

A Massive Right Atrial Myxoma

right heart failure, peripheral edema, ascites, hepatic congestion, and syncope.⁷ Cardiac auscultation in atrial myxomas may vary with the size, location, mobility, and prolapsed of the tumor through the atrioventricular valves and even body position, and therefore, detection of a murmur may or may not occur. An auscultation characteristic of myxoma is the 'tumor plop', which is an onomatopoeic representation of the heart sound caused by the presence of the tumor inside the atrial chamber that occurs in 15% of cases.⁷

Routine laboratory assessment may show non-specific changes such as anemia, increased erythrocyte sedimentation rate, increased levels of globulin and C-reactive protein, leukocytosis, thrombocytopenia, and polycythemia.⁷ Recent studies suggest that cardiac myxomas produce and release into the circulatory system an interleukin, which may be responsible for inflammatory or autoimmune phenomena.

Although transthoracic echocardiography is less invasive and presents an excellent sensitivity in detecting 95% of myxomas, the sensitivity increases to 100% when followed by transesophageal echocardiography.⁷ Computed tomography (CT) and magnetic resonance imaging may be useful to demonstrate the point of fixation and associated complications. Chest X-rays and electrocardiograms are non-specific.²

Once a cardiac myxoma is diagnosed, surgical excision should be performed without delays because of the constant risk of thromboembolic events.^{1,10} Generally, surgical treatment is definitive and recurrence is uncommon. This report describes a rare clinical case of a large RA myxoma, highlighting the difficulty of the differential diagnosis of this tumor because of its unusual presentation.

Case History

A 50 years old man, Mr. Shohrab Ali presented with shortness of breath, palpitation, weakness, gradual

loss of weight, with chronic cough and swelling of legs for one year. Initially he was diagnosed by a local registered physician as a case of COPD with cor-pulmonale and treated with diuretics, bronchodilators and antibiotics. But with this medication the patients condition was not improved rather worsening with marked weight loss, severe weakness, and progressive abdominal and leg swelling. Then the patient was referred to us. When we found the patient he appeared nervous, chronically ill, with poor nutritional status and plethoric looking. He was a non smoker, non diabetic and normotensive. He did not have any history of rheumatic fever in child hood. When we examined him he is mildly anaemic but cyanosis and clubbing was absent. He had pedal oedema and ascities. His pulse was 102 b/min, low volume, regular, BP was 100/70 mm of Hg, and JVP was raised and no abnormal venous wave seen. Precordial examination shows apex beat is in normal position without any thrill. There is epigastric pulsation with left parasternal lift and palpable P2 is present. On auscultation there is a systolic murmur in the tricuspid area with loud P2 noted. There was no chest deformity with clear lung fields and resonant throughout. The liver was enlarged felt two finger breadths from right costal margin which was smooth and tender. With the above clinical features and physical findings our clinical diagnosis was right ventricular failure due to pulmonary hypertension. Clinically the cause of pulmonary hypertension cannot be detected. The other common causes of congestive heart failure like dilated cardiomyopathy, Ebstein anomaly and mitral valvular disease could be the differential diagnosis.

His blood count shows neutrophilic leukocytosis with raised ESR but CRP, urine analysis, LFT and renal function was normal. An X-ray chest showed clear lung fields with mild cardiomegaly with RV type apex and enlarged right border suggestive of RA enlargement (Fig.1). The electrocardiogram

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showed sinus tachycardia, P-pulmonale and RBBB (Fig-2). Ultrasonograms of the abdomen showed enlarged liver with dilated hepatic veins and mild to moderate ascities suggestive of CHF. A Transthoracic echocardiogram in parasternal long axis view shows an echogenic structure within the right ventricle and in apical four chamber view showed a well defined large, mobile non-homogenous oscillatory mass with a broad base attached with the lower part of right atrial

septum, an enlarged RA and RV and dilated IVC is seen (Fig 3). Color Doppler study shows grade- III TR with PASP is about 50 mm of Hg. So finally with this report our diagnosis was a case of large right atrial myxoma with PH with right sided heart failure. A chest CT scan with I/V contrast can be done to facilitate our diagnosis. Cardiac catheterization and selective CAG should be done before surgery. We sent this case to a cardiac surgeon for emergency operation.



