CARDIAC VALVE PAPILLARY FIBROELASTOMA: SURGICAL EXCISION FOR REVEALED OR POTENTIAL EMBOLIZATION

Jean-Michel Grinda, MD
Jean Paul Couetil, MD
Sylvain Chauvaud, MD
Nicola D'Attellis, MD
Alain Berrebi, MD
Jean-Noel Fabiani, MD
Alain Deloche, MD
Alain Carpentier, MD, PhD

Objective: We have reviewed the case histories of 4 patients who underwent operations between September 1994 and November 1997 at Broussais Hospital for cardiac valvular papillary fibroelastoma. **Methods:** Diagnosis was strongly suggested by echocardiography. Tumor locations were mitral (1), tricuspid (1), and aortic (2). Indications for operation were previous stroke for the mitral tumor, prophylaxis for the tricuspid tumor, syncopal episodes for the first aortic tumor, and transient ischemic attack and mesenteric ischemia for the second aortic tumor. **Results:** Surgical excision with a conservative, valve-sparing approach was performed in all cases. For the first aortic tumor, aortic valve reconstruction was achieved with part of a cryopreserved aortic homograft cusp. Intraoperative transesophageal echocardiography showed no evidence of valvular regurgitation after excision in all cases. All patients had uneventful postoperative recoveries. No evidence of regurgitation or recurrence was seen on echocardiography at follow-up. **Conclusions:** Despite their histologically benign aspect, cardiac papillary fibroelastomas should be excised because of potential embolic complications. A conservative, valve-sparing approach is recommended, however, because of the absence of recurrence after total excision. (J Thorac Cardiovasc Surg 1999;117:106-10)

Cardiac papillary fibroelastomas (CPFEs) are rare benign tumors that can have important clinical implications. CPFEs are characteristically small and solitary, with multiple papillary fronds resembling a sea anemone, and they are most often found on the valvular endocardium.^{1,2} Before the availability of echocardiography, CPFEs were usually found incidental to autopsy or cardiac operations.¹⁻³ The first surgical excision was reported by Lichtenstein and associates⁴ in 1976. A literature review found 198 cases of CPFE. Most were described as incidental autopsy findings. Surgical excision was performed in 71 cases. The 4 cases described in this report demonstrate the complications of CPFE and the operative findings. Although these lesions are benign, early recognition and treatment are fundamental because they can cause fatal complications.

From the Department of Cardiovascular Surgery of Broussais Hospital [1], Paris, France.

Received for publication June 29, 1998; accepted for publication Aug 12, 1998.

Address for reprints: Jean-Michel Grinda, MD, Département of Cardiovascular Surgery, Broussais Hospital, 96 rue Didot, 75014 Paris, France.

Copyright © 1999 by Mosby, Inc.

0022-5223/99 \$8.00 + 0 12/193896

Clinical summaries

PATIENT 1. A 33-year-old man was referred in September 1994 for the treatment of a mitral valve tumor, which was diagnosed after the occurrence of a stroke. In July 1994 the patient had had 2 successive cerebrovascular accidents. The arising sequelae were oculomotor paralysis and aphasia. The patient's previous medical history was otherwise uneventful. At cardiac examination the patient was in sinus rhythm. The carotid and peripheral pulses were normal. Results of biologic, electrocardiographic, chest radiographic, and carotid duplex ultrasonographic studies were all within normal limits. A cerebral computed tomographic scan showed an infarct within the distribution of the left superficial sylvian artery. Angiography revealed occlusion of the distal branches of the sylvian artery, suggesting an embolic cause. Transthoracic echocardiography (TTE) revealed a mobile mass on the mitral valve. Further evaluation with transesophageal echocardiography (TEE) revealed a tumor attached to the anterior leaflet and measuring approximately 1 cm.

An operation was performed on September 2, 1994. Surgical findings consisted of a solitary verrucous tumor arising from the atrial side of the anterior mitral leaflet (Fig 1). The tumor was excised, and the valve was repaired with simple interrupted suture of the valvular edges. Histopathologic examination confirmed the diagnosis of a CPFE with an attached thrombus. The patient had an uneventful postoperative recovery. At 3 years of follow-up there was good neurologic recovery and TTE showed no valvular abnormalities.

Fig 1. Operative view of the mitral CPFE arising from the atrial side of the anterior mitral leaflet.

