sciendo
Internal Medicine 2018 vol. XV No. 4 - www.srmi.ro
10.2478/inmed-2018-0028

GIANT LEFT ATRIAL MYXOMA CAUSING MITRAL VALVE OBSTRUCTION AND PULMONARY HYPERTENSION. A CASE REPORT

Pașc Priscilla[1,2], Ioana Alexandra Coțe[1], Mircea Ioachim Popescu[1,2]
[1]Clinical County Emergency Hospital of Oradea
[2]Faculty of Medicine and Farmacy of Oradea, University of Oradea
Correspondent author: priscilla_pasc@yahoo.com

Abstract
Cardiac myxoma is the most common adult cardiac tumor, with an incidence of 1: 1,000,000 in the general population. Usually occurring between 4-6 decades of life, especially in women, most of the myxomas appear sporadically and are of unknown etiology. Rare cases are genetically determined. Sudden death can occur in 15% of cases. While papillary tumors often complicate coronary or systemic thromboembolism, solid tumors, ovoids, cause heart failure by obstructing the mitral orifice. From a clinical point of view, the atrial myxoma may mimic a valvulopathy, heart failure, dilatation cardiomyopathy, bacterial endocarditis, and may cause heart rhythm disorders, syncope, myocardial infarction and systemic or pulmonary thromboembolism. I will present the case of a giant atrial myxoma, intermittent obstructive of the mitral orifice, evolving towards global heart failure. The lack of severe acute complications during the most likely long-lasting evolution represents an undiscovered curiosity.
Keywords: *cardiac myxoma, mitral valve obstruction, pulmonary hipertension, heart failure.*

Rezumat
Mixomul cardiac reprezintă cea mai frecventă tumoră cardiacă a adultului, cu o incidență de 1:1.000.000 în populația generală. De obicei apărute între decadele 4-6 de viață, mai ales la femei, majoritatea mixoamelor apar sporadic și sunt de etiologie necunoscută. Rare cazuri sunt determinate genetic. Moartea subită poate apărea în 15% dintre cazuri. În timp ce tumorile papilare se complică adesea cu trombembolismul coronarian sau sistemic, tumorile solide, ovoide, determină insuficiență cardiacă prin obstruarea orificiului mitral. Din punct de vedere clinic, mixomul atrial poate mima o valvulopatie, insuficiență cardiacă, cardiomiopatie dilatativă, endocardită bacteriană și poate determina tulburări de ritm cardiac, sincope, infarct miocardic și trombembolism sistemic sau pumonar. În continuare, voi prezenta cazul unui mixom atrial gigant, obstructiv intermitent al orificiului mitral, cu evoluție spre insuficiență cardiacă globală. Lipsa complicațiilor acute severe pe parcursul evoluției cel mai probabil îndelungate reprezintă o curiozitate încă nedeslușită.
Cuvinte cheie: *mixom cardiac, obstrucție a valvei mitrale, hipertensiune pulmonară, insuficiență cardiacă.*

Internal Medicine

Clinical cases

Introduction

According to literature data, primary cardiac tumors constitute a rarely encountered pathology, with an incidence ranging from 0.0017% to 0.19% in the general population. Approximately 75% of these tumors are benign, most of them being myxomas. Usually occurring between 4-6 decades of life, especially in women, most of the atrial mixes appear sporadically and are of unknown etiology. Rarely, they are genetically determined and autosomal dominantly transmitted, for example, in Carney's Syndrome. Family myxoma is usually found in young men, with an average age of 21 years. Cardiac myxomas were also described in the child, where it occurs at a frequency of 6% of the child's benign heart tumors.

In terms of location, they often develop in the left atrium, only in rare cases they are detected in the right atrial cavity. Most are solitary and pediculated and have frequently as the attachment site the edges of the foramen ovale, although they can adhere to the wall of the atrium or posterior atrial. Mixes vary in size, from 1 cm to over 10 cm, with a rate of growth as variable. Cases with a growth rate of 1 cm / month were identified.

