

CARDIAC PAPILLARY FIBROELASTOMA: A CASE WITH UNPRECEDENTED SPREAD

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Abstract

Introduction: Papillary fibroelastomas (PFE) are a rare primary cardiac neoplasm, out of which multifocal PFE constitute a small minority of cases. These benign masses are commonly found on valvular surfaces, particularly the aortic valve.

Clinical case: We present a patient with a history of embolic stroke and intra-cardiac masses. Multimodal imaging revealed multiple nodules with extensive intra-cardiac distribution. All nodules were successfully removed without valve dysfunction.

Conclusion: This is a unique case of multiple PFE involving intracardiac cavities as well as all valvular structures.

Keywords: Cardiac masses; cardiac magnetic resonance; multiple papillary fibroelastomas; imaging; echocardiography.

INTRODUCTION

Papillary fibroelastomas (PFE) is a rare neoplasm, with an estimated incidence of 0.0017–0.3300 percent¹. While the majority of PFEs are solitary, previous reviews found the incidence of multiple PFE to range from 7 to 13 percent². PFEs are most commonly found on valvular surfaces, with some series reporting up to 73% of all cases, mainly in the aortic valve. In most cases, the lesions appear to be limited to one or two cardiac surfaces².

CLINICAL CASE

We present a case of a 72-year-old man who was

referred to our hospital for suspected valvular vegetations on transthoracic echocardiography (TTE; Figure 1). The patient was asymptomatic and physical examination was normal.

The patient had past history of chronic alcohol and tobacco abuse (50 pack-year). He had also history of ischemic stroke two years previously, with full neurological recovery. No valvular dysfunction or masses were found in stroke related echocardiographic studies. At the time of presentation, the patient had been receiving monthly intracavitary mitomycin chemotherapy for one year following transurethral resection of high grade urothelial papillary carcinoma (T1N0M0). He had no known history of previous heart disease.

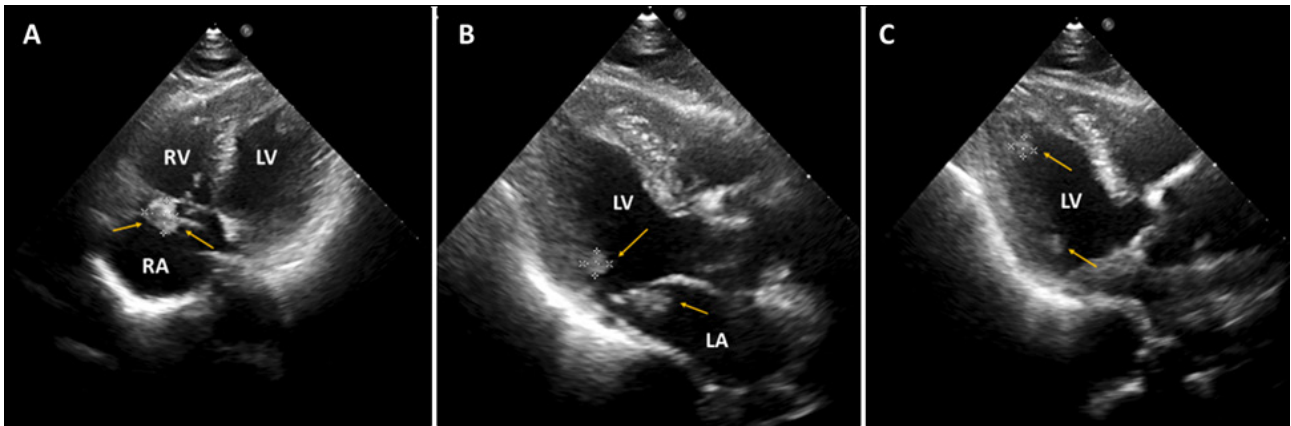


Figure 1

Transthoracic echocardiography - Nodules in the right atrium (RA) are attached to the atrial side of the posterior and septal tricuspid valve leaflets (13 × 15 mm and 7 mm) (A, arrows). A 17 × 16 mm nodule in the left atrium (LA) (B, arrow) is attached to the atrial side of the mitral valve, and there are three 9-mm nodules on the wall of the left ventricle (LV) (B-C, arrows). RV, right ventricle.

