

# Right Atrial Myxoma and Chronic Transudative Ascites: A Rare and Challenging Clinical Presentation



Mohammadbagher Sharifkazemi, MD, Gholamreza Rezaian, MD, and Firouzeh Abtahi, MD,  
*Shiraz, Iran*

## INTRODUCTION

Atrial myxomas are the most common primary cardiac tumors and account for about 40% to 50% of such masses.<sup>1</sup> Right atrial (RA) myxomas account for about 25% of cardiac myxomas<sup>2</sup> and are often difficult to diagnose. They may remain asymptomatic; however, patients will eventually develop nonspecific constitutional symptoms such as fever, weight loss, arthralgia, anemia, and clotting disturbances due to the production of interleukin-6.<sup>3</sup> Embolic complications, syncope, and sudden death have also been reported.<sup>4</sup> Although tumor fragmentation and embolization are more common in left-sided myxomas,<sup>5</sup> rarely pulmonary embolization and secondary pulmonary hypertension may occur in patients with RA myxomas as well.<sup>6,7</sup> In addition, right ventricular failure and ascites may occur as a result of tricuspid valve orifice obstruction.<sup>8</sup> Massive ascites is a highly uncommon extracardiac presentation of RA myxoma and may lead to an extensive clinical, paraclinical, and imaging investigation. Herein, we report such an unusual patient with a very large RA myxoma alongside chronic, persistent, and recurrent transudative ascites despite repeated peritoneocentesis.

## CASE PRESENTATION

A 55-year-old man with a history of diabetes mellitus and hypertension presented with a history of progressive severe abdominal protrusion, lower extremity edema, and exertional dyspnea of 2 years' duration. He also had experienced a few episodes of presyncope while standing during this time period. However, he had no fever, weight loss, or true syncopal attacks. An extensive gastrointestinal investigation including abdominal sonography, computed tomography, and even liver biopsy failed to elucidate the underlying pathology. Ascitic fluid was transudative in nature, and hematologic studies were inconclusive. Because of his condition, he was transferred to our university hospital for further evaluation. On arrival he was fully oriented. Vital signs were normal. Cardiac auscultation was unremarkable. There was marked abdominal distention associated with shifting dullness. Electrocardiographic findings were normal.

Transthoracic echocardiography and then transesophageal echocardiography revealed a bulky mobile echogenic RA mass of very

large dimensions (6.8 × 5.6 cm) protruding through the tricuspid valve into the right ventricle during diastole (Figure 1, Video 1). The attachment site was evident neither by transthoracic nor transesophageal echocardiography. The inferior vena cava was patent and free of tumor invasion. Coronary angiography showed a significant tumor blush via a sizable feeding vessel originating from the right coronary artery (Figure 2). At surgery, a large, lobulated, gelatinous RA mass was found which was attached to the lateral RA free wall just anterior to the superior vena cava orifice by a pedicle. Pathologic analysis confirmed the echocardiographic suspicion of myxoma (Figure 3). At the postoperative follow-up 10 weeks later, the patient reported a significant symptomatic improvement as well as marked weight loss.