Fig 1: X-ray chest showed clear lung fields with mild cardiomegaly with RV type apex and enlarged right border suggestive of RA enlargement

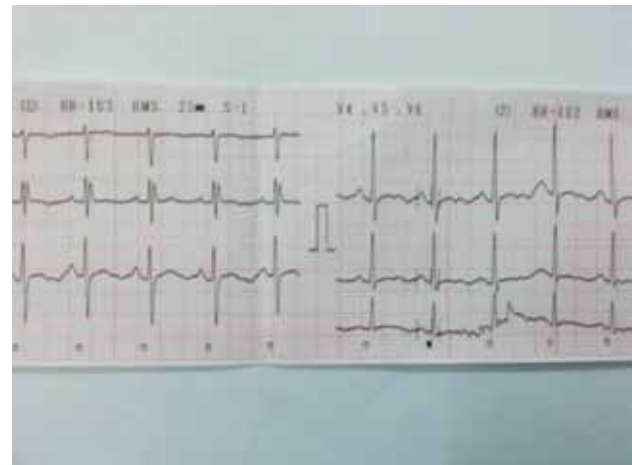
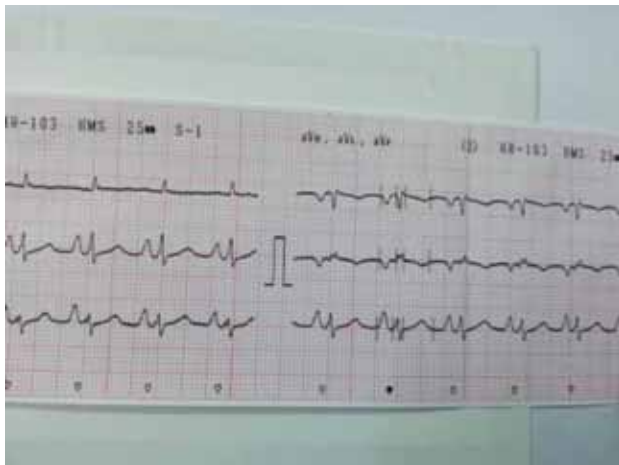


Fig 2: Electrocardiogram shows sinus tachycardia, P-pulmonale and RBBB.

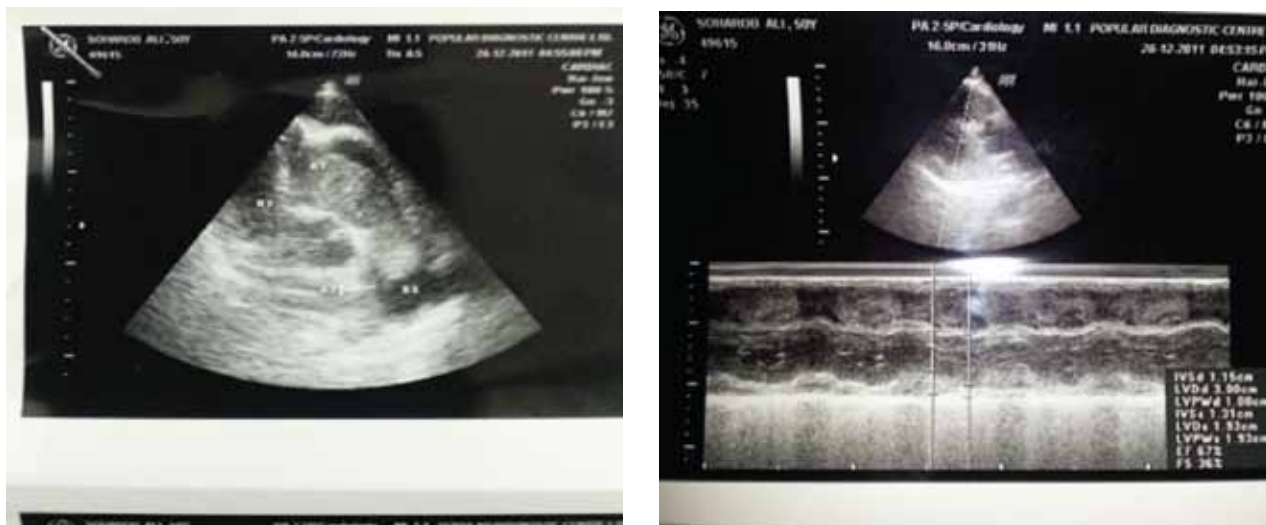


Fig 3: Transthoracic echocardiogram (2D and M-Mode) shows a large RA mass before an operation.

Discussion

Primary cardiac neoplasms are rare and occur with an estimated incidence of 0.0017% to 0.19%, representing less than 5% of all heart tumors.¹¹ Myxoma is the most prevalent primary cardiac tumor. The RA is an unusual location and is the site of 15% to 20% of cases of myxoma.⁵ A low incidence of RA myxoma has been reported for decades in several series of autopsy cases. Approximately 70% of affected patients are women,^{2,12} but our case was male. It affects predominantly between the third and sixth decades of life.^{10,13} as was the case of the 50year old patient described in this report.

