

# Pulmonary Embolism Due to the Right Atrial Myxoma

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## ABSTRACT

A 47-year-old man was admitted to the hospital with a pleuritic pain, dyspnea, nonproductive cough and low-grade fever. An ECG documented a sinus tachycardia with S<sub>1</sub>Q<sub>3</sub>T<sub>3</sub> pattern and incomplete right bundle branch block, and lung scintigraphy showed multiple perfusion defects. The initial diagnosis was pulmonary embolism. Echocardiography, undertaken before application of the anticoagulant therapy because of hematological disturbances reflecting possible coagulopathy (elevated erythrocyte sedimentation rate, increased leukocyte count, decreased platelet count), revealed a large mobile tumor in the right atrium. Tumor was surgically removed, and histological findings was supported a diagnosis of the cardiac myxoma. The right cardiac myxoma should be considered in the differential diagnosis of pulmonary embolism, particularly in cases presented in conjunction with constitutional symptoms and/or hematological disturbances. In these patients echocardiography should be undertaken early to exclude the rare but treatable diseases of the right heart.

**Key words:** echocardiography, myxoma, pulmonary embolism

## Introduction

Myxoma is the most common cardiac tumor<sup>1,2</sup>. Despite its benign pathologic nature, catastrophic results can occur because of systemic or pulmonary embolism and/or intracardiac obstruction<sup>1–3</sup>. In some cases myxoma can be accompanied with nonspecific extracardiac symptoms and hematological disturbances that fail to suggest the proper etiology<sup>4,5</sup>. We report a case of pulmonary embolism and hematological disorder secondary to a large right atrial myxoma.

## Case Report

A 47-year-old man was admitted to our hospital with a right-sided pleuritic chest pain, dyspnea, nonproductive cough and low-grade fever. Dyspnea and nonproductive cough had appeared 8 months previously but the severity and frequency of symptoms worsened in the previous 2 weeks, especially in exertion and supine position. In addition to these symptoms he has become weak and easily fatigued. He has no conventional cardiovascular risk factors.

On physical examination patient was feverish (37.6 °C) and tachypnoic (a respiratory rate of 28/min). There was no jugular venous distention. Peripheral pulse examination revealed normal carotid and extremity arterial pulsation. His arterial pressure was 140/85 mmHg, and pulse 110 beats per minute. Cardiac auscultation revealed regular rhythm, and no murmurs or extra heart sounds. Lungs were clear to auscultation. There was no lower extremity edema or evidence of peripheral venous thrombosis.

An ECG documented a sinus tachycardia (110/min) with S<sub>1</sub>Q<sub>3</sub>T<sub>3</sub> pattern, and incomplete right bundle branch block. Chest radiography demonstrated mild cardiomegaly. Radionuclide lung perfusion scan showed multiple moderately sized segmental and subsegmental defects in both lungs. The patients erythrocyte sedimentation rate (47 mm/h), total leukocyte count (16.700/mm<sup>3</sup>), C-reactive protein (32.5 mg/L), D-dimers (1375 µg/L), and serum lactate dehydrogenase level (1478 U/L) were increased, while platelet count (83.000/mm<sup>3</sup>) was decreased. Other hematological and biochemical parameters were in the normal limits. In blood gas analysis there were only

slightly decreased pO<sub>2</sub> (8.4 kPa) and O<sub>2</sub> saturation (93%). Antinuclear and anticardiolipin antibodies were negative, and no lupus anticoagulant was detected.

The initial diagnosis was pulmonary embolism. Because of the decreased platelet count and possibility of an underlying coagulopathy, patient was not treated with anticoagulant therapy. Also, patient's medical history and laboratory findings suggested possibility of cardiac myxoma. Therefore, patient was referred to echocardiography.

