

Asymptomatic Recurrence of Right Ventricular Myxoma after Excision of Four-Chamber Myxoma



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INTRODUCTION

Cardiac myxoma is the most common primary cardiac tumor. Treatment involves surgical resection and continued monitoring with echocardiography thereafter. There are no guidelines on the frequency and duration of monitoring. Presented here is the case of a 46-year-old man who underwent resection of four-chamber cardiac myxoma 15 years previously. He had stopped surveillance 3.5 years previously because the annual echocardiogram for the past 10 years did not show any evidence of recurrence. However, routine echocardiography revealed a right ventricular mass consistent with myxoma on pathology. This case not only highlights the rarity of four-chamber myxoma and its recurrence but also teaches an important lesson to continue surveillance in asymptomatic patients even after many normal echocardiograms.

CASE PRESENTATION

A 46-year-old asymptomatic man with a medical history of nonischemic cardiomyopathy and cardiac myxoma involving all four chambers (confirmed on the operative report) resected 15 years previously presented to the primary care clinic for a routine checkup. His last transthoracic echocardiogram had been obtained 3.5 years earlier and did not demonstrate recurrence of myxoma. The left ventricular ejection fraction had been stable at about 30%. There was mild tricuspid regurgitation and moderately depressed right ventricular systolic function. The patient had previously undergone annual transthoracic echocardiography for surveillance of recurrence of myxoma until it was decided to stop 3.5 years earlier, as the patient had been doing well clinically, with no evidence of recurrence on echocardiography. Routine transthoracic echocardiography was ordered at the primary care visit, which revealed a 4.6×2.9 -cm mass in the right ventricle (Figure 1, Videos 1 and 2). Maximum tricuspid and pulmonic gradients were 21 and 4 mm Hg, respectively. There was no pulmonary hypertension.

The patient was admitted to the hospital for further diagnostics. On admission, the patient was noted to have normal sinus rhythm with no cardiac murmurs and no neurologic deficits, only freckling of his skin, which raised the possibility of an autosomal-dominant syndrome called Carney complex in the context of cardiac myxoma. Cardiac magnetic resonance was performed and revealed a 4.0×3.0 -cm, rounded, mobile mass adherent to the right ventricular free wall involving the papillary muscles. The mass had a 1.0×1.5 -cm finger-like projection into the right ventricular outflow tract (Figures 2 and 3, Video 3). Mild tricuspid regurgitation was seen. The pulmonic valve was normal in appearance. Perfusion imaging revealed heterogeneous uptake in the center of the mass, in keeping with a vascular supply. These findings were consistent with recurrence of myxoma.

The patient was taken to the operating room, where redo sternotomy was performed. The patient was put on cardiopulmonary bypass and underwent right atriotomy in which the right ventricular mass was entirely excised at its attachment to the right ventricle and papillary muscle. The anterior leaflet of the tricuspid valve was then sewn back onto the papillary muscle. A tricuspid annuloplasty ring (Edwards #28; Edwards Lifesciences, Irvine, CA) was sewn onto the annulus. No damage was done to the pulmonic valve. Intraoperative transesophageal echocardiography (Figures 4 and 5, Videos 4–6) was performed and confirmed the removal of the right ventricular mass with some mild to moderate tricuspid regurgitation. Postoperative maximum tricuspid and pulmonic gradients were 4 and 2 mm Hg, respectively. The patient's postoperative course was uneventful, and the patient was discharged. Pathology of the right ventricular mass was consistent with myxoma (Figures 6 and 7). Given the freckling of the skin, the prior four-chamber myxoma, and now recurrence, Carney complex was considered, but the patient had no other history of extracardiac tumors or endocrine abnormalities. In addition, he denied any family history of myxoma or other tumors. Eight months later, repeat transthoracic echocardiography showed no evidence of recurrence.

DISCUSSION

Cardiac tumors are rare, and primary cardiac tumors are rarer still. Of the primary cardiac tumors, cardiac myxoma is the most common.¹ Myxomas are thought to arise from proliferation of the multipotent mesenchymal cells. Cardiac myxoma has a 2:1 female predominance and a mean age of presentation of 53 years.¹ Myxomas are usually sporadic, but there are familial cases.² A combination of myxoma, skin pigmentation, and hyperactivity of the adrenal or testicular glands was described as Carney complex in 1985.³

Clinical manifestations of myxoma are usually dependent on the size, location, and mobility of the tumor. They range from causing no symptoms to causing high morbidity and mortality. Patients may

