



## Massive right atrial myxoma with dyspnea at rest in an elderly patient: A case report

Veliki miksom desne pretkomore sa dispnejom u miru kod bolesnice u starijem životnom dobu

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### Abstract

**Introduction.** Primary heart tumors are extremely rare and myxoma is the most common type of these tumors. Although intra-atrial presentation is a predilection place, right atrial localization is atypical. The symptom triad is characteristic in the clinical presentation of the tumor: embolic complication, intracardiac blood flow obstruction and systemic manifestations like elevated erythrocyte sedimentation rate, fever, anemia, body weight loss. **Case report.** We presented an elderly female patient with massive myxoma in the right atrium, 77 × 44 mm in diameter, which filled the entire right atrium and spread into the right ventricle, causing the tricuspid valve obstruction and dyspnea. It was visualized by transthoracic echocardiography and small and insignificant pericardial effusion was also seen. After surgical removal of the tumor, the patient remained without any symptoms and pericardial effusion. **Conclusion.** Tumors of the right heart have to be considered in the differential diagnosis of unexplained dyspnea in elderly patients. Transthoracic echocardiography is certainly necessary and mostly available diagnostic tool that can be of great help in diagnosing heart tumor as well as planning cardiac surgery, as it provides in most cases excellent visualization of the tumor and its relationship with other parts of the heart.

### Key words:

myxoma; heart atria; dyspnea; aged; diagnosis; cardiac surgical procedures; treatment outcome.

### Apstrakt

**Uvod.** Primarni tumori srca su izuzetno retki. Najčešće su to miksomi. Lokalizacija u desnoj pretkomori je atipična. Za kliničku sliku karakterističan je trijas simptoma: embolijske komplikacije, opstruktivne tegobe i sistemske manifestacije bolesti kao što su: povišena sedimentacija eritrocita, povišena telesna temperatura, anemija, gubitak telesne težine. **Prikaz bolesnice.** U radu je prikazana starija bolesnica sa velikim miksomom desne pretkomore, dimenzija 77 × 44 mm, koji je potpuno ispinjavao desnu pretkomoru i skoro potpuno desnu komoru, uzrokujući opstrukciju trikuspidnog protoka i dispneju. Miksom je dijagnostikovao transtorakalnom ehokardiografijom. Pored toga, ehokardiografski verifikovan je manji hemodinamski nesigifikantni perikardni izliv bez značaja. Nakon hirurškog odstranjenja tumora bolesnica je bila bez simptoma i bez perikardnog izliva. **Zaključak.** Tumore desnog srca trebalo bi imati u vidu u diferencijalnoj dijagnostici prilikom pojave neočekivane dispneje kod starijih bolesnika. Transtorakalna ehokardiografija neophodna je i dostupna dijagnostička procedura koja daje pouzdane rezultate u dijagnostici tumora srca, naročito kod bolesnika kod kojih se planira hirurška intervencija. Kod većine bolesnika ova metoda omogućava dobru vizualizaciju tumora i definiše njegove odnose sa ostalim strukturama srca.

### Ključne reči:

miksom; srce, pretkomora; dispneja; stare osobe; dijagnoza; hirurgija, kardijalna, procedure; lečenje, ishod.

### Introduction

Based on data from 22 large autopsy studies, the incidence of primary tumors of the heart at 731,309 autopsied was 157 or 0.02%<sup>1</sup>. In the Mayo Clinic series, that was con-

ducted in a period 1954–1979, which including 23,673 patients, primary tumor of the heart was diagnosed in 0.17% of patients. Myxoma was present in 28 patients, 17 were localized in the left atrium and 4 in the right atrium<sup>2</sup>. In Military Medical Academy in Belgrade, overall 63 patients with

myxoma of the heart were diagnosed and treated from 1961 up to date. Among those patients, 12 had myxoma in the right atrium<sup>3</sup>. Myxoma can be asymptomatic, but clinical presentation may include nonspecific signs and symptoms: arthralgia, body weight loss, anemia, hypergammaglobulinemia, elevated erythrocyte sedimentation rate or dyspnea<sup>4</sup>.

