

A Case Report of Papillary Fibroelastoma Attached to Chorda Tendineae of Mitral Valve

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The case was a Japanese man of 62 years old. A bulbar mass attached to mitral valve anterior leaflet was discovered in an echocardiography within detailed examination of ischemic heart disease accidentally. We diagnosed him as ischemic heart disease and doubt of heart tumor. We thought about the danger of future embolism, surgical treatment was decided. We dissected the tumor together with one chorda tendineae of mitral valve, and a performed aorta - coronary bypass surgery. We diagnosed the tumor as papillary fibroelastoma by pathological diagnosis.

Papillary fibroelastoma is extremely rare with 7-9% of benign tumor of heart primary. Most of the papillary fibroelastoma is incidentally discovered by echocardiography or autopsy. Or it is discovered for systemic embolism. The tumor is benign, but there is a problem to cause embolism. Therefore, when we discovered papillary fibroelastoma, surgical resection of the tumor is the first-line therapy. A problem on surgical therapy is the range of resection area. Papillary fibroelastoma is benign tumor, but the pathological characteristic is still unidentified. Further epidemiological and pathological studies are necessary to determine the extent of surgical excision in associated with characteristics of papillary fibroelastoma.

Key words: papillary fibroelastoma, cardiac benign tumor, mitral valve

INTRODUCTION

Papillary fibroelastoma is benign tumor of extremely rare heart primary. It is often discovered in an echocardiography and autopsy accidentally. Or it is discovered by systemic embolism such as cerebral infarction, myocardial infarction, pulmonary embolism. We experienced a case of papillary fibroelastoma accidentally discovered during workup of ischemic heart disease. This tumor occurred from chorda tendineae connected to anterior cusp of mitral valve and was removed it surgically by an operation. Because it is a rare case, we add examination by documents and present it.

CASE REPORT

The case was a 62 years old Japanese man. The chief complaint was on exertional anterior chest pain. This patient was noted hypertension by health examination in 2000, but did not take treatment. When he did a laborious work since July, 2003, anterior chest pain developed and had a checkup in Tokai University Hachioji Hospital in August, 2003. His medical history was hypertension and alcoholic liver injury. He smoked 20/day for 40 years. By physical examination, the blood pressure was diastolic hypertension in 142/98 mmHg. There was no cardiac murmur by auscultation. There were no abnormal abdominal, neurological findings. By his blood examination, only mild hyperlipemia was present. Inflammation makers were negative in results of leukocyte count 5,600/mm³ CRP 0.19 mg/dl,

too. The pathogenic fungi was negative by blood culture. There are no abnormal findings in chest X-rays.

We did treadmill stress electrocardiogram to him for detailed examination of on exertion anterior chest pain. Because he appealed for lower extremities defatigation, in Modified-Bruce protocol Stage 3, the test was stopped. Negative T wave developed in II, III, aVf leads in electrocardiography, and ST depression developed in V4-6 leads. We suspected angina pectoris from this test result and performed coronary angiography. There was 75% stenosis in right coronary artery #4PD (Fig. 1).

We more performed an echocardiography for heart function test. There was a bulbar mass of 14 × 12 mm attached at mitral valve anterior leaflet (Fig. 2). The mass was seen to stick to chorda tendineae of mitral valve by transesophageal echocardiography (Fig. 3). Wall motion of left ventricle was normal, and there were not the findings of mitral valvular regurgitation.

By computed tomography, we could find the intracardiac tumor, but there was no sign of systemic embolism.

He did not show pyrexia, and inflammatory reaction and blood culture were negative. There was not the medical record which suspected infectious endocarditis. In addition, there was not any findings to suspected malignant tumors of other organs. We therefore diagnosed it as heart primary tumor and angina pectoris.

