

Biatrial Cardiac Myxoma: A Case Report

Mohammad Reza AghajankhahTamijani*

Department of Cardiology, Heshmat Cardiovascular Research Center, Guilan University of Medical Sciences, Rasht, Iran

*Corresponding author: aghajankhah@gums.ac.ir

Abstract A 37 years old man presented with acute ataxia and vertigo. An axial brain CT Scan without contrast showed multiple subcortical hyperdense lesions. Brain MRI with gadolinium showed multiple hyperintense lesions. Transthoracic echocardiography showed large mobile biatrial masses. Biatrial masses were resected surgically. Both tumors had the histopathological features of benign cardiac myxoma. The clinical diagnosis was multiple brain emboli from cardiac myxoma. Myxoma is a primary benign cardiac tumor with a reported incidence of 0.0017% among the general population. The patient may present with constitutional symptoms, sequelae of valvular obstruction, embolic events or sudden death. Early diagnosis and treatment of these life threatening neoplasms is the challenge for physicians. Myxomas which arise from two different cardiac chambers are uncommon.

Keywords: *myxoma, echocardiography, biatrial*

Cite This Article: Mohammad Reza AghajankhahTamijani, "Biatrial Cardiac Myxoma: A Case Report." *American Journal of Medical Case Reports*, vol. 4, no. 1 (2016): 22-25. doi: 10.12691/ajmcr-4-1-7.

1. Introduction

Primary cardiac tumor is accounting for less than 0.2% of all tumors found in human being [1]. Although about 75 % of intracardiac tumors are benign but they may be associated with major complications such as thrombus, valve obstruction or even sudden cardiac death [1]. cardiac myxomas are the most common type of primary intracardiac tumors and originate primarily from left atrium (LA). Myxomas originating from right atrium (RA) are less common and those arising from two cardiac chambers are extremely rare fewer than 2.5% [1,2].

2. Case Report

A 37 years old man presented with acute ataxia and vertigo. His problem begins since last evening. Other symptoms were nausea, vomiting and diplopia. He said that four months ago he had similar symptoms for three days that resolved spontaneously. One year ago, he had two episodes of transient right hemifacial paresis. He suffered from bipolar mood disorder for seven years. Except for dilated pupils and ataxic gait the remainder of the exam was normal.

Electrocardiogram showed normal sinus rhythm, right axis deviation and right bundle branch block. Chest x ray was normal. The patient had elevated ESR and CRP levels.

An axial brain CT Scan without contrast was performed in emergency department and showed multiple subcortical hyperdense lesions.

In brain MRI with gadolinium, multiple hyperintense lesions were seen in T1 (Figure 1).

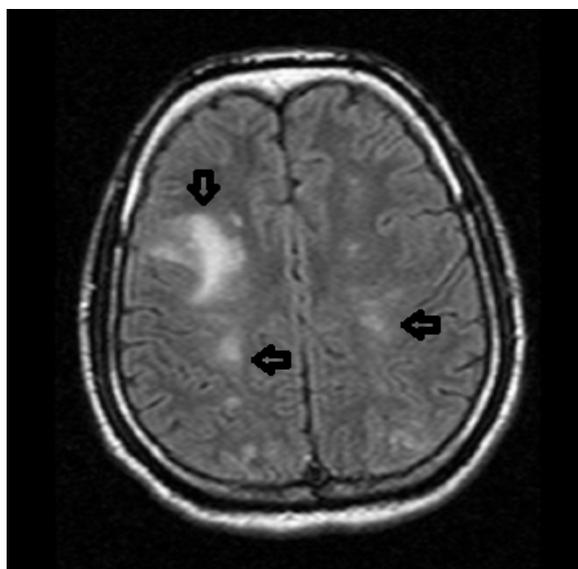


Figure 1. Brain MRI with gadolinium demonstrating multiple hyperintense lesions (arrows)

Transthoracic echocardiography showed large biatrial mobile masses (Figure 2) and in transesophageal echocardiography, a large mobile mass (60×27 mm) was seen in LA that attached to interatrial septum and aortomitral

intervalvular fibrosa which protruded into LV through mitral valve during diastole. In RA also another mobile mass (43×31 mm) was seen(Figure 3).

