CASE REPORT OPEN ACCESS

Excision of a Left Atrial Myxoma Turns Out to be a Rare Presentation of Diffuse Large B-cell Lymphoma on Histopathology

Muhammad Aasim, Raheel Khan, Attaul Muhsin, Raheela Aziz

Department of Department of Cardiac Surgery, MTI-HMC/KGMC, Hayatabad, Peshawar

Cardiac myxomas are benign primary tumors of the heart, accounting for approximately 50% of all primary heart tumors. A 50 years old female presented to the outpatient department with complaints of shortness of breath on exertion, palpitations, and easy fatigability ongoing for a period of six to eight months. Echocardiography findings revealed a large mass about 4.9 cm^2 in the left atrium, suggestive of atrial myxoma with mild mitral regurgitation. She was planned for open heart surgical excision of her LA-Myxoma. The patient tolerated the procedure well and had an uneventful post-operative course. Histological evaluation of the specimen led to the diagnosis of a Diffuse Large B-Cell Lymphoma. Patient recovered well and was referred to oncological facility, where she was started on chemotherapy sessions. At 06 months follow up patient was seen in good cardiac health and no residual cardiac dysfunction, mass or defect. No matter how classical clinical and imaging presentation in favor of benign myxoma may be, the tissue must be sent for appropriate histological examination and tissue diagnosis when excised through indicated cardiac surgery.

INTRODUCTION

Cardiac myxomas are benign primary tumors of the heart, accounting for approximately 50% of all primary heart tumors. Myxomas commonly affect patients between ages of 30 years and 70 years, with a higher prevalence in women (1). Left atrial Myxomas (LA-Myxomas) are more common than Right atrial Myxomas (RA-Myxomas). Primary tumors of the heart are rare, with a reported incidence of 0.0017% to 0.19% or 1.38 per 100,000 people per year in the general population (2,3). The cardiac lymphomas are not common accounting about 9–24% of the cardiac

tumors (4), which is classified into primary cardiac lymphoma (PCL) and secondary cardiac lymphoma (SCL) (5). Primary cardiac lymphoma (PCL) is rare, only accounting for < 2% of all primary cardiac tumors (6-8). More than 80% of PCLs are diffuse B-cell non-Hodgkin's lymphomas (NHL) (9). Although patients can be asymptomatic, symptoms may include dyspnea, angina, syncope, cough, vertigo, fatigue, fever and stroke (10).

CASE REPORT

A lady around 50 years in age normally built, coming from Chitral (Pakistan), presented to the outpatient department with complaints of shortness of breath on exertion, palpitations, and easy fatigability. These symptoms had been ongoing for a period of six to eight months, prompting her to seek medical attention. She denied fever, chills, weight or appetite changes. Her past medical history was found to be insignificant with no chronic illnesses and no history of hospitalization for cardiac or any major issues. There were no previous surgical interventions reported. At presentation patient was afebrile with BP of 130/80 mmHg, pulse 86 bpm. Her lungs were having fine basal crepts on auscultation. Cardiac examination revealed a regular heart rate and rhythm, normal S1 and S2 heart sounds, and slight diastolic murmur mimicking Mitral valve stenosis. No additional heart sounds were detected. General physical examination was unremarkable. On chest X-Ray film, her chest was normal with normal lung parenchyma, fine basal haziness and normal cardiothoracic ratio. Echocardiography findings revealed a large mass about 4.9cm2 in the left atrium, suggestive of atrial myxoma with mild mitral regurgitation and trace tricuspid regurgitation, with normal Bi-ventricular systolic functions. Her Coronary-angiography showed normal coronary arteries. All baseline blood investigations were within normal limits. She was diagnosed as a case of large left atrial myxoma and planned for open heart surgical excision of her LA-Myxoma.

The patient admitted to the cardiac surgery department was highly suspected to have cardiac myxoma based on symptoms, mass location, and size. A decision was made to undergo surgical resection via median sternotomy. Cardiopulmonary bypass was initiated and the left atrium accessed through a right atriotomy trans-septal approach. Incision at the fossa ovalis was made. The myxoma was discovered attached to the left atrial side of the interatrial septum via a stalk at the fossa ovalis level, just like the typical isolated LA-Myxomas. A wider incision was performed

at the fossa ovalis to remove the mass and its stalk, ensuring clear margins due to its significant size. The size was estimated as 4.9 cm2 on TTE, but the specimen was reported as 3 × 2.5 × 1.5 cm in the pathology report and described as firm, yellow to gray, friable cut surface, soft and gelatinized mass tissue. The right atriotomy was closed. The patient was subsequently weaned from cardiopulmonary bypass and one mediastinal drain was placed within the pericardial space. The patient tolerated the procedure well and had an uneventful post-operative course. Histological evaluation of the specimen showed a lymphoid neoplasm composed of large tumor cells arranged in sheets in a discohesive fashion with scattered lymphocytes in the background. Tumor cells have rounded vesicular nuclei, conspicuous nucleoli and appreciable amount of cytoplasm with numerous mitoses were present. Based on these features and immune-histochemical staining diagnosis of Diffuse Large B-Cell Lymphoma was established.