From a clinical point of view, the atrial myxoma symptomatology is polymorphic and may mimic a number of other cardiac conditions such as valvulopathy, heart failure, dilatation cardiomyopathy, bacterial endocarditis, atrial thrombosis. For this reason, the left atrium and ventricle myxoma should be differentiated especially by mitral stenosis, mitral failure, cardiomyopathy, subvalvular and valvular aortic stenosis.

Sudden death can occur in 15% of cases. While papillary tumors often complicate with coronary or systemic thromboembolism, solid tumors, ovoids, cause heart failure by obstructing the mitral orifice. At the time of diagnosis, over 50% of the patients found have symptoms of congestive heart failure. Other possible complications may be heart rhythm disorders, syncope, myocardial infarction.

Two-dimensional echocardiography is the diagnostic investigation. The treatment is surgical. Although atrial myxomas are benign, local recurrences due to inadequate resection or malignancy are possible. Occasionally, myxomas relapse away from the initial site, the explanation for this phenomenon being found in tumor intravascular microembolization. The risk of recurrence is greater in cases of familial myxomas.

Presentation of the case

I will present the case of a 74-year-old man with no cardiovascular history with bilateral

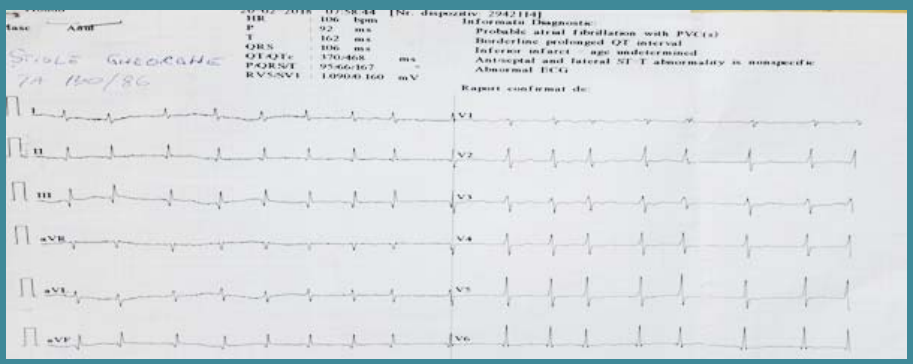

Figure 1. *The electrocardiogram performed performed at hospital admission*

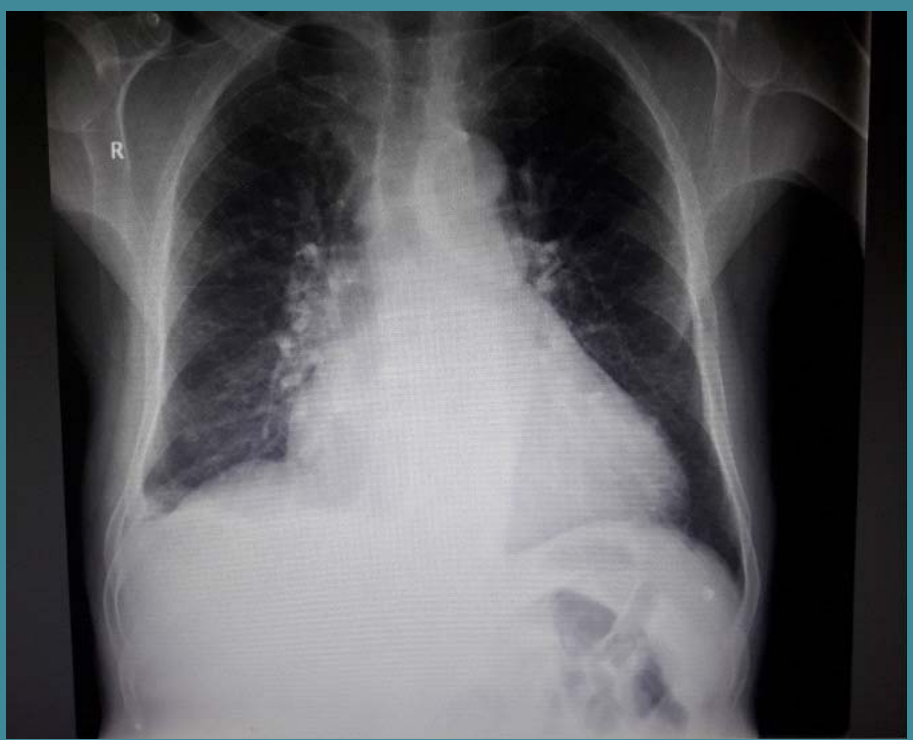

Figure 2. *Pulmonary radiography*

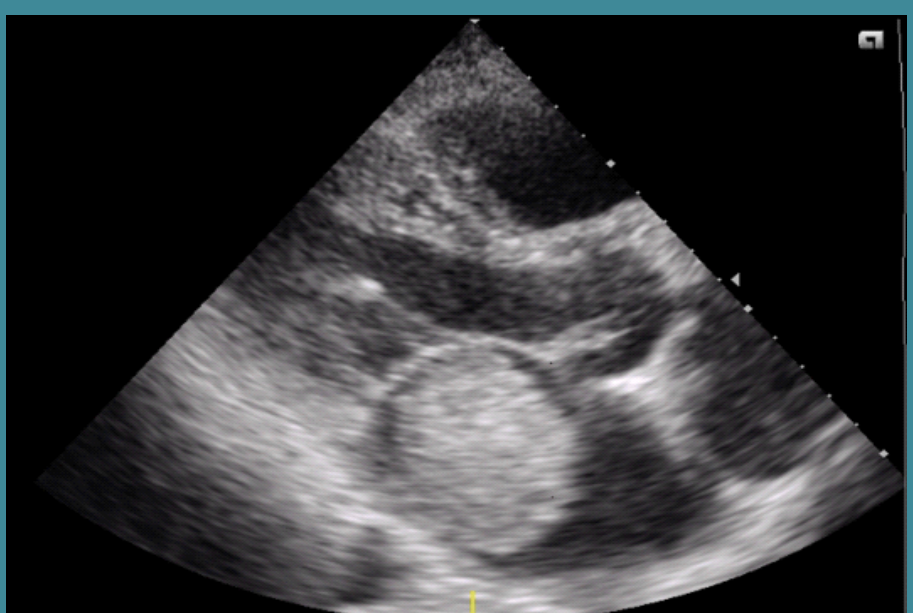

Figure 3. *Echocardiography performed at the admission-parasternal long axis section*

Internal Medicine

Clinical cases

