

## CASE REPORT

## BEGINNER

## CLINICAL CASE

# Giant Tricuspid Valve Fibroelastoma Incidentally Diagnosed During Routine Stress Testing



Christina Cardy, DNP,<sup>a</sup> Nicole Riddle, MD,<sup>b</sup> John Dunning, MD,<sup>c</sup> Allan Chen, MD<sup>a</sup>

## ABSTRACT

Papillary fibroelastomas (PFEs) are the most common valvular tumor, typically occurring on left-sided valves. We describe the evaluation and treatment of a giant tricuspid PFE in a healthy 43-year-old police officer who was referred for evaluation of frequent premature ventricular contractions during job-related treadmill stress testing. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2019;1:564–8) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Primary cardiac tumors are rare, with their frequency in autopsy ranging from 0.001% to 0.03% (1). About 75% of cardiac tumors are benign, and up to 12% of cases are found incidentally. Papillary fibroelastoma (PFE) is the most common valvular tumor, with 36% involving the aortic valve and 29% affecting the mitral valve (1,2). Less

frequently, the tumor infiltrates the pulmonic (8%) or tricuspid valve (15%) (3). We describe the evaluation and treatment of a giant PFE in a 43-year-old, otherwise healthy man who presented to cardiology after job-related treadmill stress testing revealed frequent premature ventricular contractions (PVCs). This report highlights the atypical presentation and size of this tumor and reminds us to consider this differential in healthy patients diagnosed with an intra-cardiac mass.

## LEARNING OBJECTIVES

- To describe the general diagnostic evaluation and work-up of a patient diagnosed with an intracardiac mass.
- To discuss various cardiac imaging modalities and their roles in the diagnosis and differentiation of PFE.
- To describe the etiology, basic pathology, and typical characteristics of PFE.
- To identify risk factors for PFE-related death or nonfatal embolization.
- To discuss the appropriateness of surgical excision of PFE.

## HISTORY OF PRESENTATION

A 43-year-old male police officer with no significant past medical history was referred to cardiology after having an increased PVC burden during routine job-related stress testing. The stress test was aborted, and a point-of-care echocardiogram was performed that demonstrated a possible right ventricular thrombus. The patient was referred to the emergency room where a formal echocardiogram confirmed the presence of a 13.6 mm × 10.2 mm echo-dense mass on

From the <sup>a</sup>Division of Cardiovascular Sciences, University of South Florida Health, Tampa, Florida; <sup>b</sup>Department of Pathology and Cell Biology, University of South Florida Health, Tampa, Florida; and the <sup>c</sup>Division of Cardiothoracic Surgery and Transplantation, University of South Florida Health, Tampa, Florida. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Informed consent was obtained for this case.

Manuscript received April 25, 2019; revised manuscript received July 11, 2019, accepted August 12, 2019.

the ventricular aspect of the tricuspid valve. The left and right ventricular systolic function were normal. The patient denied chest pain, palpitations, intravenous drug use, oral infections, or known coagulopathies. He was admitted for further testing.

### PAST MEDICAL HISTORY

The patient had no significant past medical history.

### DIFFERENTIAL DIAGNOSIS

Differential diagnoses included thrombus, myxoma, lymphoma, PFE, and vegetation.

### INVESTIGATIONS

The patient's electrocardiogram obtained during the initial stress test demonstrated PVCs all of right ventricular outflow tract (RVOT) origin (Figure 1). After transthoracic echocardiography, further investigation with transesophageal echocardiography (TEE) revealed an elongated, mobile mass extending from the superior vena cava to the RVOT. There were areas of potential attachments to the septum along with

chordal involvement (Figure 2). Cardiac magnetic resonance (CMR) indicated a 40 mm × 12 mm × 9 mm mass in the right ventricle that was T2-weighted hypointense and did not exhibit first pass or late gadolinium enhancement, most consistent with thrombus (Figure 3, Video 1). The mass appeared to be attached to the right ventricular side of the interventricular septum, just above the posterior leaflet of the tricuspid valve. There was a filling defect within the superior vena cava suspicious for thrombus.