DISCUSSION

It is very uncommon to encounter primary cardiac tumors, with an occurrence rate falling between 0.0017– 0.03%. In contrast, secondary cardiac tumors are 30 times more common when compared with primary cardiac tumors. Among primary cardiac tumors, a striking Seventy-five percent are benign, 50% of which are myxomas. Myxomas predominantly occur in 50- to 60-year-old females with women being 2.05 times more prone to developing this condition than men (11–13). Disseminated cancers and lymphomas rarely involve the heart, and usually have no cardiac symptoms (14). Primary cardiac lymphomas (PCL) typically are invasive large B-cell lymphoma (15,16,17). PCL most commonly diffusely invades the pericardium, epicardium, or myocardium or presents as invasive nodular masses (15,18). In our patient symptoms such as fever, chills, fatigue, malaise and weight loss were not seen.

Patient recovered well and was referred to oncological facility, where she was started on chemotherapy sessions. At 06 months follow up patient was seen in good cardiac health and no residual cardiac dysfunction, mass or defect. The treating oncologist had advised PET-CT-scan for confirming complete remission of her lymphoma, at the last follow up when this case report is completed.

CONCLUSION

It is to conclude from this case report that no matter how classical clinical and imaging presentation in favor of benign myxoma may be, the tissue must be sent for appropriate histological examination and tissue diagnosis when excised through indicated cardiac surgery.

REFERENCES

1. Demir M, Akpinar O, Acarturk E. Atrial myxoma: an unusual cause of myocardial infarction. Texas Heart Institute Journal. 2005;32(3):445.
2. Reynen K. Cardiac myxomas. New England Journal of Medicine. 1995 Dec 14;333(24):1610-7.
3. Cresti A, Chiavarelli M, Glauber M, Tanganelli P, Scalese M, Cesareo F, Guerrini F, Capati E, Focardi M, Severi S. Incidence rate of primary cardiac tumors: a 14-year population study. Journal of cardiovascular medicine. 2016 Jan 1;17(1):37-43.
4. Petrich A, Cho SI, Billett H. Primary cardiac lymphoma: an analysis of presentation, treatment, and outcome patterns. Cancer. 2011 Feb 1;117(3):581-9.
5. PJ M. Involvement of the heart by malignant lymphoma: a clinicopathologic study. Cancer. 1982;49:944-51.
6. White RW, Hirst NA, Edward S, Nair UR. Plasmacytoid lymphoma within a left atrial myxoma: a rare coincidental dual pathology. Interactive CardioVascular and Thoracic Surgery. 2010 Jan 1;10(1):140-1.
7. Wu ZR, Weng DS, Ding YQ, Han HX, Zhu MG. Primary diffuse large B-cell lymphoma of the heart: a clinicopathological study. Nan Fang yi ke da xue xue bao= Journal of Southern Medical University. 2006 Oct 1;26(10):1481-3.
8. Park SM, Shim CY, Choi D, Lee JH, Kim SA, Choi EY, Ha JW, Chung N. Coronary sinus obstruction by primary cardiac lymphoma as a cause of dyspnea due to significant diastolic dysfunction and elevated filling pressures. Journal of the American Society of Echocardiography. 2010 Jun 1;23(6):682-e5.
9. Antoniades L, Eftychiou C, Petrou PM, Bagatzounis A, Minas M. Primary cardiac lymphoma: case report and brief review of the literature. Echocardiography. 2009 Feb;26(2):214-9.
10. Wang JG, Li YJ, Liu H, Li NN, Zhao J, Xing XM. Clinicopathologic analysis of cardiac myxomas: seven years' experience with 61 patients. Journal of thoracic disease. 2012 Jun 6;4(3):272.
11. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma: a series of 112 consecutive cases. Medicine. 2001 May 1;80(3):159-72.
12. Al-Zamkan BK, Hashem AM, Alaaeldin SA, Abdel Aziz M. An exceptionally giant left atrial myxoma: a case report and literature review. European Heart Journal-Case Reports. 2020 Dec;4(6):1-7.
13. Li H, Guo H, Xiong H, Xu J, Wang W, Hu S. Clinical features and surgical results of right atrial myxoma. Journal of Cardiac Surgery. 2016 Jan;31(1):15-7.
14. McDonnell PJ, Mann RB, Bulkley BH. Involvement of the heart by malignant lymphoma: a

clinicopathologic study. Cancer. 1982 Mar 1;49(5):944-51.

15. Burke A, Virmani R. Tumors of the heart and great vessels. Afip; 1996.
16. Chalabreysse L, Berger F, Loire R, Devouassoux G, Cordier JF, Thivolet-Bejui F. Primary cardiac lymphoma in immunocompetent patients: a report of three cases and review of the literature. Virchows Archiv. 2002 Nov;441:456-61.
17. Ikeda H, Nakamura S, Nishimaki H, Masuda K, Takeo T, Kasai K, Ohashi T, Sakamoto N, Wakida Y, Itoh G. Primary lymphoma of the heart: case report and literature review. Pathology international. 2004 Mar;54(3):187-95.
18. Gl C. Primary cardiac lymphoma in immunocompetent patients. Cancer. 1997;80:1497-506.