The Journal of Tehran University Heart Center

Case Report

Biatrial Myxoma with a Shared Stalk: A Case Report

Mehrnoush Toufan, MD[1], Naser Khezerlou-Aghdam, MD[1*], Shahab Masoumi, MD[1], Mahshid Dehghan, MD[2], Aisan Akhgari, MD[2]

[1]*Cardiovascular Research Center, Shahid Madani Hospital, Tabriz University of Medical Sciences, Tabriz, Iran.*
[2]*Tabriz University of Medical Sciences, Tabriz, Iran.*

Received 19 April 2021; Accepted 27 September 2021

Abstract

Myxomas are rare cardiac neoplasms and may present as single or multiple tumors. Only a few cases of single biatrial myxomas have been reported. We report a very rare case of this condition in a middle-aged woman, presenting with exertional dyspnea and cough. The patient had a resting tachycardia of 105 beats per minute, and cardiac auscultation discovered a mid-diastolic murmur across the mitral valve, followed by a tumor plop focused on the apex and elevated levels of C-reactive protein (1+) and creatine phosphokinase in lab data. The diagnosis was made via transesophageal and transthoracic echocardiographic examinations, showing the tumor extension through a patent foramen ovale (PFO). The operation was undertaken, the myxoma was excised, and the PFO was repaired. She was discharged with no further complications. Although myxomas are rare, considering this condition before surgery is significant. The involvement of both atria via a PFO is possible.

J Teh Univ Heart Ctr 2021;16(4):174-177

This paper should be cited as: Toufan M, Khezerlou-Aghdam N, Masoumi S, Dehghan M, Akhgari A. Biatrial Myxoma with a Shared Stalk: A Case Report. J Teh Univ Heart Ctr 2021;16(4):174-177.

Keywords: *Myxoma; Cardiac neoplasm; Foramen ovale, patent*

Introduction

Primary intracardiac tumors are rare, and 75% of these tumors are benign. Even though myxomas are rare in the cardiac surgery population, with an incidence rate of 0.0017% among the general population, they account for 50% of intracardiac tumors.

Patients with myxomas commonly present with a triad of systemic embolism, mitral valve inflow obstruction, and constitutional symptoms. Symptoms are related to the size, mobility, and location of the tumor, varying from asymptomatic to constitutional symptoms (eg, dyspnea).[1-3]

Approximately, 75% to 80% of myxomas originate from the left atrium, followed by the right atrium with an incidence rate of 10% to 20%. The incidence rate of multiple myxomas among all cardiac myxomas is 5%, half of them with a bilateral origin (2.5%).[4, 5]

The available literature describes only a few cases of biatrial myxomas, usually a single tumor reaching both atria.[6] This study reports a very rare case of a single biatrial myxoma in a middle-aged woman.

Case Report

A 53-year-old woman presented with a 3-month history of exertional dyspnea and positional cough in the supine position. Physical examination revealed a resting

[*]**Corresponding Author:** ***Naser Khezerlou-Aghdam,*** *Assistant Professor of Cardiology, Shahid Madani Hospital, Daneshgah Street, Tabriz, Iran. 5166615573. Tel: +98 4133373902. Fax: +98 4133363880. E-mail: nkhezerlou90@gmail.com.*

tachycardia of 105 beats per minute. Cardiac auscultation discovered a mid-diastolic murmur across the mitral valve, followed by a tumor plop focused on the apex. There was no significant finding in the chest X-ray. The patient's electrocardiogram was normal and had no significant findings. Laboratory evaluations showed elevated levels of C-reactive protein CRP (+1) and creatine phosphokinase (250 IU/L), as well as anemia (hemoglobin = 9.5 g/dL), and leukopenia (white blood cells = 6000/μL). Other laboratory biomarkers were within normal ranges. Angiographic evaluations showed no abnormalities.

Transthoracic echocardiography revealed left atrial enlargement with evidence of a large left atrial mass, prolapsing through the mitral valve during the diastole. The left atrial mass seemed to be attached to the interatrial septum. In addition, echocardiography showed significant functional mitral stenosis, mild eccentric mitral regurgitation, and mild tricuspid regurgitation. The left ventricle had normal size and function, with an intact ventricular septum. Pulmonary artery pressure was 48 mmHg.

The patient was further evaluated using transesophageal echocardiography, which illustrated a large dumbbell-shaped, highly mobile, inhomogeneous, smooth mass (6.2 × 3.4 cm) with a stalk (1.7 × 2.4 cm) attached to the interatrial septum at the left atrial side just beside the fossa ovalis. The mass extended toward the right atrium via a PFO with a minor left-to-right shunt (Figures 1, 2, and 3 & Videos 1 and 2).

