

Papillary Fibroelastoma: Echocardiographic Characteristics for Diagnosis and Pathologic Correlation

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Objectives. We sought to determine the clinical and echocardiographic characteristics of papillary fibroelastoma (PFE).

Background. PFE is a rarely encountered cardiac tumor about which relatively little is known.

Methods. Institutional records were reviewed for the years 1980 to 1995 for patients with pathologic or echocardiographic diagnosis of PFE. Group 1 included 17 patients with the pathologic diagnosis of PFE who also underwent echocardiography. Echocardiographic features of PFE were established in group 1. Group 2 included 37 patients with only echocardiographic evidence of PFE.

Results. In group 1, 7 (41.2%) of 17 patients had symptoms related to PFE. Neurologic events occurred in 5 (29.4%) of 17 patients. All patients had the tumor surgically removed. During follow-up, no new embolic events occurred. Echocardiographic characteristics of PFE included a small tumor ($12.1 \pm 6.5 \times 9.0 \pm$

4.3 mm), usually pedunculated (14 [94%] of 17 patients) and mobile, with a homogeneous speckled pattern and a characteristic stippling along the edges. PFEs were most common on valvular surfaces (12 [60%] of 20 PFEs) but were not uncommon on other endocardial surfaces (8 [40%] of 20 PFEs). The tumor did not cause valvular dysfunction. In group 2, 16 (43%) of 37 patients were asymptomatic. Five patients (13.5%) had a previous neurologic event. During follow-up (mean 31 months, range 1 to 77), nine neurologic events occurred.

Conclusions. PFEs are associated with embolism, can be diagnosed with echocardiography, are often an incidental clinical finding and do not cause valvular dysfunction.

(J Am Coll Cardiol 1997;30:784-90)

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Papillary fibroelastoma (PFE) is an infrequently encountered, poorly characterized benign tumor found on the endocardium (1-3). Pathologically, these tumors grossly appear like a "sea anemone," because they consist of multiple frond-like projections (Fig. 1). Histologically, they are avascular tumors derived from normal components of the endocardium (Fig. 1) (1-3).

Clinical characteristics are poorly understood. A large experience including echocardiographic analysis is not available. There has not been a systematic review of the clinical characteristics, pathologic correlation with echocardiographic findings or description of long-term follow-up.

In isolated case reports and pathology series (1-3), PFEs have been suggested as a source of embolism (4-19), but the embolic potential in a continuous series of patients remains unknown. With the widespread availability of high resolution echocardiography, PFE is diagnosed more often (20-23).

To further investigate the clinical and echocardiographic

characteristics of PFEs, the Mayo Clinic's experience with these tumors from 1980 to 1995 was reviewed. Questions included 1) the potential for valvular dysfunction; 2) the potential for systemic embolization; 3) the usefulness of echocardiography in making the diagnosis; and 4) how to manage the incidental finding of PFE in an otherwise asymptomatic patient.

Methods

Patient group. The surgical, pathologic and echocardiographic records from the Mayo Clinic were reviewed for the diagnosis of PFE. Patients were separated into two groups.

Group 1. Patients in group 1 had PFE diagnosed pathologically from surgical specimens. The diagnosis was based on the microscopic appearance of the tumor—a core of dense connective tissue surrounded by elastic fibers, smooth muscle cells and loose connective tissue covered with hyperplastic endocardial cells that are avascular (Fig. 1) (1-3). Eighteen patients with histologically proven PFE were identified through the pathology data base at the Mayo Clinic. Patients were excluded if they had not had transesophageal echocardiography (TEE) or transthoracic echocardiography (TTE) (n = 1). Thus, the echocardiographic characteristics of the tumor were identified in 17 patients comprising group 1.

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Manuscript received December 11, 1996; revised manuscript received May 5, 1997, accepted May 22, 1997.

