

Right Atrial Myxoma Associated with Atrial Septal Defect: A Case Report and Review of the Literature

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We describe a case of right atrial myxoma with mild cyanosis due to a right-to-left shunt at the atrial level. The patient was a 31-year-old woman with a 10-month history of easy fatigability and shortness of breath. Echocardiography showed a right atrial tumor producing a partial dynamic tricuspid obstruction. Digital subtraction angiography via the superior vena cava disclosed a mass lesion which occupied the right atrium with early visualization of the ascending aorta. Successful excision of the tumor and repair of the atrial septal defect totally relieved her presenting symptoms. In a rare association of a right atrial myxoma with atrial septal defect, preoperative evaluation and operative management are discussed. (Ann Thorac Cardiovasc Surg 2001; 7: 166–9)

Key words: right atrial myxoma, atrial septal defect, right-to-left shunt, cyanosis, intravenous digital subtraction angiography

Introduction

Right atrial myxoma is a relatively uncommon lesion, comprising 18% of all myxomas.¹⁾ It has been emphasized that they presented with signs and symptoms of right heart failure and pulmonary embolism.^{1,2)} Here, we report a case of right atrial myxoma associated with atrial septal defect. A right-to-left shunt through the interatrial defect resulted in mild cyanosis. The coexistence of these lesions is very rare.

Case Report

A 31-year-old woman was admitted to our institution complaining of easy fatigability and shortness of breath for the previous 10 months. The patient's difficulty in breathing became particularly acute for the few months prior to hospitalization. She had a nonproductive cough

but no hemoptysis. There was no past history of cardiac or pulmonary disease. On physical examinations, the heart rate was regular at 92 beats/min, and blood pressure was 102/64 mmHg. She was afebrile. The lips and fingernails were mildly cyanotic but no clubbing. The chest was clear to auscultation. Heart sounds were normal, and there were no murmurs except for a trivial diastolic murmur at the lower left sternal border. The liver was not palpable. There was no evidence of right heart failure.

A complete blood count showed an upper limit of the normal range with a hemoglobin level of 15.7 g/dL, and a hematocrit value of 49.4%. Serum cross-reacting protein (CRP) was increased to 8.67 mg/dL and lactate dehydrogenase (LDH) to 468 IU/L. Arterial blood gas on room air showed severe hypoxia with an oxygen tension of 49.0 mmHg and oxygen saturation of 81.8%. All other routine laboratory data were within normal limits.

The chest X-ray showed moderate cardiomegaly, mainly on the right side. Electrocardiography demonstrated right atrial enlargement and incomplete right bundle-branch block. Transthoracic echocardiography revealed a large right atrial mass which moved into the tricuspid orifice during diastole. Pulmonary perfusion scintigraphy revealed no evidence of a perfusion defect. Digital subtraction angiography was performed via the

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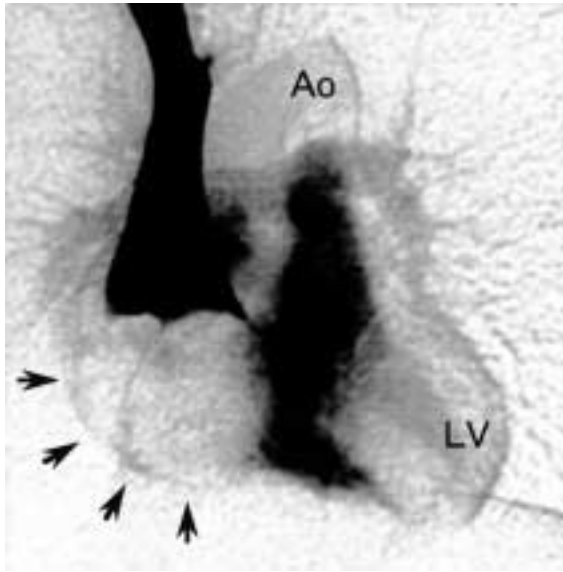


Fig. 1. Digital subtraction angiography with superior vena caval injection. Posteroanterior projection showing a large filling defect in the right atrium (arrows). Early visualization of the left heart with a partial dynamic tricuspid obstruction indicated interatrial right-to-left shunting.

right cephalic vein. A superior vena cava injection of contrast material revealed a filling defect of the right atrium. Stagnation of contrast material in the superior vena cava and early visualization of the left heart indicated obstructed flow through the tricuspid valve and a right-to-left shunting at the atrial level (Fig. 1). Further complete catheterization was not carried out, because it might have caused paradoxical or pulmonary embolization. Although repetitive echocardiography was performed, no shunt signal was seen.