RA myxomas usually originate in the fossa ovalis or base of the interatrial septum,⁸ but in this case, the myxoma was implanted in the lower part of IAS, close to the tricuspid valve. Myxomas are usually polypoid and pedunculated tumors (approximately 83% of cases).¹⁰ In this report, our patient had a 50 x 20 x 32 mm solitary pedunculated mass with polypoid areas and a lobulated surface.

In a recent publication reporting 19 years of experience with surgical treatment of primary intracardiac myxoma, seven (17%) cases out of 41

originated from the RA. However, in this series, the mean maximal diameter of the tumor was 5.1 ± 1.8 cm.¹⁴ To the best of our knowledge our case is one of the large RA myxomas described in the literature.

The signs and symptoms of RA myxomas are atypical and highly variable, depending on the size, position, and mobility of the tumor, and are modified according to physical activity and body position of the patient.⁷ RA myxomas may remain asymptomatic¹⁵ or eventually causes constitutional signs and symptoms, including fever, weight loss, arthralgias, Raynauds phenomenon, anemia, hypergammaglobulinemia, and an increased erythrocyte sedimentation rate due to the production of interleukin-6.¹⁰ These symptoms disappear after the tumor is removed.¹⁰ In this report, our patient also had fever, anemia and complained of weight loss as a constitutional sign during his illness.

The patients may also present with atypical chest pain, syncope, lethargy, malaise, palpitation, peripheral edema, pulmonary embolism, and haemoptysis. However, the most common manifestation is dyspnea (in 80% of patients), and

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right heart failure has been reported. Our patient had all the features of right heart failure including dyspnoea, leg edema, ascites and congested liver. Echocardiography remains the best diagnostic method for locating and assessing the extent of myxomas and for detecting their recurrence, with a sensitivity of up to 100%. However, transthoracic echocardiogram may not identify tumors smaller than 5 mm in diameter, and a transoesophageal echocardiogram is required when there is suspicion of a very small tumor.¹² In this case, an echocardiogram suggested the hypothesis of RA myxoma, which should be confirmed by a histopathological exam after surgery.

Although echocardiography is the modality of choice for screening cardiac masses, magnetic resonance imaging and CT provide information regarding tissue characteristics and allow an excellent overview of cardiac and paracardiac morphology but unfortunately we could not do the CT scan of our patient.

The treatment of choice for myxomas is surgical removal.^{1,10} Complete resection of the tumor and its implantation base with a good safety margin is essential to cure the disease, preventing recurrence and subsequent reoperations, which exposes the patient to other complications such as bleeding and the need for blood products. Myxomas are usually removed with a large resection of their pedicle or attachment to prevent recurrence.

The recurrence rate of sporadic tumors is very low: between 1% and 3%.⁷ The operative mortality ranges from 0% to 3% in multiple series.⁷ The survival rate after surgery is elevated.¹⁰ The surgical technique follows the basic concepts of cardiac surgery. However, some aspects should be taken into consideration in the surgical treatment of myxoma. Before resection, it is fundamentally important to clamp both the aorta and pulmonary trunk to avoid embolizations of fragments because the myxomas are gelatinous and friable masses.⁸ Bahnsen and Newman¹⁶ in 1953 reported the first

removal of a right atrial myxoma and since that time 4 others have been operated upon. Ripstein¹⁷ in 1953 attempted unsuccessfully to remove a right atrial myxoma through the open right atrium under hypothermia; this tumor extended through the septum into the left atrium.

Recently Coates and Drake¹⁸ reported the successful removal of a right atrial myxoma under open-heart conditions. This patient had a variable right-to-left shunt through a patent foramen ovale. Lyons and his group¹⁹ were unsuccessful in removal of a similar tumor under open-heart surgery. They thought that death was due to a flabby myocardium and the production of total tricuspid insufficiency upon removal of the myxoma, which had previously caused tricuspid stenosis. We have some limitation to describe this case report including we did not perform CT scan and surgical aspects of myxomas could not be discussed.

Conclusion

Since myxomas of the atria can be successfully removed, it is important to diagnose the lesion correctly and treated accordingly to prevent fatal complication as surgical removal of myxoma is almost curative. As the RA myxoma is a rare location it should be considered in the differential diagnosis of any patients presented with the features of right sided heart failure of uncertain etiology.

