Research Paper Medical Science

# Giant Right Atrial Myxoma: a Case Report

* Adrian Molnar, MD ** Diana Sacui, MD *** Traian Scridon, MD

*,**,*** Cluj-Napoca "N. Stancioiu" Heart Institute, Cardiovascular Surgery Clinic

**ABSTRACT**

*Atrial myxoma is the commonest (20–30% of all) primary intra-cardiac tumour in adults and two thirds of these arise in the left atrium. Other locations are right atrium (next commonest), ventricles, superior vena cava or pulmonary veins (1). Although they are benign, it is recommended their immediate removal as soon as the diagnosis is confirmed, since they are associated with tumour embolization and their harmful consequences (2).*

*We present the case of a 73 year old female with a 4-month history of progressive exertional dyspnoea accompanied by palpitations and peripheral edema. Trans-esophageal echocardiogram revealed a large mobile mass in the right atrium prolapsing into the right ventricle. We performed the tumour resection in association with direct repair of the inter-atrial septum defect.*

**Keywords : atrial myxoma, embolization, removal, transesophageal echocardiogram**

**Introduction**

Metastatic tumours are the commonest cardiac tumours being found in 1-3% of patients dying of cancer while primary tumours are unusual and have an incidence of 0.02-0.5%. The majority (80%) of all primary cardiac tumours are benign with myxomas accounting for 50%. Myxomas arising from the right atrium are uncommon [3]. The tumour consists of myxoid stroma in which are embeded single stellate cells in addition to larger clumps of similar cells often arranged as collar around open spaces. The lesion can be peeled away from the underlying myocardium from which there is no obvious evidence of an origin [4].

It could be seen in patients between 3–83 years of age, with the majority presenting in fifth decade of life as sporadic cases (90%) and second decade as familial cases (10%). It is an important source of central nervous system embolism. Elderly patients often present with non specific symptoms that are often overlooked in the absence of a supporting cardiac history which makes an early diagnosis challenging [1].

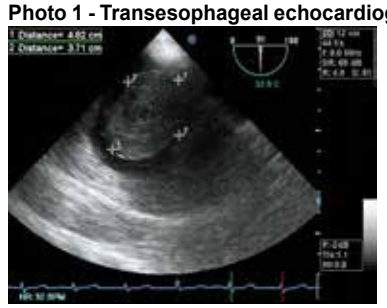
Taking into account these considerations, we present the clinical observation encountered in our experience.

**Case Presentation**

A 73 year old female patient presented with a four month history of shortness of breath accompanied by peripheral edema and palpitations. She had also an ischemic parietal stroke, and a radical surgical treatment for duodenal adenocarcinoma in the year prior to her current admission. The cardiovascular, respiratory and abdominal examination was unremarkable and a detailed neurological assesment did not reveal any significant abnormality. The electrocardiogram showed sinus rhythm, with a frequency of 80 bpm, and minor T wave changes in anterior leads.

An in-patient transesophageal echocardiography (TEE) demonstrated a giant mass (approximately 50 / 40 mm, with a volume of 70 cm) which occupies almost entirely the right atrium, prolapsing into the right ventricle, with intermittent obstruction of the tricuspid valve and a very high risk of embolization; the mass was attached to the ostium secundum area of the inter-atrial septum, and also a patent foramen ovale was present (photo 1).

**Photo 1 - Transesophageal echocardiography (TEE) image.**



The patient accepted the surgical treatment in view of the risks involved, and we performed the complete tumour resection and direct repair (suture) of the inter-atrial septum defect. The surgery was performed under extracorporeal circulation, the case characteristic being consistent with the fact that, due to the presence and the large dimensions of the mixoma, the inferior vena cava couldn't be canulated, the venous drainage being accomplished by superior vena cava and right femoral vein cannulation. In order to have a clean surgical field, withot blood, we clamped the inferior vena cava above the diaphragm (photo 2).

**Photo 2 - Intraoperative image. Right atrium opened, inferior vena cava clamped, the mixoma was excized, showing the implantation base in the inter-atrial septum.**

The mixoma have been relatively easily removed, without fragmentation, the base of implantation with a diameter of approximately 1.5 cm. being excised and the resulting defect was closed with continuous sutured. In the image below can be seen the mixoma after its removal eith the dimensions of 11/7 cm (photo 3).