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Abbreviations and Acronyms

CAD = coronary artery disease
CVA = cerebrovascular accident
PFE = papillary fibroelastoma
TEE = transesophageal echocardiography
TIA = transient ischemic attack
TTE = transthoracic echocardiography

Group 2. Since the original description of the two-dimensional features of PFE by Shub et al. (20) at the authors' institution, all patients with the echocardiographic diagnosis of PFE listed in the Echocardiography and Hemodynamic Laboratory data base from January 1, 1980 to January 1, 1995 were identified. During this time, 290,239 patients having echocardiography were enrolled in the data base. Forty-one patients were identified with the potential diagnosis of PFE. All echocardiographic studies previously stored on 3/4-in. videotape were reviewed by the authors (K.W.K., M.E.S.). The presumptive diagnosis of PFE was then confirmed or rejected, based on a review of the echocardiographic videotape and application of the echocardiographic criteria determined in the analysis of group 1. Clinical histories were reviewed. Exclusion criteria included bacterial endocarditis infection or bacteremia ($n = 1$), systemic connective tissue disease ($n = 2$) and failure to meet the echocardiographic criteria established in group 1 ($n = 1$). Therefore, group 2 consisted of 37 patients.

In both groups 1 and 2, the stored videotapes were reviewed and the dimensions of the PFEs were measured using off-line measurement calipers on a Microsonic digitizer (Nova Microsonic). The position, attachment characteristics, movement and valvular consequences were evaluated. Clinical information was obtained from review of the patients' medical records. Neurologic events were attributed to the PFE if patients had

normal carotid arteries, a normal aorta, no concurrent atrial fibrillation and no significant valvular heart disease. Cardiac ischemic symptoms were attributed to the tumor if the patient had a negative coronary angiographic study ($\leq 50\%$ diameter stenosis of a coronary artery). Follow-up data were obtained for 53 patients (98%) from a mail survey or telephone interview of the patients. End points were clinical embolic events, myocardial infarction and valvular dysfunction at presentation and death at follow-up.

Statistical analysis. Results are summarized as the mean value \pm SD. Comparisons between selected variables in group 1 and group 2 were tested by calculating the Student *t* test; $p < 0.05$ was considered significant.

Results

The total study group (groups 1 and 2) consisted of 54 patients (27 men and 27 women, mean age 58.4 ± 14.7 years), comprising 0.019% of all patients who had an echocardiographic evaluation during the study period.

Echocardiographic characteristics of PFE. *Group 1.* The echocardiogram clearly showed PFE at the time of initial examination in 15 patients (88%). Analysis of the echocardiograms showed a reproducible spectrum of characteristics typical of PFEs.

ATTACHMENT. Three patients had multiple PFEs (total of 20 tumors). The PFEs on the atrioventricular valve most often (5 of 6 tumors) were on the ventricular surface of the valve. The PFEs on the semilunar (aortic) valve were equally distributed between the ventricular (3 of 6 tumors) and aortic (3 of 6 tumors) surfaces. In all cases, the PFE was attached to the endocardium. The tumors were commonly pedunculated, and a stalk attachment was identified echocardiographically in 17 (94%) of the 18 PFEs identified by echocardiography. All tumors demonstrated independent mobility.

Figure 1. Pathologic features of PFE of mitral valve. Gross specimen (left) shows multiple frond-like structures (photographed underwater), giving the appearance of a pom-pom or sea anemone. Low power (top right) and higher power views (bottom right) show microscopic features. At the core of each frond is dense elastin (black) coated by collagen (red-pink) and lined by flat endocardial cells. Elastic-van Gieson. Top right $\times 18$, bottom right $\times 54$, both reduced by 30%. (Courtesy of Dr. William D. Edwards, Division of Anatomic Pathology, Mayo Clinic.)