References

1. Arruda MV, Braile DM, Joaquim MR, Soares MJ, Alves RH. Resection of left ventricular myxoma after embolic stroke (in english and portuguese). *Rev Bras Cir Cardiovasc.* 2008; 23:578-580. doi:10.1590/S0102-76382008000400022. [PubMed] [Cross Ref]
2. Jang KH, Shin DH, Lee C, Jang JK, Cheong S, Yoo SY. Left atrial mass with stalk: thrombus or myxoma? *J Cardiovasc Ultrasound.* 2010;18:154-156. doi: 10.4250/jcu.2010. 18.4.154. [PMC free article] [PubMed] [Cross Ref]

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3. Vale Mde P, Freire Sobrinho A, Sales MV, Teixeira MM, Cabral KC. Giant myxoma in the left atrium: case report (in english and portuguese). *Rev Bras Cir Cardiovasc.* 2008;23:276-278. doi:10.1590/S0102-76382008000200022. [PubMed] [Cross Ref]
4. Hermans K, Jaarsma W, Plokker HW, Cramer MJ, Morshuis WJ. Four cardiac myxomas diagnosed three times in one patient. *Eur J Echocardiogr.* 2003;4:336 – 338. doi: 10.1016/S1525 – 2167(03)00018 – 0. [PubMed] [Cross Ref]
5. Diaz A, Di Salvo C, Lawrence D, Hayward M. Left atrial and right ventricular myxoma: an uncommon presentation of a rare tumor. *Interact Cardiovasc Thorac Surg.* 2011; 12:622 – 623. doi:10.1510/icvts.2010.255661. [PubMed] [Cross Ref]
6. Croti UA, Braile DM, Souza AS, Cury PM. Right ventricle and tricuspid valve myxoma (in Portuguese) *Rev Bras Cir Cardiovasc.* 2008;23:142–144. doi: 10.1590/S0102 – 76382008000100026. [PubMed] [Cross Ref]
7. Oliveira R, Branco L, Galrinho A, Anreus J, Fiarresga A, Mamede A, et al. Cardiac myxoma: a 13-year experience in echocardiographic diagnosis (in english and portuguese). *Rev Port Cardiol.* 2010;29:1087 – 1100. [PubMed]
8. Stolf NA, Benicio A, Moreira LF, Rossi E. Right atrium myxoma originating from the inferior vena cava: an unusual location with therapeutic and diagnostic implications. *Rev Bras Cir Cardiovasc.* 2000;15:255–258. doi: 10.1590/S0102 – 76382000000300009. [Cross Ref]
9. Murayama H, Tamaki S, Kato N, Yuji N, Yokote J, Mutsuga M, Okada M. Right atrial myxoma associated with atrial septal defect: a case report and report review of the literature. *Ann Thorac Cardiovasc Surg.* 2001;7:166 –169. [PubMed]
10. Guhathakurta S, Riordan JP. Surgical treatment of right atrial myxoma. *Tex Heart Inst J.* 2000;27:61–63. [PMC free article] [PubMed]
11. Azevedo O, Almeida J, Nolasco T, Medeiros R, Casanova J, Bartosch C et al. Massive right atrial myxoma presenting as syncope and exertional dyspnea: case report. *Cardiovasc Ultrasound.* 2010;8:23. doi: 10.1186/1476 – 7120 – 8 – 23. [PMC free article] [PubMed] [Cross Ref]
12. Manfroio W, Vieira SR, Saadi EK, Saadi J, Alboim C. Multiple recurrences of cardiac myxomas with acute tumoral pulmonary embolism [in portuguese] *Arq Bras Cardiol.* 2001;77:161–163. [PubMed]
13. Ojji DB, Ajidukun SS, Omonua OO, Abdulkareem LL, Parsonage W. A probable right atrial myxoma prolapsing through the tricuspid valve into the right ventricle: a case report. *Cases J.* 2008;1:386. doi: 10.1186/1757 – 1626 – 1 – 386. [PMC free article] [PubMed] [Cross Ref]
14. Samanidis G, Perreas K, Kalogris P, Dimitriou S, Balanika M, Amanatidis G et al. A surgical treatment of primary intracardiac myxoma: 19 years of experience. *Interact Cardiovasc Thorac Surg.* 2011;13:597–600. doi: 10.1510/icvts.2011.278705. [PubMed] [Cross Ref]
15. Yuce M, Dagdelen S, Ergelen M, Eren N, Caglar N. A huge obstructive myxoma located in the right heart without causing any symptom. *Int J Cardiol.* 2007;114:405–406. doi 10.1016/j.ijcard.2005.11.086. [PubMed] [Cross Ref]
16. Bahnson, H. T, And Newman, E. G. Diagnosis and removal of intracavitary tumor of the right atrium. *Bull. Johns Hopkins Hosp* 1953;93:150.
17. Ripstein, C. B, From Bahnson H T. Personal communication.
18. Coates E. O. Jr, Drake H E. Myxoma of the right atrium with variable right to left shunt. *New England J. Med.* 1958;259:165.
19. Lyons H A, Kelly J J, Nussbaum, N, Dennis, C. Right atrial myxoma. *Am. J. Med.* 1958 ;25:321.