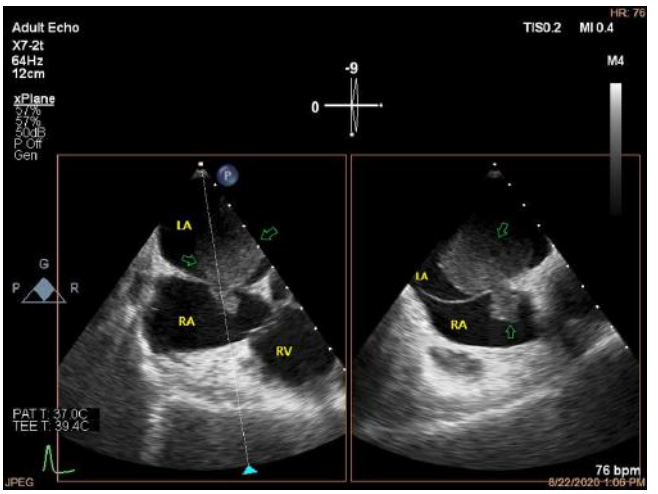

Figure 1. Transesophageal echocardiography shows a mass (arrows) in the LA and the RA.
LA, Left atrium; RV, Right ventricle; RA, Right atrium

She was referred to the surgery department for surgery, which was undertaken the following day. During the operation, examination of the heart revealed an extensively dilated left atrium. A longitudinal right atriotomy was performed, and the myxoma was delivered into the right atrium, leaving the left atrium intact. The mass was dumbbell-shaped, and its stalk appeared to originate from the left atrial septum. A large portion of the mass was inside the left atrium, and the smaller part was inside the right atrium, which was connected across the PFO. The myxoma was excised along with the stalk, and the oval hole was enlarged by cutting around the mass. The resultant septal defect was repaired. Histopathological evaluations reported fragments of tissue with a myxoid background. The tissue contained cells with round nuclei and many eosinophilic nucleoli arranged in complex structures resembling cords, nests, rings, or poorly formed glands. A final diagnosis of cardiac myxoma was established.

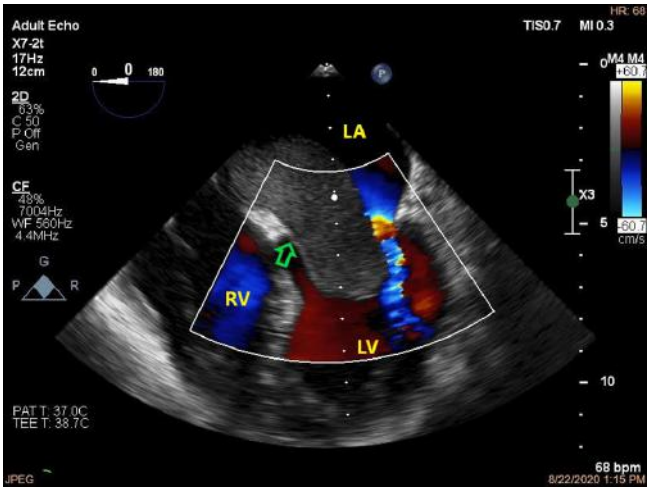

Figure 2. Transesophageal echocardiography illustrates a mass (arrow) in the LA with dynamic mitral valve stenosis.
LA, Left atrium; RV, Right ventricle; LV, Left ventricle

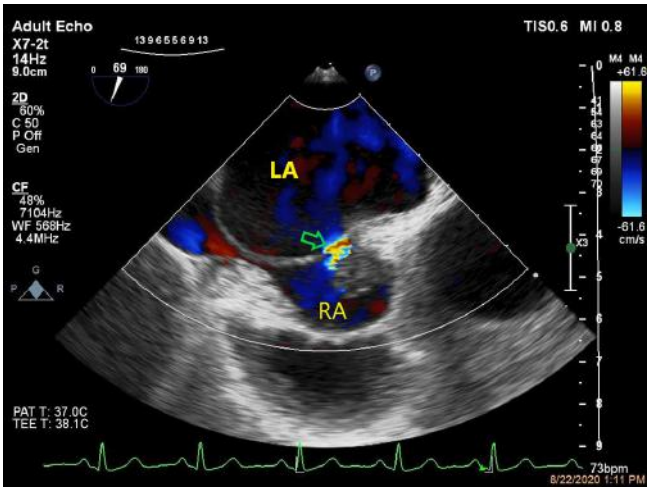

Figure 3. Transesophageal echocardiography shows a left-to-right shunt through a patent foramen ovale (arrow).
LA, Left atrium; RA, Right atrium

During surgery, it was confirmed that there was a single myxoma with a common stalk that extended through the PFO. The postoperative course was uneventful, and the

patient was discharged with no further complications. Considering the probability of recurrence, follow-up echocardiography will be done a year after the surgery.

Discussion

Myxomas are the most common type of adult primary cardiac tumors, and they originate from multipotent mesenchymal cells.[7] Most myxomas are caused sporadically, and 5% are multiple due to an inherited autosomal dominant disorder called "Carney's syndrome".[4, 5, 8] Transthoracic echocardiography alone is a sufficient diagnostic modality in many patients; nonetheless, in patients with myxoma, further examinations of these patients before surgery with transesophageal echocardiography are needed to diagnose cases of multiple myxomas or myxomas that have spread to other cavities.