congenital blight who is presenting in the emergency room for phenomena of global heart failure manifested by dyspnea with orthopnea, decreased tolerance to effort, fatigue, edema of lower limbs, oliguria.

The general objective examination reveals an altered general condition, important edema of the lower limbs, increased abdomen in volume, diffuse diffuse rallies in the respiratory system exam. The targeted examination of the cardiovascular system detects tahiaritmic heartbeats, sistolic murmur at the cardiac apex and a blood pressure of 130/81 mmHg.

Laboratory tests reveal an increased NTproBNP that advocates cardiac decompensation, accompanied by a mild hepatocytosis syndrome (due to liver blood stasis), renal impairment and mild electrolyte disturbances.

Emergency **electrocardiogram** records atrial fibrillation, intermediate ORS axis and without significant ST-T changes (figure 1).

Pulmonary radiography, recommended in patients with suspected heart failure to exclude lung pathology, highlights a slightly increased bilateral peribronhovascular drawing, minimal basal pleural effusion (figure 2).

Echocardiography on admission reveals the presence of a polilobate left atrial myxoma (figure 3, 6) attached to the lateral wall of the left atrium(figure 5), below the posterior mitral valve, of large size, reaching 4/5 cm in diameter, which prolates intermittently through the mitral valve with obstruction of the intracardiac blood flow during diastole(figure 4). Also, the left and right atrium dilated, the left ventricle is undilated but with a moderately depressed ejection fraction (33%). Continuous wave Doppler reveals a third degree tricuspid regurgitation with 2 jets and a moderate secondary pulmonary hypertension.