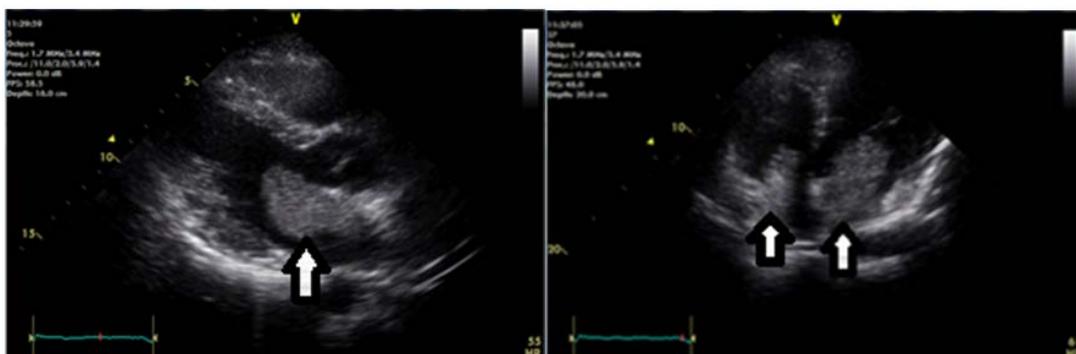


Figure 2. Transthoracic echocardiography of left atrial tumor prolapsing in to left ventricle in diastole from parasternal long axis view at left and biatrial tumors from apical four chamber view at right (arrows)

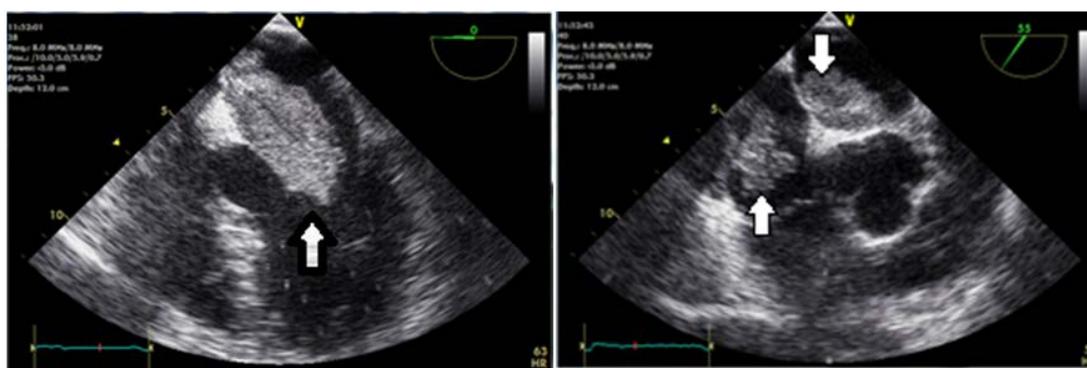


Figure 3. Transesophageal echocardiography showing left atrial tumor(left) and biatrial tumors(right)

Surgical consult was performed and biatrial masses were resected using median sternotomy under cardiopulmonary bypass (Figure 4). RA mass was resected via right atriotomy and LA mass was resected from interatrial

septal approach. A pericardial patch was used to reconstruct the interatrial septum. Both tumors had the histopathological features of benign cardiac myxoma (Figure 5).

