Decision-Making in a Pulmonary Valve Fibroelastoma: The Role of Intraoperative Transesophageal Echocardiography

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INTRODUCTION

Cardiac tumors are uncommon pathologies.[1] Echocardiography is a key to their identification, assessment of their local expansion, and hemodynamic consequences,[2] as well as to surgical decision-making. In most cases, treatment consists of immediate resection to avoid complications such as embolic events.[3] In adult patients, most cardiac tumors are myxomas (80%).[4,5] Cardiac fibroelastomas (CFEs) are the second most common primary tumors of the heart and are commonly attached to the aortic valve. In rare cases, CFEs may be attached to the pulmonary valve.[6]

We herein present the case of a 74-year-old patient with a tumor located on the pulmonary valve. After resection, it was identified as a CFE. We will discuss how transesophageal echocardiography (TEE) has been a mainstay for diagnosis, decision-making, and postoperative evaluation of this unusual condition.

CASE PRESENTATION

This is a case of a 74-year-old man who presented to the emergency department 3 months before surgery for a syncopal episode. The patient has a medical history of transient ischemic attack, a newly discovered indeterminate nodule in the left lung, and cardiovascular risk factors: type 2 diabetes, hypertension, and dyslipidemia. There was no identifiable cause for the syncope. In this context, a transthoracic echocardiography (TTE) was done and showed an otherwise normal study except for an incidental mass attached to the ventricular side of the pulmonary valve measuring 1.3 × 1.1 cm. It was mobile with heterogeneous echogenicity and regular edges. There was a trace pulmonary regurgitation (PR). Due to these physical characteristics, the main hypothesis was a myxoma with a risk of enlargement or embolic complications. However, the simultaneous existence of a lung nodule and the unusual location raised doubts about the origin of the mass. The preoperative coronary angiogram showed an 80% stenosis of the obtuse marginal 2 (OM2) and a 50% stenosis of the OM1. The surgical plan for this patient was a coronary artery bypass graft and pulmonic valve mass removal, with consideration of a pulmonic valve replacement in case it was needed. A preoperative TEE was performed to better assess the extent of the valve involvement and the physiologic and hemodynamic changes with the mass relative to the pulmonary valve.

On the day of surgery, the TEE showed normal biventricular systolic function with no wall motion abnormality and normal functioning mitral, aortic, and tricuspid valves. The exam confirmed the presence of a mass measuring 1.2 × 1.1 cm on the ventricular side of the pulmonary valve (Figures 1-3, Video 1). It was a heterogeneous, pedunculated mass with smooth edges that protruded toward the main pulmonary artery during systole and rested on the right ventricular (RV) outflow tract (RVOT) during diastole (Figure 4 and Video 2). It was not associated with any significant degree of PR or flow acceleration across the valve (mean gradient, 1.4 mm Hg).

The consideration for valve replacement will depend on the TEE post–mass removal in the post–cardiopulmonary bypass period. The surgery proceeded unremarkably. To prevent embolism, the surgeon started with the tumor resection. The tumor was attached to the undersurface of the anterior leaflet of the pulmonary valve (Figure 5), which was shaved to remove it. The pulmonary valve grossly appeared normal. The coronary artery bypass graft was then performed (vein graft to OM2). After the cardiopulmonary bypass period, the TEE showed normal left ventricular systolic function with no new wall motion abnormality and no significant aortic, mitral, or tricuspid valve pathology. The right ventricle (RV) had a normal size, but the systolic function was visually mildly reduced with a fractional area

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TEE, midesophageal, RV inflow-outflow (69°) view, demonstrates a mobile, round 1.2-cm mass in the RVOT in diastole and protruding into the main pulmonary artery in systole.

Video 2: Two-dimensional TEE, upper esophageal, ascending aorta short-axis (0°) view, demonstrates a pediculated mass attached to the pulmonary valve protruding into the main pulmonary artery in systole.