A prompt operation was indicated, and the patient was taken to the operating theater. After institution of cardiopulmonary bypass, utilizing a direct cannulation into the superior vena cava and a femoral approach to the inferior vena cava, the aorta was cross-clamped with cardioplegic arrest. The right atrium was opened carefully. A large lobulated tumor almost filling the entire right atrium was encountered (Fig. 2a). It had a broad-based attachment to the dorsal free wall of the right atrium and the interatrial septum on the inferolateral aspect of an atrial septal defect. The size of the defect was 2.0×1.5 cm (Fig. 2b). Because it was extensively attached to both the right atrium and interatrial septum, the tumor was completely excised together with the atrial wall and the interatrial septum. The left atrium was inspected through the surgically enlarged interatrial septum defect, and there was no evidence of further tumors. The interatrial

communication was closed with a bovine pericardial patch, and the excised atrial free wall was closed directly. The tumor measured 7.5×6.3×5.6 cm and weighed 86 g (Fig. 2c), and its microscopic appearance was consistent with myxoma.

The postoperative course was quite uneventful. The arterial blood gas showed normal oxygenation. The patient has returned to normal activity with total resolution of the preoperative symptoms. She remains completely asymptomatic 3 years after surgery with no evidence of recurring tumor.

Discussion

Primary cardiac tumors are uncommon, with an incidence of between 0.001 and 0.28% in reported or collected autopsy series.¹⁾ Cardiac myxoma is the most common, comprising approximately 50% of all benign cardiac neoplasms in adult. Approximately three fourths of them are situated in the left atrium, and 18% in the right atrium.¹⁾ They may be multicentric within a single chamber or biatrial. An association of right atrial myxoma and atrial septal defect is very rare. To date, six cases of right-to-left shunt caused by this combination of lesions have been reported in the English-language literature (Table 1).³⁻⁸⁾ Most of them were in a single case report or in only a small series of myxomas. Of the 351 patients included in the largest published series of myxomas reviewed by Abenoza and Sibley, none were complicated by atrial septal defect.⁹⁾ Thus the incidence of the coexistence of these two lesions remains unclear. Nine additional cases of right atrial myxoma with patent foramen ovale have also been reported.¹⁰⁻¹⁸⁾ Such cases may also have interatrial right-to-left shunting.

The most common symptom of cardiac myxoma is congestive heart failure, followed by embolization.^{1,2)} These symptoms, usually based on the location of the tumors, vary with their size, shape, and also with the physical activity and position of the patient. In right atrial myxoma the presentations may include ascites, hepatomegaly or peripheral edema due to right heart failure. Pulmonary embolization may also occur. Other clinical symptoms are vague constitutional ones such as malaise, low grade fever and weight loss.^{1,2)} In this case of right atrial myxoma, the patient showed mild cyanosis without any remarkable symptoms of right heart failure. Pulmonary perfusion scintigraphy was normal, which excluded the possibility of pulmonary embolization. Interatrial right-to-left shunt was suggested when a patient without pulmonary disease showed cyanosis. This

