An Atypical Right Atrial Myxoma With Spontaneous Rupture
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Summary

Typically, cardiac myxomas arise from the interatrial septum at the border of the fossa ovalis in the left atrium, whereas atypical right atrial myxoma, especially with spontaneous rupture, is extremely rare. Here we report the diagnostic evaluation and successful surgical resection of an atypical myxoma with spontaneous rupture arising from the posterior wall of the right atrium in a 34-year-old male. (Int Heart J 2016; 57: 262-264)

Key words: Cardiac tumor, Cardiac magnetic resonance

Primary cardiac tumors are very rare, with an autopsy incidence ranging from 0.001% to 0.030%.1 Myxomas account for 50% of all benign tumors of the heart. They occur mainly in the third to sixth decade of life, affecting women more frequently than men. The majority of cardiac myxomas are located in the left atrium (75%), and rarely in the right atrium (RA, 10% to 20%).2 Most atrial myxomas have a stalk frequently attached to the interatrial septum at the border of the fossa ovalis and then in descending order of frequency from the posterior wall, anterior wall, and atrial appendage.3 Echocardiograms and cardiac magnetic resonance (CMR) might be of important significance in the diagnosis and guiding treatment of cardiac tumors.

Case Report

A previously healthy 34-year-old man was admitted to our hospital due to chest distress and polypnea which continued for 1 month. There was no history of coronary artery disease or trauma, and his family history was negative. On physical examination, a systolic grade II-VI blowing murmur was detected on the apex. His blood pressure was 120/60 mmHg and he had a regular pulse of 70 beats/min. The results of an examination of other systems were normal. The ECG revealed incomplete right bundle branch block and V2-V6 lead ST-segment elevation. The laboratory findings were unremarkable.

Transthoracic echocardiography (TTE) revealed the following: biatrium and right ventricular enlargement, moderate tricuspid valve regurgitation, a large echogenic mobile mass (5.3 cm × 4.2 cm) could be found in the RA, and a tortuous banded isoechoic structure attached to the mass protruding into the right ventricle during diastole (Figure 1B) and entering into the RA during systole (Figure 1A). However, left ventricular systolic function was normal with an ejection fraction of 61%.

Cardiac magnetic resonance (CMR) was performed to further characterize the mass and confirmed a mass with spontaneous rupture arising from the posterior wall of the RA on a broad base (Figure 2 A, B, C). CMR could clearly show the location of the rupture (Figure 2, white arrow). The mass showed isointense signal intensity on T1-weighted imaging (T1WI, Figure 2D) and high signal intensity on T2-weighted imaging (T2WI, Figure 2E). Late gadolinium enhancement (LGE) images showed heterogeneous enhancement of the mass (Figure 2F). The blood clot had higher signal intensity on T1WI and T2WI and nonenhancement on LGE (Figure 2, yellow arrow).

The patient underwent resection of the right atrial mass with cardiopulmonary bypass, and implantation of the tumor could be seen at the posterior wall of the RA on a broad base (Figure 1D). Postoperative TTE demonstrated the cardiac myxoma was completely resected and the right atrium was smaller than before (Figure 1C).

Macroscopically, the tumor showed an irregular and gelatinous shape with a bright red blood clot inside and was 5 cm × 4.2 cm × 3.5 cm (Figure 1E). On microscopy, there was abundant myxoid stroma with stellate and polygonal cells (Figure 1F). Necrosis, atypia, and pleomorphism were not detected. A pathological examination confirmed the diagnosis of myxoma.

