

Case Report

A Rare Case of Papillary Fibroelastoma Involving The Tricuspid Valve. A single Center experience over a period of 22 years (1999-2021)

Running head: Papillary Fibroelastoma and Tricuspid Valve

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ABSTRACT

Background and aim. Papillary fibroelastoma (PFE) represents only 16% of the benign cardiac tumor and approximately 15% of these are located on tricuspid valve. **Materials and Methods** Over a period of 22 years (1999-2021) we observed 75 pts with cardiac tumors at our Center over 9650 pts operated on. Most of them were mixoma but in 10 cases histology showed a PFE. We describe a rare case (1/75 of cardiac tumors) of a tricuspid valve PFE in a 69-year-old patient. Trans-thoracic echocardiography demonstrated a mobile mass (20 x 10 mm), adhering to the atrial side of the septal leaflet of the tricuspid valve. In consideration of the mobility of the mass and the consequent high embolic risk, surgical removal was made. Patient underwent surgery through a median sternotomy on CPBP. A “gelatinous” mass adhering to the tricuspid leaflet was found and completely removed. The postoperative course was uneventful. The pathological evaluation confirmed the diagnosis of PFE. **Conclusions.** PFE of the tricuspid valve is rare entities being in most cases found incidentally. In our experience the incidence of this tumor in this location is 1/10000 cases of cardiac surgery. Although most patients are asymptomatic, surgical treatment is nevertheless recommended in consideration of the high embolic risk.

INTRODUCTION

Primary cardiac tumor are rare occurrence and the majority of these are benign with heterogeneous range of clinical presentation ¹.

Over a period of 22 years since the starting of the activity of Cardiac Surgery at our Hospital in 1999, 75/9650 patients (0,78%) had had surgery for cardiac tumors but only one case of papillary fibroelastoma (PFE) on the tricuspid valve was observed and surgically treated. Demographic data are reported in Tab 1.

PFEs represent only 16% of the benign cardiac tumor ¹⁻² and approximately 15% of these are located on tricuspid valve ³. Although they are frequently incidental discovered on imaging tests ordered for other reasons, their identification as a possible etiology of vascular embolism, stroke and cardiac arrest is well known ⁴. In consideration of the high embolic risk, surgical tumor removal is therefore recommended ¹⁻⁴.

Informed consent was obtained from the patient but not ethics approval statement regarding IRB because of it's a case report that does not meet the DHHS definition of "research".

CASE REPORT

A 69-year-old female was referred to our Department for an incidental finding of a tilting mass adhering to the tricuspid valve. No significant pathological history, nor symptoms related to the mass. Trans-thoracic echocardiography demonstrated a normal left ventricular ejection fraction and no valvular dysfunction. A mobile mass (20 x 10 mm), adhering to the atrial side of the septal leaflet of the tricuspid valve that crosses the valve and projecting into the ventricular side, was seen (Fig 1). Pre-operative coronarography revealed no coronary artery disease.

In consideration of the high mobility of the mass and the high embolic risk, surgical removal was recommended. The patient underwent surgery through a median sternotomy with cardiopulmonary bypass established via aorto-bicaval cannulation and hematic cardioplegic arrest (aortic X clamp 13'; CPBP time 44'). The right atrium was opened and a "gelatinous" mass adhering to the tricuspid leaflet was found looking at first time as a small myxoma (Fig 2).

The mass was completely removed and the valvular leaflet was curetted.

By putting the tumor in a specimen for pathologist examination, this in the water acquired a distinctive "anemone-like" shape (Fig 3).

The implant-base surface was cauterized without any injury to the valve leaflet. After weaning from CPBP the intra-operative TEE showed absence of intra-cavitary mass and no tricuspid valve regurgitation. Patient was extubated within 6 hours after surgery and postoperative course was uneventful. Discharge from the hospital at home was on 6th day. The pathological evaluation confirmed the diagnosis of PFE.