## DISCUSSION

There are some unusual features about this case that make it an interesting one: (1) the presenting symptoms and signs were extracardiac and nonconstitutional, (2) the tumor size was quite impressive, and (3) the tumor had an uncommon attachment site. Myxoma is the most frequently encountered primary cardiac tumor, which predominantly involves the left atrium. RA myxomas account for about 25% of all cardiac myxomas<sup>2</sup> and are mainly (70%) seen in women<sup>8</sup> between the third and sixth decades of life.<sup>9,10</sup> The main findings in patients with myxoma are characterized by the classic triad of constitutional symptoms, intracardiac blood flow obstruction, and thromboembolic complications.<sup>11</sup> Many patients with RA myxoma, however, may remain asymptomatic for a long period of time, which may delay the correct diagnosis and lead to the development of unusual extracardiac manifestations.<sup>12</sup> Our patient's main presentation was long-standing, chronic, unexplained, and massive transudative ascites, requiring repeated peritoneocentesis and frequent empiric drug therapies. Nonspecific constitutional symptoms such as fever, elevated erythrocyte sedimentation rate, Raynaud's phenomenon, weight loss, myalgia, arthralgia, and anemia may be seen in 20% to 60% of patients with myxoma.<sup>2,13</sup> Constitutional symptoms, including fever, however, were not prominent in our patient. It should be emphasized, however, that constitutional symptoms are most frequently encountered in patients with left atrial, rather than RA, myxomas. These symptoms disappear after surgical removal of the tumor.<sup>9</sup> Although our patient's symptoms were nonconstitutional, he reported significant symptomatic improvement and weight loss at postoperative clinical follow-up at 10 weeks.

The average tumor size at the time of diagnosis has been reported to be about 50 to 60 mm.<sup>14</sup> Our patient's tumor size was about 68 × 56 mm by transesophageal echocardiography at the time of diagnosis and had an unusual attachment site—namely, the lateral RA free wall just anterior to the orifice of the superior vena cava. Also, his large tumor size was in favor of the long-standing disease duration. Frequently, when myxomas become bulkier, patients' constitutional symptoms become more evident, and obstructive

From the Cardiology Department, Shiraz University of Medical Sciences, Shiraz, Iran.

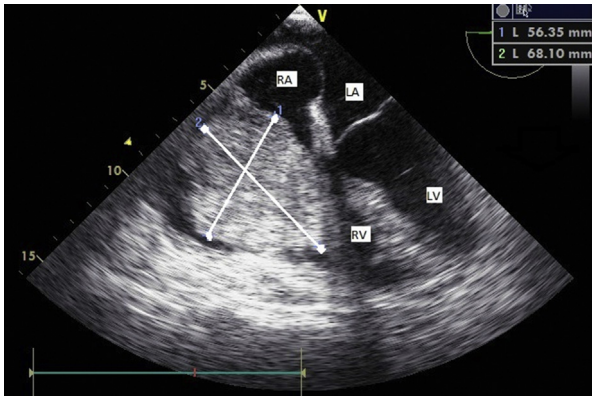
Keywords: Right atrial myxoma, Transudative ascites, Syncope, Echocardiography

Conflict of interest: The authors reported no actual or potential conflicts of interest relative to this document.

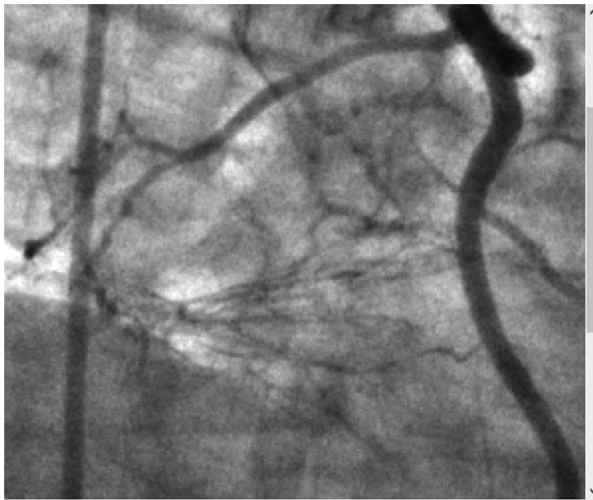
Copyright 2018 by the American Society of Echocardiography. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