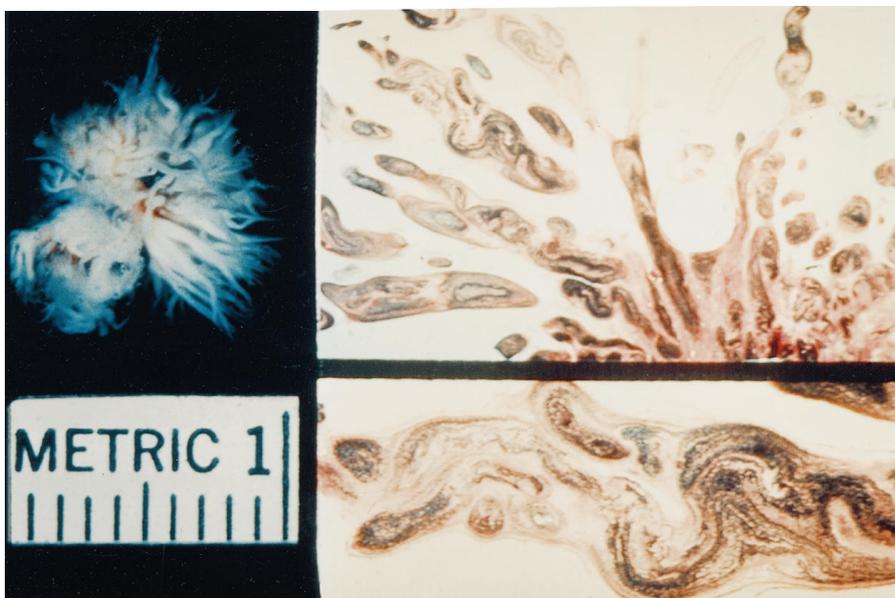


Table 1. Location of Papillary Fibroelastomas in Groups 1 and 2

Location	Group 1 [no. (%)]	Group 2 [no. (%)]
Aortic valve	6 (30)	17 (41.2)
Mitral valve*	4 (20)	16 (39.0)
Chordae	1	1
Papillary muscle	0	1
Tricuspid valve	2 (10)	4 (9.8)
Pulmonary valve	0	1 (2.4)
Septum/LVOT	6 (30)	3 (7.3)
Left ventricular free wall	1 (5)	0
Left atrium	1 (5)	0
Total no. of papillary fibroelastomas	20	41

*Papillary fibroelastomas associated with the papillary muscle or chordae were grouped with mitral valve tumors for formal counts. LVOT = left ventricular outflow tract.

LOCATION. Of the 20 PFEs in the 17 patients in group 1, 12 (60%) were on the valvular endocardium (6 on aortic valve, 4 on mitral valve and 2 on tricuspid valve; none were on the pulmonary valve) and 8 (40%) on the nonvalvular endocardium (6 on septum/left ventricular outflow tract, 1 on ventricular free wall and 1 on left atrium) (Table 1).

SIZE AND SHAPE. PFEs tended to be relatively small (mean size $12.1 \pm 6.5 \times 9.0 \pm 4.3$ mm in longest and widest dimension, respectively). The size ranged from 1 to 21 mm in length and 1 to 17 mm in width. The shape varied from ones with a well developed "head" (Fig. 2) to ones with elongated strand-like projections (Fig. 3).

TISSUE CHARACTERISTICS. PFEs appeared speckled with echolucencies, especially near the edges. A characteristic stippled edge, with shimmer or vibration at the tumor-blood interface, was seen and was especially notable in larger tumors as they moved in the blood flow. This was best appreciated on TEE at high frequency and with the high resolution (zoom) function, which often allowed individual finger-like projections to be distinguished. These finger-like projections are consistent with the fronds that are described pathologically and commonly referred to as "sea anemone" (Fig. 1).

ECHOCARDIOGRAPHICALLY FAILED DIAGNOSIS. The echocardiographic examination did not show PFE in two patients. One patient with the diagnosis of severe dilated cardiomyopathy had only TTE examination, and the images were not optimal. A small PFE was found in the left atrium in the explanted heart after transplantation. A retrospective review of the stored videotape did not reveal the PFE. The other patient had emergency TTE and TEE for dissection of the ascending aorta. The tumor was found on the excised aortic valve. Review of the stored videotape of this patient revealed a mass (0.6×0.6 mm) on the aortic valve, consistent with PFE.