Pulmonary embolism as a right atrial myxoma complication is accompanied with dyspnea, hemoptysis, syncope, chest pain, right heart failure or sudden death<sup>5</sup>. Transthoracic echocardiography is a standard diagnostic procedure in revealing myxomas, and cardiac surgery procedure in most cases is the treatment of choice.

### Case report

A 77-year-old female presented with 4 month lasting symptoms of progressive dyspnea on exertion with palpitations and gradual reduction in exercise tolerance and with symptoms of heart failure, NYHA class II/III. Chest pain and orthopnea were not present. The patient had a previous history of hypertension and diabetes mellitus type II.

The patient was dispnoic on the first clinical examination, and auscultation showed weakened respiratory sounds in the basal portions of the lungs. Cardiac auscultation was unremarkable, with normal blood pressure (BP) 110/70 mmHg. The patient had 5 cm hepatomegaly and bilateral pedal edema. Electrocardiogram showed a reduced amplitude of the R-wave in precordial leads (Figure 1).

Hematological and biochemical investigations demonstrated normal hemoglobin of 142 mg/L, a mildly elevated erythrocyte sedimentation rate (23 mm/h). Brain natriuretic peptide was not measured. Chest radiography showed a distorted right atrial border with small bilateral pleural effusion (Figure 2). Transthoracic echocardiography (TTE) showed a huge mobile mass, of 77 × 44 mm, occupying the entire right atrium. The mass was connected to the lower portion of the interatrial septum, and protruding with its floating part through the tricuspid valve into the normal-sized right ventricle (Figures 3 and 4). The inferior vena cava was dilated with minimal collapse on inspiration suggesting raised right

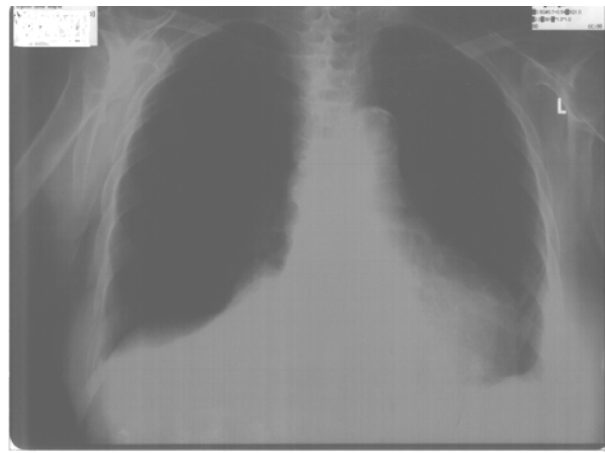


Fig. 2 – Chest radiography showing a distorted right atrial border with small bilateral pleural effusion.



Fig. 3 – Transthoracic echocardiography (TTE) – 4 chamber view with a small pericardial effusion behind the posterior left ventricular wall and in front of the right ventricle (white arrows) and massive myxoma (black arrow).

atrial pressure. The left atrium, left ventricle, mitral valve, aortic valve, and pulmonary valve all appeared normal. Insignificant circular pericardial effusion of 10 mm, was also seen in TTE. Doppler ultrasonography of the major arteries