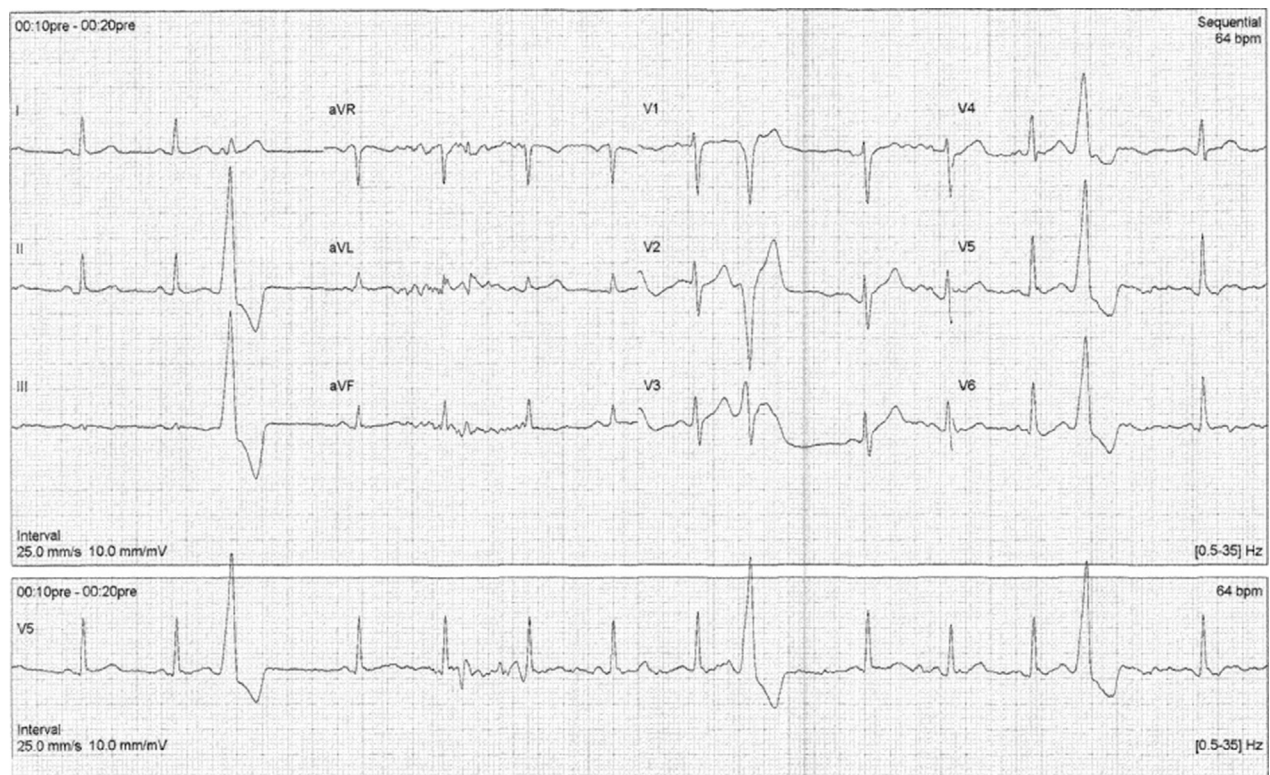
### MANAGEMENT

Based on the TEE and CMR findings described previously, the patient was initiated on a heparin infusion, and cardiothoracic surgery was consulted. CMR was repeated after 4 days of therapeutic anticoagulation to assess for resolution of the mass. Repeat CMR revealed an 11 mm × 7 mm intraventricular mass significantly reduced in size. The patient was discharged home on warfarin with instructions to pursue

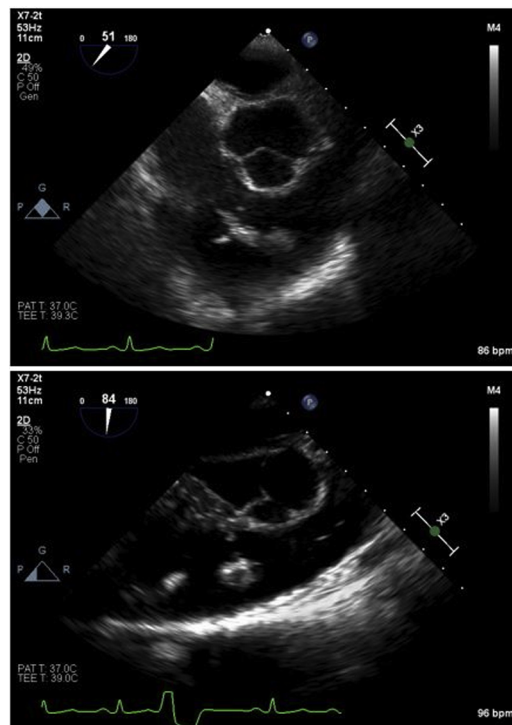
### ABBREVIATIONS AND ACRONYMS

- CMR = cardiac magnetic resonance
- PFE = papillary fibroelastoma
- PVC = premature ventricular contraction
- RVOT = right ventricular outflow tract
- TEE = transesophageal echocardiogram

