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Slowly Growing Cardiac Tumor : A Case of Fibroelastoma

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Abstract

Echocardiographic follow-up for 16 years in an asymptomatic patient with mitral stenosis showed very slow growth of a mass attached to the mitral valve. The tumor doubling time was estimated to be 3.6 years. Surgical excision of the mass was performed when the patient eventually developed dyspnea on exertion, and histopathological examination revealed papillary fibroelastoma. Echocardiographic follow-up and anti-coagulation may be sufficient treatment for asymptomatic patients with papillary fibroelastoma.

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Key Words

■ **Neoplasm（fibroelastoma）** ■ **Valvular disease** ■ **Echocardiography**
■ **Cardiac surgery**

INTRODUCTION

Papillary fibroelastoma accounts for 8% of benign cardiac tumors in the text book of Armed Forces Institute of Pathology Series and is usually found incidentally at surgery, autopsy, or routine echocardiography[1, 2]. Here, we report a case of this tumor that showed very slow growth.

CASE REPORT

A 46-year-old woman was referred to our hospital 16 years before the current admission for evaluation of rheumatic heart disease. She had mitral stenosis, aortic stenosis, aortic regurgitation, and atrial fibrillation. The mitral valve area was 1.9 cm^2 on two-dimensional（2D）echocardiograms in 1988, at the age of 37（**Fig. 1−left**）. As her valvular heart disease was not severe enough to produce any symptoms, she was not willing to undergo surgery. Serial 2D echocardiography demonstrated a solitary ventricular mass that arose from the chordae tendineae of the mitral valve. The mass showed gradual enlargement over the subsequent 16-year period. **Fig. 1−right** was taken in 1994, at the age of 43.

On admission in 1997, she had mild dyspnea on exertion. A diastolic rumble was heard at the apex, and a grade III/VI systolic ejection murmur followed by a grade III/VI diastolic blowing murmur was heard along the left sternal border. Neither pulmonary congestion nor peripheral edema was noted. Neurological examination showed no abnormal findings. The 2D echocardiograms obtained on admission are shown in **Fig. 2**. A mass of 28 mm diameter was attached to the chordae tendineae and papillary muscles（**Figs. 2−a, b**）. A smaller mass was present on the interventricular septum, and another mass was noted on the posterior mitral leaflet（**Figs. 2−c, d**）. The mitral valve area was 1.4 cm^2 on 2D echocardiograms and 1.1 cm^2 on Doppler echocardiograms. There was severe aortic stenosis with a Doppler pressure gradient of

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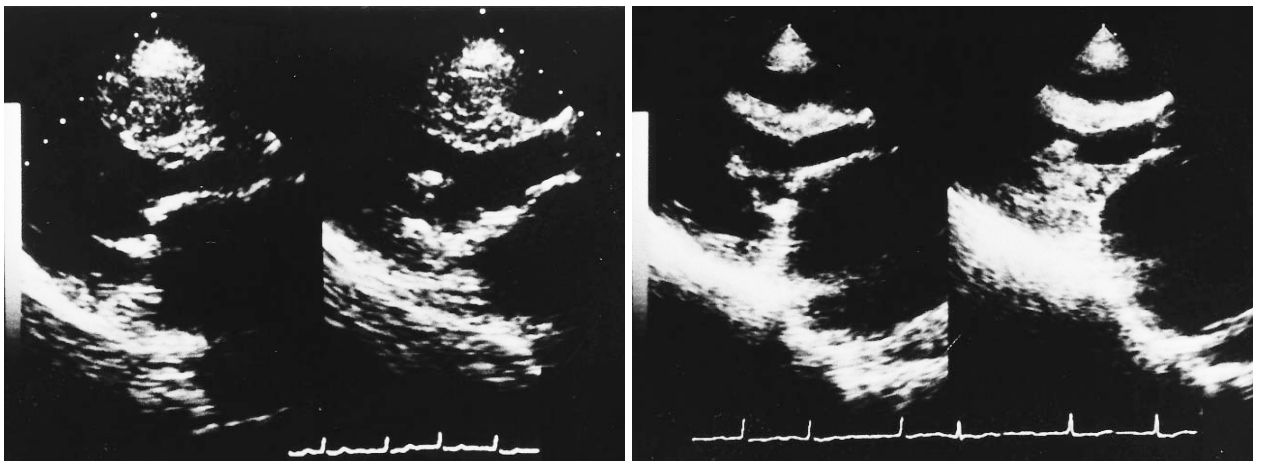

Fig. 1 Two-dimensional echocardiograms
The mass attached to the mitral valve increased in size between Oct. 24, 1988(*left*)and Oct. 5, 1994(*right*).

