Double Primary Left Ventricular and Aortic Valve Papillary Fibroelastoma

Hiroyuki Tanaka, MD; Takashi Narisawa, MD; Takanobu Mori, MD;
Yoshiro Masuda, MD; Daijiro Kishi, MD

Papillary fibroelastomas of the heart are relatively rare benign tumors and although they can be symptom free, symptoms such as cerebral ischemia and cardiac infarction can be lethal. It is important to diagnose this tumor using echocardiography, because lethal embolisms can be prevented by surgical resection. Recurrence of this tumor has not been reported and multiple tumors are rare. The present report is a case of double primary aortic valve fibroelastoma with aortic regurgitation diagnosed 4 years after surgery for a left ventricular fibroelastoma with mitral valvular disease. (*Circ J* 2004; 68: 504–506)

Key Words: Aortic valve; Echocardiography; Left ventricle; Papillary fibroelastoma

Papillary fibroelastomas, which are generally found on the valvular endocardium, are relatively uncommon benign tumors and the next most common of the primary cardiac tumors after myxomas and lipomas.^{1,2} However, they can cause lethal embolisms resulting in myocardial infarction, cerebral infarction and pulmonary embolism.² The number of patients diagnosed with papillary fibroelastomas has increased since the advent of echocardiography, but cases of recurrence have not been reported and multiple occurrence is rare.^{3–11} We present a case of double papillary fibroelastoma of the aortic valve with aortic regurgitation and mitral periprosthetic leakage occurring 4 years after resection of an initial left ventricular papillary fibroelastoma with mitral and tricuspid valvular disease.

Case Report

In 1997, a 52-year-old man presented to hospital with dyspnea and a medical history that included atrial fibrillation in 1993. Cardiac catheterization revealed mild pulmonary hypertension of 45/22 mmHg, elevated wedge pressure of 22 mmHg, and obstruction of the posterior descending artery. Transthoracic echocardiography confirmed the presence of a left ventricular tumor (Fig 1A), and revealed moderate mitral stenosis and regurgitation, tricuspid regurgitation and trivial aortic regurgitation. Mitral valve replacement (29 mm Carbomedics Valve), tricuspid annuloplasty (32 mm Carpentier Ring) and tumor excision without blood transfusion were performed. The tumor, found on the anterior wall of the left ventricle near the apex of the heart, was pale yellow and 7 mm in maximum length. It was excised through the mitral valve (Fig 2A). Pathology showed papillary growth with myxoid stroma and the

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Department of Thoracic Cardiovascular Surgery, Showa University Fujigaoka Hospital, Yokohama, Japan

Mailing address: Hiroyuki Tanaka, MD, Department of Thoracic Cardiovascular Surgery, Showa University Fujigaoka Hospital, 1-30 Fujigaoka, Aoba-ku, Yokohama 227-8501, Japan. E-mail: h.tanaka@showa-university-fujigaoka.gr.jp

Fig 1. (A) Transthoracic echocardiography before the first operation shows a friable mass (arrow) attached to the left ventricular wall near the apex. LV, left ventricle; RV, right ventricle. (B) Transesophageal echocardiography before the second operation shows 2 friable masses (arrow) attached to the aortic valve.

Fig 2. (A) Left ventricular tumor at first operation. (B-1) Aortic valve tumor at the second operation. (B-2) One tumor originates from the commissure of the right coronary leaflet.

Fig 3. (A) Left ventricular tumor showing myxoid matrix in the papillary projection and the tumor surface covered by endocardial cells ($\times 10$; hematoxylin and eosin). (B) Aortic valve tumor showing almost the same findings, but there is less elastic fiber compared with the original left ventricular tumor ($\times 10$; hematoxylin and eosin).

surface of the tumor was covered by endocardial cells, findings consistent with a diagnosis of papillary fibroelastoma (Fig 3A). The mitral valve showed evidence of rheumatic disease. The patient returned to normal life on warfarin therapy.

In 2001, the patient was readmitted because of exertional dyspnea. A transesophageal echocardiograph disclosed aortic regurgitation, 2 mobile gelatinous masses, which appeared vegetative, at the aortic valve (Fig 1B), and mitral periprosthetic leakage. Infectious endocarditis was suspected and he underwent a second operation in August 2001. The aortic valve leaflet was shortened, thickened and calcified, and there was a 3 mm hole in the right coronary cusp. Two, soft, rose-pink, friable masses approximately 11 mm in size, were identified on the commissure of the right and non-coronary cusps, and were removed together with the aortic valve (Fig 2B). The aortic valve was replaced by a 19-mm St Jude Medical HP valve and the 5 mm mitral periprosthetic leakage on the anterior side of

the commissure was closed from the left atrial trans-aortic left ventricular (LV) approach. The operation was completed without blood transfusion. Pathologic examination of both lesions confirmed papillary fibroelastomas; however, there was relatively little elastic fiber in these tumors compared with the previous LV tumor (Fig 3B). The aortic valve showed rheumatic change. The postoperative course was uneventful, and the patient has been free of cardiovascular or cerebral events since the second operation.

Discussion

Papillary fibroelastomas are rare benign cardiac tumors of which there have been fewer than 100 cases reported worldwide.¹ They are the third most common of the primary cardiac tumors after myxomas and lipomas, and account for 7% of the total cases.² They can occur anywhere in the heart, but approximately 80% occur on the valvular endocardium² and can be associated with mechanical valve

thrombus³ and infection⁴ According to Howard et al, 55% of these tumors are found on the mitral or aortic valves¹ they are usually singular, and only 7 cases of multiple tumors have been published in the literature.^{5–11} The etiology of these tumors is unknown, but reaction to mechanical trauma, neoplasms, hamartomas or inflammatory nodules has been considered.^{2,12} In the present case, the papillary fibroelastoma occurred in the LV cavity in association with mitral regurgitation, and the second tumors were on the aortic valve and were associated with aortic regurgitation. As these tumors occurred with the retrograde turbulent blood flow, they may have developed from the proliferation of endocardial tissue in response to shear stresses.² Because of their very soft, friable characteristics, often associated with adherent thrombus, they tend to embolize;² therefore, the common presenting symptoms are embolism to brain and heart resulting in transient ischemic attack or stroke, and myocardial infarction. It is the location of the tumor rather than its size that causes the symptoms and outcome. In order to prevent these lethal embolisms, identification of the tumor (using transthoracic and transesophageal echocardiography) and its surgical removal are very important.^{6,10} Recurrence has not previously been reported; in the present case another tumor occurred in the heart 4 years after the first was excised, and although the second tumors had slightly different characteristics and location we cannot rule out recurrence. This may be the first case of multifocal, double primary papillary fibroelastoma.

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