

DIAGNOSIS AND DESCRIPTION OF MITRAL VALVE PAPILLARY FIBROELASTOMA BY MRI: A CASE REPORT

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ABSTRACT

Papillary fibroelastoma is a rare primary tumor of the heart and is usually found incidentally at autopsy. The tumor is considered as benign but it may cause potential fatal event such as stroke and cardiac obstruction. In this case report, we describe a mitral valve papillary fibroelastoma detected by magnetic resonance imaging. A 28 year-old-female presented with cardiac syncope and chest pain and a small mass was detected by transthoracic echocardiography. The mass was further characterized by MRI and was described here. Complete surgical excision was performed. A mitral papillary fibroelastoma was diagnosed histologically. This case illustrated the value of MRI as an alternative non-invasive diagnostic tool in establishing the diagnosis.

Cardiac papillary fibroelastoma is a rare primary cardiac tumor, which is considered as endocardial in origin and is usually found incidentally at autopsy.¹ It is the third most common primary cardiac tumor but is the most common primary tumor of the heart valve.² Characteristically, it is small, solitary and usually not resulting in clinical problem. However, it can cause important clinical implications.³⁻⁵ Echocardiography, particularly with transesophageal echocardiography, is the primary diagnostic procedure. It provides information regarding characteristic, location and other associated abnormality.⁶ Magnetic resonance imaging (MRI) is useful as an adjunct diagnostic tool. It has the potential to provide the more precise delineation in term of size, shape, location and surface as well as some tissue characteristic.⁷

In this case report, we described the mitral valve papillary fibroelastoma diagnosed by MRI.

CASE REPORT

A 28 year-old-female with no previous medical history presented 4- week history of intermittent chest pain and one episode of syncope while she was walking. Chest pain was characterized as tightness at substernal area with radiation to left mandible and left arm, lasting approximately 15 minutes. The pain was relieved by rest. She did not experience any dyspnea or fever.

Physical examination revealed only a grade I-II diastolic murmur at left upper sternal border.

Chest X-ray showed mild cardiomegaly without pulmonary venous congestion. Electrocardiogram revealed symmetrical inverted T wave in lead V1 to V5 with occasional paroxysmal ventricular contraction. Transthoracic echocardiography demonstrated a 2x2x2 cm. mass of homogeneous echogenicity attached to the ventricular aspect of the anterior leaflet of the

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mitral valve (Fig.1). Doppler examination revealed mild mitral regurgitation secondary to the mass interfering with the valve function. There was mild left ventricular enlargement with poor left ventricular ejection function (35%).

Further evaluation with MRI revealed a 2x2x2 cm. mobile, multilobulated frond-like mass attached to the ventricular side of the anterior leaflet of mitral valve. The mass has a central core of hyposignal on and peripheral frond-like appearance of iso to slight hypersignal on T1W, hypersignal on T2W (Fig.2) with moderate peripheral enhancement after intravenously administered gadolinium (Fig3). On GRE T2W, the mass had the signal higher than the myocardium, which suggested tumor rather than thrombus. Mild mitral regurgitation and enlargement of left ventricle

were also noted. There was no other cardiac mass.

Patient did not undergo for the cardiac catheterization due to the less likelihood of coronary artery disease in this age group.

Laboratory evaluation revealed no leukocytosis.

The patient underwent surgery with excision of a bead-liked mass from ventricular side of anterior leaflet of mitral valve and the reconstruction of the mitral valve was performed. Histology confirmed the diagnosis of papillary fibroelastoma. The patient did not have any post-operative complications. She remained asymptomatic in a 5-month followed-up period.