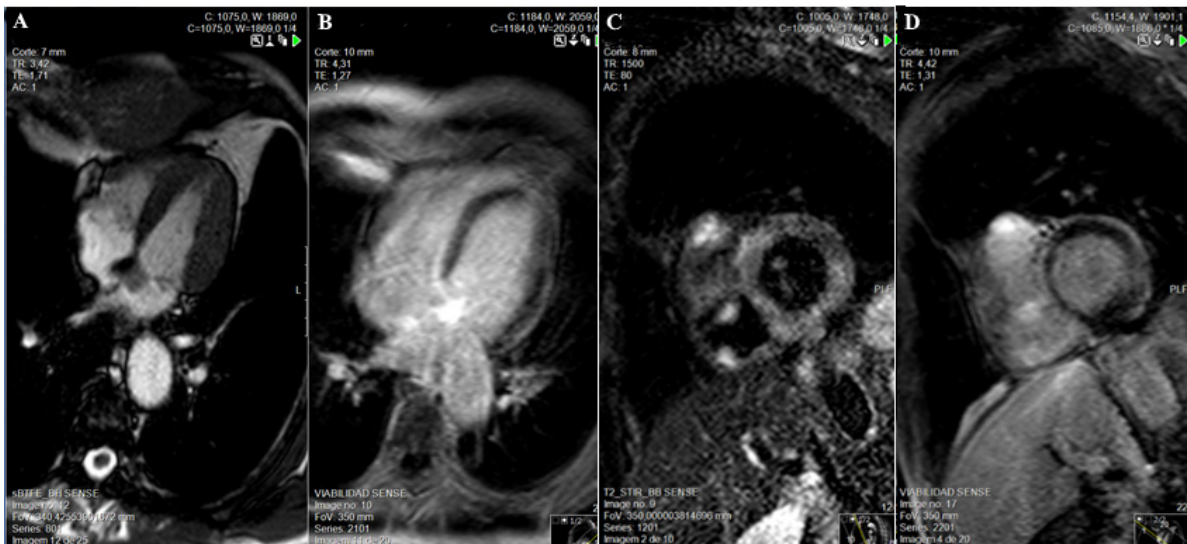


Figure 2

Cardiac magnetic resonance imaging - The 4-chamber cine SSFP study shows isointense masses attached to the interatrial septum (A). T2-weighted STIR showing signal hyperintensity of the masses (C). In B and D, the nodules show late enhancement after gadolinium.

Transesophageal echocardiography (TEE) showed multiple masses of various sizes affecting every valve, multiple chambers, and the interatrial septum. The masses displayed intermediate heterogeneous echogenicity. The largest nodules (each approximately 10 × 15 mm) were located on the atrial side of the posterior leaflet of the tricuspid valve and on the atrial side of the posterior leaflet of the mitral valve; the latter was more heterogeneous and friable. Other small masses were seen at the aortic and pulmonary valves, near the fossa ovalis, the right atrial roof, and the subvalvular apparatus of the mitral valve. No valve dysfunction was observed and left ventricular systolic function was preserved.

Laboratory evaluation for tumor, inflammatory, and autoimmune biomarkers was negative. Cerebral magnetic resonance imaging showed a sequelae lesion in the right hemisphere, probably of cardioembolic origin, although a clear correlation with an embolic phenomenon related to the masses could not be established. PET scan was performed with no significant results.

Cardiac magnetic resonance (CMR) also showed multiple, mobile, rounded nodular masses involving both atrioventricular valves, the subvalvular apparatus, the aortic valve, the pulmonary valve (Figure 2), and the atrial septum. The nodules were isointense on the steady-state free precession (SSFP) cine sequence and they showed

a homogeneous hyperintense signal on T2-weighted short-tau inversion recovery (T2w-STIR) sequences, not suppressing with fat saturation, and demonstrating late enhancement after gadolinium. These findings pointed to the possibility of PFE, excluded thrombus, and made metastasis less likely.

Considering the high embolic risk, complete surgical-view resection was performed under cardiopulmonary bypass and cardioplegic arrest through left and right atriotomies, an aortotomy, and a longitudinal pulmonary artery incision (Figure 3). Nodules were excised from the left and right atria, the left ventricle, the aortic side of the aortic valve, the atrial side of the mitral valve, the atrial side of the tricuspid valve, and the pulmonary side of the pulmonary valve by using a simple shave technique. All native valves were preserved, and intraoperative TEE

thermore, to the authors' knowledge, no other cases of full valvular as well as extensive intra-cavity involvement have been reported.

The most common complication of PFE is thromboembolic stroke, with an incidence as high as 53%³. In the present case, we cannot directly associate the patient's history of neurological symptoms and embolic stroke with our findings, however smaller nodules with embolic potential could have eluded detection by echocardiography at the time.

PFE management remains controversial with some series reporting a high success rate with simple tumor shave resection³, while other authors recommend valve replacement⁵. Due to the extensive distribution of the nodules and the involvement of all cardiac valves with no functional impact, simple resection and valve preserva-