Video 3: Two-dimensional TEE with color flow Doppler, midesophageal RV inflow-outflow (52°) view, after removal of the mass demonstrates a centrally directed, long jet of PR with a narrow origin consistent with mild to moderate grade.

View the video content online at www.cvcasejournal.com.

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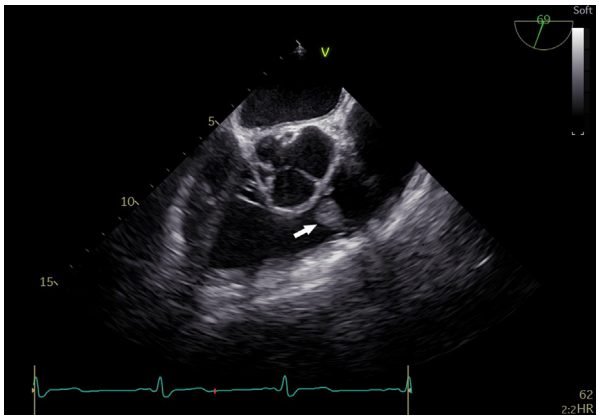

Figure 1 Two-dimensional TEE, midesophageal, RV inflow-outflow (69°) view, early diastolic phase, demonstrates a round, heterogeneous echogenicity, smooth-edged mass (*arrow*) in the RVOT.

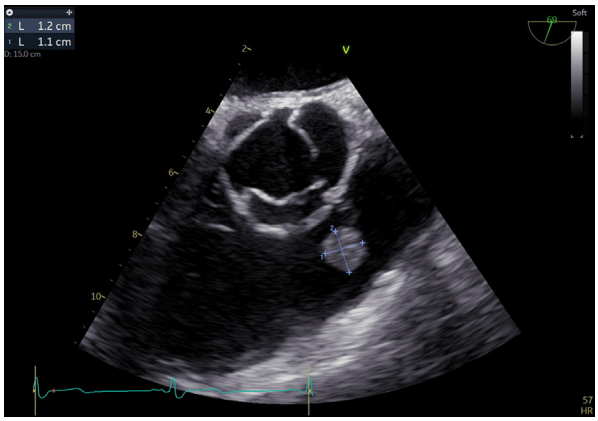

Figure 3 Two-dimensional TEE, midesophageal, RVOT (69°) view in midsystole demonstrates the pulmonary valve mass, which measured 1.2 × 1.1 cm in maximal dimensions.

change at 34%. The previous mass associated with the pulmonary valve was no longer seen. The septum and cavitary walls were intact. There was mild-moderate, centrally directed PR with a long jet (50 mm), narrow origin, and dense continuous-wave Doppler with a pressure half-time >200 ms (Figures 6 and 7, Video 3).[7] After discussion with the surgical team, it was decided to not replace the valve.

The postoperative period was marked by a quick recovery. On postop day 3, the TTE showed a normal pulmonary valve without residual mass or stenosis (mean gradient, 2 mm Hg). The RV was normal in size and function. The PR jet was narrow with a very small penetration in the RVOT; its envelope in continuous-wave Doppler was faint, not allowing measurement, and was classified as mild. The rest of the study was otherwise normal. The patient developed persistent atrial fibrillation and was treated medically. The patient was finally discharged from the hospital on postoperative day 6. The final pathology analysis of this tumor resulted in the diagnosis of papillary fibroelastoma.

DISCUSSION

Cardiac tumors are uncommon, and management guidelines are based on a small number of reported cases.[8] Most cardiac tumors are benign, and 87% of primary benign cardiac tumors are myxomas.