The investigations are completed with abdominal ultrasound showing the presence of small amount of ascites (perihepatic, perisplenic fluid , also in Morrison space).

As for the treatment followed, the patient is treated with diuretics at increased doses, resulting in increased diuresis and decreased body weight, clinically, with improvement in dyspnea and the remission of edema. In addition, digital drugs are associated for cardiac frequency control, and anticoagulants (low molecular weight heparin), as the patient has a moderate-high risk of stroke (ChadsVasc 3, Hasbleed 2).

After stabilizing the patient, he is transferred to the cardiovascular surgery clinic for surgical treatment. The myxoma is approached by a transeptal technique and is removed (figure 7, 8, 9). Durrring the same procedure, the tricuspid valve repair is done using “De Vega” technique (figure 10). The histopathological examination confirms the diagnosis of the

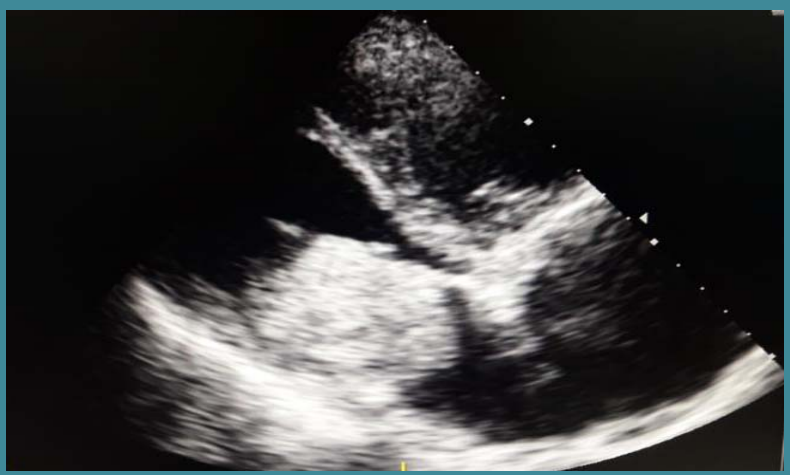

Figure 4. *Echocardiography- parasternal long axis section, showing the myxoma prolabating through the mitral orifice*

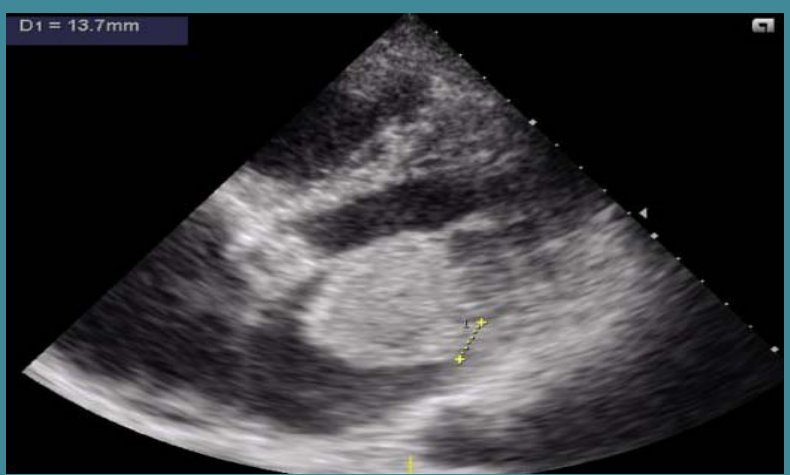

Figure 5. *Echocardiography- apical section- indicates the attachment site to the lateral wall of the left atrium through a pedicle that measures 13,7 mm*