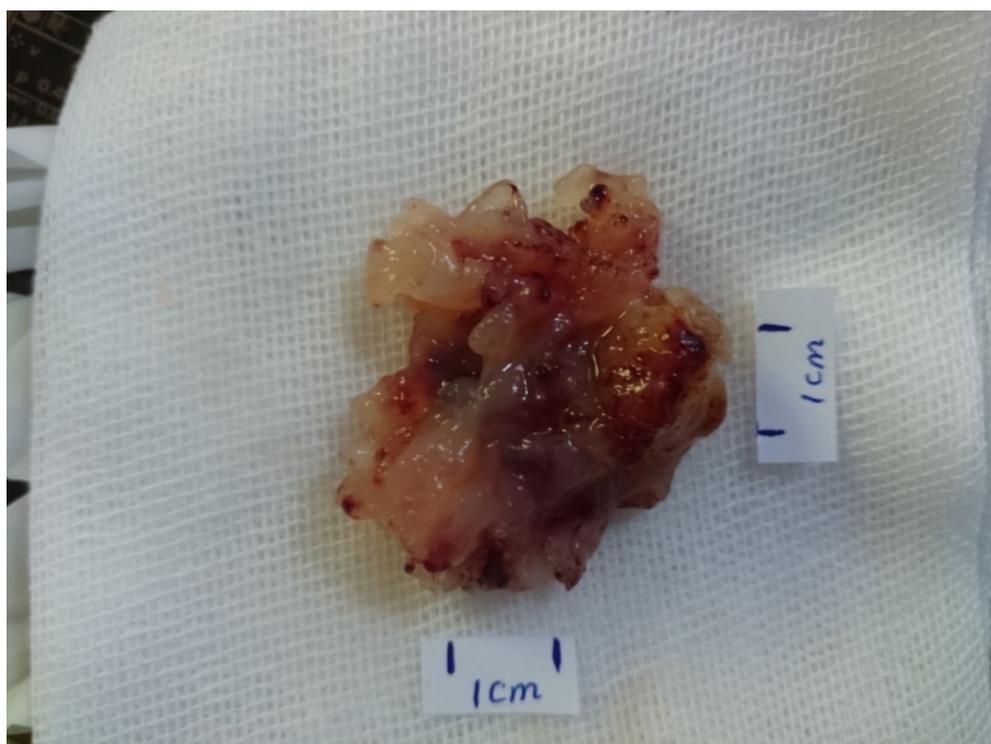


Figure 4. Gross photograph of the surgically excised variegated right atrial myxoma with a friable and gelatinous texture

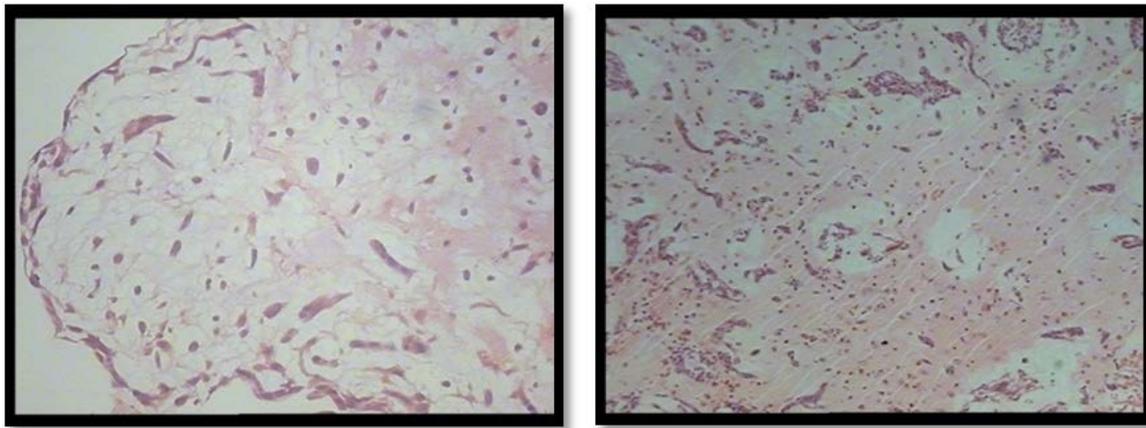


Figure 5. Microscopic view of resected cardiac tumors

3. Discussion

The first myxoma diagnosed clinically was by Kirkeby and Leren in 1952 [3]. Subsequently, the first successful removal of a cardiac myxoma using cardiopulmonary bypass was performed in Stockholm at the Karolinska Institute by Clarence Crafoord on July 16, 1954 [4]. In 1967, Yipintsoi described the first successful removal of a biatrial myxoma using cardiopulmonary bypass in a 37-year-old Thai policeman [3]. Biatrial myxomas continue to be rare tumors. A report by Imperio et al. in 1980 described a total of three cases of successful removal of biatrial myxomas in the literature [14] and approximately 19 cases were reported by Peachell in 1998 in the literature at that time [11] and finally a review of English literature by Irani et al. revealed ten other entries in 2008 [2].