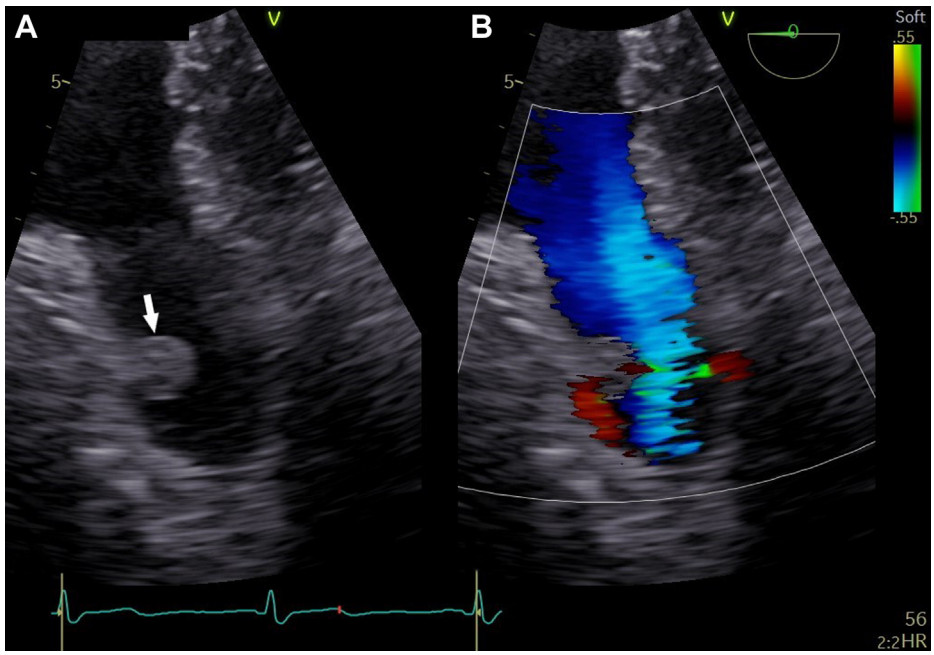

Figure 2 Two-dimensional TEE, deep transgastric RV outflow (0°) view without **(A)** and with **(B)** color flow Doppler in late systole demonstrates a nonobstructive mass (*arrow*) with laminar, nonturbulent flow in the RVOT attached to the pulmonary valve.

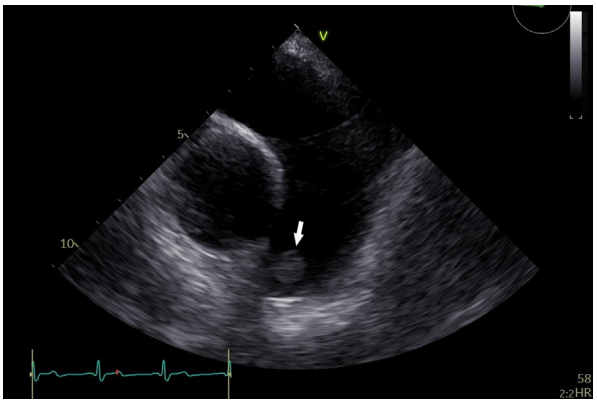

Figure 4 Two-dimensional TEE, upper esophageal, ascending aorta short-axis (0°) view, demonstrates a pediculated mass attached to the pulmonary valve protruding in the main pulmonary artery in systole (*arrow*).

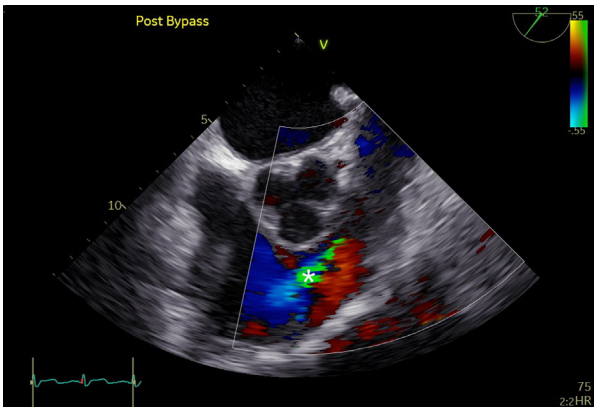

Figure 6 Two-dimensional TEE with color flow Doppler, midesophageal RV inflow-outflow (52°) view, late diastolic phase after removal of the mass, demonstrates a centrally directed, long jet of PR (*) with a narrow origin consistent with mild to moderate grade.