Group 2. In comparison with group 1, the PFEs in group 2 were significantly ($p < 0.01$) smaller (mean $7.7 \pm 3.2 \times 5.1 \pm 3.9$ mm, range 1 to 21 in length and 1 to 17 in width). Four patients had two or more PFEs, for a total of 41 tumors. As in group 1, the most commonly affected valves were the aortic (17 [41.2%] of 41 tumors) and mitral valves (16 [39%] of 41

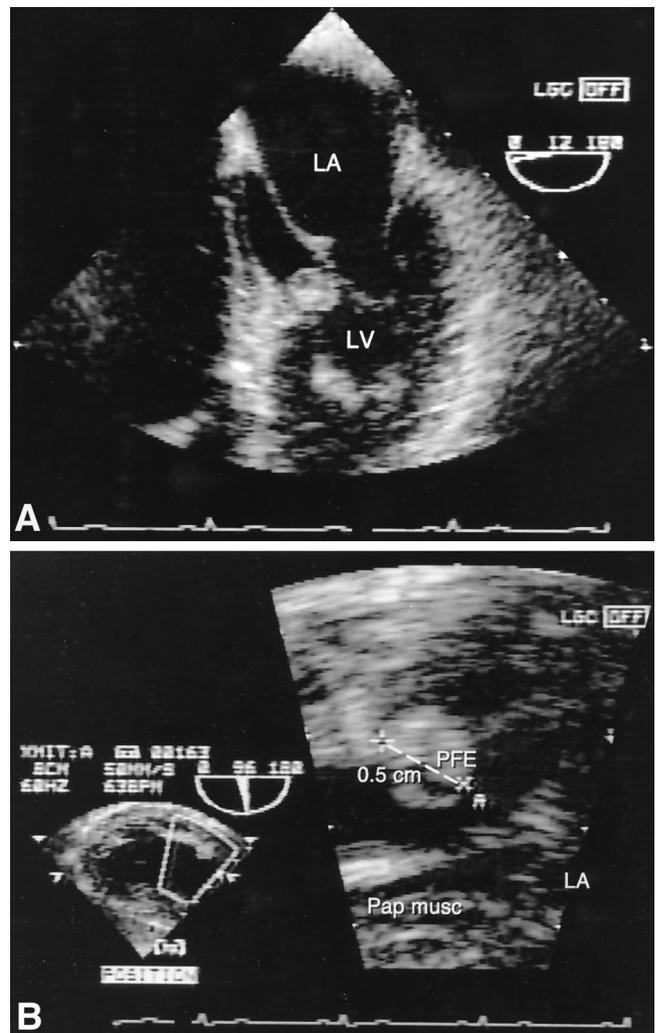


Figure 2. A, Echocardiogram showing a PFE on the ventricular septum. Note its rounded, bulky shape. The patient previously had patch repair of a ventriculoseptal defect. B, Zoom view of the same PFE. LA = left atrium; LV = left ventricle; Pap musc = papillary muscle.

tumors) (Table 1). The tricuspid valve was involved in four patients, the pulmonary valve in one and the septum/left ventricular outflow tract in three (Table 1).

Clinical Presentation. *Group 1.* Embolic events occurred in six patients in group 1 (35%). In five patients, there was no other potential source of embolism (Table 2). Three of the patients had a transient ischemic attack (TIA), and two had retinal emboli, with normal carotid arteries, no debris in the aortic arch on ultrasound examination and no evidence of atrial fibrillation. One patient treated with warfarin had recurrent TIAs.

Chest pain occurred in five patients (29%) and was attributed to PFE in two. Both had normal coronary arteries and negative ergotamine testing. The PFEs were on the aortic valve and left ventricular outflow tract. Three other patients with chest pain had significant coronary artery disease (CAD).

All other symptoms of patients in group 1 were attributed to other underlying diseases.

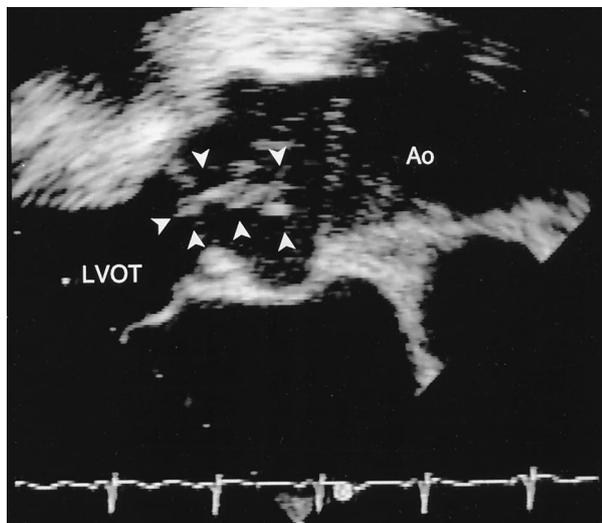


Figure 3. Zoom view of an aortic valve PFE (arrowheads), showing the linear morphology, with strands visible. Ao = aorta; LVOT = left ventricular outflow tract.