PATIENT 2. A 65-year-old man was referred in November 1994 for coronary artery bypass grafting. His previous medical history was significant for an anterior myocardial infarction in October 1994. The patient underwent coronary artery bypass grafting (4 grafts) and was discharged from the hospital on postoperative day 9. During his convalescence, TTE revealed a mass on the tricuspid valve. TEE demonstrated a solitary tumor attached to the atrial aspect of the tricuspid valve and measuring approximately 2 cm, without abnormalities of the other valves (Fig 2). Results of pulmonary scintigraphy were within normal limits.

The patient was reoperated on in July 1995. Surgical findings consisted of a CPFE (confirmed by histopathologic examination) attached to the atrial side of the septal tricuspid leaflet. The tumor was excised, and the tricuspid valve repair was achieved with simple sutures at the edge of the resection. The patient's postoperative course was uneventful. TTE at 3 years of follow-up showed no valvular abnormalities.

PATIENT 3. A 62-year-old man was referred in September 1995 for a tumor of the aortic valve. The patient had an uneventful previous medical history except for a hiatal hernia. He had been previously admitted to his referring hospital because of an unexplained loss of consciousness while walking on August 28, 1995. At admission, results of clinical examination, electrocardiography, and chest radiography were all unremarkable, but TTE and TEE detected a mobile echodense mass (15×15 mm) arising from the aortic valve. Differentiation of this mass as a tumor, a thrombus, or vegetation was not possible before the operation; however, there was no evidence of endocarditis and multiple blood cultures yielded negative results. This solitary tumor was attached to the right aortic cusp by a short pedicle. The tumor was mobile on the aortic surface of the cusp, with a tendency toward prolapse into the right coronary ostium, suggesting the possibility of an intermittent right coronary ostial obstruction. No other evidence of valve pathology was found.

The operation was performed on September 20, 1995. Surgical findings consisted of a CPFE (confirmed by histopathologic examination) attached near the free edge of the right aortic cusp and mobile within the right coronary sinus of

Fig 2. Preoperative TEE view of the CPFE adherent to the atrial side of the tricuspid leaflet.

Valsalva. The tumor and a third of the right aortic cusp were removed. Restoration of normal right cusp architecture was achieved with part of a cryopreserved aortic homograft cusp. Intraoperative TEE showed no evidence of valvular regurgitation. The patient's postoperative course was uneventful. TTE at 3 years of follow-up showed no valvular abnormalities.

PATIENT 4. A 55-year-old woman was referred in November 1997 for an aortic valve tumor. The patient's previous medical history was notable for a transient ischemic attack in April 1997. Results of medical examination, coagulation tests, electrocardiography, cerebral computed tomographic scan, and TTE were normal. In June 1997 the patient had an acute mesenteric infarction, necessitating a small-bowel resection followed by anticoagulant treatment. Histologic analysis of the resected bowel did not contribute to a precise diagnosis but did suggest an embolic cause. Results of medical examination, laboratory test, electrocardiography, chest radiography, and abdominal echocardiography were normal. TTE did not reveal any valvular abnormalities; however, TEE showed a mobile mass (<1 cm) attached to the noncoronary cusp of the aortic valve (Fig 3).

The patient underwent surgical resection in November 1997. The intraoperative finding consisted of a CPFE (confirmed by histologic examination) attached to the free edge of the aortic valve cusp facing the noncoronary sinus. Simple excision of the tumor was performed. Intraoperative TTE showed no aortic valve regurgitation. The patient's postoperative course was uneventful.

Discussion

CPFEs are classified as primary benign endocardial tumors.^{1,2} Nomenclature can be confusing, because many names have been used to describe this tumor.¹⁻³ *Papillary fibroelastoma* is the most widely accepted term. Primary cardiac neoplasms are rare, with an inci-

Fig 3. Preoperative TEE view of the CPFE adherent to the noncoronary cusp of the aortic valve.

Fig 4. Histologic section of a resected CPFE. The papillary fronds are lined by endothelial cells. The matrix consists of mucopolysaccharide, elastic fibers, and rare spindle cells. (Hematoxylin and eosin stain; original magnification $\times 25$.)

dence at autopsy ranging between 0.002% and 0.33%.^{1,5} Eighty percent of these primary cardiac neoplasms are benign tumors, with CPFE as the third most common type, after myxoma and lipoma but before heman-gioma.^{1,5} In 1971 Abu Nassar and Parker² reported on 39 cases in a literature review of CPFE, and McAllister and Fenoglio¹ collected 42 cases from the Armed Forces Institute of Pathology database in 1977. To date 198 cases of CPFE have been reported in the literature.

