Romanian Journal of Cardiology | Vol. 27, No. 4, 2017

CASE PRESENTATION

Large left atrial myxoma with left ventricle inflow obstruction

Teodora Nita[1], Paula Gocan[1], Ioana Dregoesc[1], Svetlana Encica[2], Lucian Stan[3], Ruxandra Beyer[1]

Abstract: Cardiac myxomas are rare benign tumors generally located at the border of fosa ovalis in the left atrium. Clinical presentation may vary and overlap with other cardiovascular disease, so echocardiography is necessary for diagnosis. We describe the clinical case of a 58-year-old woman with chronic exertional dyspnea diagnosed with left atrial myxoma based on transthoracic echocardiography. Severe mitral valve obstruction and regurgitation by the large tumoral mass had important hemodynamic consequences leading to severe pulmonary hypertension. Complete surgical resection was performed and histopathological examination confirmed the diagnosis.
Keywords: left atrial myxoma, dyspnea, echocardiography, mitral valve

Rezumat: Mixoamele cardiace sunt tumori benigne rare, situate în general la nivelul fosei ovale, în atriul stâng. Prezentarea clinică poate varia și se poate suprapune cu alte boli cardiovasculare, astfel încât ecocardiografia este necesară pentru diagnostic. Prezentăm cazul clinic al unei femei de 58 de ani cu dispnee la efort, diagnosticată cu mixom atrial stâng ecocardiografic. Obstrucția severă la nivelul valvelor mitrale și regurgitarea mitrală produse de către formațiunea tumorală au avut consecințe hemodinamice importante care au condus la hipertensiune pulmonară severă. S-a efectuat rezecția chirurgicală completă și examen histopatologic care a confirmat diagnosticul.
Cuvinte cheie: mixom atrial stâng, dispnee, ecocardiografie, valvă mitrală

INTRODUCTION

The heart is the source of both primary and secondary tumors. Any form of cancer can determine metastases in the heart, 20 to 40 fold more frequent than primary tumors[1]. About 75 percent of primary tumors are benign, the most frequent are myxomas with typical localization in the left atrium[2]. Other localization of myxomas have been described: right atrium[3], ventricles[4], inferior and superior cava veins[5], Eustachian valve[6], femoral vein[7].

CLINICAL MANIFESTATIONS

Clinical manifestation depend on the size and position of the tumor and can be classified as follows: symptoms due to mechanical obstruction and/or valve involvement, as well as due to the direct invasion of the myocardium (cough, dyspnea, thoracic pain, dizziness, palpitations, syncope, circulatory collapse), constitutional symptoms (weight loss, pyrexia of unknown origin, malaise) and embolic symptoms mainly in the central nervous system[8].

CASE REPORT

A 58 year-old woman with medical history of hypothyroidism presented in the outpatient department for chronic exertional dyspnea. Clinical examination revealed blood pressure 100/80 mmHg, pulse rate 101 bpm and a holosystolic murmur at the apex of the hearth. Auscultation of the lung was normal bilaterally and no jugular vein distention was presented. Pulses were intact bilaterally with no peripheral oedema and there were no neurological deficits. Electrocardiogram revealed normal sinus rhythm with minor right bundle branch block (Figure 1). Transthoracic echocardiogram showed a large echogenic mass of 53/31mm in the enlarged left atrium (54/81 mm) attached to interatrial septum (Figure 2). The tumor area was 16.8 cm^2, homogeneous and mobile with protrusion into

[1] Department of Cardiology, Heart Institute "Niculae Stancioiu", Cluj-Napoca, Romania
[2] Department of Anatomic Pathology, Heart Institute "Niculae Stancioiu", Cluj-Napoca, Romania
[3] Department of Cardiovascular Surgery, Heart Institute "Niculae Stancioiu", Cluj-Napoca, Romania

Contact address:
Teodora Nita, Heart Institute "Niculae Stancioiu", Cluj-Napoca, Romania.
E-mail: caleanteodora@yahoo.com

