SAGE Open Medical Case Reports
Volume 11: 1–4

DOI: 10.1177/2050313X221149829
journals.sagepub.com/home/sco
SAGE

A rare case of rapidly progressive atrial myxoma masquerading as cough syndrome

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Abstract

Primary cardiac tumors are rare; however, atrial myxoma is one of the most common benign cardiac tumors. Myxomas may arise from any of the cardiac chambers but have a predilection for the left atrium. Its manifestations may mimic left heart failure or pulmonary hypertension either by causing blood flow obstruction or by mitral regurgitation. Patients can also present with thromboembolic features. Often, the diagnosis may be missed or delayed due to subclinical cardiac manifestations. Prompt diagnosis and surgical resection is necessary for favorable prognosis. Often, mitral valve replacement may be needed. We report a rare case of rapidly growing atrial myxoma in a 39-year-old female who presented with progressively worsening shortness of breath and cough with a previously normal echocardiogram 8 months prior. Her symptoms resolved after surgical resection and mitral valve replacement.

Keywords

Intracardiac tumors, primary cardiac malignancy, atrial myxoma, cough syndrome, cardiovascular, rare tumors, rapidly progressive tumors

Date received: 4 November 2022; accepted: 20 December 2022

Introduction

Primary cardiac tumors are rare, affecting less than 0.1% of the population.[1] Atrial myxomas are the most common primary tumors of the heart, often originating in the left atrium. Individuals between the ages of 30 and 60 are at a higher risk, with a greater prevalence among women relative to men.[2] Although it is a primarily benign tumor, in about 30%–40% of cases, it is associated with systemic arterial embolization.[3] The clinical presentation varies, ranging from an incidental finding on imaging in asymptomatic patients to shock or sudden cardiac death.[3,4] The growth rate of myxomas has been reported to be between 1.3 and 6.9 mm/month with an average rate of 4.9 mm/month. The diagnosis is confirmed with echocardiography or cardiac magnetic resonance imaging.[4] Prompt surgical resection is necessary following diagnosis, often with a favorable prognosis. We report a young female presenting with cough symptoms who was subsequently diagnosed with rapidly growing atrial myxoma with a growth rate of approximately 7.5 mm/month.

Case description

A 39-year-old female presented to our emergency department (ED) for the evaluation of progressive shortness of breath and dry non-productive cough that began 4 months prior to presentation. In the months prior to presentation, she had several presentations to her primary care provider with cough symptoms and had been managed symptomatically with antitussives without improvement or resolution of her symptoms. She had no fever, chills or night sweats, arthralgia, myalgia, or bipedal edema. A review of environmental, work and living arrangements could not explain her symptoms. She underwent extensive outpatient investigations including serial chest radiographs, tests for common respiratory viruses, and laboratory tests, which were all negative. Her symptoms subsequently progressed to shortness of breath on exertion, and then at rest, associated with orthopnea and paroxysmal nocturnal dyspnea. Cardiac etiology was not pursued initially as the patient had a cardiac evaluation by her cardiologist and had a normal echocardiogram (except for findings of a patent foramen ovale) 4 months prior to onset of symptoms (Figure 1). Two weeks prior to

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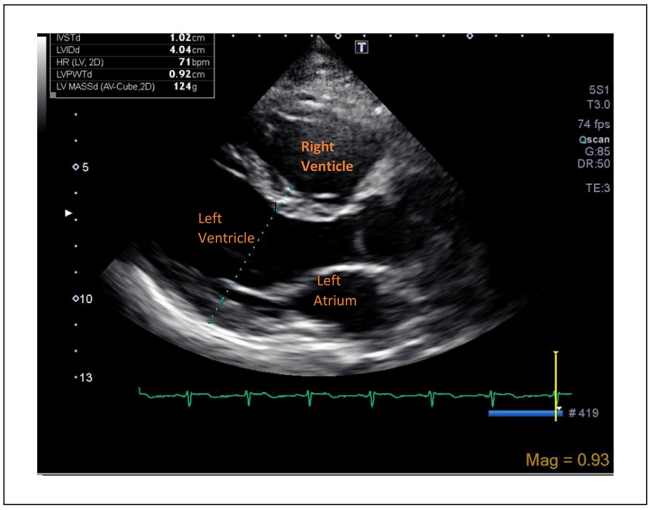

Figure 1. Parasternal long axis view of the transthoracic echocardiogram of the patient performed 8 months prior showing a normal left atrium and ventricle without any mass.

