A Giant Right Atrial Mass with Concomitant Pulmonary Embolism

Fatemeh Mirrazeghi, Mohammad Esmaeil Rezaei, Anita Sadeghpour, Alireza Alizadeh Ghavidel, and Kambiz Mozaffari

1 Echocardiography Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University Medical Sciences, Tehran, IR Iran
2 Heart Valve Disease Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, IR Iran
3 Rajaie Cardiovascular Medical and Research Center, Iran University Medical Sciences, Tehran, IR Iran

*Corresponding author: Mohammad Esmaeil Rezaei, Echocardiography Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University Medical Sciences, Tehran, IR Iran, E-mail: mohammad.rezaei@yahoo.com

Received 2016 July 30; Accepted 2016 August 15.

Abstract

Myxomas are the most common primary cardiac tumors. They are mostly located in the left atrium, but 15% to 20% of them arise from the right atrium (RA). We herein describe a 22-year-old man with a giant RA mass and simultaneous pulmonary embolism. The mass was detected accidentally on transthoracic echocardiography in preoperative workup for an elective noncardiac surgery and was confirmed with multimodality imaging. The patient underwent surgical removal of the mass and concomitant pulmonary artery embolectomy. Histopathological examination confirmed the diagnosis of an RA myxoma. He had an uneventful recovery and was asymptomatic after 24 months of follow-up. This case is a villous RA myxoma, which is a very rare subtype with a high tendency to pulmonary embolism. We report this case as an unusual location of a very large myxoma and emphasize the role of multimodality imaging in the preoperative management of this patient as cardiac computed tomography angiography confirmed the associated pulmonary embolism.

Keywords: Echocardiography, Cardiac Mass, Cardiac CT, Cardiac MR

1. Introduction

Cardiac tumors are classified into primary and secondary or metastatic. Secondary or metastatic tumors are 20 - 40 times more common than primary tumors (1).

Primary cardiac neoplasms are uncommon and occur with an incidence rate of 0.001% - 0.2%. Approximately 50% of benign cardiac tumors are myxomas; they are mostly polypoid and are located in the left atrium. The right atrium (RA) is an unusual location inasmuch as it is observed only in 15% - 20% of myxoma cases, and occasionally the ventricles are involved (2, 3). Commonly, these tumors arise from the atrial septum. They rarely originate from the junction of the inferior vena cava (IVC) and the RA or from the Eustachian valve (4). The villous RA myxoma is a very rare subtype with a high tendency to pulmonary embolism (2).

In the familial form of cardiac myxomas, there is an increased risk of the multiple masses and recurrence after surgical excision (5-7).

The embolic phenomena (as a result of fragmentation and thromboembolism) occur from the tumor surface; however, sudden death, syncope, and hemoptysis may also occur. Transthoracic (TTE) and transesophageal (TEE) echocardiographic examinations have 95% and 100% sensitivity for the diagnosis, respectively (7). In addition, cardiac magnetic resonance (CMR) or cardiac computed tomography (CT) may be useful to demonstrate the point of fixation and associated complications.

In cine CMR with low signal intensity, the mass’s origin, size, mobility, extension into the different cardiac chambers, and hemodynamic obstruction can be depicted. On cardiac CT, myxomas usually have heterogeneous low attenuation. Calcification can also be found frequently (8-12).

We report a giant RA myxoma with simultaneous pulmonary embolism in a 22-year-old man, assessed via multimodality imaging.

2. Case Presentation

A 22-year-old man was admitted for elective hemorrhoid surgery. The patient was healthy looking, nondiabetic, normotensive, but he was a smoker and an oral opium user. There were no specific symptoms or findings in the examinations of the heart and the other systems. He was referred to a cardiologist for nonspecific repolarization change in the right precordial leads on ECG. On transthoracic echocardiography, a large mass in the RA was detected and the patient was referred to our center for complementary evaluation. The lab tests revealed increased ESR and CRP.