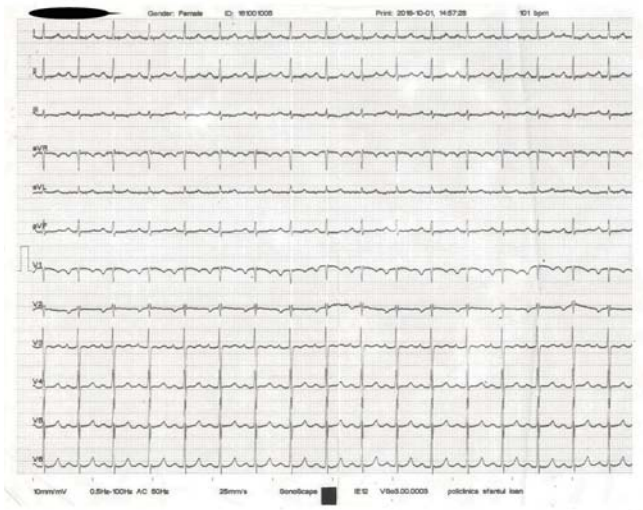

Figure 1. Sinus tachycardia.

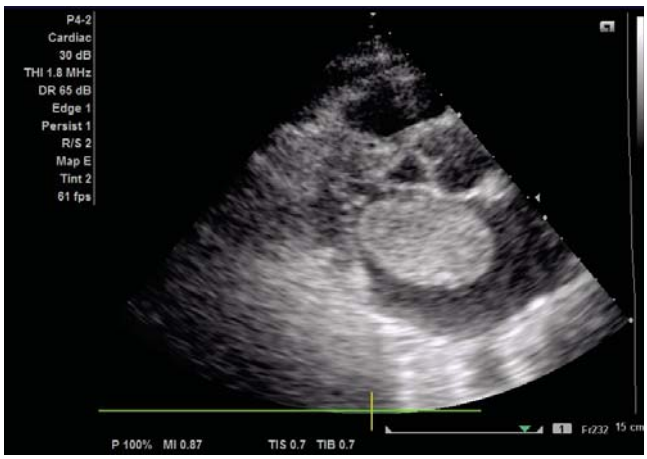

Figure 2. Transthoracic echocardiography shows a large mass filling left atrium (parasternal long axis).

the left ventricle during diastole, causing severe mitral regurgitation and stenosis (Figure 3). The left ventricle ejection fraction was normal. The right ventricle was slightly enlarged, the systolic pulmonary pressure was estimated at 70 mmHg.

The patient was referred to another hospital for further investigations and surgical treatment. On admision, laboratory findings were near normal: WBC 11510/µl, Hb – 13.3 g/dl, Hct – 39.6%, Platelets – 161000/µl, Na – 145 mmol/l, K – 5 mmol/l, Urea – 31.1 mg/dl, Creatinine – 0.75 mg/dl, ALT – 15UI/l, AST – 23UI/l, TSH – 1.45 µUi/ml, FT4 – 0.98 ng/dl, prothrombin time – 12.4 sec. The coronary angiography was normal. The patient was promptly transferred to Cardiovascular Surgery Department and underwent resection of the left atrium tumor. Intraoperative the heart motion was adequate with enlarged left atrium. The tumor was friable, implanted on the interatrial septum, had 60/40 mm diameters, suggesting an atrial myxoma (Figure 4). The atrial wall was reconstructed with a bovine pericardial patch. The diagnosis of myxoma was confirmed by the histological exams (Figure 5 and 6). Early postoperative recovery was good, with removal of the drainages the following day. Five days later the patient presented rapid onset of palpitations, dyspnea with orthopnea. Electrocardiogram showed atrial fibrillation. Conversion to normal sinus rhythm was obtained using intravenous amiodarone. Chest X-Ray reveled moderate right pleural effusion and pleural drainage was made for 24 hours. After 13 days the patient was discharged asymptomatic on beta-blocker, propafenone, diuretics, H2-blocants, levothyroxine and vitamin K antagonists.

DISCUSSION

Clinical presentation, diagnosis and treatment of cardiac tumors can be very challenging.