CPFEs are small, avascular, benign tumors. They are derived from the normal components of endocardium (fibrous tissue, elastic fibers, and smooth muscle cells). Characteristically they have multiple papillary fronds, resembling a sea anemone, and usually are attached to the endocardium by a short pedicle.^{1,3} Histologically

Fig 5. Locations of 173 CPFEs.

the papillary fronds consist of a central core of dense connective tissue surrounded by a layer of loose connective tissue and covered by hyperplastic endothelial cells (Fig 4).¹⁻⁴

CPFEs can arise from virtually any endocardial surface, but they most often originate from the valvular endocardium.^{1,3,4} Eighty-four percent of the tumors in the 198 collected cases of CPFE ($n = 171$) arose from cardiac valves. CPFE is by far the most common tumor arising from cardiac valves. Eighty-eight percent of the 132 valvular tumors and 71% of the 56 valvular tumors studied by Ryan and associates⁶ and by Edwards and associates,⁷ respectively, were CPFEs. CPFE affect each of the 4 cardiac valves with nearly equal frequencies.^{1,3} Precise data concerning locations of the tumor were available for 173 patients in the literature (Fig 5). Among these, the aortic valve was the site of involvement in 51 cases (29%), the mitral valve was the site in 43 cases (25%), the tricuspid valve was the site in 29 cases (17%), and the pulmonary valve was the site in 23 cases (13%, Fig 1). CPFEs arising from semilunar valves are located with nearly equal frequencies on the ventricular and arterial sides of the valve and may be situated anywhere on the valve.¹⁻³ In the case of the atrioventricular valves, the tumor usually arises on the middle portion of the valve and generally projects into the atrium.¹⁻³ Nonvalvular locations, observed in 15.7% of the 198 collected cases, include left and right ventricular mural and septal endocardial walls,^{1-5,8-10} right and left atrial endocardium,^{1,2,11} papillary muscle,^{2,12} chordae tendinae,^{2,12} and intima of the right coronary ostium.¹³ In anatomic series, CPFEs occur with equal frequency on the left and right sides of the heart,^{1,3,5,7} but 81% of the CPFEs reportedly surgically excised were located on the left side. In fact, CPFEs are usually solitary tumors, and multiple CPFEs have been found in only few instances (7.5% of the collected cases).^{1,2,6,7,12,14,15}

Generally a CPFE is a small lesion, ranging in size from 0.1 to 5.7 cm and mostly measuring less than 1.5 cm.³ Most cases of CPFE have been encountered in

adults, usually in patients in the sixth decade or beyond (ranging from a 6-day-old neonate to a 92-year-old woman).³ They occur equally in male and female patients.³ Initially described as an incidental finding at autopsy (63% of the collected cases) or surgery, the clinical significance of CPFEs has been debated at length.² The evidence that CPFEs can cause cardiac or neurologic symptoms is now well established. Among the 71 patients whose operations for CPFE removal are reported on in the literature, neurologic accidents were the most common symptoms (48%, Table I). CPFEs have a significant propensity toward embolization, causing transient ischemic attack, stroke, angina, myocardial infarction, pulmonary embolism, congestive heart failure, near syncope, and sudden death.^{3,16-30} Embolization may occur from fragments of the papillary fronds of the tumor itself or from a thrombus that frequently forms around the tumor. In fact, the tumor can be a nidus for platelet and fibrin aggregates that subsequently embolize.^{3,16} Because results of physical examination are generally normal in patients with CPFEs, 2-dimensional echocardiography is necessary in young patients with transient ischemic attacks or strokes, even in the absence of clinical or electrocardiographic evidence of heart disease. Furthermore, it is of interest that CPFE may also cause obstructive cardiac symptoms,^{3,11,13} being responsible of left or right ventricular outflow tract obstruction and dynamic ostial coronary obstruction, as described for patient 3 in our series.