FIGURE 1 Electrocardiogram Obtained Prior to Admission



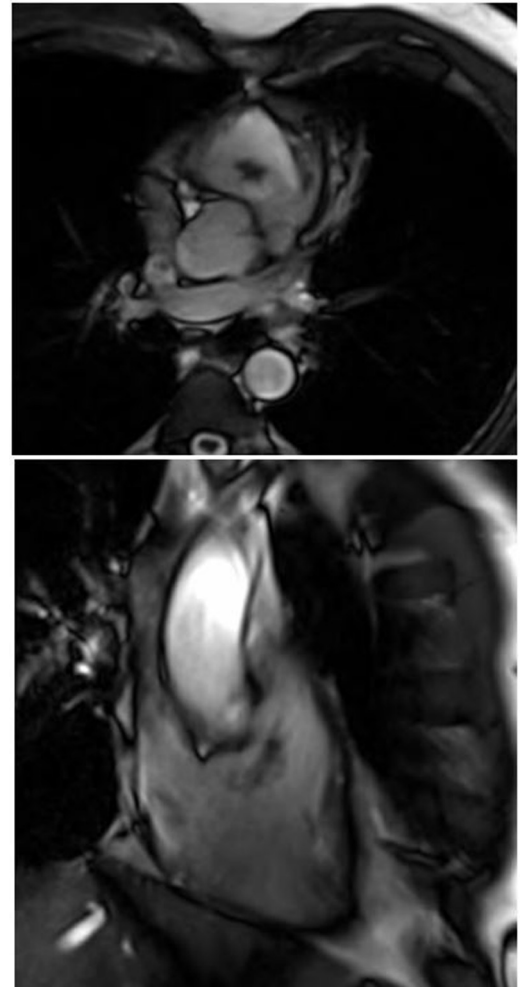
Electrocardiogram obtained during initial stress test demonstrated premature ventricular contractions of right ventricular outflow tract origin.

**FIGURE 2** Transesophageal Echocardiography

Mobile elongated echodensity demonstrated in the right ventricle and outflow tract. See [Video 2](#).

hypercoagulable work-up and repeat echocardiography in 3 months.

At the 3-month follow-up, hypercoagulable work-up was negative, and echocardiography demonstrated no resolution of the mass despite uninterrupted therapeutic anticoagulation. The patient was referred for surgery because of inconclusive TEE and CMR findings, the size and mobility of the mass, the perceived risks of superior vena cava obstruction and embolization, and the mass persisted after anticoagulation. On entering the brachiocephalic vein for cardiopulmonary bypass cannulation, an extension of the mass was visible in the lumen lying free from adhesion to the wall; the mass was amputated. In the right atrium, there was no adherent mass. There was a nodular, multilobed mass adherent to and appearing to take origin from the ventricular aspect of the tricuspid valve measuring up to 150 mm when fully elongated ([Figure 4](#)). The tricuspid valve was not salvageable for repair. The mass was removed en bloc with the valve leaflets, and a tissue prosthesis was implanted. Histological examination of the resected tissue revealed a benign PFE ([Figure 5](#)).

**FIGURE 3** Cardiac Magnetic Resonance Imaging (Steady-State Free Precession)

Irregular mobile mass seen in the right ventricular outflow tract that was T2-weighted hypointense and did not exhibit first pass or late gadolinium enhancement. See [Video 1](#).

## DISCUSSION

The majority of PFEs are found incidentally at the time of echocardiography, cardiac surgery, or autopsy (2). Males comprise 55% of patients, and the valvular surface is the predominant location of the tumor (3). The aortic and mitral valves are most commonly involved; in a retrospective review of 725 cases, the incidence of tricuspid valve PFE was 15% (3). Clinically, PFEs have presented with stroke, myocardial infarction, sudden death, heart failure, syncope, and pulmonary or peripheral embolism (3,4). It is possible that the usually asymptomatic right-sided tumors do

not necessitate medical attention and are thereby underreported (5).

