

A Report of a Surgical Case of Papillary Fibroelastoma in the Left Ventricular Outflow Tract

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While a 74-year-old man with a past history of cerebral infarction was undergoing surface echocardiography due to transient atrial fibrillation, an unstable, pedunculated tumor was detected in the left ventricular outflow tract. As the results of transesophageal echocardiography suggested a left ventricular tumor, semi-emergency surgery was performed. Based on pathological findings, the tumor was diagnosed as papillary fibroelastoma. This tumor mainly affects the cardiac valves and is often discovered during open-heart surgery or autopsy. This report presents a very rare case of preoperatively identified papillary fibroelastoma in the left ventricular outflow tract. (Ann Thorac Cardiovasc Surg 2003; 9: 270–3)

Key words: cardiac surgery, papillary fibroelastoma, left ventricular tumor, transesophageal echocardiography

Introduction

Papillary fibroelastoma is a rare primary cardiac tumor usually affecting the cardiac valves. The introduction and widespread use of echocardiography has led to this tumor being more frequently reported; nevertheless, it is often discovered incidentally during open heart surgery or autopsy. Since papillary fibroelastoma is associated with such severe embolism-related complications, it is generally removed surgically. Along with a brief review of the literature, we herein present the case of a patient who underwent surgery after a pedunculated tumor in the left ventricular outflow tract was identified by echocardiography.

Case Report

The patient was a 74-year-old man with incomplete pa-

ralysis of the left upper arm due to a cerebral infarction that occurred in 1999. In 2001, the patient was commenced on medication for transient atrial fibrillation. On April 15, 2002, the patient experienced chest discomfort and was admitted to the internal medicine department at another hospital with a diagnosis of transient atrial fibrillation. As a left ventricular tumor was suspected on echocardiography, the patient was admitted to the First Department of Internal Medicine at Fukushima Medical University Hospital on April 18.

Physical examination on admission revealed no abnormalities of blood pressure, heart sounds, respiratory sounds or neurological findings; height was 160 cm, weight 58 kg. In terms of laboratory findings on admission, general hematological and biochemical tests showed no abnormalities, and inflammatory markers, such as C-reactive protein (CRP) and erythrocyte sedimentation rate, were normal. Moreover, results of coagulation and fibrinolytic screens were within normal limits. Electrocardiography showed sinus rhythm, heart rate of 72 beats per minute, with no ST-T changes evident. Chest X-rays demonstrated a cardiothoracic ratio (CTR) of 52% and no abnormal shadows. Head computed tomographic (CT) scans confirmed signs indicative of an old cerebral infarction in the right frontal lobe. Surface echocardiography identified an 8×6 mm unstable tumor in the left ventricu-

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Received February 13, 2003; accepted for publication March 18, 2003.

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Fig. 1. Transesophageal echocardiography showed a pedunculated tumor (white arrow) attached to the free wall of the left ventricular outflow tract. The tumor had an uneven internal structure and a bright tumor center and a papillary structure (AO, aorta; LV, left ventricle).

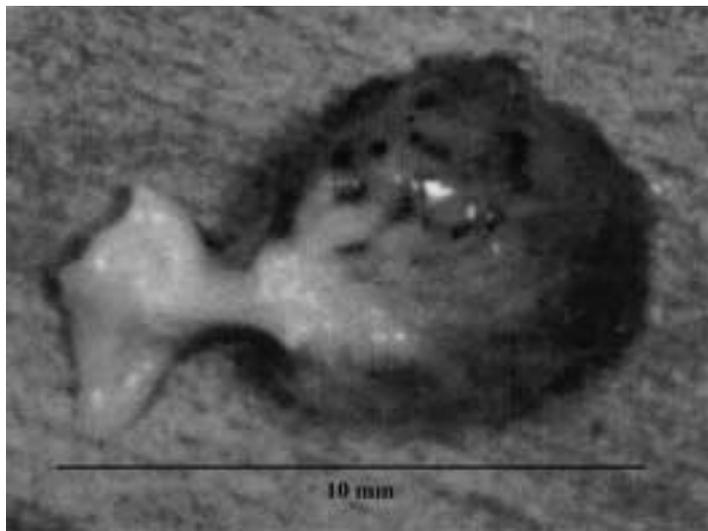


Fig. 2. The resected tumor showed a 6.7×6.5×4.8 mm jelly-like appearance with a 1 mm peduncle.

lar outflow tract, while transesophageal echocardiography showed a pedunculated tumor attached to the free wall of the left ventricle, 18 mm from the aortic valve annulus. The tumor had an uneven internal structure and measured 9.1×5.9 mm in size (Fig. 1). Although the tumor was movable and unstable, it was solitary, and no abnormality was seen in the cardiac valves. A cardiac catheterization procedure showed no significant coronary constriction or feeding vessels to the tumor. Furthermore, the tumor could not be visualized by pulmonary arteriography.