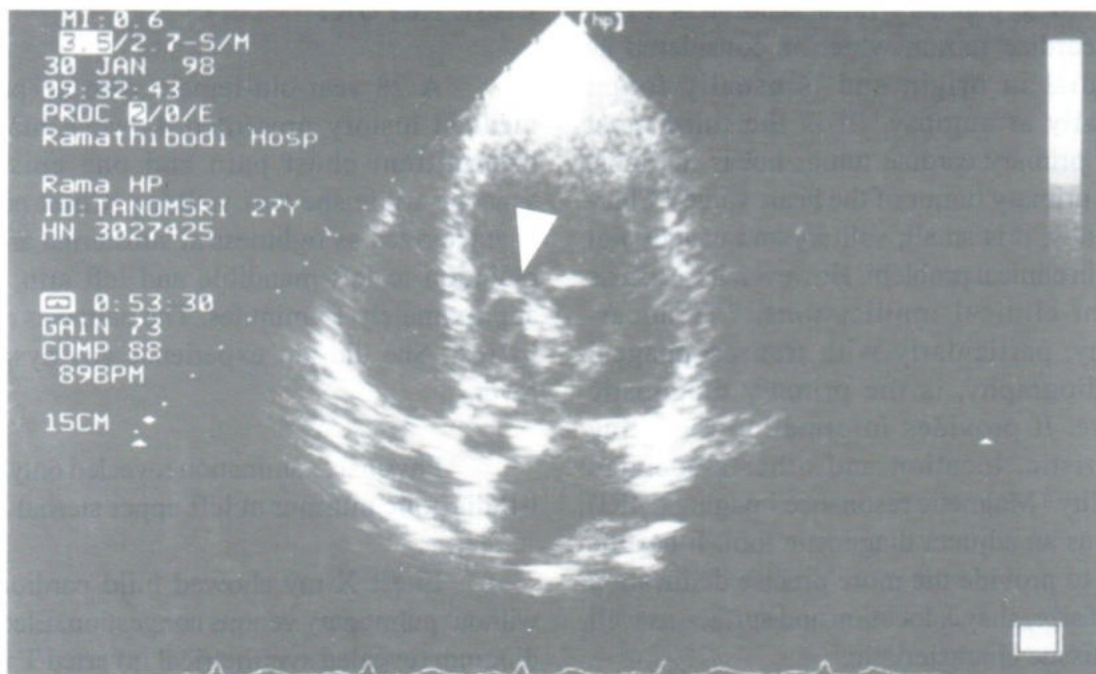


Fig. 1. Transthoracic echocardiography demonstrated an echogenic mass at anterior leaflet of mitral valve.

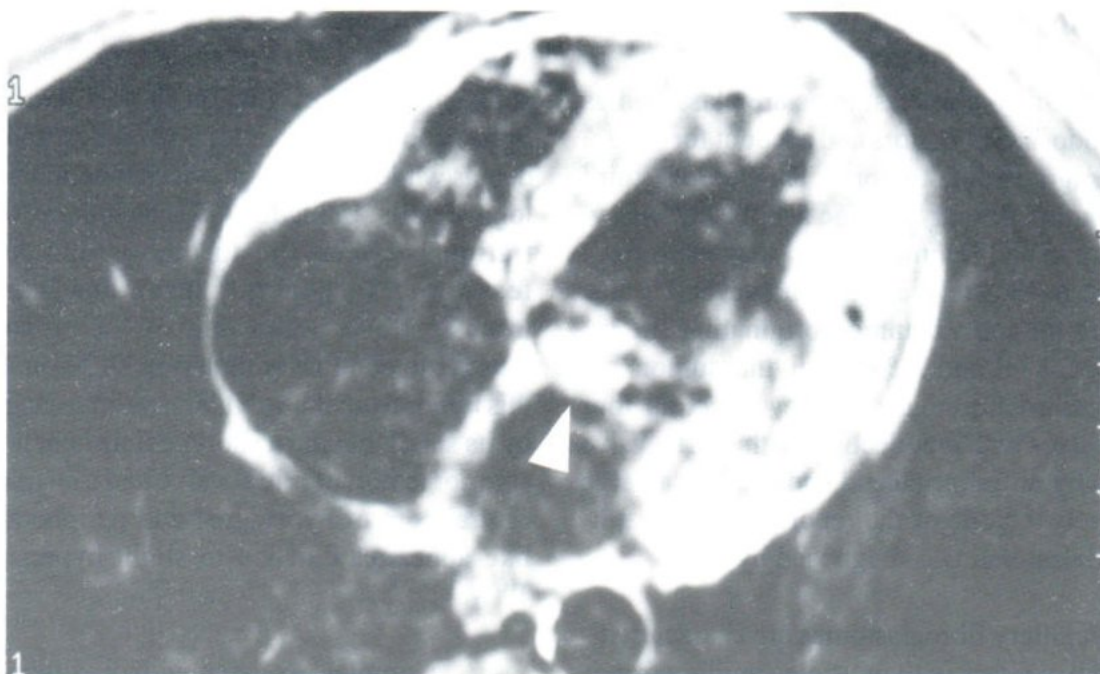


Fig. 2. MRI SE T1W in axial plane revealed a small, well-defined mass of isosignal to slight hypersignal, attaching to the anterior leaflet of the mitral valve. Note the characteristic of the frond-like appearance of the tumor.