TTE and TEE showed normal left ventricular size and function, severe right ventricular enlargement with mild
systolic dysfunction, moderate RA enlargement, and a large highly mobile multilobulated heterogeneous mass (6.5 × 5 cm in size) in the RA with attachment to the interatrial septum (IAS) at the IVC-RA junction site, with a relatively narrow stalk. Moreover, there was moderate tricuspid regurgitation and mild tricuspid stenosis due to a protruding RA mass across the tricuspid valve in diastole. No visible mass was detected in the main pulmonary artery and the proximal portions of its branches. Additionally, there was a tiny patent foramen ovale. 3D echocardiography was thereafter done for a precise evaluation of the size of the mass and its relation to the cardiac chambers (Figures 1 – 3).

Figure 1. Transthoracic Echocardiography Shows a Large Multi-Lobulated Right Atrial (RA) Mass, Which Has Filled Most of the RA.

Figure 2. Transesophageal Echocardiography in the Bicaval View Shows a Large Multilobulated Right Atrial (RA) Mass, Attached to the Lower Portion of the Interatrial Septum.

CT angiography of the pulmonary artery was performed for possible pulmonary embolism and showed a filling defect in the right lower lobe pulmonary artery branch, in favor of chronic or subacute thromboemboli, and a round heterogeneous mass in the RA, abutting the IAS (Figure 4).

Figure 3. 3D Echocardiography of the Right Atrial (RA) Mass.

Figure 4. Cardiac Computed Tomography Shows a Large Round Heterogeneous Right Atrial (RA) Mass (Arrow Head), Abutting the Interatrial Septum.

CMR revealed a large and relatively heterogeneous mobile mass lesion in the RA attached to the IAS-IVC junction and the RA site, measuring about 63 × 56 mm in size and protruding through the tricuspid valve. Small low-signal foci were seen in this lesion due to tiny calcification. These features were highly suggestive of a large myxoma. Further, a filling defect was noted at the distal portion of the right pulmonary artery, suggestive of pulmonary thromboembolism.

Surgery was performed with the aid of standard car-
diopulmonary bypass. The large tumor with a narrow stalk (6 mm in size), adherent to the IAS near the IVC, was completely removed with about a 1-cm margin of the IAS through right atriotomy. Right pulmonary artery embolectomy was performed, and 2 masses (2 × 1 cm and 1 × 1 cm in size, correspondingly) were removed. The tricuspid valve was repaired, and ring annuloplasty was done. The surgery was performed without complications. Intraoperative TEE showed no residual RA mass and successful tricuspid valve repair. The patient remained hemodynamically stable, had an uneventful recovery, and was discharged from the hospital after 5 days.

Histopathological examination showed an RA mass, consisting of a gelatinous polypoid creamy brown piece with scattered stellate cells, and smooth muscle cells in a myxoid background rich in extracellular matrix (Figure 5) compatible with an RA myxoma.

3. Discussion

In our patient, a myxoma was diagnosed accidentally together with pulmonary embolism as a rare but important complication of RA myxomas.

The complete removal of both atrial and pulmonary tumors is important to avoid pulmonary complications and failure to resolve the symptoms (4).

Although TTE and TEE provide a good visualization of the mass with sensitivity of 95% and 100%, respectively, 3D TEE may be helpful in surgery planning insofar as it confers a better definition of the mass’s real shape, position, and attachment. In our patient, an RA mobile mass originating from the IAS was detected via TTE. The mass’s size, boundaries, and relationship with the septum were assessed via 3D TEE in detail.

Magnetic resonance imaging and CT provide information regarding the tissue’s characteristics and allow an excellent overview of cardiac and paracardiac morphology as well as an evaluation of possible pulmonary embolism. In our patient, CT revealed pulmonary embolism in the distal portion of the right pulmonary artery.

The treatment of choice for a myxoma is surgical removal. Complete resection of the tumor and its margin is essential to cure and prevent recurrence. Furthermore, in our case, pulmonary embolectomy was indicated to prevent right ventricular dysfunction in the future due to chronic pulmonary hypertension.

The recurrence rate of sporadic tumors is very low and stands between 1% and 3%, and the operative mortality rate ranges from 0% to 3% in multiple series. The survival rate after surgery is high.

3.1. Conclusions

In our patient, the type and location of the RA mass were unusual for a myxoma. Be that as it may, it should always be considered an important differential diagnosis. Echocardiography is the 1st-line imaging modality for cardiac masses, while cardiac CT and CMR can provide more information regarding the associated complications such as pulmonary embolism and tissue characterization of the mass, which has an important impact on the type of cardiac surgery.

References