Transthoracic echocardiography (TTE) using Vivid 3, General Electric, Milwaukee, WI, USA, machine demonstrated normal left ventricular size and function, normal left atrial size, and normal function of the aortic and mitral valves. The right atrium (RA=56 mm) and right ventricle (RV=45 mm) were enlarged. A large mobile mass was identified in the right atrium. There is no pericardial effusion. Transesophageal echocardiography (TEE) using a 5-MHz multiplane imaging transducer revealed a tumors mass in the right atrium, attached with a stalk close to the entry of the inferior vena cava. The tumor extended during diastole through the tricuspid valve into the right ventricle almost to the right ventricular apex causing severe relative tricuspid valve stenosis (Figure 1). Also, a moderate tricuspid regurgitation (angiographic grade +2/+3) was registered by color Doppler. Right ventricular systolic pressure estimated from tricuspid regurgitant jet (RVSP=45 mmHg), as well as shorter pulmonary valve acceleration time (PVaccT = 89 ms), suggested moderate pulmonary hypertension.

Immediate surgical treatment was indicated because of the high risk of recurrent, potentially fatal, embolism. The surgeon completely removed a tumor with the surrounding endocardium. Macroscopically, the resected tumor was a well-defined encapsulated mass with a smooth contour weighted 129 g and measured 98 x 56 x 44 mm in size (Figure 2). Hystologically, the tumor consisted of a

hypocellular mass of a myxoid matrix, rich in acid mucopolysaccharides, with a supporting structure of spindle-like, elongated or stellate cells scattered in an abundant stroma. These findings supported diagnosis of myxoma.

Excision of the tumor resulted in marked symptomatic and hematological improvement. The patient was discharged 10 days after the operation. Six months later the patient was asymptomatic. Follow-up echocardiography showed normal cavities dimensions and ventricular function. However, mild to moderate tricuspid regurgitation (angiographic grade +1/+2) was still evident. Also, there was a slightly elevated pulmonary artery pressure (RVSP=33 mmHg, PVaccT 106 ms). One year later, the patient remains well, with normal echocardiographic finding and pulmonary artery pressure.

### Discussion

Primary cardiac tumors are uncommon with the incidence between 0.0017 % and 0.33%<sup>1,2</sup>. About two-thirds of these tumors are myxoma that typically arises in the left atrium (80%) along the interatrial septum near the fossa ovalis. Occasionally, myxomas arise in the right atrium (15%), the ventricles (3–4%), or valves. Rarely, the tumor is present in more than one cavity<sup>1,2</sup>. Cardiac myxomas arise more frequently in women and usually present between the ages of 50 and 70 years<sup>1,2</sup>. Sporadic cases of myxoma are almost always single. However, approximately 7% of cardiac myxomas are a component of a complex hereditary syndrome that affects multiple organs<sup>1</sup>.

Clinical manifestations of cardiac myxomas are most often determined by tumor size, location and mobility, and can be separated into three sessions: (1) symptoms of mechanical valvular obstruction, (2) embolization to the pulmonary or systemic circulation and (3) nonspecific constitutional symptoms and/or hematological findings<sup>1–5</sup>.