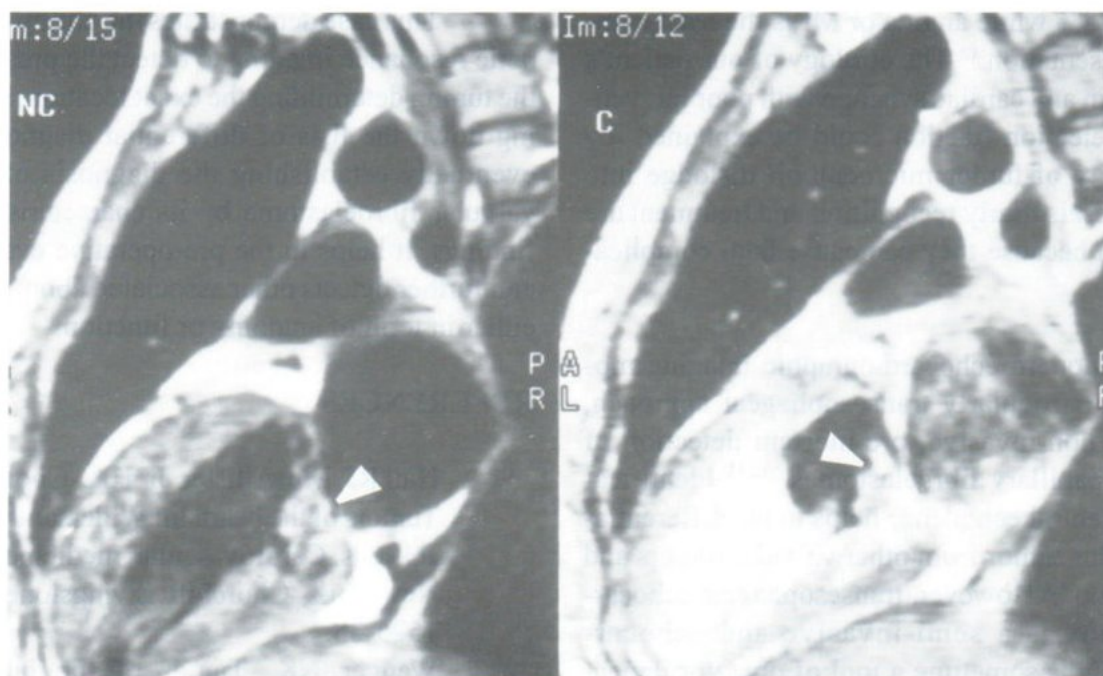


Fig. 3. MRI SE T1W pre gadolinium (left) and post gadolinium (right) in sagittal oblique plane demonstrated further details of the mass which showed a central core of hyposignal and peripheral frond-like appearance of iso to slight hypersignal on T1W (left). Gadolinium SE T1W revealed a peripheral enhancing frond-like peripheral part (right).

DISCUSSION

Papillary fibroelastomas are classified as benign endocardial tumors and account for 5-10% of all benign cardiac tumors.¹ They are the third most common primary cardiac neoplasm, following myxoma and lipoma.^{3,5} They may arise from anywhere in the endocardium, however, cardiac valves are the most common originating site.^{3,8,9} In addition, 88% and 71% of valvular tumors are papillary fibroelastomas.^{10,11} They affect all four cardiac valves with predilection on the aortic valve followed by mitral, tricuspid and pulmonic valve respectively.¹² None of the reported mitral papillary fibroelastoma was diagnosed by MRI.

Papillary fibroelastomas are characteristically small, solitary with multiple papillary fronds and usually are attached to the endocardium by a short pedicle.^{3,8} Neurologic symptoms such as stroke are the most common symptoms, accounting for 75% while angina or MI is the least common presentation.¹² The etiology of our patient's chest pain and cardiac syncope with poor left ventricular ejection fraction could be explained by possibility of tumor microemboli dislodge into great vessels. Early recognition and treatment are essential because they can cause fatal complication.^{12,13}

Modern echocardiographic imaging modalities, particularly transesophageal approach, allow a non-invasive, pre-mortem detection of cardiac papillary fibroelastomas.^{6,12,13} Identification of central echodense helps in the differentiation of this tumor from other valvular tumors and vegetation.^{6,8} However, transesophageal echocardiography is a semi-invasive and echocardiography is sometime a tool of operator dependence.