Myxomas are benign primary cardiac tumors that most often (about 80%) are found in left atrium and in decreasing frequencies in right atrium (18%), right ventricle, left ventricle [5], and fewer than 2.5% are biatrial [1,2,5]. The incidence of cardiac myxoma peaks at 40 to 60 years of age with a female-to-male ratio of approximately 3:1. Most myxomas occur sporadically but may be familial.

Early diagnosis for intracardiac tumor is difficult because the symptoms are frequently nonspecific [6]. The patient can remain asymptomatic for a long period of time before diagnosis [7]. The clinical manifestations have been classified by Goodwin as consisting of constitutional symptoms, sequelae of valvular obstruction and embolic phenomena [8]. Sudden death may occur from coronary embolization of tumor or sudden obstruction of the mitral valve orifice [9]. The heart auscultation may result in abnormal findings in 50% to 85% of patients [9,10]. These abnormalities may include a holosystolic murmur of mitral insufficiency, a systolic ejection murmur, a diastolic murmur of mitral stenosis, a loud S1 or S2, an S4, or a tumor plop [1,10,12,13].

Physical examination abnormalities may be found in patients with Carney's syndrome, a rare disease described by J. Aidan Carney in 1985. It includes the triad of cardiac myxomas, spotty skin pigmentation and endocrine overactivity (also known as the NAME Syndrome: nevi, atrial myxoma, myxoid neurofibroma, ephelides). The Carney complex can be spontaneous or can be inherited as an autosomal dominant syndrome with variable penetrance [14]. The Carney complex occurs at a younger age and

should be considered when cardiac myxomas are discovered in atypical location in heart [5].

Myxomas typically form a pedunculated mass with a short broad base (85% of myxomas), but sessile form can also occur and appear yellow, white or brown and are frequently covered with thrombus [15].

Histologically, myxomas are composed of spindle- and stellate-shaped cells with myxoid stroma that may also contain endothelial cells, smooth muscle cells, and other elements surrounded by an acid mucopolysaccharide substance [16] (Figure 5).

Early diagnosis for intracardiac tumor is difficult because the symptoms are frequently nonspecific [6]. The patient can remain asymptomatic for a long period of time before diagnosis [7]. The clinical manifestations have been classified by Goodwin as consisting of constitutional symptoms, sequelae of valvular obstruction and embolic phenomena [8]. Sudden death may occur from coronary embolization of tumor or sudden obstruction of the mitral valve orifice [9]. The heart auscultation may result in abnormal findings in 50% to 85% of patients [9,10]. These abnormalities may include a holosystolic murmur of mitral insufficiency, a systolic ejection murmur, a diastolic murmur of mitral stenosis, a loud S1 or S2, an S4, or a tumor plop [1,10,12,13]. None of the characteristics of Carney complex were present in our patient and he had no family history. He had no constitutional symptoms and cardiac examination was normal.

Anemia, elevated ESR, elevated C-reactive protein (CRP), and increased gamma globulin are of associated laboratory abnormalities [9,10].

The most useful examination in the diagnosis is the echocardiogram which could confirm the location and extension of myxomas with a highly diagnostic sensitivity. Plain chest X-ray or CT scan is not diagnostic except for the pulmonary metastasis in cases of malignancy or the metastatic tumor [15].

The only definitive treatment is surgical removal [5]. Life-long follow-up is needed because myxomas have some tendency to recur. Recurrence rates vary but range from 5% to 14%. The time to recurrence in different series has varied from 0.5 to 6.5 years [17].