In comparison, the remaining 10% to 15% include lipomas, fibromas, rhabdomyomas, hemangiomas, teratomas, papillary fibroelastomas, pericardial cysts, or cystic tumors of the atrioventricular node.[5] There is a limited reported incidence of CFEs, which usually arise from valve leaflets, most commonly in the aortic and mitral valves.[9] Clinically, CFEs have presented with sudden death, heart failure, and embolic events. These reports show that tumor mobility is the only predictor of mortality and embolization-related morbidity. Left-sided CFE is more clinically reported since they present more symptomatically. Right-sided CFE is less documented and is primarily an incidental finding from autopsies and imaging studies.[10] Few cases were reported to have cyanotic spells from RVOT obstruction or pulmonary embolism from tricuspid fibroelastoma[11,12]; however, the causative relationship was not described. Our patient was asymptomatic, and imaging was concerning for a myxoma. Symptomatic patients should be treated surgically because the successful complete resection of CFE is curative, and the long-term postoperative prognosis is excellent.[9]

Saric *et al.*[3] recommended TEE over TTE for CFE assessment. Better evaluation of the tumor size, shape, attachment, and relation with surrounding structures is key to estimating the embolic risk. In this regard, sweeps in all directions, three-dimensional echocardiography with multiplanar reconstruction, and echocardiography with an ultrasound-enhancing agent are advised for a close study of the tumor.[3] Transesophageal echocardiography is also instrumental in giving real-time data to facilitate its removal. Our patient had mild to moderate PR thought to be secondary to spatial conformation changes and transient postbypass pulmonary hypertension. On follow-up, there was mild PR on TTE postoperative day 3. Reported CFE recurrence is typically considered rare,[9] but more recently, the recurrence rate was noted to be >10%.[13]

This case is an example of how TEE is able to contribute to the active management of a patient treated for a rare case where questions about the diagnosis and surgical management are present.

CONCLUSION

Although rare, depending on their size and location, cardiac tumors can have significant hemodynamic consequences. It is essential to understand physiology to anticipate management. Transesophageal echocardiography is a critical tool for making management-defining decisions in perioperative patient care. In a way, it helps predict the success of the surgery, assisting surgeons in deciding whether or not revision should be made at that crucial moment.

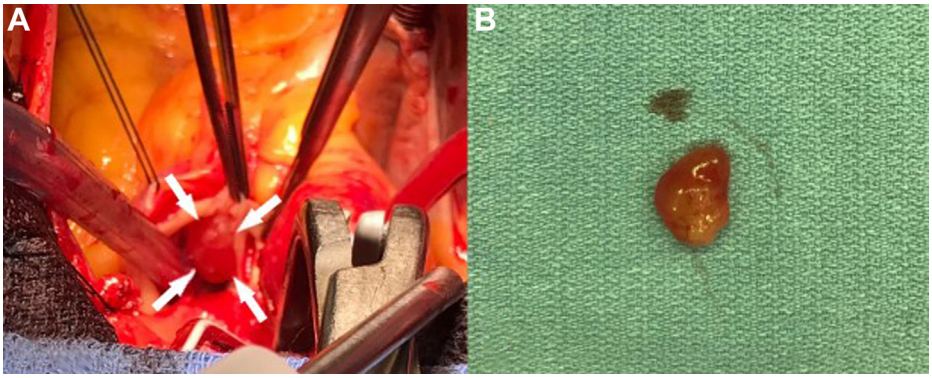

Figure 5 **(A)** Intraoperative photograph of the in situ mass attached to the ventricular side of the pulmonary valve (*arrows*). **(B)** Intraoperative photograph of the excised mass.