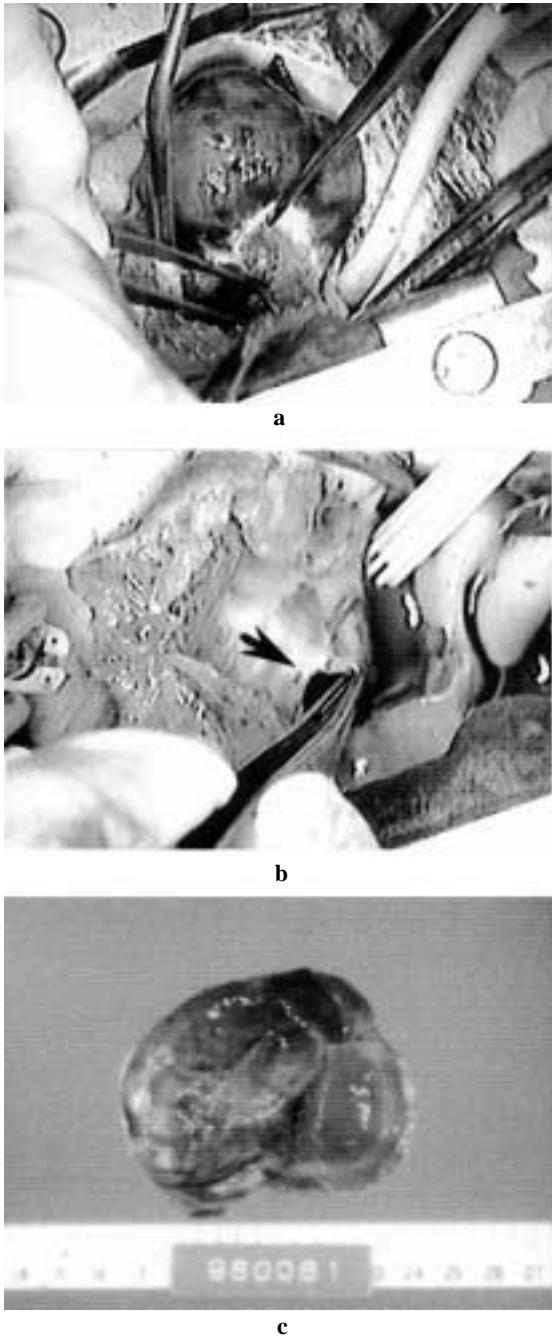


Fig. 2. Right atrial myxoma with atrial septal defect. Intraoperative photographs showing the right atrial mass (a), and the atrial septal defect (b: arrow). Gross specimen of the tumor removed from the right atrium (c).

decompression of the right atrium due to a right-to-left shunt may explain the lack of right heart failure in the present case.

Recent progress in diagnostic modalities in cardiology, including echocardiography, computed tomography, and magnetic resonance imaging, is now allowing diagnosis of primary cardiac tumors without cardiac cath-

eterization or angiography. However, the latter two methods remain effective to diagnose an interatrial shunt. In this case, echocardiography readily showed a right atrial mass lesion, but no shunt signal could be demonstrated. We then employed intravenous digital subtraction angiography. A venous catheter was inserted using a right cephalic venous approach and placed in the superior vena cava. It showed stasis of contrast material in the superior vena cava and early visualization of the aorta as well as a right atrial filling defect. We concluded that the tricuspid partial obstruction due to the right atrial tumor and subsequent elevation of the right atrial pressure had resulted in significant right-to-left shunt through an interatrial communication.

In none of the previously reported six cases of right atrial myxoma with atrial septal defect, echocardiography could detect an interatrial shunt signal preoperatively.³⁻⁸⁾ In all of these earlier patients, cardiac catheterization and angiography were employed to confirm the existence of a myxoma and an interatrial right-to-left shunt. Cardiac catheterization in patients with known myxoma is potentially hazardous for embolization.⁵⁾ Intravenous digital subtraction angiography is completely safe, because only the tip of the catheter is moved forward within the superior vena cava. Fluoroscopy is used to confirm the tip position so that it does not reach the tumor. We emphasize that an interatrial shunt should be demonstrated preoperatively in order to exclude other disorders which cause cyanosis. Intravenous digital subtraction angiography is a quite easy and effective diagnostic modality in patients with this combination of lesions.

Excision of atrial myxomas using cardiopulmonary bypass has been established with generally good clinical results. However, one must use a special technique based on the location of the tumors to control embolization. In a case of right atrial myxoma, Kabbani et al. recommended retrograde cannulation of the inferior vena cava through the femoral vein to avoid any possible fragmentation of the tumor.¹⁹⁾ Lee et al. indicated that the right atrium could be opened first of all using a suction device and the inferior vena cava then cannulated under direct vision after most of the tumor had been removed.¹⁶⁾ In this case, we employed a femoral venous approach for the inferior vena caval cannulation. There was no evidence of tumor embolization during or after the operation.

Resection of the tumor constitutes the definitive treatment for cardiac myxomas. There is controversy as to whether excision of the area of the atrial wall or interatrial septum where the atrial myxoma attaches is really

Table 1. Cases of right atrial myxoma associated with atrial septal defect

Author	Age	Sex	O ₂ sat (PaO ₂)	Cardiac catheterization, angiography	Postoperative course
Taber and Lam	51	F	76%	yes	asymptomatic
Willman, et al.	61	F	66-88% varying with position	yes	asymptomatic
Marpole, et al.	73	M	64%	yes	cerebral embolism
Talley, et al.	49	F	43 mmHg	yes	cerebral embolism
Powers, et al.	48	M	-	yes	asymptomatic
Natarajan, et al.	63	F	77%	yes	asymptomatic

O₂ sat: oxygen saturation; PaO₂: arterial oxygen partial pressure.

necessary.^{6,16)} Some surgeons believe complete resection of the cardiac wall at the site of tumor attachment is necessary to prevent recurrence of the tumor.^{14,19)} However, there is no evidence in the literature that radical resection of the atrial wall or interatrial septum reduces the possibility of recurrence.²⁰⁾ Right atrial myxomas tend to have a broad-based attachment to the atrial wall or septum.²⁾ In this case, because of the extensive attachment of the tumor to the right atrial wall and the interatrial septum, the tumor was resected together with the underlying atrial wall and interatrial septum. No myxoma has recurred during the three-year follow-up period.

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