Biatrial myxoma is rare and initial presentation of the patient with biatrial myxoma and CNS embolic symptoms is rare too. Although this is not the first case, we presented it to highlight the importance of careful examination of all cardiac chambers seeking additional tumors for early diagnosis and treatment of this life threatening condition

to prevent irreversible complications or death. Because the recurrence rate is somewhat higher with biatrial myxoma, serial follow-up echocardiography and family screening is necessary. After surgery our patient had no symptom at six months follow up and repeat echocardiogram was normal.

Competing Interest

The author have no competing interests.

References

- [1] Zhenghua, X., Wei M., Da Z., and Eryong Z., "A Typical Bilateral Atrial Myxoma: A Case Report," *Case Reports in Cardiology*, Article ID 460268, 3 pages.2012.
- [2] Irani, A., Estrera, A., Buja, M., Safi, H., "Biatrial Myxoma: A Case Report and Review of the Literature". *J Card Surg*, 23. 385-390. 2008.
- [3] Yipintsoi, T., Donavanik, L., Bhamarapravati, N., Prachaubmoh, K., "Bilateral atrial myxoma with successful removal Report of a case," *Dis Chest*, 52. 828-834. 1967.
- [4] Crafoord, C., "Discussion on mitral stenosis and mitral insufficiency," In: *Proceedings of the International Symposium on Cardiovascular Surgery*, Philadelphia, Saunders, 1955, 202-211.
- [5] Lenihan, DJ., Yusuf SW., "Tumors Affecting the Cardiovascular System," in: Douglas, L., Douglas, P.Z., Peter, L., Robert, O., Eugene B., *Braunwald's heart disease A textbook of Cardiovascular medicine*, tenth edition, Saunders, Philadelphia, 2015, 1865-6.
- [6] Kuon, E., Kreplin, M., Weiss, W., Dahm, J. B., "The challenge presented by right atrial myxoma", *Herz*, 29(7). 702-709.
- [7] Burke, A.P., Virmani, R., "Cardiac myxoma: A clinicopathologic study", *Am J Clin Path*, 100, 671-680, 1993.
- [8] Goodwin, J.F., "Diagnosis of left atrial myxoma", *Lancet*, 1, 464-468, 1963.
- [9] Burke, A., Virmani, R., "Cardiac myxoma", In R. J., (ed), *Atlas of tumor pathology. Tumors of the heart and great vessels*, 3rd ed., Washington: Armed Forces Institute of Pathology 1996, 21-47.
- [10] Acebo, E., Val-Bernal, J.F., Gomez-Roman, J.J., "Clinicopathologic study and DNA analysis of 37 cardiac myxomas: A 28-year experience", *Chest* 123, 1379-1385, 2003.
- [11] Peachell, J.L., Mullen J.K., Bentley, M.J., "Biatrial myxoma: A rare cardiac tumor", *Ann Thorac Surg*, 65, 1768-1769, 1998.
- [12] B. KimK., ChoJ, N., Park, H.J., "Reversible pulmonary hypertension in adolescent with left atrial myxoma", *Journal of Cardiovascular Ultrasound*, 19, 221-223, 2011.
- [13] Attar, S., Lee, Y.C., Singleton, R., et al, "Cardiac myxoma", *Ann Thorac Surg*, 29, 397-405, 1980.
- [14] Imperio, J., Summers, D., Krasnow, N., et al., "The distribution patterns of biatrial myxoma", *Ann Thorac Surg*, 29, 469-473, 1980.
- [15] Nina, V.J., Silva, N.A., Gaspar, S. F., "Atypical size and location of a right atrial myxoma: a case report", *Journal of Medical Case Reports*, 23, 6-26, 2012.
- [16] McManus, B., "Primary tumors of the heart", In Bonow, R.O., Mann, D.L., Zipes, D.P., Libby, P. (eds) *Braunwald's Heart Disease*, 9th ed, Philadelphia, Elsevier Saunders, 2011, 1638-1650.
- [17] Pinede, L., Duhaut, P., Loire, R., "Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases", *Medicine*, Baltimore, 80, 159, 2001.