Discussion

Cardiac myxomas are the most common benign primary tumor of the heart. Typically, myxomas arise from the interatrial septum at the border of the fossa ovalis in the left atrium. Myxomas arising from other sites are designated as “atypical myxomas.” Atypical myxomas arising from the free wall of the atrium are very rare (approximately 10% of the myxomas), and seldom appear in the literature.4,7 There are two main gross anatomical types of atrial myxomas, a solid type that is firm and smooth and a gelatinous type that is soft and friable.3

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Received for publication August 18, 2015. Revised and accepted September 28, 2015. Released in advance online on J-STAGE March 11, 2016. All rights reserved by the International Heart Journal Association.
Figure 1. A, B, C: Transthoracic echocardiographic images (TTE). TTE demonstrated a large mobile echogenic mass (5.3 cm x 4.2 cm) in the right atrium (RA). A tortuous banded isoechoic structure attached to the mass protruding into the right ventricle during diastole (B, arrow) and entering into the RA during systole (A, arrow). Postoperative TTE demonstrated the cardiac myxoma was completely resected and the RA was smaller than before (C). D: Intraoperative photograph showed the giant right atrial heterogeneous mass (arrow). E: Resected specimen photograph showing the tumor (5 cm x 4.2 cm x 3.5 cm) has an irregular and gelatinous shape with a bright red blood clot inside. F: High-powered image showing a tumor composed of small polygonal and stellate cells surrounded by myxoid stroma. (hematoxylin-eosin, 100 x magnification).

Figure 2. Cardiac magnetic resonance (CMR) images. Cine images confirmed the myxoma arising from the posterior wall of the right atrium (RA) on a broad base (A, B, C), and the location of the rupture was clearly shown (white arrow). The myxoma showed a predominantly isointense signal intensity on T1WI (D) and high signal intensity on T2WI (E). Late gadolinium enhancement (LGE) showed heterogeneous enhancement of the mass (F). The blood clot appeared to have higher signal intensity on T1WI and T2WI and nonenhancement on LGE (white arrow).
The probable reason for the spontaneously myxoma rupture in our case was that the tumor was gelatinous and squeezed by the heart for a long time. Also the large size of a tumor, irregular surface, and atypical location may lead to a higher risk of cardiac myxoma rupture,\(^8\) and only isolated case reports have described myxoma rupture.\(^8\) Thus, an atypical myxoma arising from the posterior wall of the RA with spontaneous rupture in our case is extremely rare.

Clinical manifestations of cardiac myxomas are often very confusing, and may present with obstructive, embolic, or constitutional symptoms. Research shows that atypical myxomas are associated with a higher risk of embolism,\(^8\) and myxoma rupture can result in significant embolic complications. The patient in our case had no embolic complications. Therefore, early detection of atypical cardiac myxomas, especially with tumor rupture, may be essential to reduce the occurrence and risk of embolic complications.

Medical imagery plays an important role in the diagnosis of myxomas. TTE is the mainstay imaging technique for cardiac tumor investigation and characterization. Although generally adequate to assess the size, location, and attachment of the lesion, echocardiography carries several limitations, including the dependence on an adequate acoustic window, a suboptimal visualization of extracardiac extension, and poor soft-tissue characterization.\(^10\) These limitations make it less effective at identifying atypical myxomas.

As a useful adjunctive modality, CMR has its own advantages. By using various sequences, CMR can provide superior soft tissue contrast which distinctly detects the internal details of myxomas, such as rupture and hemorrhage. Cine images clearly showed the location of rupture in our case. The myxomas appeared isointense on T1WI, and high signal intensity on T2WI and heterogeneous enhancement on LGE were observed, and thus the appearance corresponded with that of a previous report in the literature.\(^11\) The blood clot had high signal intensity on T1WI and T2WI and nonenhancement on LGE. On the other hand, by arbitrary angle and plane scanning in a large field of view, CMR can more sensitively and clearly display the relationship between myxomas and the large blood vessels, which can offer more useful information for surgical procedures. Above all, CMR can help us to further understand atypical myxomas and with clinical therapeutics.

**Conclusion:** In this case report, we documented a patient with an atypical myxoma arising from the posterior wall of the RA with spontaneous rupture who was diagnosed by TTE and CMR. Given the embolic potential of myxomas, surgical excision was the best treatment option.\(^12\) Prior to surgery, imaging techniques could offer comprehensive and accurate anatomic information to surgeons to help identify a precise surgical strategy.

**REFERENCES**