DISCUSSION

PFEs are rare entities represent the second most common benign cardiac neoplasm of adulthood. In most cases, their localization is on the cardiac valves but infrequently affecting the tricuspid valve. ⁴⁻⁵ PFEs identification in recent years has increased thanks to the development of diagnostic techniques, being in most cases found incidentally. Trans-thoracic echocardiography is generally the test of choice for the initial evaluation of the patient. PFEs commonly present as a relatively small (from 2 mm to 40 mm), mobile, pedunculated mass, attached to the endocardium ⁶. In consideration of the reduced sensitivity and specificity of the trans-thoracic echocardiogram in the identification of PFEs smaller than 2 mm, in case of a high suspicion for cardio-embolic source of stroke, an echocardiogram trans-esophageal evaluation could be considered ⁷. Cardiac MRI could potentially offer better spatial resolution, especially in valve involvement, while remaining a more expensive and not always available exam ⁸. Most patients with PFEs are asymptomatic at the time of the diagnosis, but surgical treatment is nevertheless recommended in consideration of the

embolic risk even correlated with right heart neoplasm⁸⁻¹¹. The goal of the intervention is to be as radical as possible, resecting the mass in correspondence of the peduncle. It may be necessary to combine a valve repair surgery in the case of insufficiency associated or secondary to valve damage caused by removal of the mass. In our case, the mass was pedunculated, and it was therefore possible to radically remove it with cauterization of the implant-base surface. The mass was easily removed without any damage to the tricuspid valve, no valvular surgery was therefore necessary.

BIBLIOGRAPHY

1. Centofanti, P., Di Rosa, E., Deorsola, L., Actis Dato, GM., Patanè, F., La Torre, M., Barbato, L., Verzini, A., Fortunato, G., Di Summa, M. Primary cardiac tumors: Early and late results of surgical treatment in 91 patients. *Annals of Thoracic Surgery* 1999;68:1236-1241.
2. Poterucha, TJ., Kochav, J., O'Connor, DS. and Rosner, GF. Cardiac Tumors: Clinical Presentation, Diagnosis, and Management. *Curr. Treat. Options Oncol.* 2019;20(8):66.
3. Edwards, FH., Hale, D., Cohen, A., Thompson, L., Pezzella, AT., and Virmani, R. Primary cardiac valve tumors. *Ann. Thorac. Surg.* 1991;52:1127–1131.
4. Gowda, RM., Khan IA., Nair, CK., Mehta, NJ., Vasavada, BC. Cardiac papillary fibroelastoma: A comprehensive analysis of 725 cases. *Am. Heart J.* 2003;146:404–410.
5. Tamin, SS., Maleszewski, JJ., Scott, CG., Khan, SK., Edwards, WD., Bruce, CJ., Oh, JK., Pellika, PA., Klarich, KW. Prognostic and bioepidemiologic implications of papillary fibroelastomas. *J Am Coll Cardiol.* 2015;65:2420–2429.
6. Klarich, KW., Enriquez-Sarano, M., Gura, GM., Edwards, WD., Tajik, J., Seward, JB. Papillary Fibroelastoma: Echocardiographic Characteristics for Diagnosis and Pathologic Correlation. *J Am Coll Cardiol* 1997;30(3):784-90
7. Grote, J., Mügge, A., Schäfers, H. J., Daniel, WG. and Lichtlen, PR. Multiplane transoesophageal echocardiography detection of a papillary fibroelastoma of the aortic valve causing myocardial infarction. *Eur Heart J.* 1995;16:426–429.
8. Araoz, PA., Mulvagh, SL., Tazelaar, HD., Julsrud, PR. and Breen, JF. CT and MR imaging of benign primary cardiac neoplasms with echocardiographic correlation. *Radiographics* 2000;20:1303–1319.
9. Neerukonda, SK., Jantz, RD., Vijay, NK., Narrod, JA. and Schoonmaker, FW. Pulmonary embolization of papillary fibroelastoma. Arising from the tricuspid valve. *Texas Heart Inst. J.* 1991;18:132–5.
10. Gabbieri, D., Rossi, G., Bavutti, L., Corghi, F., Zacà, F., Sarandria, D., Pierangeli, A., Ghidoni, I. Papillary fibroelastoma of the right atrium as an unusual source of recurrent pulmonary embolism. *J Cardiovasc Med (Hagerstown)* 2006;7:373–378.
11. Mastroberto, P., Olivito, S., Onorati, F., di Virgilio, A., Merola, S., Renzulli, A. Papillary fibroblastoma of tricuspid valve with pulmonary embolization. *Asian Cardiovasc Thorac Ann.* 2006;14(3):e53-4.