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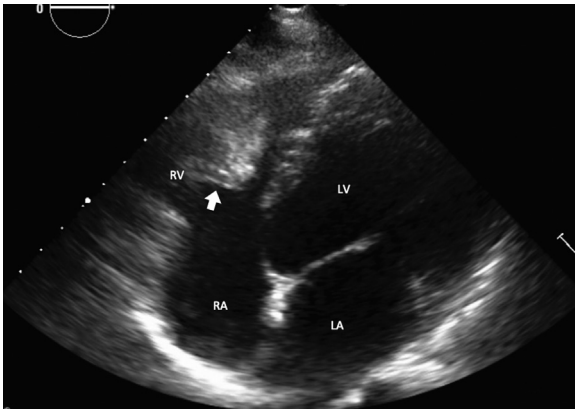


Figure 1 Four-chamber view of right ventricular mass (arrow) on transthoracic echocardiography. LA, Left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

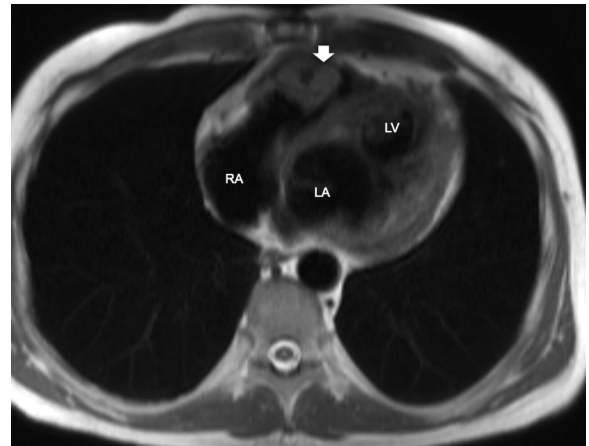


Figure 3 Cardiac magnetic resonance image (cross-sectional view) showing right ventricular mass (arrow). LA, Left atrium; LV, left ventricle; RA, right atrium.

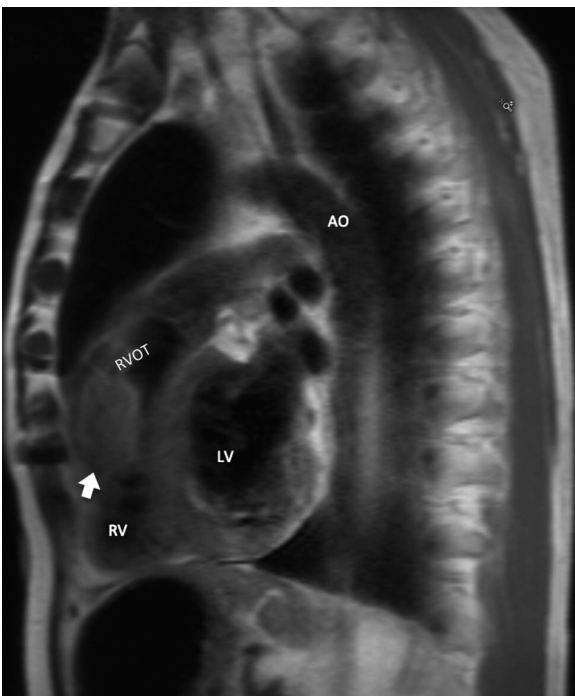


Figure 2 Cardiac magnetic resonance image (sagittal view) showing right ventricular mass (arrow) projecting into the right ventricular outflow tract (RVOT). AO, Aorta; LV, left ventricle; RV, right ventricle.

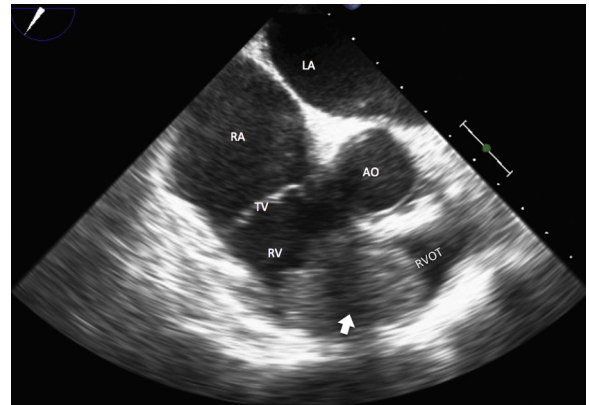


Figure 4 Transesophageal echocardiogram (right ventricular inflow and outflow view) showing a right ventricular mass (arrow). AO, Aorta; LA, left atrium; RA, right atrium; RV, right ventricle; RVOT, right ventricular outflow tract; TV, tricuspid valve.

experience various sequelae of systemic embolization when the tumor is on the left side. When located on the right side, myxoma can also embolize, causing pulmonary hypertension and acute right ventricular failure. Tumors may also cause obstructive symptoms, including dyspnea, recurrent pulmonary edema, syncope, and even sudden death. Additionally, patients may experience constitutional symptoms, including fatigue, fever, arthralgia, and myalgia.²