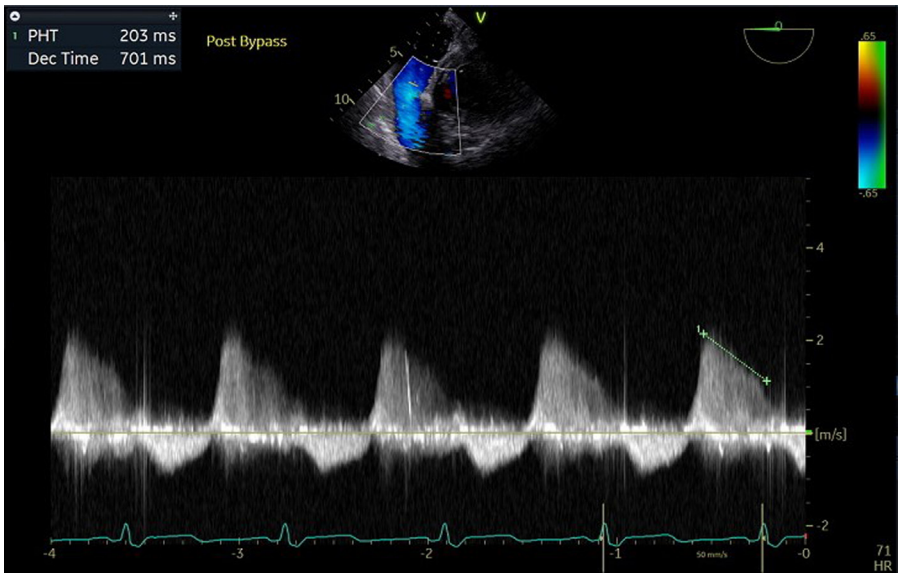

Figure 7 Two-dimensional TEE, deep transgastric (0°) view with continuous-wave Doppler cursor aligned with postoperative PR demonstrates a nonsevere pressure half-time of 203 ms.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

SUPPLEMENTARY DATA

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REFERENCES

1. Reynen K. Frequency of primary tumors of the heart. Am J Cardiol 1996;77:107.
2. Bhattacharyya S, Khattar RS, Gujral DM, et al. Cardiac tumors: the role of cardiovascular imaging. Expert Rev Cardiovasc Ther 2014;12:37-43.
3. Saric M, Armour AC, Arnaout MS, et al. Guidelines for the use of echocardiography in the evaluation of a cardiac Source of embolism. J Am Soc Echocardiogr 2016;29:1-42.
4. Odim J, Reehal V, Laks H, et al. Surgical pathology of cardiac tumors. Two decades at an urban institution. Cardiovasc Pathol 2003;12:267-70.
5. Yu K, Liu Y, Wang H, et al. Epidemiological and pathological characteristics of cardiac tumors: a clinical study of 242 cases. Interact Cardiovasc Thorac Surg 2007 Oct;6:636-9.
6. Sun JP, Asher CR, Yang XS, et al. Clinical and echocardiographic characteristics of papillary fibroelastomas. Circulation 2001;103:2687-93.
7. Zoghbi WA, Adams D, Bonow RO, et al. Recommendations for noninvasive evaluation of native valvular regurgitation. J Am Soc Echocardiogr 2017;30:303-71.
8. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. Medicine (Baltimore) 2001;80:159-72.
9. Gowda RM, Khan IA, Nair CK, et al. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. Am Heart J 2003;146:404-10.
10. Shahian DM, Labib SB, Chang G. Cardiac papillary fibroelastoma. Ann Thorac Surg 1995;59:538-41, ISSN 0003-4975.
11. Karapanagiotidis GT, Lees N, Howlett P, et al. Tricuspid valve papillary fibroelastoma: an unusual case of dizzy spells. Perfusion 2012;27:156-9.
12. Neerukonda SK, Jantz RD, Vijay NK, et al. Pulmonary embolization of papillary fibroelastoma. Arising from the tricuspid valve. Tex Heart Inst J 1991;18:132-5.
13. Sorour AA, Kurmann RD, El-Am EA, et al. Recurrence of pathologically proven papillary fibroelastoma. Ann Thorac Surg 2022;113:1208-14.