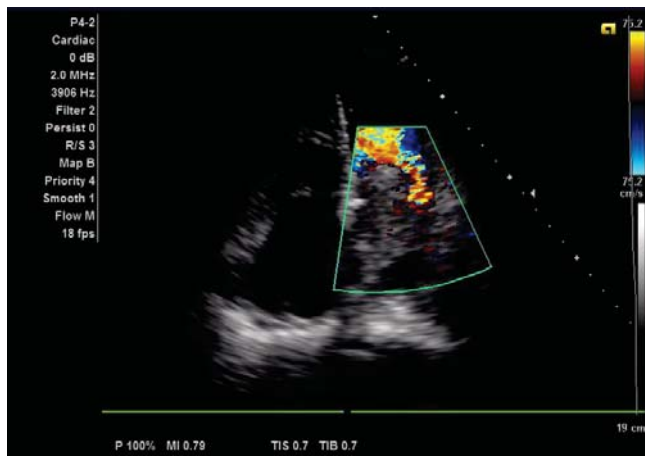

Figure 3. Apical 4 cambers. Left atrial tumor and mitral regurgitation.

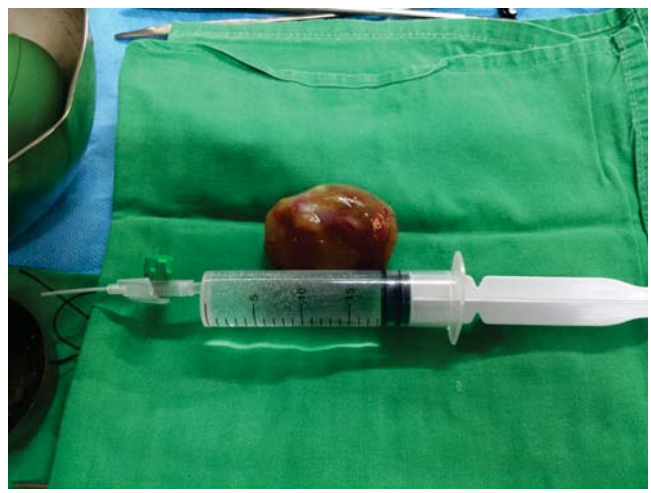

Figure 4. Post-operative view of the tumor.

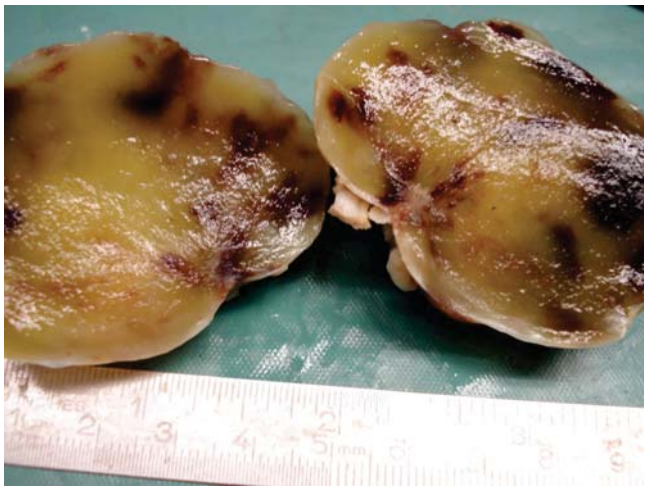

Figure 5. Macroscopic view of myxoma. The tumor is very well delimited, oval-shaped, elastic consistency, smooth surface and brownish yellow with purplish areas color.