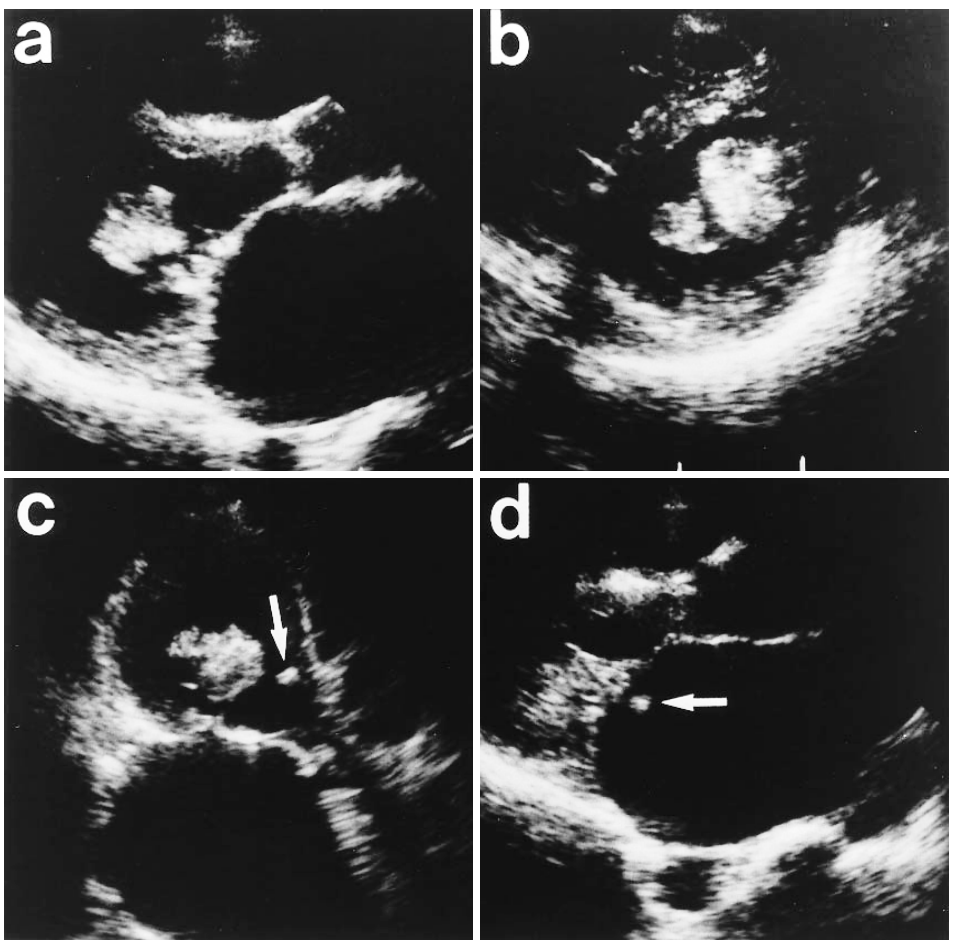

Fig. 2 Two-dimensional echocardiograms
A round mass is attached to the papillary muscles and the chordae tendineae of the mitral valve in the long-axis view(*a*)and the short-axis view(*b*). Additional masses can be seen on the interventricular septum(*c*; *arrow*)and on the posterior mitral valve leaflet(*d*; *arrow*).