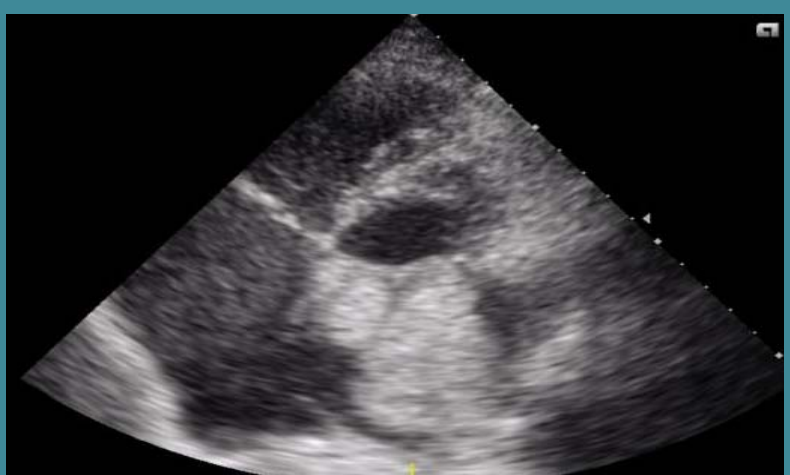

Figure 6. *Echocardiography - apical section - highlights the polilobate form of the myxoma*

myxoma. At subsequent medical checkups, the patient is clinically ameliorated, with increasing effort capacity and increasing quality of life. Echocardiography reveals a slightly improved ejected fraction.

Discussions

Atrial myxomas are the most common primary cardiac tumors. Because of polymorphic and non-specific symptoms, diagnosis is a challenge for the cardiologist. The left atrial mixem can be completely asymptomatic in the early stages. In advanced stages, however, the clinical findings of cardiac mixes are characterized by the triad formed by obstruction of intracardiac blood flow, embolism and systemic manifestations.

Congestive heart failure is the most common form of clinical presentation. Depending on size, location, and characteristics, represented by the shape and type of implantation, the cardiac myxoma may cause changes in cardiac activity. The main mechanism underlying these disorders is considered to be the obstruction of the intracardiac blood flow produced by the tumor.

The mobility and size of the cardiac myxoma are characteristics that condition the intracardiac obstructive phenomena. The left atrial location, which is the most common, affects the functioning of the mitral valve apparatus and generates stenosis or mitral insufficiency, creating the conditions of atrial and pulmonary pressure-overloading. Thus, dyspnea is the dominant symptom that appears the earliest, initially at the effort, then becoming constant.

The case presented is a special one from the point of view of its clinical similarity with a closely related mitral stenosis with natural evolution to heart failure. Clinical-pathological correlations have shown that the effects of mitral stenosis occur when the diameter of the tumor exceeds 5 cm. The giant dimensions of the myxoma, which almost completely occupies the left atrium, almost completely obstructs the mitral orifice during diastole, when it prolapses through the mitral valve. Its long pedal allows for extensive movement, favoring obstruction of intracardiac blood flow. Secondary, increases backward the pressure of the intracardiac cavities, causing progression to dilatation of the left atrium, then to the right cavities, eventually with the occurrence of congestive heart failure phenomena.

Factors favoring embolism in cardiac myxomas depend on both the tumor itself and the particularities of cardiac function. It has been found that some myxomas are more emboligenic than others, depending on the characteristics of the tumor. The soft, friable, with small implantation base (pediculous tumors) and with perivalvular low septal