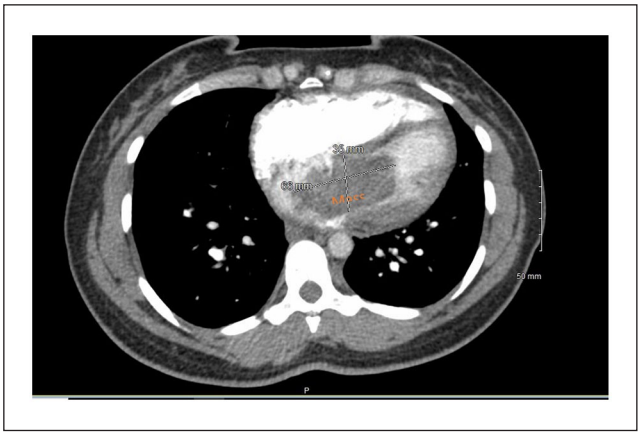

Figure 2. CT angiogram of the chest demonstrated a large intracardiac mass.

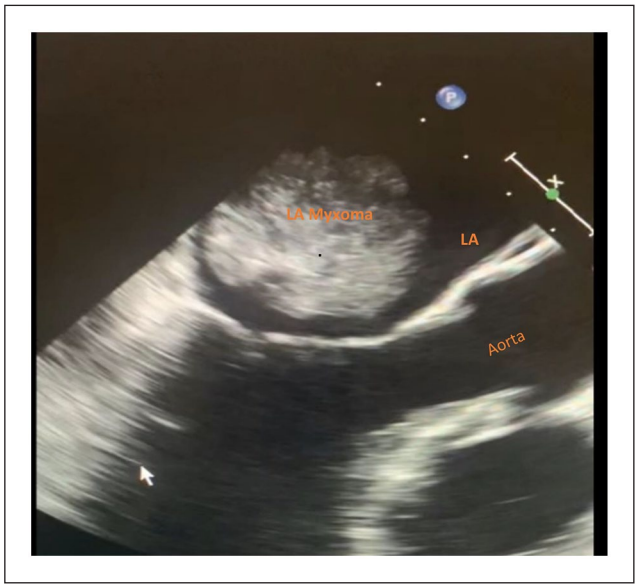

Figure 3. Transesophageal echocardiogram of the same patient showed a large echodense mass, the myxoma in the left atrium (LA).

current presentation, an outpatient chest computed tomography scan showed a 4.6 cm mass in the left atrium with an enlarged left subcarinal lymph node. She was then referred to a pulmonologist. She, however, had significant worsening of symptoms with severe limitation in her daily activities prior to the scheduled appointment with the pulmonologist prompting her presentation to the hospital ED for evaluation.

She had a past medical history of essential hypertension, patent foramen ovale on the echocardiogram done 8 months prior, and gastroesophageal reflux disease. Home medications included amlodipine, metoprolol, and pantoprazole. She was a non-tobacco smoker, consumed alcohol socially, and denied the use of illicit substances.

At presentation, physical examination revealed an uncomfortable young female, tachycardic (103 beats/min), borderline normotensive (95/63 mm Hg), she was afebrile, not tachypneic (18 cycles/min), oxygen saturation on room air was 98%. Cardiovascular exam revealed sinus tachycardia, no murmurs nor abnormal heart sounds, peripheral pulses were normal, there was no jugular venous distension, and she had no bipedal edema. Lung auscultations revealed bibasilar crackles; other physical examinations were normal.