Various MRI techniques allow a non-invasive diagnosis of papillary fibroelastomas. MRI

is also useful in evaluation of valvular function. Spin echo T1W technique provides anatomical details while cine MRI provides functional physiologic details. Using the GRE technique, differentiation between the thrombus and tumor is feasible.¹⁴ In general, thrombus has signal intensity lower than that of the surrounding muscle, in contrast with tumor. Differentiation of this tumor from the valvular myxoma can be made by its small, having a short pedicle attached to the anterior leaflet of mitral valve and a characteristic central core of hyposignal and peripheral frond-like appearance of iso to slight hypersignal on T1W.

Complete excision with or without valvular repair is the mean of eliminating the source and potential of fatal embolization.¹⁵

This case demonstrates the ability of magnetic resonance imaging to detect the presence of the tumor, determining the exact location, narrowing down the lists of differential diagnosis and eventually establishing the diagnosis of mitral valvular fibroelastoma by its characteristic MR findings. It helps in the pre-operative evaluation and further detects other associated abnormalities either in term of anatomy or function.

REFERENCES

1. Hall RJ, Mc Allister Jr HA, Cooley DA. Tumors of the heart. In: Willerson JT, Cohn JN, eds. Cardiovascular medicine. 1st ed. New York, Churchill Livingstone, 1995; 1525-38.
2. Wenger NK. Tumor of the heart. In: Gravanis MB, eds. Cardiovascular disorders: pathogenesis and pathophysiology. 1st ed. St Louis, Mosby, 1993;270-98.

3. Mc Allister HA Jr, Fenoglio JJ Jr. Tumors of the cardiovascular system. In: Atlas of tumor pathology. Fasc 15, 2nd series. Washington DC: Armed Forces Institute of Pathology, 1978;20-5.
4. Abu Nassar SG, Parker JC Jr. Incidental papillary endocardial tumors: its potential significance. Arch Pathol 1971;92:370-6.
5. Chitwood WR Jr. Cardiac neoplasm: current diagnosis, pathology and therapy. J cardiac Surg 1988;3:119-54.
6. Klarich KW, Enriquez-Sarano M, Gura GM, Edwards WD, Tajik AJ, Seward JB. Papillary fibroelastoma: echocardiographic characteristics for diagnosis and pathologic correlation. J Am Coll Cardiol 1997;30:784-90.
7. Link KM, Lesko NM. MR evaluation of cardiac/juxtacardiac masses. Topics Magn Reson Imaging 1995;7(4):232-45.
8. Shahian DM, Labib SB, Chang G. Cardiac papillary fibroelastoma. Ann Thorac Surg 1995;59:538-41.
9. Lichtenstein HL, Lee JC, Stewart S. Papillary tumor of the heart: incidental finding at surgery. Hum Pathol 1979;10:473-5.
10. Ryan PE Jr, Obeid AL, Parker FB Jr. Primary cardiac valve tumors. J Heart Valve Dis 1995;2:222-6.
11. Edwards FH, Hale D, Cohen A, Thompson L, Pezzella AT, Virmani R. Primary cardiac valve tumors. Ann Thorac Surg 1991;52:1127-31.
12. Grinda J-M, Couetil JP, Chauvaud S, et al. Cardiac valve papillary fibroelastoma: surgical excision for revealed or potential embolization. Cardiovasc Surg 1999;117:106-10.
13. Bhagwandien NS, Shah N, Costello JM Jr, Gilbert CL, Blankenship JC. Echocardiographic detection of pulmonary valve papillary fibroelastoma. J Cardiovasc Surg 1998;39:351-4.
14. Seelos KC, Caputo GR, Carrol CL et al. Cine gradient refocused echo (GRE) imaging of intravascular masses: differentiation between tumor and nontumor thrombus. J Comput Assist Tomogr 1992;16:169-75.
15. McFadden PM, Lacy JR. Intracardiac papillary fibroelastoma: an occult cause of embolic neurologic deficit. Ann Thorac Surg 1987;43:667-9.