If his coronary lesion is isolated, it is indication of

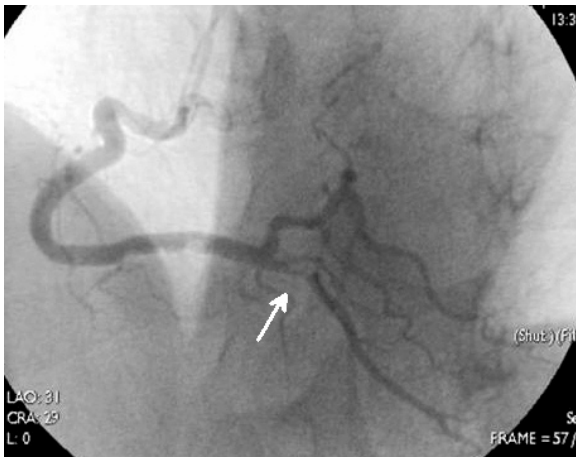


Fig. 1 By coronary angiography, there was a significant stenosis of 75% in right coronary artery #4PD. (a white arrow)

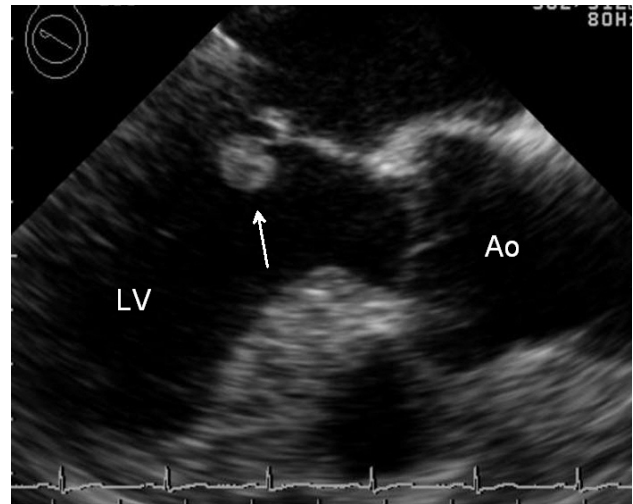


Fig. 3 Transesophageal echocardiography demonstrated the mass was seen to stick to chorda tendineae of mitral valve. (a white arrow)
Ao: Ascending aorta, LV: Left ventricle

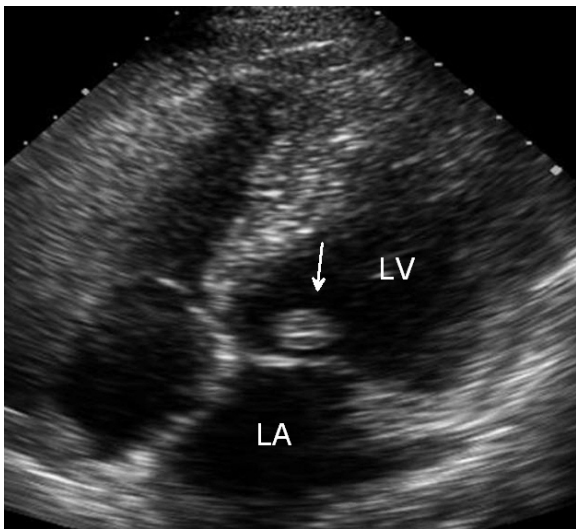


Fig. 2 Echocardiography (four chamber image) demonstrated a bulbar mass of 14 × 12 mm attached at mitral valve anterior leaflet. (a white arrow)
LA: Left atrium, LV: Left ventricle



Fig. 4 Macroscopic findings of resected the tumor together with one chorda tendineae of mitral valve. The tumor was soft in color tone of milk yellow with size of 12 mm. The tumor did not have the adhesion of thrombus.

a catheter treatment. He does not have medical history of embolism till now. However, we thought about the danger of future embolism, surgical treatment was decided. In addition, we administered antiplatelet drug to prevent thrombus adhesion to a tumor.

We performed an operation on October 9, 2003. We did median sternotomy and approached a heart. We used a cardio-pulmonary bypass device and we went via atrial septum and approached at mitral valve. There was 12 mm great opalescent tumor to stick to chorda tendineae of mitral valve anterior leaflet. We dissected the tumor together with one chorda tendineae of mitral valve. The tumor was soft in color tone of milk yellow, and did not have the adhesion of thrombus (Fig. 4). There were no malignant findings by quick pathology examination of a tumor. The mitral valve cusp did not have the abnormal findings. We removed one mitral valvular chorda tendineae surgically, but the mitral val-

ular regurgitation did not occur. We performed aorta - coronary bypass surgery (aorta - right coronary artery #4PD) by using a great saphenous vein graft.