Cardiac myxoma is most commonly found in the left atrium (75%), followed by the right atrium (20%), right ventricle (3%–4%), and left ventricle (3%–4%).⁴ They are rarely multiple and multifocal. There are only two reports of four-chamber myxoma in the literature.^{4,5} When there are multiple, multifocal masses, especially in ventricular

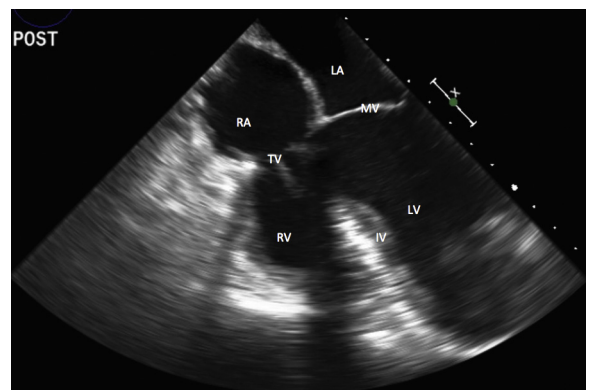


Figure 5 Transesophageal echocardiogram (four-chamber view) after right ventricular mass was resected. IV, Interventricular septum; LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle; TV, tricuspid valve.



Figure 6 Gross specimen of right ventricular myxoma, consisting of multiple fragments of soft, myxoid, gelatinous tissue, with loosely adherent blood clot.

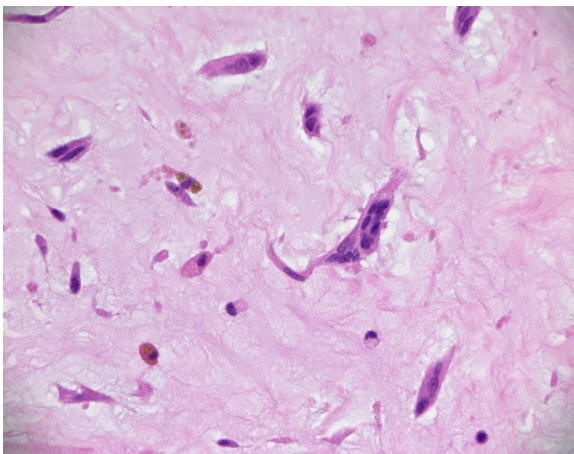


Figure 7 Hematoxylin and Eosin stain of specimen (200× original magnification). Cardiac myxoma cell morphology can be variable, with irregular spindle and stellate shapes and abundant eosinophilic cytoplasm.

chambers, one should consider Carney complex, which has an autosomal-dominant inheritance pattern.⁶ This patient did not have a family history or extracardiac manifestations to suggest Carney complex. Previous case reports have attempted to estimate the growth rate of myxoma, which has been highly variable. The rate of growth ranges from no growth in 15 years to 1.36 cm/mo.⁷⁻¹⁰

Diagnosis relies mainly on different imaging modalities, including echocardiography, computed tomography, and cardiac magnetic resonance. Two-dimensional transthoracic echocardiography is inexpensive and allows easy visualization of the tumor. Transesophageal echocardiography has better image quality and can visualize the atria, which are inadequately seen on transthoracic echocardiography. Both computed tomography and cardiac magnetic resonance provide information on the extent of the tumor and other anatomic information, which is valuable for operative planning. The diagnosis of myxoma is confirmed only on pathology, which usually contains an acid-mucopolysaccharide-rich stroma mixed with polygonal cells.²

Cardiac myxoma should be resected promptly after the diagnosis is made on imaging given the myriad complications that may occur with delay. Patients need to undergo sternotomy and be put on cardiopul-

monary bypass for surgery, with resection of the tumor generally done through atriotomy. The root of the pedicle should be completely excised to prevent recurrence. Surgery is generally curative and has a low operative mortality rate (3%).^{11,12} Recurrence is thought to be due to incomplete resection or de novo genesis. The recurrence rate of myxoma has been reported to range from 1% to 5%.¹¹⁻¹³ On the basis of a review by Lone *et al.*,¹⁴ long-term echocardiography is recommended for follow-up. However, Bjessmo and Ivert¹⁵ recommended surveillance echocardiography to be limited after uncomplicated resection. Given the potential for catastrophic morbidity and mortality of untreated myxoma, it would be most prudent to continue yearly surveillance indefinitely.

CONCLUSION

Cardiac myxoma can be sporadic or familial. The definitive treatment involves surgical resection, but there is a low risk for recurrence. This case of myxoma recurring in an asymptomatic patient even after a decade of normal results on surveillance transthoracic echocardiography highlights that in fact indefinite monitoring is necessary.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.case.2017.07.002>.

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