Group 2. The primary reason for referral for echocardiography was a neurologic event in five patients (13.5%). For all patients presenting with neurologic events, clinical screening tests revealed coexistent conditions capable of causing neurologic events: one patient had aortic arch disease, two had small vessel cerebrovascular disease, one had carotid stenosis and one had a patent foramen ovale with right to left shunting.

Papillary fibroelastoma was an incidental finding in 16 patients (43%). The indications for TTE or TEE are shown in Table 3.

Valvular dysfunction. Of the 54 patients in the study, seven (13%) had severe valvular disease, and in only one of these patients was the PFE on the dysfunctional valve. In no case was the PFE thought to be responsible for valvular dysfunction.

Associated endocardial abnormalities. In 10 patients (59%) in group 1, the PFE was in association with a previous endocardial injury: hypertrophic cardiomyopathy in three, aortic valve thickening/sclerosis in three, mitral valve prolapse in one, previous cardiac surgery (patch closure of ventricular

Table 2. Presenting Symptoms of Patients With Papillary Fibroelastoma

Symptom	Group 1	Group 2
Asymptomatic	1	16
TIA/CVA	4 (3*)	5
Peripheral emboli	2 (2*)	0
Chest pain	5 (2*)	3
Congestive heart failure	2	5
Dyspnea	3	4
Arrhythmia	0	1
Syncope/presyncope	0	3

*Symptoms thought to be due to papillary fibroelastoma. Data presented are number of patients. CVA = cerebrovascular accident; TIA = transient ischemic attack.

Table 3. Indications for Echocardiography

Indication	Group 1 (n = 17)	Group 2 (n = 37)
TIA/CVA	3	5
Congenital heart disease	1	1
Murmur	1	11
Ischemic heart disease	1	6
Prosthetic valve/intraop	3	2
Cardiomyopathy	2	4
Aortic disease	1	0
Chest pain	5	3
Study protocol	0	2
Precardioversion	0	3

Data presented are number of patients. intraop = intraoperative; other abbreviations as in Table 2.

septal defect) in one, rheumatic heart disease in one, and mantle radiation in one.

Diagnostic value of echocardiography. The present study was not designed to determine the sensitivity and specificity of echocardiography for the detection of PFE. Among pathologically confirmed cases, the sensitivity of echocardiography was 88% (15 of 17 patients).

Patient follow-up. **Group 1.** Mean follow-up was 34.2 months (range 0.2 to 179.9). All 17 patients underwent surgical removal of PFE. Indications for operation were removal of tumor in asymptomatic patients (n = 5) or in those with embolic phenomena (n = 5), and other cardiovascular operations were required (n = 7). Three perioperative deaths occurred in the last group <30 days postoperatively: dissecting aortic aneurysm in one patient, intractable arrhythmia after a myectomy in one patient with hypertrophic cardiomyopathy and low output failure in one patient with marked apical hypertrophic cardiomyopathy. No recurrent embolic events, chest pain or myocardial infarctions occurred during follow-up. The two patients with angina and normal coronary arteries remained free of chest pain.

Of 12 patients with valvular PFEs, one clinically required valve replacement of the mitral valve because of flail anterior leaflet. Among the 11 other patients, the PFE was successfully removed with native valve preservation in 10 (91%), and one patient required valve replacement (aortic valve).

Group 2. Mean follow-up was 30.7 months (range 1.1 to 78.5). During follow-up, nine new neurologic events (five cerebrovascular accidents [CVAs], three TIAs and one retinal embolism), one peripheral embolus (femoral) and one myocardial infarction occurred. Two patients had multiple events. One patient who was originally asymptomatic had a CVA with no other identifiable source of embolism. All the other patients had other disease processes capable of causing neurologic sequelae.