Based on the above findings, the mass was diagnosed as a left ventricular tumor, and the patient was referred to our department to undergo surgery. On May 2, after a median sternotomy, extracorporeal circulation was initiated between the ascending aorta and the right atrium. Under cardiac arrest, an incision was made in the ascend-

ing aorta in order to examine the aortic valve, but no abnormality was seen, and when the left ventricle was examined through the aortic valve, a 6.7×6.5×4.8 mm jelly-like tumor with a 1 mm peduncle (Fig. 2) was observed on the left ventricular wall, 15 mm from the right coronary cusp. Using a scalpel, the tumor was removed together with the left ventricular endocardium by wedge resection. As pathological analysis of intraoperative frozen section confirmed papillary fibroelastoma, the chest was closed and the operation was completed. Extracorporeal circulation was maintained for 40 minutes and the duration of aortic clamping was 25 minutes.

Figure 3 shows the pathological findings for the resected tumor. The tumor exhibited a papillary structure, and elastic fibers were found at the periphery of the papillary peduncle, surrounding central collagen fibers. Con-

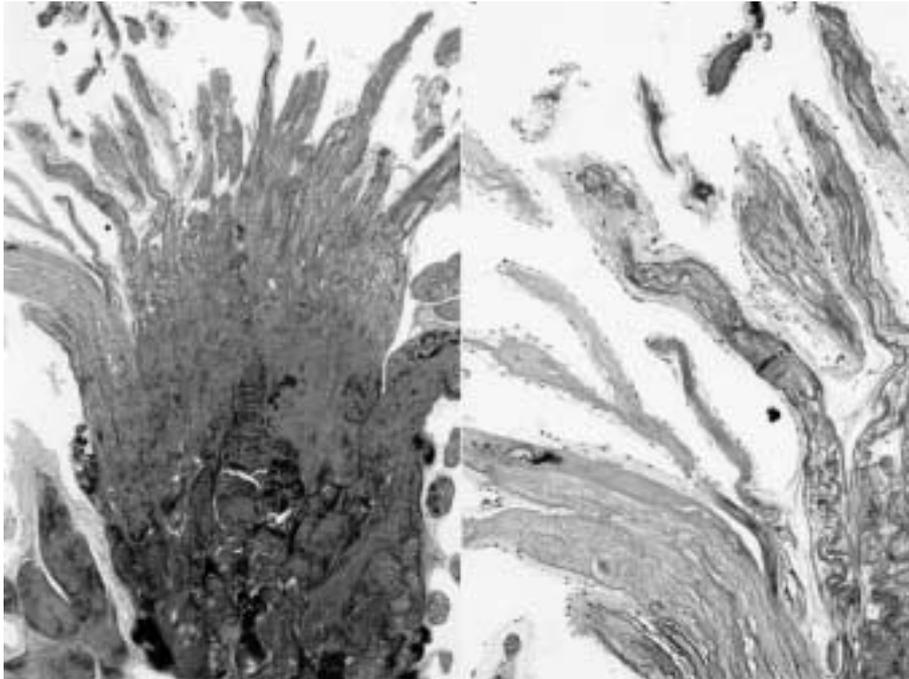


Fig. 3. The pathological findings for the resected tumor showed a papillary structure (left: Elastica Masson $\times 2$ magnification) and elastic fibers found at the periphery of the papillary peduncle which was coated with a layer of endothelial cells, surrounding central collagen fibers (right: Elastica Masson $\times 4$ magnification).

sequently, the tumor was diagnosed as papillary fibroelastoma.

Discussion

Papillary fibroelastoma accounts for about 8% of all cardiac tumors, and is the third most common benign cardiac tumor after myxoma and lipoma.¹⁾ However, as the incidence of primary cardiac tumors is low at 0.002-0.33%,^{1,2)} papillary fibroelastoma is correspondingly very rare. Since papillary fibroelastoma affects the cardiac valves in 84% of cases and 88% of all cardiac valve tumors are papillary fibroelastoma,^{3,4)} this tumor can be seen to be closely related to the cardiac valves. Furthermore, although the largest reported case measured 57 mm, papillary fibroelastomas are generally smaller than 10 mm,⁵⁾ and because these tumors are often asymptomatic, they are frequently discovered during open heart surgery or autopsy. However, in recent years, due to the introduction and widespread use of echocardiography, papillary fibroelastoma is often discovered during investigation of other heart conditions, and the number of reports is on the rise.^{3,4)}