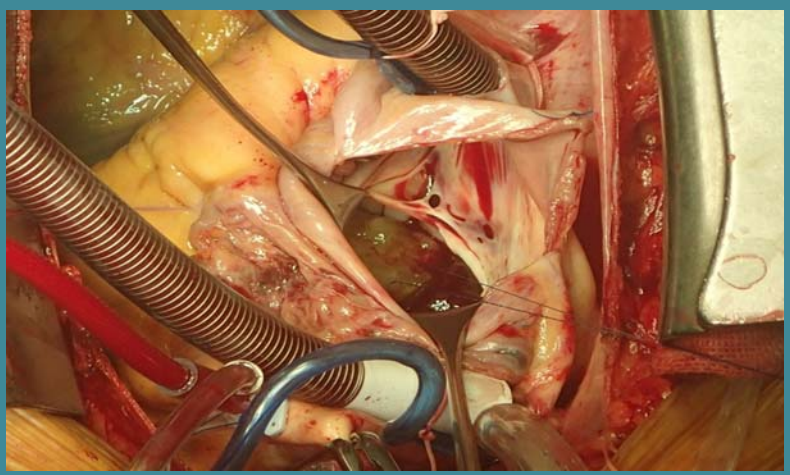

Figure 7. *Transeptal approach of the myxoma*

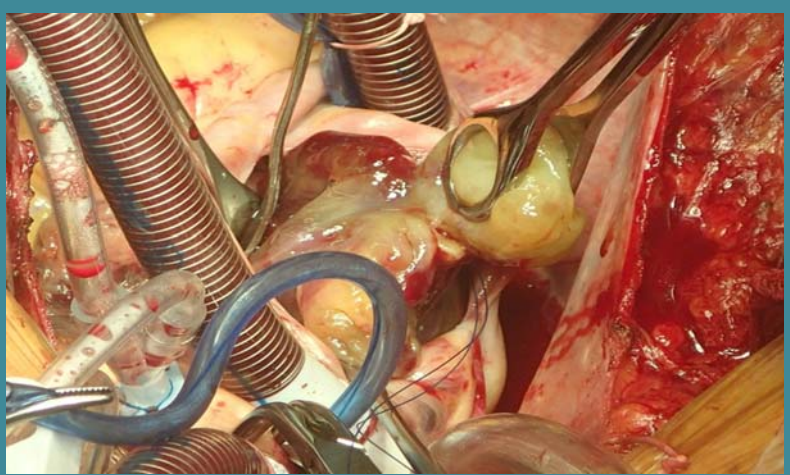

Figure 8. *Surgical removement of the cardiac myxoma*

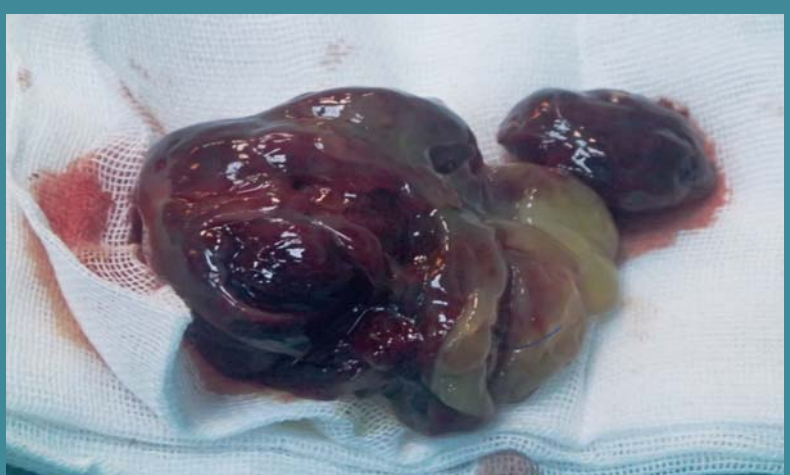

Figure 9. *Surgical resection piece showing the polilobate form of the myxoma*

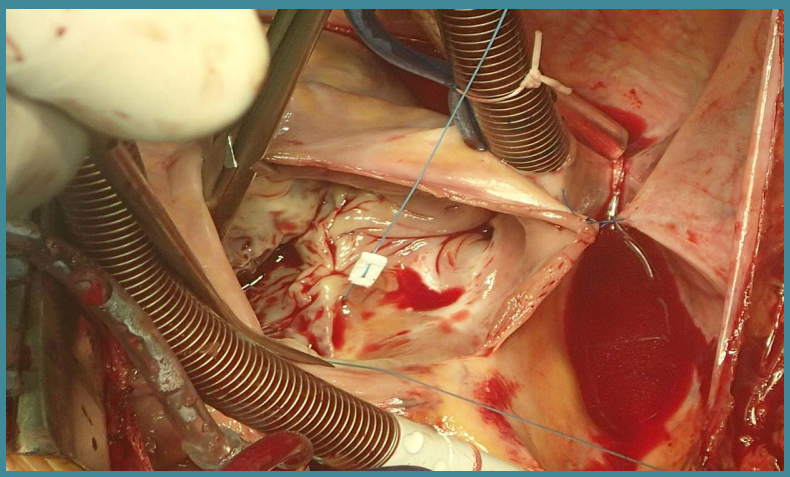

Figure 10. *Tricuspid annuloplasty using De Vega technique*

localization allowing large amplitude movements is most frequently complicated with cerebral or systemic embolization. Embolic events more frequently affect cerebral arteries, including retinal arteries. Embolisms in the pulmonary arteries, causing pulmonary infarction or acute pulmonary heart disease, in the visceral, renal or coronary arteries, manifested by angina or infarction, are also possible. The multiple embolism or the detachment of the entire tumor mass and the passage into the circulatory torrent are serious gravity situations with fatal prognosis in the short term. In our case, a curiosity of the case is the lack of severe acute complications during the most likely long evolution. No other episodes of sudden health alterations that require hospitalization have been reported ,as the patient presented for the first time in his life in the emergency room.However, admission revealed an atrial fibrillation, most likely secondary to the dilatation of the excessively overloaded left atrium. Studies in this regard have shown an incidence of approximately 20% of atrial fibrillation associated with large atrial myxomas.

Extremely diverse general manifestations, such as fatigue, fever, weight loss, night sweats, arthralgia, are supposed to be secondary to pulmonary hypertension, haemolytic anemia or systemic

microembolies. Early echocardiography plays an essential role in the diagnosis and clinical management of these patients. It allows differential diagnosis with valvular diseases and can estimate the transvalvular gradient.