Three patients had cardiac surgery for other cardiovascular reasons, and the PFE was not identified. One patient was operated on (4 years and 8 months after diagnosis) at another institution to remove a 0.5 × 0.7 cm PFE on the posteromedial papillary muscle. Intraoperatively, the tumor could not be

identified. Intraoperative TEE was not performed. Another patient had a 1.3×0.8 cm septal PFE that had been confirmed on three separate echocardiographic examinations. Intraoperative TEE (2.5 months later) could not be used to identify the tumor. Lastly, a patient with severe stenosis of the aortic valve had valve replacement (23 months after diagnosis of PFE), and the 0.2×0.4 cm aortic valve PFE was not seen surgically; intraoperative TEE was not done.

Among the 16 patients who were asymptomatic at presentation (mean follow-up 2.9 ± 1.5 years), five developed symptoms. One had an embolic CVA not attributable to another cause that caused hemiparesis and aphasia 1 year after the discovery of the 2.5×1.5 -cm tumor on the posteromedial papillary muscle. In three patients with chest pain, two did not have significant CAD. Both of these patients had PFEs on the mitral valve. The other patient had progressive aortic stenosis and required an operation, but no PFE was found. Thus, 11 (69%) of 16 patients remained asymptomatic. One patient had a neurologic event possibly related to PFE.

In both groups 1 and 2, patients died of comorbid conditions, and no single cause of death could be attributed to the PFE. Long-term survival was similar between the two groups.

Discussion

To our knowledge, the present study included the largest series of patients with PFEs on echocardiography with pathologic correlation. This study demonstrated that PFEs 1) can be identified on the echocardiogram with a high degree of certainty; 2) are associated with embolic events in a high percentage of patients; 3) do not cause significant valvular dysfunction despite the tendency for being located on the valvular endocardium; 4) tend to occur in areas of endocardial damage (nine patients [53%] in group 1); and 5) can be removed, if valvular, with the usual salvage of the native valve, and after removal, there are no recurrent embolic events.

Echocardiographic characteristics. The most characteristic ultrasound features that identify a tumor as a PFE are small size (usually <1.5 cm); pedicel or stalk attachment to endocardium, with high mobility; and refractive appearance and, oftentimes, areas of echolucency within the tumor. The stippled edge and vibration or shimmer of the peripheral edge of the tumor are appreciated best under high resolution on TEE. These characteristics have not been observed previously in a large series of patients but are in agreement with the case descriptions in the published data (4,5,8,9,12,20-22,24-33).

TEE is helpful intraoperatively in guiding the operation and postoperatively in evaluating valve competency (4,11,25,34,35). In group 2, three patients had an operation, during which the PFE could not be identified. However, because of the retrospective nature of this study, it is unclear whether this was due to the time lapse between the diagnosis and cardiac surgery, technical limitations of the operations, embolization of the tumor, false positive diagnosis on the echocardiographic exam-

ination or lack of intraoperative TEE guidance at the time of operation ($n = 2$).

Differential diagnosis. The differential diagnosis of PFE includes myxoma (36), fenestrations (37) (when located on the aortic valve), strands (38), giant Lambl's excrescence (39,40), vegetation and thrombus and fibroma (1-3). In pathology series, primary cardiac tumors are uncommon, present in $<0.3\%$ of autopsies (2,3). Three-fourths of cardiac tumors are benign, and of the benign tumors, PFE is the third most common tumor (accounting for 7% to 8% of benign tumors), after myxoma and fibroma (2). Papillary fibroelastoma is the most common of the primary valvular cardiac tumors, accounting for 73% of these tumors (1). Echocardiographically, the distinction between PFE and Lambl's excrescence (39,40), strands (38) and fenestrations (37) is primarily size; of these, the PFE is largest (12.1×9.0 mm). Lambl's excrescences have a mean size of 8×1 mm (40). Histologically, PFEs and Lambl's excrescence are similar (39). Myxoma can usually be distinguished from PFE by location; myxoma is an atrial tumor (75% in the left atrium and 15% to 20% in the right atrium) (36,41). Fibromas are generally highly refractive ovoid masses that usually occur in children and young adults and typically involve the left ventricle, right ventricle and septum (41,42).