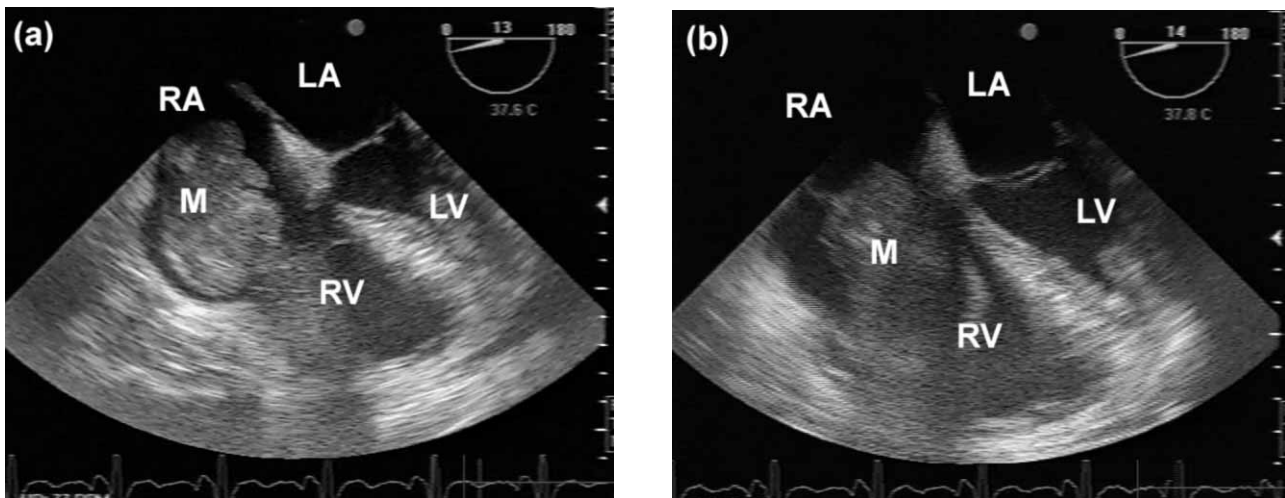


Fig. 1. Midesophageal four-chamber echocardiographic study in (a) systole and (b) diastole, revealing a right atrial (RA) mass (M) with a stalk (arrow) extended through the tricuspid valve into the right ventricle (RV). LA denotes left atrium, LV – left ventricle.

Obstruction to blood flow may occur at the orifice of any valve mimicking the clinical picture of valve stenosis. If the tumor is large enough, soft and easily deformable, and if it has a long stalk, temporary complete valve obstruction may occur, resulting in syncope or sudden death. However, the most frequently obstructive symptom is dyspnea that occurs in approximately 80% of patients with atrial myxoma<sup>1,2</sup>. Left atrial myxoma produces symptoms when it reaches approximately 7 cm in size. Those in the right atrium that produce symptoms are usually approximately twice as large and sometimes several-fold larger<sup>6</sup>.



Fig. 2. The tumor weighed 129 g and measured 98 x 56 x 44 mm in size.

Embolic phenomenon in cardiac myxoma is common, with the incidence ranging from 30% to 40%<sup>1,2</sup>. In left-sided cardiac myxomas the emboli generally display a predilection for the central nervous system, but also can affect other organs such as the liver, spleen, kidney, retina, coronary vessels, abdominal aorta, and distal arterial tree<sup>1,2,5,7</sup>. In right-sided myxomas, clinically evident embolism is uncommon. Nevertheless, in these cases, there have been reports not only of repeated microembolization into the pulmonary vessels with subsequent pulmonary hypertension, but also of lethal pulmonary embolism<sup>3</sup>. The myxoma size is a significant determinant of valvular obstruction and constitutional symptoms, whereas the villous or papillary type and irregular surface is a risk factor for embolism<sup>4,5</sup>. Also, males are statistically at greater risk than females of developing embolic complications<sup>4,5</sup>. Our case is unusual because a large solid myxoma with a smooth surface cause pulmonary embolization.

Constitutional symptoms (fever, weight loss, weakness, fatigue, Raynaud's phenomenon, erythematous rash, digital clubbing, arthralgias) and laboratory findings (anemia, elevated erythrocyte sedimentation rate, elevated leukocyte count, decreased platelet count, positive seroreactive protein, and abnormal serum proteins)

are reported in 10–45% of patients<sup>1,2,5,6</sup>. These symptoms may mimic infective endocarditis, collagen vascular disease, or occult malignancy, and may be due to the release of the acute phase reactant interleukin-6 from the tumor leading to inflammatory and autoimmune manifestations<sup>5</sup>. Plasma interleukin-6 concentration correlated positively with both the myxoma size and constitutional symptoms<sup>4,5</sup>. Although nonspecific constitutional symptoms and laboratory findings are most often in patients with left atrial myxoma<sup>4</sup>, our case serves to remind the clinician that these symptoms can also be found in patients with a right-sided myxoma.

Cardiac myxomas range from small (< 1 cm or < 10 g) to large (> 10 cm or > 100 g)<sup>1,5</sup>. Lazarides was reported a right atrial myxoma that weighed 450 g<sup>6</sup>! However, how fast atrial myxomas grow has never been clarified. It was estimated that recurrent atrial myxomas grow an average of 0.24–1.6 cm per year<sup>1</sup>. Therefore, the large tumors size in our patient assumed that this intracardiac pathology existed for some years prior to diagnosis.