Only in the case of left atrial myxomas, the transmitral blood flow is reduced, and only the large myxomas may resemble severe mitral stenosis. In this case, echocardiography is essential, and can highlight the presence of intracavitary masses. It can also make differential diagnosis with other heart tumors or intracardiac thrombi. In addition, it allows to highlight indirect signs of pulmonary hypertension.

Surgical treatment is usually curative, as in our case, the patient underwent cardiac surgery with a good result. Recurrent myxoma is rare, between 1% and 5% of patients undergoing surgery develop tumor recurrence.

There are very rare cases published in the literature with two recurrences, which generally occur 15-20 years after the first intervention. However, the rarity of these examples is not an argument for a superficial postoperative follow-up, on the contrary, it advocates the requirement of regular echocardiographic observation of these patients, throughout their lives.

References

1.Najat N Mouine, Ilyass I Asfalou,Giant left atrial myxoma mimicking severe mitral valve stenosis and severe pulmonary hypertension, Int Arch Med. 2013; 6: 13.

2.Praneet Iyer, Myo Myo Aung, A case of large atrial myxoma presenting as an acute stroke, J Community Hosp Intern Med Perspect. 2016;

3.Maryam Sotoudeh Anvari, Mohammad Ali Boroumand , Histopathologic and Clinical Characterization of Atrial Myxoma: A Review of 19 Cases, 01 October 2009, Laboratory Medicine, Volume 40, Issue 10, 1 October 2009, Pages 596599

4.Deva Japa, Mahpaekar Mashhadi, Giant left Atrial Myxoma Induces Mitral Valve Obstruction and Pulmonary Hypertension, J Clin Diagn Res. 2016 Jan; 10(1): ED08ED09.

5.Reynen K: Cardiac myxomas. N Engl J Med 1995, 333:16101617.

6. Swartz MF, Lutz CJ. Atrial Myxomas: Pathologic Types, Tumor Location and Presenting Symptoms. J Card Surg. 2006;21(4):43540.