Location. PFEs have been reported on almost all endocardial surfaces, including the left ventricular septum (11,22,30,32,43-45), the papillary muscles (16,31,46), the chordae (9,14), the aortic root, the right ventricular outflow tract (21), the left ventricular mural endocardium (47), the right atrium (48), the atrial septum (49) and the Chiari network (50), with a preponderance in valvular endocardium. In the present study, most PFEs were on the valves (81.5%); however, a significant minority were on the nonvalvular endocardium (18.5%), including the ventricular septum, left atrium, papillary muscle and ventricular free wall.

Associated endocardial damage. In the present series, PFEs tended to arise in areas of endocardial irritation (58% in group 1). The associated cardiac diseases were degenerative (aortic valve sclerosis), mitral valve prolapse, previous operation for congenital ventriculoseptal defect repair, systolic anterior motion of the mitral valve and hypertrophic cardiomyopathy. This is consistent with the results of a review of the published data, in which several case reports have described PFEs in patients with underlying cardiac disease, such as rheumatic heart disease (29,31,32,44), hypertrophic cardiomyopathy (30,33) and mitral valve prolapse (47). Another patient in group 1 had mantle radiation therapy and a tricuspid valve PFE. However, it should be noted that 290,239 echocardiograms were performed during the study period and that potential endocardial injury is frequent; yet, PFE is rare.

Valvular dysfunction. Although PFEs are commonly valvular, they are rarely associated with valvular dysfunction (28,30,51,52). Of the seven patients in the present series with severe valvular disease, only one had a PFE located on the malfunctioning valve. (In contrast, case reports have suggested valve obstruction or regurgitation secondary to a PFE [28,30,51,52].)

Clinical presentation. Neurologic (4-19), embolic (6,27,53,54) and coronary ischemic symptoms (6,16,19,20,24-26,55-65) have been reported in association with PFEs. In one case, portions of a PFE were recovered in a coronary artery (1), suggesting that the tumor itself is friable and can embolize. Other investigators have identified platelet fibrin accumulations on a PFE, suggesting embolism (24,26,65,66), and others have postulated a ball valve phenomena of PFEs on the coronary ostia (57,64), thought to cause intermittent coronary obstruction, the mechanism of chest pain and sudden cardiac death.

Symptoms were common in both groups, with embolism being the most common symptom in group 1. In group 2, 13.5% of the patients had neurologic events and 8% had chest pain, but routine causes for these events and symptoms were identified.

Follow-up. Group 1 patients who survived cardiac surgery were symptom-free at follow-up. One patient required prosthetic valve placement after removal of the tumor, but 91% of the native valves were preserved. In both groups, mortality was not related directly to PFE but to comorbid conditions. In contrast to group 1, patients in group 2 had nine neurologic events, one femoral embolus and one myocardial infarction. One patient who was originally asymptomatic had a confirmed CVA 1 year after the PFE was identified, with no other source of embolism identified.

Study limitations. This study is retrospective and relies on the data base to identify patients. Although it is the largest series of its kind, it included a relatively small number of patients compared with the >290,000 patients studied during the period. Furthermore, the association with clinical events is based largely on indirect evidence.

Conclusions and clinical implications. PFEs are uncommon benign tumors of the endocardium, primarily valvular. However, they may occur on any endocardial surface, especially on endocardium irritated by concurrent cardiac disease. PFEs are associated with embolic events and may account for other cardiovascular symptoms, such as chest pain and syncope. They rarely cause valvular dysfunction. They can readily be recognized by their standard echocardiographic appearance, with an incremental value of TEE over TTE. The natural history still needs to be defined.

To date, conservative treatment for the patients in group 2 has not led to increased mortality, but may be associated with greater morbidity. There is strong circumstantial evidence from group 1 patients that the presence of a PFE is associated with neurologic events, possibly with cardiac ischemia. As shown in group 1, successful surgical resection can be accomplished with frequent preservation of the native valve. Management issues remain difficult because of the rarity of PFEs, and clinical decisions must be individualized, but the options include surgical removal of the tumor, anticoagulation with warfarin, antiplatelet drugs or a combination. Further study is needed.

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