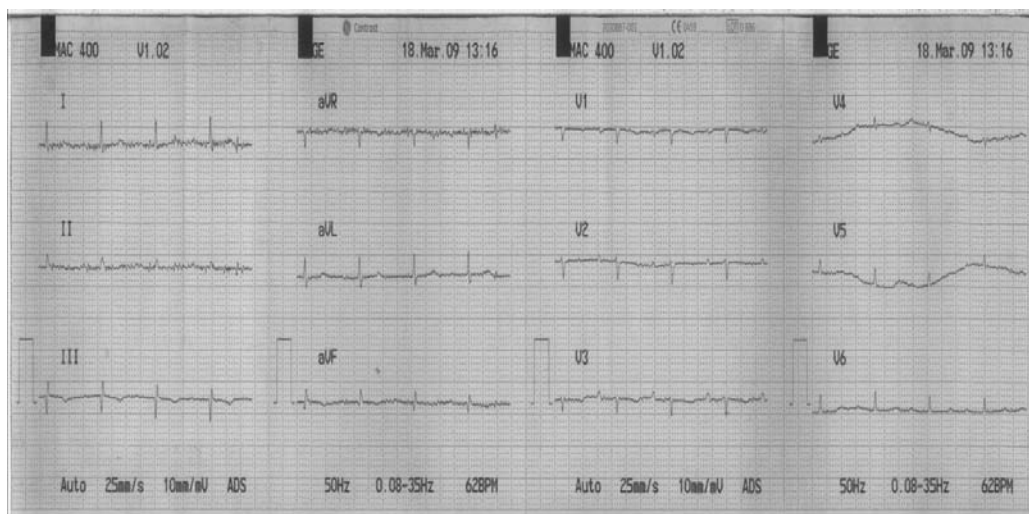


Fig. 1 – A reduced R-wave amplitude in precordial leads in electrocardiogram.



**Fig. 4 – Transthoracic echocardiography (TTE ) short axis parasternal view, with a small circular pericardial effusion behind the posterolateral wall and a bigger one in front of the right ventricle.**

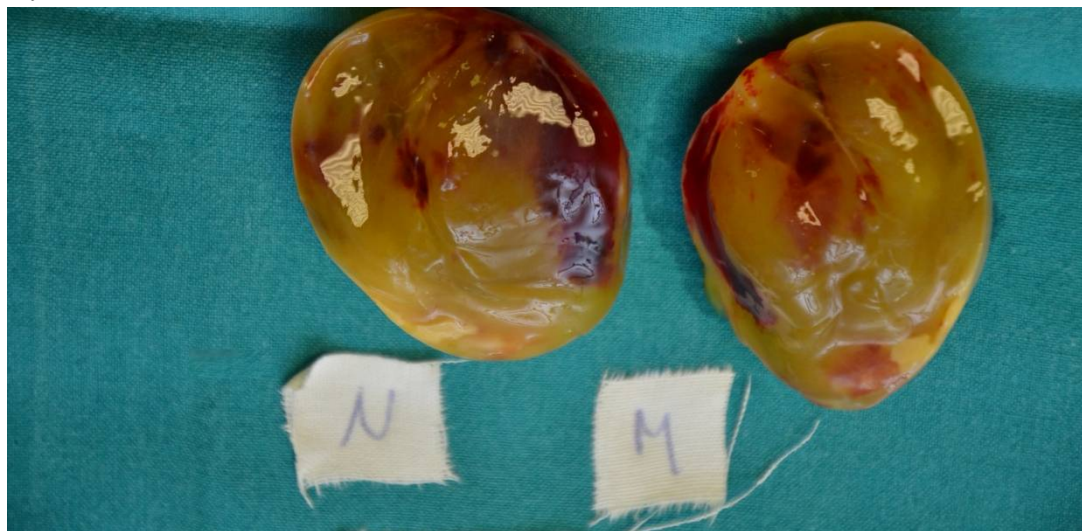


**Fig. 6 – Transthoracic echocardiography (TTE) after a year: the normal right atrium and ventricle, with no tumor, nor pericardial effusion.**

in the neck showed hemodynamically insignificant atherosclerotic changes, and on selective coronary angiography, only mild insignificant atherosclerosis was seen.

At surgery, during cardioplegia and with extracorporeal blood flow, right atrium was open and the tumor on a thin stalk connected to interatrial septum was identified. It filled the entire right atrium protruding with its floating part through tricuspid valve into the majority right ventricular space. The tumor was extirpated together with the connection part of endocardia.