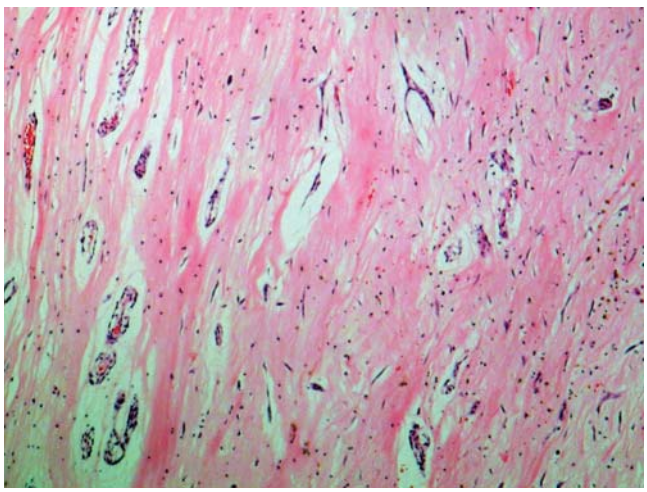

Figure 6. Photomicrograph of myxoma. The fragment is represented by a mixoid tissue which consist of elongated cells with reduced paled eosinophilic cytoplasm and hyperchromatic nuclei. The cells are disposed in groups and rows. The mixoid stroma consists of liniar deposits of calcium and iron (Gandy-Gamna bodies) and reduced lymphocytic inflamatory infiltrate (Hematoxylin and eosin).

In our patient the symptoms were caused by severe pulmonary hypertension due to the prolapse of the tumor in to the left ventricle during diastole with severe mitral obstruction and to the mitral regurgitation. Transthoracic echocardiography (TTE) is the initial test in a patient suspected with cardiac tumor. Left atrial myxomas are usually hyperechoic, well delimited tumors located at the border of the fossa ovalis. Echocardiography identifies the tumor, the implantation base and may be helpful to determine the mobility and the relation with the mitral valve. Transoesophageal echocardiography has better sensitivity and specificity than transtoracic echocardiography for

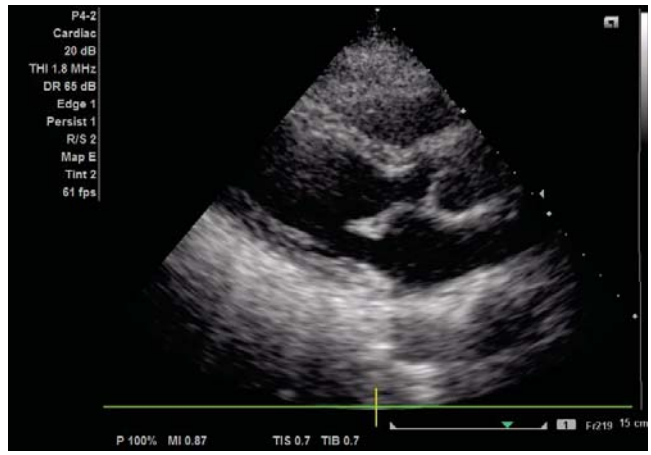

Figure 7. Parasternal long axis.

cardiac myxoma. In our patient, the TTE was sufficient in confirming the diagnosis. Differential diagnosis must be primarily made with thombus. Contrast computed tomography (CT), cardiac magnetic resonance imaging (MRI) and positron emission tomography-CT may be useful in this situation[9]. Once the presumptive diagnosis of atrial myxoma has been made prompt surgical resection is the best option because of the risks of serious complications like embolization or sudden death. Preoperative coronary angiography is indicated in selected patients (coronary artery disease suspected, age over 40 years, etc.) to assess the coronary arteries[10]. Postoperative recovery is usually good. However, early complications can occur. Supraventricular arrhythmias (premature heart beats, short-lasting atrial fibrillation), atrio-ventricular blocks, pneumonia, pleural effusion, wound infection have been described[11,12].

Postoperative early and late mortality ranges from 0 to 7.5%[12,13]. Damaged valve may require repair or replacement, but it was not necessary in this case. One month after discharge the anterior mitral valve was thickened possibly due to chronic tumoral injury, with moderate regurgitation and slight pulmonary hypertension (the right ventricle to an atrium gradient of 29 mmHg) (Figure 7). Three months after the operation the patient is recovered with no signs of recurrence. The prognosis in our patient is good, but follow-up is necessary because of the risk of recurrence which can be as high as 13%[14,15].

The particularity of this case is the high impact given by the tumor obstruction of the left ventricle inflow, resulting in very poor exercise capacity with chronic exertional dyspnea which was initially considered to be due to hypothyroidism.

CONCLUSIONS

In summary, cardiac myxomas are rare condition in clinical practice. High index of suspicion is mandatory for diagnosis and prompt therapy. Urgent surgical resection is indicated because of the high risk of complications and of sudden death, with low procedural mortality and morbidity.