Grossly, PFE is an avascular, gelatinous mass that consists of papillary fronds containing fibrous tissue, elastic fibers, and smooth muscle cells in a mucopolysaccharide matrix covered with hyperplastic endocardial cells (3). It is best visualized when immersed in water and is classically described as having a sea anemone-like appearance (5).

The origin of PFE is unknown, and, to date, no clear risk factors for the development of PFE have been reported (5). They have been considered as hamartomas, organized thrombi, and inflammatory foci caused by unusual endocardial responses to infection or trauma. Interestingly, this patient experienced right chest trauma during a motor vehicle collision 2 years before diagnosis. The patient was T-boned from the right side at a high speed. Airbags were deployed, and his right chest took impact. A transthoracic echocardiogram completed following that accident revealed no evidence of cardiac tumor.

Most PFEs are <10 mm in diameter; giant examples such as the case described in this report, especially in the absence of symptoms, are considered rare. Transthoracic echocardiography is useful for the initial evaluation and demonstrates a small, mobile, pedunculated valvular or endocardial mass, which may prolapse into the cardiac chambers (6). Occasionally, TEE is required for further assessment.

The growing clinical use of cardiac computed tomography and CMR may lead to an increased discovery of this tumor entity (5). CMR offers soft tissue characterization, evaluation of valvular function, and differentiation between a tumor and thrombus. Electrocardiographic findings are nonspecific, but occasionally patients may have atrial arrhythmias. This patient's electrocardiogram obtained during the initial stress test demonstrated PVCs all of RVOT origin (Figure 1). Post-operatively, no PVCs were captured on numerous follow-up electrocardiograms. Because a portion of the PFE was constantly mobile within the RVOT, it is indeterminate if the PVCs were directly related to the PFE entering the RVOT region (Video 2). Cardiac catheterization can be associated with an added risk because the catheter may dislodge a fragment of tumor or adherent thrombi, resulting in embolism (5).

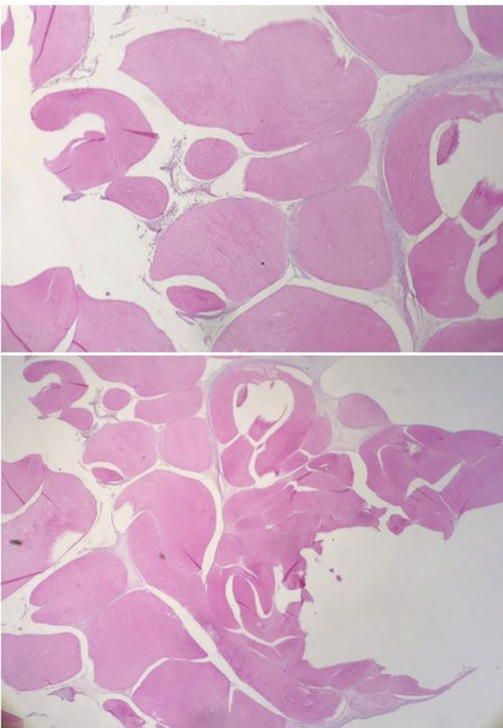
A survey of the literature did not reveal any reports of tricuspid valve PFE in a healthy adult coming to a clinician's attention because of increased PVC burden or any other cases of giant, right-sided PFEs. Additionally, the largest PFE reported measured 70 mm long, making this 150-mm PFE the largest on record (7). The literature noted that tumor mobility is the

FIGURE 4 Gross Examination



Nodular, multilobed mass adherent to and appearing to take origin from the ventricular aspect of the tricuspid valve and its associated chordae.

FIGURE 5 Histologic Examination



Characteristic 3-layered pattern of narrow to widened, elongated, and branching papillary fronds composed of central avascular collagen and variable elastic tissue surrounded by acid mucopolysaccharides and lined by hyperplastic endothelial cells.

only independent predictor of PFE-related death or nonfatal embolization (3). Depending on size and mobility, a PFE involving an atrioventricular valve can cause obstruction to filling of the right ventricle or left ventricle, mimicking tricuspid or mitral stenosis. If the tumor is large enough, mobile, and has a long stalk, complete obstruction of the RVOT or left ventricular outflow tract may occur, resulting in syncope or sudden death (3).