2468-6441

<https://doi.org/10.1016/j.case.2018.01.003>



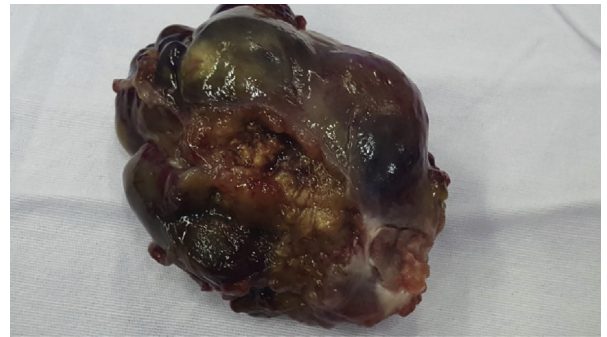
**Figure 1** Transesophageal echocardiography (*midesophageal view*) demonstrating the presence of a massive RA mass protruding into the right ventricle (6.8 × 5.6 cm). LA, Left atrium; LV, left ventricle; RV, right ventricle.



**Figure 2** Selective right coronary angiography demonstrating the atrial branch of the right coronary artery supplying the tumor and producing a “tumor blush.” At surgery, a large lobulated, gelatinous RA mass was found, which was attached to the lateral RA free wall just anterior to the superior vena cava (SVC) orifice by a pedicle. Pathologic analysis confirmed the echocardiographic suspicion of myxoma (Figure 3).

symptoms may develop. It has been reported that signs and symptoms of myxomas usually occur when the tumor assumes a minimum size of 50 mm.<sup>15</sup>

RA myxomas may obstruct the tricuspid valve, leading to the development of presyncope, right-sided heart failure symptoms such as peripheral edema, and ascites, very similar to what was found in our patient. We assume that these clinical signs and symptoms were the result of obstruction of blood flow through the right heart by the tumor because of its massive size.<sup>16</sup> Therefore, it is wise to consider RA myxoma in the differential diagnosis of different causes of unexplained transudative ascites. Metastatic invasion of cardiac chambers by renal cell carcinoma should be considered in the differential diagnosis of RA myxoma as well. However, there was no inferior vena cava involvement, and results of abdominal sonography and computed tomography were normal in our patient. RA myxoma should also be differentiated from angiosarcoma, which has a predilection to involve



**Figure 3** Photograph of the explanted tumor, a lobulated, gelatinous mass. At the postoperative follow-up 10 weeks later, the patient reported significant symptomatic alleviation as well as marked weight loss.

the right atrium, contrary to myxoma. These malignant tumors have no stalks, tend to have direct involvement with the pericardium, causing pericardial effusion, and usually invade the RA wall and fill its chamber.<sup>17</sup> However, a high index of suspicion is needed to arrive at the correct diagnosis. Thus, early diagnosis and surgical resection could be lifesaving. In this regard, echocardiography has proved to be the most reliable, efficient, and cheapest noninvasive diagnostic test of choice, which can lead to early diagnosis and subsequent surgical tumor resection. Two-dimensional and transesophageal echocardiography have diagnostic sensitivity of 93.0% and 97.0% for evaluation of cardiac mass lesions, respectively. The potential diagnostic role of three-dimensional echocardiography and use of contrast echocardiographic perfusion imaging in the assessment of such lesions, however, are also worth mentioning. Three-dimensional echocardiography can yield important and precise information regarding the attachment site, vascularity, and homogeneity of intracardiac tumors as well as the presence of calcification in such masses.<sup>18</sup> Contrast echocardiographic imaging can help differentiation of the vascular and malignant tumors from thrombi and nonmalignant masses like myxoma.<sup>19</sup>

## CONCLUSIONS

RA myxomas are quite rare and difficult to diagnose. Once sufficiently enlarged, they can obstruct the tricuspid valve, leading to the development of signs and symptoms of right heart failure, peripheral edema, and ascites. Thus, RA myxoma should be considered a possible underlying cause of unexplained chronic transudative ascites. Echocardiography is the standard diagnostic evaluation of choice, which can lead to early diagnosis and aid in surgical tumor resection. We report this patient because of his unusually rare clinical manifestations and late diagnosis but with a fruitful, optimal surgical outcome.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2018.01.003>.