**Photo 3 – Macroscopic aspect of the tumor (after removal).**


The recovery after surgery was uneventful, with the exception of an episode of transient atrial fibrilation for which she was treated with continuous i.v. amiodarone.

Pathological analysis of the removed atrial mass revealed it to be 11/7 / 3 cm attached to a 1.5 / 1 cm stalk of atrial septal tissue.

The patient was discharged in the 8th postoperative day in good conditions, and was asymptomatic after 3 months of follow-up, with no evidence of clinical or echocardiographic changes.

**Discussion**

Right atrial myxoma accounts for only about 25% of all myxomas. It usually arises from the inter-atrial septum [5]. In our patient, the atrial mass was found to prolapse through the tricuspid valve into the right ventricle, causing a certain degree of tricuspid valve stenosis. The reason for such prolapse is the attachment of the myxoma on a long stalk.

Myxomas can present at any age group but occurs more often between the 3rd and 6th decades of life as is the case with our patient who was 51 years [5].

The most common symptoms of right atrial myxoma have been reported to be those of congestive heart failure, while other symptoms vary from constitutional to thrombo-embolic and obstructive [5]. Our patient presented with features of congestive heart failure and constitutional symptoms of exertional dyspnoea, palpitations, and recurrent pedal oedema. Constitutional symptoms in patients with cardiac myxomas have been attributed to cardiac myxomas releasing the cytokin, interleukin-6 which causes inflammatory and auto-immune manifestations [5].

TEE has nearly 100% sensitivity for cardiac myxoma. The tumor tissue manifests as spherical/pedunculated mass attached to the endocardial surface with hypoechoic areas. TTE has less specificity than the TEE. Contrast CT demonstrates a well defined spherical or ovoid intracavitary mass. Magnetic resonance imaging (MRI) can visualise the point of attachment and helps differentiate a thrombus from a tumour. Differences in signal intensity between myocardium, tumor/thrombus is very helpful, especially with the use of contrast agent like Gadolinium-DTPA. A cine MRI sequence is a very sensitive technique to distinguish between an thrombus and a tumor, intra-cardiac or intravascular [1].

The only definitive treatment for atrial myxoma is considered to be the surgical excision. Therefore, if a polypoid or multilobular shaped myxoma is suspected on preoperative echocardiography, urgent surgical intervention is necessary to prevent an embolism. Recent studies on the resection of cardiac myxoma with normal cardiac tissues indicated that there was no difference in recurrence rates between patients who underwent an en bloc resection and resection with only endocardial tissue attached with the tumor. However, the type of resection depends largely on factors such as the location of the tumor and the shape of tumor stalks [6]. In relatively small tumors, TTE/TEE can be used to monitor the growth of the tumor, to decide the timing of the surgery. Conservative management is of limited value in symptomatic patients with large myxomas. However, a conservative strategy with TTE/TOE monitoring, and anticoagulation is favoured in high operative risk patients, asymptomatic patients, and slow growing atrial myxomas [1]. In the presented case, we performed a complete resection of the tumour by median sternotomy, under moderate hypothermia and arrested heart with cardiopulmonary bypass; the procedure required a right atrial approach. For the repair of the defective area in the inter-atrial septum we used the direct closure technique. Postoperatively, the patient showed no evidence of tumour recurrence clinically or in the echocardiograms during the 3 months follow-up period.

The diagnosis of atrial myxoma can be elusive, especially when symptoms are suggestive of other diagnoses. In the case of our patient, the significance of this patient's past medical history of an ischemic stroke only became apparent when the patient presented with new symptoms of shortness of breath and pedal oedema, which led to various investigations looking for a source of cardiogenic cerebral embolism. This seems to be an unusual case due to the age at presentation of the patient. Right atrial myxoma presenting in seventh decade is rare, with only few published case reports. Bire et al studied the number of myxoma cases in patients over 75 years of age between 1962 and 1997 and found only 19 confirmed cases [1].