Conservative, nonsurgical management of PFE treated with long-term anticoagulation and periodic echocardiography has been described, although no randomized controlled data are available on efficacy (3,8). With regard to this patient's profession, the position of the U.S. Department of Defense is that a police officer taking anticoagulation is not fit to perform the normal duties of a police officer because of the risk of incurring serious injury during arrest and restraint and may seek early retirement (9). The risks of surgery, which include but are not limited to degenerative valve disease, infection, reoperation, and conduction abnormalities, and the benefits of surgery versus long-term anticoagulation were discussed. Through an interdisciplinary, shared decision-making approach, the patient elected to proceed with surgical resection of the mass.

#### FOLLOW-UP

The patient's recovery was complicated by third-degree heart block requiring pacemaker placement. The presence of a lead traversing the tricuspid valve can result in regurgitation; the potential for device

leads to interfere with tricuspid valve function has gained increasing recognition as having hemodynamic and clinical consequences associated with incremental morbidity and death (10). The patient was educated on the signs and symptoms of right-sided heart failure before being discharged home in stable condition. After 2 months of recovery, the patient was able to return to work as a police officer without restrictions. At the 6-month follow-up, he had no acute complaints.

#### CONCLUSIONS

Cardiac tumors represent an important cause of morbidity and mortality and affect patients of all ages. Although the majority of cardiac tumors are benign and curable with surgery, health care providers should be aware that benign tumors such as PFE still may be fatal because of their size and location. As cardiac imaging modalities and techniques continue to improve, the identification and prevalence of these tumors have increased. To the best of our knowledge, this case of a giant tricuspid valve PFE is the largest PFE reported to date.

**ACKNOWLEDGMENTS** The authors thank the University of South Florida Cardiology, Cardiothoracic Surgery, and Pathology.

**ADDRESS FOR CORRESPONDENCE:** Dr. Christina Cardy, University of South Florida Health, Division of Cardiovascular Sciences, 2 Tampa General Circle, 5th Floor, Cardiovascular Sciences, Tampa, Florida 33606. E-mail: [ccardy@usf.edu](mailto:ccardy@usf.edu).

#### REFERENCES

- Maraj S, Pressman G, Figueredo V. Primary cardiac tumors. *Int J Cardiol* 2009;2:152-6.
- Edwards F, Hale D, Cohen A, Thompson L, Pezzella T, Virmani R. Primary cardiac valve tumors. *Ann Thorac Surg* 1991;52:1127-31.
- Gowda R, Khan I, Nair C, Mehta N, Vasavada B, Sacchi T. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. *Am Heart J* 2003;146:404-10.
- Patel J, Sheppard M. Pathological study of primary cardiac and pericardial tumours in a specialist UK centre: surgical and autopsy series. *Cardiovasc Pathol* 2010;19:343-52.
- Sydow K, Willems S, Reichenspurner H, Meinertz T. Papillary fibroelastomas of the heart. *Thorac Cardiovasc Surg* 2008;56:9-13.
- Tsukube T, Ataka K, Taniguchi T, Yokoyama M, Hanoika K. Papillary fibroelastoma of the left atrial appendage: echocardiographic findings. *Ann Thorac Surg* 2000;70:1416-7.
- Jha NK, Khouri M, Murphy DM, et al. Papillary fibroelastoma of the aortic valve—a case report and literature review. *J Cardiothorac Surg* 2010;5: 84-9.
- Seol SH, Kim DS, Han YC, et al. Nonsurgical management of a tricuspid valvular pedunculated papillary fibroelastoma. *Cardiovasc Ultrasound* 2009;7:44.
- Secretary of Defense. C3.4.2.2. Police officers and security guards: occupational medical examinations and surveillance manual. 2018;40. Available at: <https://www.esd.whs.mil/Portals/54/Documents/DD/issuances/dodm/605505mp.pdf?ver=2017-08-14-111314-123>. Accessed March 19, 2019.
- Chang M, Manning W, Ebrille E, Zimmerman P. Tricuspid valve dysfunction following pacemaker of cardioverter-defibrillator implantation. *J Am Coll Cardiol* 2017;69: 2331-41.

**KEY WORDS** cardiac magnetic resonance, cardiac tumor, echocardiography, papillary fibroelastoma, pathology, tricuspid valve

**APPENDIX** For supplemental videos, please see the online version of this paper.