Even though these tumors are benign, they may lead to complications such as mitral stenosis, cardiac arrhythmias, infections, embolic events, rupture, myocardial infarction, and sudden death. Massive atrial myxomas cause the mechanical obstruction of the valves by prolapsing into them.[9] Left atrial myxomas lead to functional mitral stenosis, which in this case presented with a mid-diastolic murmur across the mitral valve during auscultation and electrocardiographic abnormalities.

Azari et al[4] presented a case of a biatrial myxoma, in which 2 lobulated hypermobile masses were attached to the atrial septum in the region of the fossa ovalis. Vijan et al[5] also reported a biatrial myxoma in a young woman. They demonstrated a left atrial mass attached to the interatrial septum and another separated mass in the right atrium without any attachment to the interatrial septum. In contrast to the described cases, the present case describes a large dumbbell-shaped, highly mobile myxoma that extends toward the right atrium via a PFO with a minor left-to-right shunt.

Regarding cardiovascular complications and embolization risks, the treatment of choice for myxomas is surgical excision and the complete resection of the tumor and its implantation base with a good safety margin. A large resection of the myxoma pedicle or stalk is essential to prevent recurrence and the subsequent need for reoperation. Based on the location of the tumor, special techniques are used to control embolization.[10] In this case, the myxoma was delivered into the right atrium through a longitudinal right atriotomy, leaving the left atrium intact. The myxoma was excised along with the stalk, and the septal defect was repaired.

Concerning the presence of multiple tumors in patients with Carney's syndrome and cases such as our patient, in addition to the limitations of transthoracic echocardiography in diagnosing multiple myxomas and myxomas with spread to the other cavities, esophageal echocardiography is a complementary method in these patients. We believe that examination of all cardiac chambers via transesophageal echocardiography before surgery in patients with cardiac tumors is significant to investigate the extension of the tumor and the concurrent involvement of multiple chambers.

Conclusion

Even though multiple myxomas and single biatrial myxomas are rare, it is crucial to consider them before surgery. Transesophageal echocardiography has a paramount role in this matter. The right atrium should always be evaluated in the case of left atrial masses, especially when the patient's echocardiography shows an intracardiac shunt through the foramen ovale. The findings in our case report suggest that cardiologists and surgeons need to make a timely diagnosis and prompt treatment to improve the prognosis and prevent further complications.

To watch the following videos, please refer to the relevant URLs:

https://jthc.tums.ac.ir/index.php/jthc/article/view/1447/992
Video 1. This transesophageal echocardiography video shows that the myxoma stalk is located next to a patent foramen ovale, via which the myxoma has access to both the left and right atrial cavities.

https://jthc.tums.ac.ir/index.php/jthc/article/view/1447/993
Video 2. This transesophageal echocardiography video shows a large myxoma passing through a patent foramen ovale with a small left-to-right shunt in a color Doppler study.

References

1. Nina VJ, Silva NA, Gaspar SF, Rapôso TL, Ferreira EC, Nina RV, Lages JS, Silva FA, Filho NS. Atypical size and location of a right atrial myxoma: a case report. J Med Case Rep 2012;6:26.
2. Reynen K. Cardiac myxomas. N Engl J Med 1995;333:1610-1617.
3. Toufan M, Jodati A, Safaei N, Kazemi B, Pourafkari L. Myxomas in all cardiac chambers. Echocardiography 2012;29:E270-272.
4. Azari A, Moravvej Z, Chamanian S, Bigdelu L. An unusual biatrial cardiac myxoma in a young patient. Korean J Thorac Cardiovasc Surg 2015;48:67-69.
5. Vijan V, Vupputuri A, Chandrasekharan Nair R. An unusual case of biatrial myxoma in a young female. Case Rep Cardiol 2016;2016:3545480.
6. Schneider SR, Dell'Aquila A, Martens S, Rukosujew A. Biatrial recurrence of two independently growing cardiac myxoma in a patient with multiple tumor disease. Thorac Cardiovasc Surg Rep 2014;3:35-37.

7. Pucci A, Gagliardotto P, Zanini C, Pansini S, di Summa M, Mollo F. Histopathologic and clinical characterization of cardiac myxoma: review of 53 cases from a single institution. Am Heart J 2000;140:134-138.
8. Vidaillet HJ, Jr, Seward JB, Fyke FE, 3rd, Su WP, Tajik AJ. "Syndrome myxoma": a subset of patients with cardiac myxoma associated with pigmented skin lesions and peripheral and endocrine neoplasms. Br Heart J 1987;57:247-255.
9. O'Brien-Connors M. Biatrial myxoma: rare incidence in cardiac surgery. Can J Cardiovasc Nurs 2004;14:21-23.
10. Murayama H, Tamaki S, Kato N, Yuji N, Yokote J, Mutsuga M, Okada M. Right atrial myxoma associated with atrial septal defect: a case report and review of the literature. Ann Thorac Cardiovasc Surg 2001;7:166-169.