During diagnostic workup of patients with suspected CPFE, these tumors have been detected as filling defects on cardioangiographic studies.³ Even in an 83-year-old woman who could not be operated on, a diagnosis was established by percutaneous transfemoral biopsy of a right atrial tumor seen on angiography.¹¹ At present conventional TTE and TEE with multiplanar studies are the best means in establishing the diagnosis.^{3,12,14,21,31-33} The high resolution of TEE can locate the central collagenous core of the tumor and distinguish it from the rest of the mass by its bright echocardiographic appearance.^{25,26} If clearly identified, this central echodense contrast should allow differentiation of CPFEs from other intracardiac tumors, especially myxomas, but also from vegetations or mural thrombi.^{3,15}

The potential for life-threatening complications of CPFE is an indication for their surgical excision. It is recognized that even small CPFEs (3 mm) can be responsible for strokes,³ and we therefore consider that these tumors, even in patients without symptoms, should be surgically managed as long as there are no major contraindications to the operation. At diagnosis,

Table I. Seventy-one surgical cases of CPFE: Clinical symptoms encountered

Symptoms	Aortic valve (n = 15)	Mitral valve (n = 33)	Tricuspid valve (n = 10)	Non-valvular location (n = 13)
Angina or MI (n = 7)	4	2	—	1
Stroke or TIA (n = 36)	7	25	1	3
Syncope (n = 4)	1	—	3	—
CHF (n = 6)	1	3	—	2
Pulmonary embolism (n = 2)	—	—	2	—
Systemic embolization (n = 1)	1	—	—	—
Incidental finding (n = 17)	3	3	4	7

MI, Myocardial infarction; TIA, transient ischemic attack; CHF, congestive heart failure.

prophylactic anticoagulation should be initiated. Intraoperative TEE can be a useful adjunct in eliminating multiple tumor locations and in assessing the adequacy of valve repair.^{15,26} Intracardiac videothoracoscopic techniques have been reported in the treatment of left ventricular tumors.¹⁰ A conservative surgical approach is recommended and is generally possible (86% of the 71 surgical procedures of the collected cases), with standard valvular repair techniques used after tumor resection. Nevertheless, we used part of a cryopreserved aortic homograft cusp to achieve restoration of normal right cusp architecture in our third patient. This excellent tissue provided a better repair than an autologous pericardial patch would have done. Recurrence after surgical excision is as yet unknown, and the first 3 patients reported on here have no evidence of recurrence at 3 years of follow-up.

Despite their benign pathologic aspect, CPFEs should be surgically managed, even in patients without symptoms. A conservative, valve-sparing approach is generally possible, and the use of homograft tissues can be of benefit. Recurrence after surgical excision is as yet unknown; however, careful follow-up is warranted.

REFERENCES

- McAllister HA, Fenoglio JJ. Tumors of the cardiovascular system. In: Atlas of tumor pathology. 2nd series. Washington DC: Armed Forces Institute of Pathology; 1978. Fascicle 15. p. 20-5.
- Abu Nassar SG, Parker JC Jr. Incidental papillary endocardial tumor: its potential significance. Arch Pathol 1971;92:370-6.
- Shahian DM, Labib SB, Chang G. Cardiac papillary fibroelastoma. Ann Thorac Surg 1995;59:538-41.
- Lichtenstein HL, Lee JC, Stewart S. Papillary tumor of the heart: incidental finding at surgery. Hum Pathol 1979;10:473-5.

