atrium (arrows). RV – right ventricle, LV – left ventricle

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