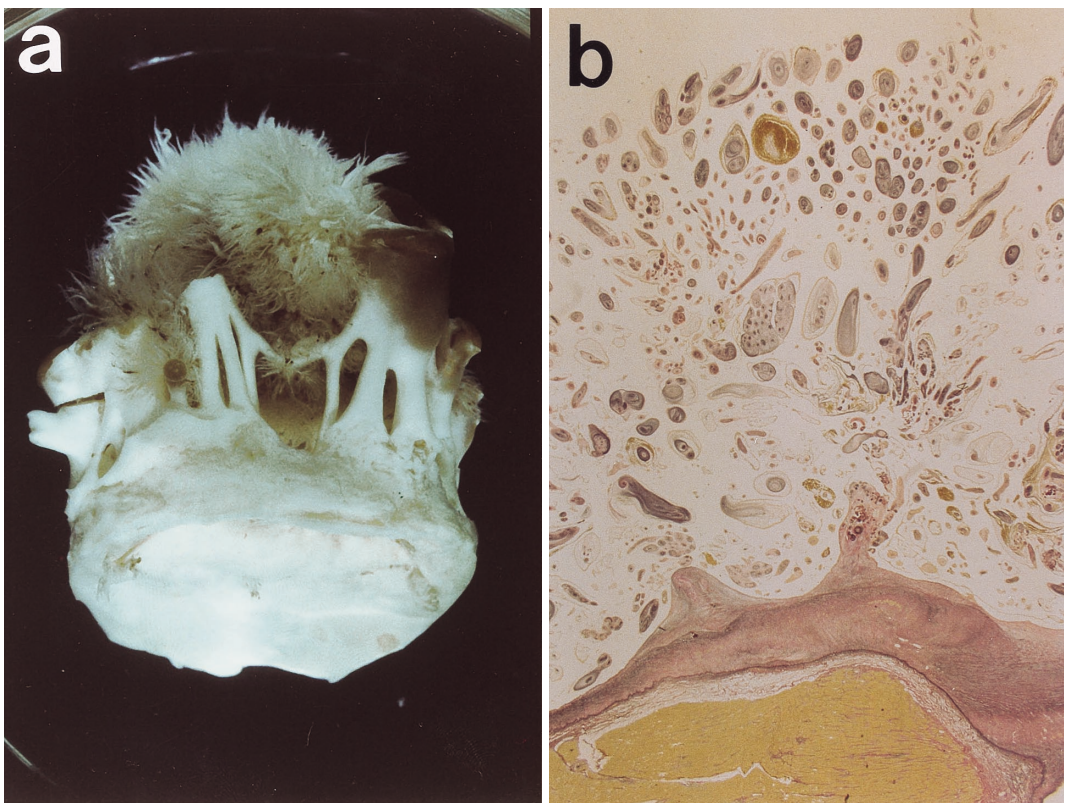

Fig. 3 Photograph of the gross appearance of the main tumor after removal (*a*) and high-power photomicrograph of a portion of the tumor (*b*; elastica van Gieson stain, × 400)
Note the linings of endocardial cells and the central core of elastic fibers.

83 mmHg, as well as moderate tricuspid regurgitation. No abnormalities of the left ventricular wall motion were observed. Left ventricular dimensions were within normal limits on M-mode echocardiography (left ventricular end-diastolic diameter 53 mm, left ventricular end-systolic diameter 38 mm, interventricular septal thickness 12 mm, and posterior wall thickness 12 mm). The fractional shortening was 28%.

Surgical excision of these masses revealed soft tumors covered with gelatinous substance and numerous thrombi. The main mass measured 30 × 25 × 15 mm and had developed between the anterior and posterior papillary muscles. After being placed in saline, the tumor showed a distinctive cluster of hair-like projections resembling a "sea anemone" (**Fig. 3−a**). Microscopic examination demonstrated papillary fibroelastoma characterized by a lining of endocardial cells and a central core of elastic fibers (**Fig. 3−b**). Both mitral and aortic valve replacement was performed using the St. Jude medical valve together with annuloplasty of the tricuspid valve. The patient has shown no tumor recurrence in the 2 years since discharge.

DISCUSSION

We retrospectively analyzed the echocardiographic findings of the main tumor. The maximum diameter was measured in the parasternal long-axis view on serial 2D echocardiograms that were recorded on videotape. The tumor grew very slowly from 13 to 28 mm diameter over the 16-year period before surgery (**Fig. 4**). The tumor doubling time was estimated to be 3.6 years using the method of Schwartz[3].

Operative excision rarely appears to be warranted when papillary fibroelastomas are detected by echocardiography and whether operative excision or valve replacement is beneficial for papillary fibroelastomas in patients with cerebral or myocardial ischemic events is unsettled[4]. During the 16-year follow-up of our patient, acute myocardial infarction or stroke did not occur and the tumor grew very slowly. Therefore, echocardiographic

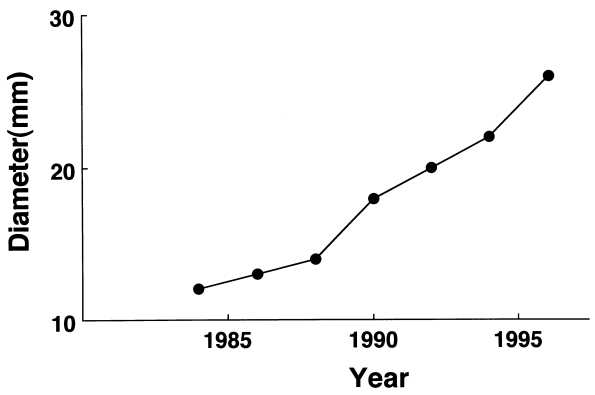

Fig. 4 Maximum diameter of the main tumor over the 16-year follow-up (from 1982 to 1997)

follow-up and anticoagulation might be sufficient treatment for asymptomatic patients with slowly growing tumors suspected to be papillary fibroelastomas. However, earlier reports demonstrated an association between this tumor and sudden death due to embolism[5−7]. Cardiac tumor is sometimes difficult to accurately identify without surgery, so early resection should be performed if the patient has any symptoms.

要　　約

長い時間をかけ徐々に増大した線維弾性腫の1例

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無症状の僧帽弁狭窄患者の心エコー図において，僧帽弁に付着する腫瘤が16年間に非常にゆっくりと増大するのが観察された．腫瘤の倍加時間は3.6年であった．患者に労作時の呼吸困難が出現した後に，腫瘤切除と弁置換術が行われた．組織病理学的所見から乳頭状線維弾性腫と診断された．この腫瘍の無症状の患者に対する治療としては，抗凝固療法と心エコー図法の経過観察の方針でよいと考えられた．

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