Incidental finding of a large pulmonary valve fibroelastoma: A case report

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Abstract Papillary fibroelastoma (PFE) is an uncommon primary neoplasm of cardiac origin. They are solitary neoplasms that historically were an incidental finding at the time of autopsy. With the advent of two-dimensional echocardiography, symptomatic cases have been reported in current literature, thus causing a paradigm shift in the management of these tumors. Although the majority of PFE are benign, because of their potential risk for complication related to embolic and obstructive phenomena, they are now considered hazardous and require tumor excision even in asymptomatic patients. We report a case of an asymptomatic incidental large papillary fibroelastoma within the right ventricular outflow tract.

Background

Primary intracardiac tumors are extremely rare with an overall incidence of <0.3%.1 Papillary fibroelastomas comprise only 8% of all primary cardiac tumors, of which the vast majority are benign.2,3 Our understanding of papillary fibroelastosomas (PFE) has changed from an innocuous post-mortem finding to a potentially life threatening pathology. Historically, small right-sided PFEs discovered on routine echocardiogram in asymptomatic patients were observed clinically.4 Two-dimensional echocardiography on symptomatic patients, however, has demonstrated that these tumors have a tendency for embolization and obstruction resulting in cerebral vascular accidents, pulmonary embolism and myocardial infarction.5

It is now suggested that all PFE, regardless of size and location, should be resected to avoid embolic or occlusive-related complications.1,6,7 To
our knowledge, only four cases of pulmonary valve PFE have been reported in previous literature. Our case demonstrates a large right-sided PFE in an asymptomatic man that historically could have been observed without intervention, but due to its potential for life threatening complications surgical intervention was carried out with excellent outcome.

Case report

The patient, an asymptomatic 52-year-old African-American male with known hypertension and dyslipidemia, was found to have a large mass in the right ventricular outflow tract on routine echocardiogram. Transesophageal echocardiogram performed for further evaluation, confirmed a mobile, 1.5 cm by 1.4 cm mass attached to the ventricular aspect of the pulmonic valve above the right ventricular outflow tract as seen in long (Fig. 1) and short axes (Fig. 2) of the valve. A coronary angiogram demonstrated normal coronary anatomy without significant disease. Surgical resection was recommended due to the tumor’s large size and mobility, resulting in high risk for embolization. A transverse arteriotomy of the main pulmonary artery was performed revealing a 1.4 × 1.0 × 1.3 cm friable, gelatinous mass fixed onto the ventricular aspect of the pulmonic valve. Gross and microscopic histopathology revealed multiple frond-like structures comprising dense elastin at the core of each frond, coated with collagen and lined by flat endocardial cells—confirming the diagnosis of papillary fibroelastoma (Fig. 3). The patient had an uncomplicated postoperative course and one and half months follow up revealed no complications.

Discussion

PFE represent 8% of all primary cardiac tumors. They are the third most common cardiac primary tumor after myxomas (30%) and lipomas (10%), but are the most common valvular tumor as up to 90% occur on the valvular endocardium.2 PFEs have a predilection for the left side of the heart. The review of current literature demonstrates that symptomatic PFEs are more likely to be located on the left side of the heart.1,2,8 Similar investigations confirm that the most commonly affected valvular site is the aortic valve, followed by the mitral and tricuspid valves, respectively. The pulmonic valve is rarely involved.1,2,8 PFEs are usually small tumors, and it has been reported by Sun et al.4 that 99% of all PFEs were 20 mm or less. The mean age of diagnosis was approximately 60 years, although they have been reported in children and teenagers.9–11 The pathophysiology of these tumors remains unknown. Several possible theories have been reported including previous endothelial damage followed by thrombosis and organization, iatrogenic

Figure 1  Transesophageal echocardiogram short axis view of the base of the heart obtained with multiplane transducer at 40° demonstrating a 1.4 by 1.5 cm mass on the ventricular surface of the pulmonic valve (arrow). LA = left atrium, RVOT = right ventricular outflow tract, AoV = aortic valve, PV = pulmonic valve.
factors and infectious processes related to cytomegalovirus. Cytomegalovirus has been recovered from PFEs suggesting a viral induction of the tumor.

PFEs have a tendency to embolize due to their soft and friable anatomy, often associated with adherent thrombus. The common presenting symptoms are related to embolic phenomena to the brain, heart and lung resulting in stroke, myocardial infarction or pulmonary embolus. Therefore, its location as well as size and mobility determine potential risks and complications.

Even though the majority of cases are asymptomatic because of their potential for life threatening complications, current literature suggests that even small PFEs should be excised.

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**Figure 2** Transesophageal echocardiogram short axis view of the pulmonic valve obtained with multiplane transducer at 0° demonstrating a spherical mass attached to the pulmonic valve (arrow). LA = left atrium, Ao = aorta, PV = pulmonic valve.

**Figure 3** Histology. Surgical specimen demonstrating multiple papillary fronds with a central core of dense fibroelastic tissue (arrow), and loose connective tissue with mucopolysaccharides (hematoxylin–eosin staining).
Surgery is strongly indicated for patients with embolic events, occlusive-related symptoms due to left or right ventricular outflow tract obstruction, and as in our case, those with highly mobile or large tumors.15–18 The aforementioned case demonstrates a rare neoplasm with an unusual size and location that underwent successful resection avoiding potential catastrophic embolic consequences.

References