Investigations done at presentation revealed mildly elevated brain natriuretic peptide (303 pg/mL), and normal troponin (12 pg/mL); EKG revealed sinus tachycardia with a ventricular rate of 106 beats/min. Previous chest CT scan was reviewed and a repeat CT chest not only showed a large intracardial mass measuring at 6.3 cm × 6 cm × 3 cm centered at the mitral valve but also present in the left atrium, with persistent left hilar lymph node, diffuse groundglass opacities in the lung parenchyma and pulmonary edema (Figure 2). She was admitted for the management of acute congestive heart failure secondary to an intracardiac mass. The cardiologist was consulted and she underwent transthoracic echocardiography, which showed a left atrial mass measuring 6.0 cm × 3.2 cm, pedunculated, extending from the left inferior interatrial septum prolapsing through the mitral valve and left ventricular ejection fraction of 55%. Preoperative planning with a transesophageal echocardiogram (Figure 3) revealed a large echo-dense mass in the left atrium bouncing into the mitral valve into the upper part of the left ventricle.

Differential diagnoses at the time were atrial myxoma, cardiac sarcoma (considering the hilar lymphadenopathy) or intramural blood clot, given the rapid growth rate. The cardiothoracic surgeon was consulted, and she underwent excision of the left atrial mass and closure of the previously diagnosed patent foramen ovale. Intraoperatively, the mitral valve appeared organically normal without leaflet prolapse but had annular dilation as well as some fixation and

Table 1. Previous studies documenting growth rates of atrial myxomas.

Study	Age, sex	Size on initial echo/ cardiac MRI (mm)	Size on ultimate echo (mm)	Growth rate (mm)	Time interval (months)
Alvarez et al.[12]	60, F	Absent	10 × 10	6.6	3
Iga et al.[13]	57, M	15 × 13	38 × 36	1.3 × 1.3	18
Ullah and MaGovern[14]	89, M	140.6	160	0.18	79
Karlof et al.[15]	58, M	Absent	150 × 30	13.6 × 3	11
Marinissen et al.[16]	65, M	No echo done	60 × 40	3.3 ×2.2	18
Roudaut et al.[17]		No echo done	5.05 × 40	6.9 × 5	8
Present case	39, F	Absent	60 × 32	7.5	8

MRI: magnetic resonance imaging; F: female; M: male.

restriction of the P3 segment of the posterior leaflet. Given these findings and the patient's young age, an attempt was made at mitral valve repair but was unsuccessful. This was primarily due to the restriction of the P3 segment of the posterior leaflet causing persistent mitral regurgitation following repair. Therefore, a cord-sparing mechanical valve replacement was completed. The excised mass appeared grossly irregular and dark brown gelatinous without a surface thrombus measuring 6.6 cm × 6 cm × 4.6 cm. Histopathology showed tumor cells within a myxoid stroma with areas of hemorrhage and inflammatory infiltrates consistent with a myxoma. She made a good recovery and was subsequently discharged home on metoprolol 12.5 mg twice daily and Coumadin. She was seen in the outpatient setting at 1, 3, and 7 months following discharge with no new symptoms or recurrence.

Discussion

Atrial myxoma has been referred to as a great mimicker.[3] They constitute the most frequent primary cardiac tumors, with a frequency of about 0.5 per million. It is more common in younger patients. About 10% of all myxomas are hereditary, autosomal dominant as part of the Carney complex syndrome.[5] This syndrome is associated with endocrine tumors, breast and skin myxomas, neurofibromatosis, and patchy pigmentation.[5]

Clinical signs of cardiac myxoma are diverse and typically ambiguous, which frequently leads to delayed therapy rather than prompt surgical removal. The classic triad of presentation is embolism, obstruction of ventricular filling, and constitutional symptoms. Embolic events are present in 30%–40% of myxoma cases.[6,7] The tissue is exceedingly friable with an increased tendency for easy detachment and embolization or adherence to a new site. Syncope or sudden cardiac death occurs due to pulmonary or systemic circulatory compromise or even due to atrioventricular valve obstruction.[3] Constitutional symptoms of fever, fatigue, joint pain, and weight loss are mediated by the tumoral release of interleukin 6.[2] Atypical presentation is not uncommon. In our case, the patient presented with nonproductive cough symptoms in the 4 months prior to her presentation to the hospital. This initially led to work up for respiratory illness. Her symptoms progressed to shortness of breath, paroxysmal nocturnal dyspnea, and orthopnea mimicking heart failure presentation. Her symptoms resolved after surgery, supporting the fact that they were related to the cardiac myxoma. A similar case by Bowman et al.[8] reported on a patient presenting with shortness of breath and non-productive cough associated with post-tussive syncopal episodes. These atypical presentations are noteworthy, as this knowledge could direct prompt evaluation in the minority of cases and improve patient mortality and morbidity outcomes.