Before the introduction of echocardiography, the time interval between the onset of symptoms and diagnosis was 5.5–12 months and there has been a trend toward shortening of that interval to approximately 3 months<sup>8</sup>. Although the incidence of myxoma did not change between the early decade and the latest 10 years, small-sized and asymptomatic myxomas were more frequently found during the later decade<sup>8</sup>. It can be explained by the development and widespread introduction of echocardiography. However, as in our patient, many patients with myxoma experience a significant delay in diagnosis that can be attributed to the absence or misleading of cardiac symptoms, or to the presence of extracardiac symptoms that fail to suggest the proper etiology.

Although it is usually possible to detect intracardiac tumors with TTE the TEE examination produces spectacular images of a small tumors (1 to 3 mm in diameter), especially in patients with poor TTE images, and makes diagnosis, particularly of atrial masses, relatively easy<sup>9</sup>. The TEE also permits a clearer picture of the attachment or stalk of the tumor and more precise characterization of the size, shape, and location of the mass<sup>10</sup>. However, although TEE is a semi-invasive diagnostic technique with a very low incidence of significant complications, catastrophic pulmonary embolism during TEE examination it has been reported<sup>11</sup>.

In cases in which the echocardiography characterization of intracardiac mass is incomplete, cardiac magnetic resonance imaging (CMR) and multislice spiral computed tomography (MSCT) are particularly helpful in determining the relationship to normal intracardiac structures and tumor extension to adjacent vascular and mediastinal structures, infiltration into the pericardium, influence on cardiac function and surgical planning<sup>12,13</sup>. Additionally, superior to echocardiography, CMR and MSCT could strengthen the diagnostic accuracy by additional information on tissue characterization using different imaging sequences<sup>12,13</sup>. These two imaging techniques can differentiate tissue composition, making it

possible to identify solid, liquid, hemorrhagic, and fatty, space-occupying tumors. Typically for cardiac myxomas, contrast enhancement is moderate and delayed enhancement can be found in the outer circumferential tumor margins only<sup>12</sup>.

The accepted treatment of cardiac myxoma is operative excision<sup>14</sup>. Surgical removal of a tumor is important for preventing valvular obstruction, eliminating systemic or pulmonary emboli, maintaining systolic function, and restoring biventricular diastolic function. Surgical treatment leads to complete resolution with low rates of recurrence and good long-term survival<sup>14</sup>. The overall risk of recurrence is about 1–3% for sporadic myxoma of-

ten because of inadequate resection<sup>1,14</sup>. Annual TTE or TEE review is suggested for a period of 3 to 4 years when the risk of recurrence is greatest. No chemotherapeutic or radiotherapeutic approach has been shown to be effective in preventing the recurrence of myxoma.

In conclusion, the right cardiac myxoma should be considered in the differential diagnosis of pulmonary embolism, particularly in cases presented in conjunction with constitutional symptoms and/or hematological disturbances. In these patients echocardiography should be undertaken early to exclude the rare but treatable diseases of the right heart.

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## PLUĆNA EMBOLIJA UZROKOVANA MIKSOMOM DESNE PREDKLIJETKE

### SAŽETAK

47-godišnji muškarac je hospitaliziran zbog desnostrane pleuritične boli, zaduhe, neproduktivnog kašlja i blago povišene tjelesne temperature. EKG-om je potvrđena sinusna tahikardija sa S<sub>1</sub>Q<sub>3</sub>T<sub>3</sub> obrascem i nepotpuni blok desne grane, a scintigrafijom pluća su pokazani višestruki ispadi perfuzije. Početna dijagnoza je bila plućna embolija. Ehokardiografskim pregledom učinjenim prije primjene antikoagulantne terapije, zbog hematoloških otklona koji su ukazivali na moguću koagulopatiju (ubrzana sedimentacija eritrocita, leukocitoza, trombocitopenija), potvrđen je veliki, pomični tumor u desnoj predklijetki. Tumor je kirurški odstranjen, a histološki nalaz je potvrdio dijagnozu miksoma. U diferencijalnoj dijagnozi plućne embolije, osobito ukoliko je praćena i konstitucionalnim simptomima i/ili hematološkim otklonima, potrebno je razmatrati i miksom desne predklijetke ili klijetke. U tih je bolesnika potrebno učiniti rani ehokardiografski pregled kako bi se isključile rijetke ali izlječive bolesti desnih šupljina srca.