**Conclusions**

Transthoracic or transesophageal echocardiography is the best diagnostic method for myxoma, because it enables determination of the tumour exact location and evaluation of valvular status and cardiac contractile function [7]. Three-dimensional echocardiography proved useful in surgery planning, allowing a better definition of the tumor outline and attachment [8]. In addition, transthoracic echocardiography remains an invaluable tool in the diagnosis of this uncommon condition, while prompt diagnosis and treatment is necessary to avoid fatal outcome [5].

Diagnosis is often difficult due to the wide array of presenting symptoms. Atrial myxomas are associated with systemic embolization in 30 to 40% of cases. These intracardiac growths may masquerade as mitral stenosis, infective endocarditis, and collagen vascular disease, which can further impede accurate diagnosis [9].

Atrial myxoma is the most common cardiac tumor and is curable with complete surgical resection [10], and postoperative survival for patients with myxoma is usually excellent. Recently, well developed preoperative diagnostic techniques, especially echocardiography, have enabled earlier detection than was previously possible. Therefore, the number of patients who undergo surgery has increased, and long-term results have improved [6]. Adequate excision of the entire mass, along with resection of normal tissue surrounding the base, prevents recurrence, except in cases of familial myxoma. In addition, careful handling of the myxoma itself may prevent intracardiac implantation or peripheral embolization of the tumor fragments [11].

Although atrial myxomas are usually benign or asymptomatic [9], they should always be considered when patients present with symptoms of heart failure with uncertain aetiology [5]. Also, there is the possibility of diastolic embolization, conduction alterations and disturbances, and lethal valve obstruc-

tions occurring. Since surgical excision has been reported to alleviate symptoms associated with cardiac myxomas, early identification and removal is preferable [9].

## REFERENCES

1. Biswas, A and Thakur, AK. An unusual presentation of atrial myxoma in an elderly patient: a case report. Cases J. 2008; 1: 384. | 2. Lorentz, MN., Vrandecic, EC., Drumond, LF., Soares, RR. Right atrial myxoma associated with acute cor pulmonale. Case report. Rev Bras Anestesiol. 2008 Jan-Feb;58(1):69-72. | 3. Bilku, RS., Loubani, M., Been, M., Patel, RL. Massive right atrial myxoma causing exertional dyspnoea. Eur J Echocardiogr. 2008 Jan;9(1):130-2. | 4. Krikler, DM., Rode, J., Davies, MJ., Woolf, N., and Moss, E. Atrial myxoma: a tumour in search of its origins. Br Heart J. 1992 January; 67(1): 89–91. | 5. Ojji, DB., Ajiduku, SS., Omonua, OO., Abdulkareem, LL., and Parsonage, W. A probable right atrial myxoma prolapsing through the tricuspid valve into the right ventricle: a case report. Cases J. 2008; 1: 386. | 6. Yu, SH., Lim, SH., Hong, YS., Yoo, KJ., Chang, BC., and Kang MS. Clinical Experiences of Cardiac Myxoma. Yonsei Med J. 2006 June 30; 47(3): 367–371. | 7. Lijoi, A., Scoti, P., Faveto, C., Canale, C., Parodi, E., Passerone, GC., Dottori, V., and Venere, G. Surgical management of intracardiac myxomas. A 16-year experience. Tex Heart Inst J. 1993; 20(3): 231–234. | 8. Azevedo, O., Almeida, J., Nolasco, T., Medeiros, R., Casanova, J., Bartosch, C., Almeida, J., Pinho, P. Massive right atrial myxoma presenting as syncope and exertional dyspnea: case report. Cardiovasc Ultrasound. 2010 Jun 18;8:23. | 9. Vicari, RM., Polanco, E., Schechtmann, N., Santiago, JO., Shaurya, K., Halstead, M., Marszal, D., Grosskreutz, T., and Thareja, S. Atrial myxoma presenting with orthostatic hypotension in an 84-year-old Hispanic man: a case report. J Med Case Reports. 2009; 3: 9328. | 10. Matebele, MP, Peters, P, Mundy, J, Shah, P. Cardiac tumors in adults: surgical management and follow-up of 19 patients in an Australian tertiary hospital. Interact Cardiovasc Thorac Surg. 2010 Jun;10(6):895. | 11. Guhathakurta, S., and Riordan, JP. Surgical Treatment of Right Atrial Myxoma. Tex Heart Inst J. 2000; 27(1): 61–63.