Pathologically, the macroscopic specimen showed a nodular formation measuring  $77 \times 43 \times 34$  mm and containing hemorrhagic areas (Figure 5). Benign myxoma was confirmed histologically, but no residual tumor at the base of the stalk.



**Fig. 5 – Macroscopic longitudinal tumor cut.**

At 12-month follow-up, the patient's exertional capacity much improved, free of other preoperative signs and symptoms. TTE after 12 month showed normal echocardiogram without signs of tumor recurrence, tricuspid insufficiency or dilatation of the right heart cavities (Figure 6).

### Discussion

Right atrial myxoma is extremely rare, and accompanied with pericardial effusion has not been published so far. Three-quarters of neoplasms of the heart are benign tumors, and among them the most common is myxoma, representing about 50% of all primary tumors of the heart. The largest series of patients have shown that females, with the average age of 50 years, are more commonly involved with this type of tumors<sup>1</sup>.

The presented patient had myxoma in a less typical localization – the right atrium. The patient presented with the symptoms in her elderly age of 77 years. Although the oldest described patient with myxoma was 95 years old, these tumors are extremely rare after the age of 60<sup>2</sup>.

Our hospital has been treated a large series of patients for tumors of the heart since 1961. The presented patient was the oldest in the group of 63 patients within more than 50 years long history of the treatment of patients with heart tumors in the Military Medical Academy in Belgrade<sup>3</sup>. In 75%

of cases myxomas are located in the left atrium, while 23% in the right atrium. The remaining myxomas are of the ventricular origin<sup>6</sup>.

The average tumor size at the time of diagnosis is about 50–60 mm<sup>2</sup>. It remains unclear whether the tumor size is directly related to the presence of symptoms, but it has been reported that signs and symptoms usually occur with the minimal tumor size of 50 mm<sup>2, 7</sup>. The presented patient's myxoma measured 77 mm in diameter.

The clinical presentation of patients with myxoma can be quite different. Symptoms of heart obstruction, embolic complications and systemic manifestations are the components of the classical triad, but rarely are present all. However, at least one of the triad symptoms is present.

In a study on 116 patients with left atrial myxoma 65% of patients had signs of intracardiac obstruction, 28% had signs of embolic complications and 33% presented with systemic events. All patients had at least one of the characteristic manifestations of myxoma<sup>8</sup>. In our group of 63 patients with myxoma in any side of the heart, 62% had symptoms and signs of intracardiac obstruction, 26% had signs of embolization, and 30% had systemic symptoms. All patients had at least one component of the classic triad<sup>9, 10</sup>. The presented patient had symptoms and signs of obstructive syndrome of the right heart cavities and dyspnea which is the most common manifestation of myxoma

logged in the right atrium. In addition, the presented patient had a small, hemodynamically insignificant pericardial effusion, which disappeared immediately after surgical removal of the tumor. Pericardial and pleural effusion as a systemic manifestation was previously seen in only one 46-year-old woman with left atrial myxoma. These effusions disappeared soon after the surgical removal of the tumor<sup>11</sup>. The most common differential diagnostic problem after discovering a mass formation in the heart cavities is to differentiate myxoma from the thrombus. The morphology and motility of myxoma and thrombus may be similar and difficult to distinguish by echocardiography, and surgical excision may be necessary for certain diagnosis<sup>4</sup>.

### Conclusion

Right atrial myxoma is extremely rare, and accompanied with pericardial effusion has not been published so far. Tumors of the right heart have to be considered in the differential diagnosis of unexplained dyspnea in elderly patients. Transthoracic echocardiography is certainly necessary and most available diagnostic tool that can be of great help in diagnosing heart tumor, as well as planning cardiac surgery, as in most cases it provides excellent visualization of the tumor and its relationship with other parts of the heart.

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