In the present patient, although papillary fibroelastoma was discovered unexpectedly during investigation of sudden atrial fibrillation, it could not be preoperatively differentiated from myxoma, infectious or verrucous endocarditis or clot. Furthermore, the tumor was located in

the left ventricular outflow tract, and according to AFIP studies^{1,6)} involving 76 tumors in 72 patients, papillary fibroelastoma affecting tissues other than the cardiac valves is rare, with this series describing only two left ventricular tumors (2.6%). To the best of our knowledge, there have been a total of 31 reported cases of surgically treated papillary fibroelastoma in Japan, but of these, nine patients had left ventricular tumors (29%) and five patients had left ventricular outflow tract tumors (16.1%).⁷⁻¹¹⁾ Hence, the incidence of these tumors was greater when compared to overseas studies. In the present patient, transesophageal echocardiography identified a short peduncle⁶⁾ and a bright tumor center,^{5,12)} which are considered relatively characteristic findings, and since a papillary structure¹³⁾ was observed, papillary fibroelastoma was strongly suspected. As reported in the past,^{3,12)} transesophageal echocardiography was the most useful imaging technique.

Although papillary fibroelastoma is benign, it is known to induce such severe complications as cerebral and myocardial infarction, regardless of its size.^{5,10)} The cause of such severe complications may be that a fragile tumor itself could break off and act as an embolus or that a fibrin clot forming on the surface of the papillary tissue could embolize. When a tumor develops in the left atrium or the left ventricle, papillary fibroelastoma is likely to facilitate fibrin and platelet coagulation, thus leading to an embolism.^{4,14)} According to a report published by

Grinda and colleagues,³⁾ of 71 patients who underwent surgery for this condition, complications that developed were cerebral infarction (n=36), myocardial infarction (n=7), syncope (n=4), chronic heart failure (n=6), and pulmonary infarction (n=2). The tumor was discovered unexpectedly in 17 of these patients. In another report,¹⁵⁾ about half of the patients had cerebral complications. As a result, papillary fibroelastoma should be surgically removed regardless of its size when it forms in the left side of the heart due to the increased risk of complications. The present patient also had a history of cerebral infarction, and although the cause of the infarction could not be identified, since the tumor could have been implicated, semi-emergency surgery was performed. In terms of prognosis, there have been no reports of recurrence following simple tumor excision,³⁾ but it would be prudent to follow up the present patient carefully.

References

1. McAllister HA Jr, Fenoglio JJ. Tumors of the cardiovascular system. In: Atlas of Tumor Pathology, 2nd series, Armed Forces Institute of Pathology, Washington DC, 1978; Fascicle 15: pp 20–5.
2. Chitwood WR Jr. Cardiac neoplasm: current diagnosis, pathology, and therapy. *J Card Surg* 1988; **3**: 119–54.
3. Grinda JM, Chauvaud S, D'Attellis N, et al. Cardiac valve papillary fibroelastoma: surgical excision for revealed or potential embolization. *J Thorac Cardiovasc Surg* 1999; **117**: 106–10.
4. Ryan PE Jr, Oberid AL, Parker FB Jr. Primary cardiac valve tumors. *J Heart Valve Dis* 1995; **4**: 222–6.
5. Shahian DM, Labib SB, Chang G. Cardiac papillary fibroelastoma. *Ann Thorac Surg* 1995; **59**: 538–41.
6. Burke A, Virmani R. Tumors of the heart and great vessels. In: Atlas of Tumor Pathology, 3rd series, Armed Forces Institute of Pathology, Washington DC, 1995; Fascicle 16: pp 47–54.
7. Shigenobu M, Kurosumi K, Imai S, et al. Experience of surgical treatment for a case of cardiac fibroelastoma. *Geka* 1989; **51**: 315–7. (in Japanese)
8. Kawada M, Oda K, Hirose K, et al. A case report of surgical treatment for left ventricular papillary fibroelastoma. *Nippon Kyobu Geka Gakkai Zasshi* 1996; **44**: 1159–62. (in Japanese)
9. Minatoya K, Okabayashi H, Yokota T, Hoover EL. Cardiac papillary fibroelastomas: rationale for excision. *Ann Thorac Surg* 1996; **62**: 1519–21.
10. Taniyasu N, Akiyama K, Iba Y, et al. Papillary fibroelastoma in association with thrombosis on a mechanical valve. *Jpn Circ J* 2000; **64**: 797–9.
11. Uchida S, Obayashi N, Yamanari H, et al. Papillary fibroelastoma in the left ventricular outflow tract. *Heart Vessels* 1992; **7**: 164–7.
12. 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.
13. Ong LS, Nanda NC, Barold SS. Two-dimensional echocardiographic detection and diagnostic features of left ventricular papillary fibroelastoma. *Am Heart J* 1982; **103**: 917–8.
14. Topol EJ, Biern RO, Reitz BA. Cardiac papillary fibroelastoma and stroke. Echocardiographic diagnosis and guide to excision. *Am J Med* 1986; **80**: 129–32.
15. Colucci V, Alberti A, Bonacia E, et al. Papillary fibroelastoma of the mitral valve. A rare cause of embolic events. *Tex Heart Inst J* 1995; **22**: 327–31.