Conflict of interest: none declared.

References

1. Abraham KP, Reddy V, Gattuso P. Neoplasms metastatic to the heart: review of 3314 consecutive autopsies. Am J Cardiovasc Pathol. 1990; 3: 195–198
2. Yu K, Liu Y, Wang H, Hu S, Long C. Epidemiological and pathological characteristics of cardiac tumors: a clinical study of 242 cases. Interact Cardiovasc Thorac Surg. 2007 Oct:6(5):636-9.
3. Bjessmo S, Ivert T. Cardiac myxoma: 40 years' experience in 63 patients. Ann Thorac Surg. 1997;63:697–700
4. Ermek T, Aybek N, Zhang WM, Guo YZ, Guo S, Mamataly A, Chang DQ, Liu J, Zhang ZG. A rare case of biventricular myxoma. J Cardiothorac Surg. 2017 Mar 27;12(1):17. doi: 10.1186/s13019-017-0584-6.
5. Sabzi F, Nasiri B. Myxoma of the Superior Vena Cava Origin Presented as a Right Atrial Mass. J Teh Univ Heart Ctr 2013;8(4):202-204.
6. Karimi VM, Anushiravani A, Dabbaghmanesh MH, Hosseinzadeh M, Rasekhi AR, Zamirian M, Anushiravani A. Myxoma Immediately above the Junction of the Inferior Vena Cava and the Right Atrium: A Rare Cause of Budd-Chiari Syndrome. J Tehran Heart Cent. 2016 Jul 6;11(3):139-142.
7. Ramphal PS, Spencer HW, Mitchell DI, Denbow CJ. Myxoma of right femoral vein origin presenting as a right atrial mass with syncope. Thorac Cardiovasc Surg. 1998 Oct;116(4):655-6.
8. Larrieu AJ, Jamieson WR, Tyers GF, Burr LH, Munro AI, Miyagishima RT, Gerein AN, Allen P. Primary cardiac tumors: experience with 25 cases. J Thorac Cardiovasc Surg. 1982 Mar;83(3):339-48.
9. Johnson TR, Becker CR, Wintersperger BJ, Herzog P, Lenhard MS, Reiser MF. Images in cardiovascular medicine. Detection of cardiac metastasis by positron-emission tomography-computed tomography. Circulation. 2005 Jul 26;112(4):e61-2.
10. Dumitrasciuc G, Gaspar M, Ionac A, Jusca C, Deutsch P, Petrescu L, Mut-Vitcu B, Stefan I. Dragulescu SI. Left atrial myxoma associated with coronary artery disease - uncommon challenge. TMJ 2004:54(4): 370-73
11. Lee KS, Kim GS, Jung Y, Jeong IS, Na KJ, Oh BS, Ahn BH, Oh SG. Surgical resection of cardiac myxoma-a 30-year single institutional experience. J Cardiothorac Surg. 2017 Mar 27;12(1):18. doi: 10.1186/s13019-017-0583-7.
12. Perek B, Misterski M, Stefaniak S, Ligowski M, Puślecki M, Jemielity M. Early and long-term outcome of surgery for cardiac myxoma: experience of a single cardiac surgical centre. Kardiol Pol. 2011;69(6):558-64.
13. Keeling IM, Oberwalder P, Anelli-Monti M, Schuchlenz H, Demel U, Tilz GP, Rehak P, Rigler B. Cardiac myxomas: 24 years of experience in 49 patients. Eur J Cardiothorac Surg. 2002 Dec;22(6):971-7.
14. Iliescu VA, Dorobantu LF, Stiru O, Iosifescu AG, Coman I, Marin S, Filipescu D. A doua recidiva a unui mixom cardiac aparuta la 7 ani de la prima operatie. Chirurgia. 2008:103(2): 239-241.
15. Elbardissi AW, Dearani JA, Daly RC, Mullany CJ, Orszulak TA, Puga FJ, Schaff HV. Survival after resection of primary cardiac tumors: a 48-year experience. Circulation. 2008 Sep 30;118(14 Suppl):S7-15.