5. Chitwood WR Jr. Cardiac neoplasm: current diagnosis, pathology, and therapy. *J Cardiac Surg* 1988;3:119-54.
6. Ryan PE Jr, Obeid AL, Parker FB Jr. Primary cardiac valve tumors. *J Heart Valve Dis* 1995;4:222-6.
7. Edwards FH, Hale D, Cohen A, Thompson L, Pezzella AT, Virmani R. Primary cardiac valve tumors. *Ann Thorac Surg* 1991;52:1127-31.
8. Almagro UA, Perry LS, Choi H, Pintar K. Papillary fibroelastoma of the heart: report of six cases. *Arch Pathol Lab Med* 1982;106:318-21.
9. Schuetz WH, Welz A, Heymer B. A symptomatic papillary fibroelastoma of the left ventricle removed with the aid of transesophageal echocardiography. *Thorac Cardiovasc Surg* 1993;41:258-60.
10. Espada R, Talwalker NG, Wilcox G, Kleiman NS, Verani MS. Visualization of ventricular fibroelastoma with a video-assisted thoracoscope. *Ann Thorac Surg* 1997;63:221-3.
11. Schwinger ME, Katz E, Rotterdam H, Slater J, Weiss EC, Kronzon I. Right atrial papillary fibroelastoma: diagnosis by transthoracic and transesophageal echocardiography and percutaneous transvenous biopsy. *Am Heart J* 1989;118:1047-50.
12. Lund GK, Schroder S, Koschyk DH, Nienaber CA. Echocardiographic diagnosis of papillary fibroelastoma of the mitral and tricuspid apparatus. *Clin Cardiol* 1997;20:175-7.
13. Boone S, Higginson LA, Walley VM. Endothelial papillary fibroelastomas arising in and around the aortic sinus, filling the ostium of the right coronary artery. *Arch Pathol Lab Med* 1992;116:135-7.
14. Li Mandri G, Homma S, Di Tullio MR, et al. Detection of multiple papillary fibroelastomas of the tricuspid valve by transesophageal echocardiography. *J Am Soc Echocardiogr* 1994;7:315-7.
15. Lee KS, Topol EJ, Stewart WJ. Atypical presentation of papillary fibroelastoma mimicking multiple vegetations in suspected bacterial endocarditis. *Am Heart J* 1993;125:1443-5.
16. Mc Fadden PM, Lacy JR. Intracardiac papillary fibroelastoma: an occult cause of embolic neurologic deficit. *Ann Thorac Surg* 1987;43:667-9.
17. Mann J, Parker DJ. Papillary fibroelastoma of the mitral valve: a rare cause of transient neurological deficits. *Br Heart J* 1994;71:6.
18. Zamora RL, Adelberg DA, Berger AS, Huettner P, Kaplan HJ. Branch retinal artery occlusion caused by a mitral valve papillary fibroelastoma. *Am J Ophthalmol* 1995;132:5-9.
19. Muir KW, McNeish I, Grosset DG, Metcalfe M. Visualization of cardiac emboli from mitral valve papillary fibroelastoma. *Stroke* 1996;27:1133-4.
20. Colucci V, Alberti A, Bonacina E, Gordini V. Papillary fibroelastoma of the mitral valve: a rare cause of embolic events. *Texas Heart Inst J* 1995;22:327-31.
21. Brown RD Jr, Khandheria BK, Edwards WD. Cardiac papillary fibroelastoma: a treatable cause of transient ischemic attack and ischemic stroke detected by transesophageal echocardiography. *Mayo Clin Proc* 1995;70:863-8.
22. de Menezes IC, Fragata J, Martins FM. Papillary fibroelastoma of the mitral valve in a 3-year-old child: case report. *Pediatr Cardiol* 1996;17:194-5.
23. Meyer AH, Clements SD Jr, Sadel SM, Shanewise JS, Gravanis MB. Papillary fibroelastoma of the cardiac valves. *Heart Dis Stroke* 1994;3:318-9.
24. Zull DN, Diamond M, Beringer D. Angina and sudden death resulting from papillary fibroelastoma of the aortic valve. *Ann Emerg Med* 1985;14:470-3.
25. Etienne Y, Jobic Y, Houel JF, et al. Papillary fibroelastoma of the aortic valve with myocardial infarction: echocardiographic diagnosis and surgical excision. *Am Heart J* 1994;127:443-5.
26. Grote J, Mügge A, Schäfers HJ, Daniel WG, Lichtlen PR. Multiplane transesophageal echocardiography detection of a papillary fibroelastoma of the aortic valve causing myocardial infarction. *Eur Heart J* 1995;16:426-9.
27. Fine G, Pai SR. Cardiac papillary fibroelastoma: a source of coronary artery emboli and myocardial infarction. *Henry Ford Hosp Med J* 1984;32:204-8.
28. Eckstein FS, Schäfers HJ, Grote J, Mügge A, Borst HG. Papillary fibroelastoma of the aortic valve presenting with myocardial infarction. *Ann Thorac Surg* 1995;60:206-8.
29. Ganjoo AK, Johnson WD, Gordon RT, Jain DP, Lang GE, Shankar VS. Tricuspid papillary fibroelastoma causing syncopal episodes. *J Thoracic Cardiovasc Surg* 1996;112:551-3.
30. Waltenberger J, Thelin S. Images in cardiovascular medicine: papillary fibroelastoma as an unusual source of repeated pulmonary embolism. *Circulation* 1994;89:2433.
31. Watanabe T, Hosoda Y, Kikuchi N, Kawai S. Papillary fibroelastoma of the tricuspid valve in association with atrial septal defect: report of a case. *Surg Today* 1996;26:831-3.
32. Lee CC, Celik C, Lajos TZ. Excision of papillary fibroelastoma arising from the septal leaflet of the tricuspid valve. *J Cardiac Surg* 1995;10:589-91.
33. Gopal A, Li Mandri G, King DL, Marboe C, Homma S. Aortic valve papillary fibroelastoma: a diagnosis by transthoracic echocardiography. *Chest* 1994;105:1885-7.