Echocardiography is a non-invasive imaging modality capable of detecting atrial myxoma and assessing its location and size, especially for preoperative planning.[4] Cardiac magnetic resonance imaging may also be used but presents cost constraints compared to echocardiography.[4] Evidently, not all patients presenting with cough syndrome require cardiac evaluation. However, patients presenting with progressive cough symptoms refractory to usual antitussives and conservative management especially if associated with dyspnea and orthopnea should undergo further evaluation. Compared to transthoracic echocardiogram (TTE), transesophageal echocardiography (TEE) has greater sensitivity in the diagnosis of atrial myxoma, particularly for smaller tumors but is rarely required except in preoperative planning.[4]

Roskell and Biddolph investigated if the tumor growth rate was precipitated by cellular proliferation by examining five clinical cases based on immunohistochemical assessments.[9] The authors noted that rapidly growing myxomas are likely due to extracellular matrix alterations and mesenchymal differentiation and not cellular growth.[9] Gewehr et al. in their case report suggested that multiple foci of hemorrhage and thrombosis inside the myxoma could have contributed to the high tumor volume. Thrombus is a differential to consider in cardiac tumors and has been reported admixed in myxomas with the potential to overestimate the tumor size.[10,11] The histopathology in our case revealed extensive areas of hemorrhage, and inflammatory infiltrates with no thrombus. The exact mechanisms promoting rapid growth in myxomas are based on a few subjects but reports imply that both cellular proliferation and hemorrhagic foci may be responsible.

Myxomas are generally thought to be slow-growing cardiac tumors. Previous studies have made attempts to provide

growth rate estimates as shown in Table 1. Alvarez et al. reported a 6.6 mm growth rate per month. In their case, the patient was found to have atrial myxoma, which was absent on a two-dimensional echocardiogram obtained 3 months prior to a radiofrequency ablation procedure for atrial fibrillation.[12]

Two case reports noted a monthly growth rate of 13 and 0.18 mm per month, respectively, surveilled on serial echocardiography.[13,14] Furthermore, three reports described atrial myxoma after coronary artery bypass graft surgery at a rate of 4.4, 3.3, and 13.6 mm/month.[15–17] In our case, the growth rate was estimated at 7.5 mm/month. We even think the rate may be more rapid than 7.5 mm/month because she had florid symptoms within a short time frame, as well as chest CT obtained 2 weeks apart showing a 4.6 cm mass compared to 6.3 cm seen on repeat CT at her presentation. This further correlated with the size determined by TTE and confirmed intraoperatively.

In general, myxoma growth rate estimates are made with the assumption that it develops linearly, while in fact, growth may be exponential. Furthermore, patients diagnosed with left atrial myxoma should undergo surgical excision as the risk of cardioembolic and related complications outweighs the risks associated with surgery.

Conclusion

This case highlights the nonspecific presentation of atrial myxoma making diagnosis sometimes elusive. We also report on the potential for rapid growth. In the absence of a history of cardiac disease, cardiac investigations for the etiology of cough symptoms may not be pursued. This potentially delays diagnosis with patients ultimately presenting with profound signs of cardiac dysfunction. Although not all patients presenting with cough syndrome require cardiac evaluation, however, we recommend that an echocardiogram should be considered in a young patient presenting primarily with refractory cough symptoms in the right clinical setting.

Author contributions

I.T.S., G.E., and S.C.A. drafted, edited, and revised the article. N.A. revised and edited the article. The authors participated in the care of the patient. All authors approved the submitted article.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethical approval

Our institution does not require ethical approval for reporting individual cases.

Informed consent

Written informed consent was obtained from the patient for her anonymized information to be published in this article.

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