## REFERENCES

1. Azevedo O, Almeida J, Nolaso T, Medeiros R, Casanova J, Bartosch C, et al. Massive right atrial myxoma presenting as syncope and exertional dyspnea; case report. *Cardiovascular Ultrasound* 2010;8:23.

2. Reynen K. Cardiac myxomas. *N Engl J Med* 1995;333:1610-7.
3. Mendoza CE, Rosado MF, Bernal L. The role of interleukin-6 in cases of cardiac myxoma. Clinical features, immunologic abnormalities, and a possible role in recurrence. *Tex Heart Inst J* 2001;28:3-7.
4. Patrianakos AP, Parthenakis FI, Nyktari E, Kochiadakis GE, Koutsopoulos AV, Vardas PE. Right atrial myxoma: echocardiographic appearance. *Eur J Echocardiogr* 2008;9:422-3.
5. Sabolek M, Bachus-Banaschak K, Bachus R, Arnold G, Storch A. Multiple cerebral aneurysms as delayed complication of left cardiac myxoma: a case report and review. *Acta Neurologica Scandinavica* 2005;111:345-50.
6. Daga Calejero B, González Carretero M, Ortas Nadal R, Ferreira Montero I. Pulmonary embolism and myxoma of right atrial. *An Med Interna* 2005;22:478-80.
7. Fabijanić D, Rudez I, Kardum D, Radić M, Glavas D, Lozo P. Pulmonary embolism due to the right atrial myxoma. *Coll Antropol* 2006;30:933-6.
8. Jang KH, Shin DH, Lee C, Jang JK, Cheong S, Yoo SY. Left atrial mass with stalk: thrombus or myxoma? *J Cardiovasc Ultrasound* 2010;18:154-6.
9. Guhathakurta S, Riordan JP. Surgical treatment of right atrial myxoma. *Tex Heart Inst J* 2000;27:61-3.
10. Ojji DB, Ajiduku SS, Omonua OO, Abdulkareem LL, Parsonage W. A probable right atrial myxoma prolapsing through the tricuspid valve into the right ventricle; a case report. *J Med Case Reports* 2009;3:9328.
11. Oliveira R, Branco L, Galrinho A, Abreu A, Abreu J, Fiarresga A, et al. Cardiac myxoma; a 13- year experience in echocardiographic diagnosis. *Rev Port Cardiol* 2010;29:1087-100.
12. Novendstem SL, Silliman SL, Booth RP. Cardiovascular complications of atrial myxoma. *Hosp Physicians* 2001;8:39-42.
13. Burke AP, Virmani R. Cardiac myxoma. A clinicopathologic study. *Am J Clin Pathol* 1993;100:671-80.
14. Wold LE, Lie JT. Cardiac myxomas: a clinicopathologic profile. *Am J Pathol* 1980;101:219-40.
15. Bilku RS, Loubani M, Been M, Patel RL. Massive right atrial myxoma causing exertional dyspnoea. *Eur J Echocardiogr* 2008;9:130-2.
16. Sabageh D, Odujoko OO, Komolafe AO. Right atrial myxoma as a possible cause of hemorrhagic stroke and sudden death. *Niger Med J* 2012;53:102-4.
17. Kupsky DF, Newman DB, Kumar G, Maleszewski JJ, Edwards WD, Klarich KW. Echocardiographic features of cardiac angiosarcomas: the Mayo Clinic experience (1976–2013). *Echocardiography* 2016;33:186-92.
18. Zaragoza-Marcias E, Chen MA, Gill EA. Real time three-dimensional echocardiography evaluation of intracardiac masses. *Echocardiography* 2012;29:207-19.
19. Kirkpatrick JN, Wong T, Bednarz JE, Spencer KT, Sugeng L, Ward RP, et al. Differential diagnosis of cardiac masses using contrast echocardiographic perfusion imaging. *J Am Coll Cardiol* 2004;43:1412-9.