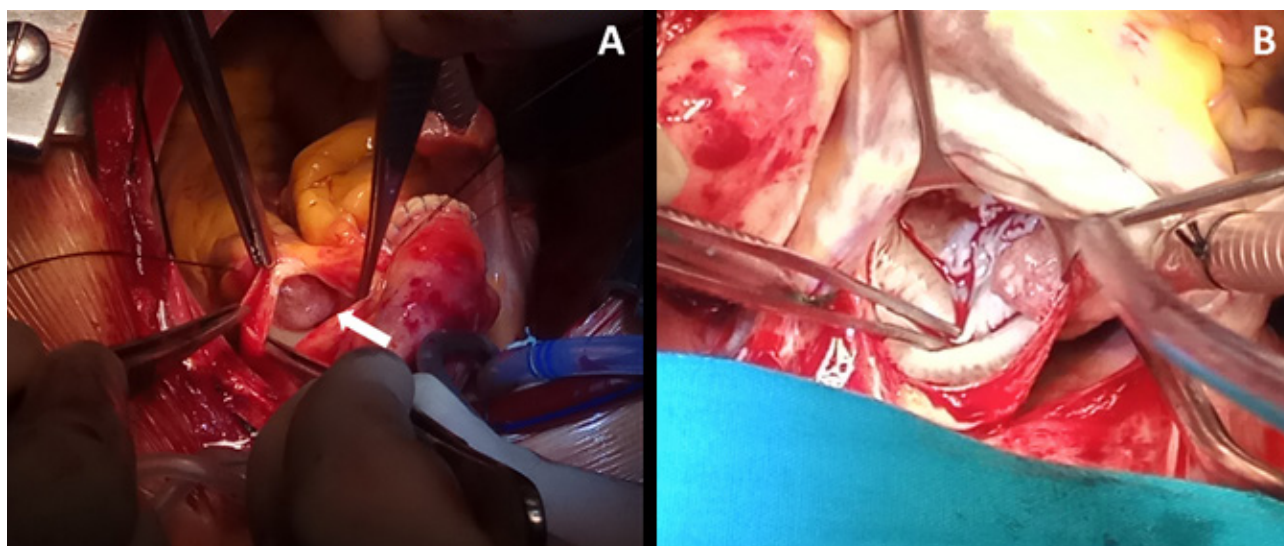


Figure 3

Surgeon's View - The pulmonary valve nodule is seen after longitudinal incision of the pulmonary artery (A, arrow). Nodule in the left atrium (B).

showed no valve dysfunction and no residual nodules.

Histopathological evaluation of the specimens were compatible with multifocal PFE (Figure 4).

The patient had an uneventful recovery and was discharged on the seventh postoperative day. At 6-month follow-up TTE showed no recurrence of the masses.

DISCUSSION

Panvalvular PFE has been reported previously³, however, only one previous case⁴ with simultaneous involvement of all cardiac valves has been reported. Fur-

tion were chosen.

CONCLUSION

To our knowledge, no other cases of both all valvular and most cardiac chambers involvement have been previously reported. The complexity of the case made the diagnosis challenging, as other, more common sources of intracardiac masses were also plausible. Surgical resection was feasible and safe. In conclusion, this case emphasizes the necessity for a well-structured and multidisciplinary approach, keeping in mind that rarer entities may coexist even in the most unlikely scenarios.

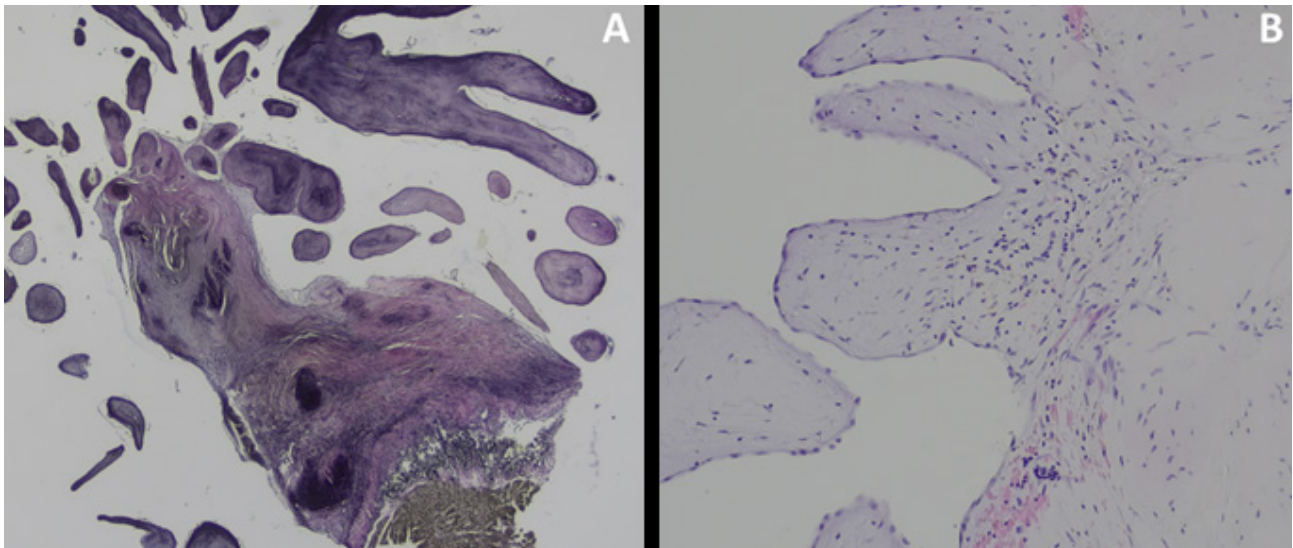


Figure 4

Histological imaging - The findings are compatible with papillary fibroelastoma (A. Elastin staining at 200× magnification; B. Hematoxylin-eosin at 200× magnification).

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