Double Primary Left Ventricular and Aortic Valve Papillary Fibroelastoma

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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

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...
Papillary fibroelastoma of LV and Aortic Valve

The 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. They are the third most common of the primary cardiac tumors after myxomas and lipomas, and account for 7% of the total cases. They can occur anywhere in the heart, but approximately 80% occur on the valvular endocardium and can be associated with mechanical valve replacement.
thrombus and infection. According to Howard et al, 55% of these tumors are found on the mitral or aortic valves; they are usually singular, and only 7 cases of multiple tumors have been published in the literature. The etiology of these tumors is unknown, but reaction to mechanical trauma, neoplasms, hamartomas or inflammatory nodules has been considered. 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. Because of their very soft, friable characteristics, often associated with adherent thrombus, they tend to embolize; 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. 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.

References