Postoperative course was fine. There was not a recurrence of tumor and mitral valvular regurgitation of a tumor by echocardiography for postoperative 24 months.

Pathologic findings of the tumor were as follows:

In hematoxylin and eosin staining, the tumor was papillary configuration. The center was elastic fiber, which was surrounded by mucous layer with covered by mono-layer cells (Fig. 5). CD 31, 34 CD staining demonstrated the cells were endothelial cell. The tumor did not have any blood vessel (Fig. 6). By alcian blue staining and digestion processing by hyaluronidase, we confirmed deposition of acid mucopolysaccharid in mucous blanket of a tumor (Fig. 7).

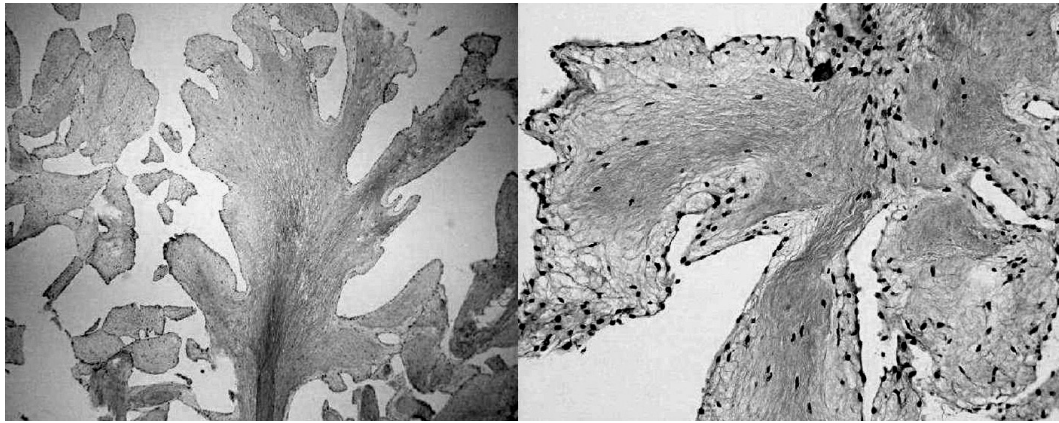


Fig. 5 Hematoxylin and eosin staining of the tumor. The tumor was papillary configuration, and center was elastic fiber, which was surrounded by mucous layer with covered by mono-layer cells.

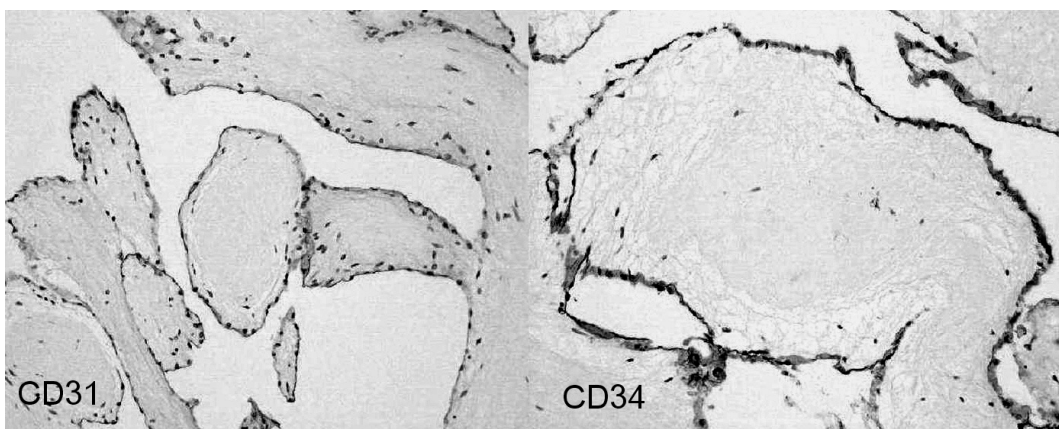


Fig. 6 CD 31, 34 CD staining of the tumor. The lateral stratum of the tumor was built with mono-layer cells, and they were demonstrated that the cells were endothelial cell in those stainings. The tumor did not have any blood vessel.

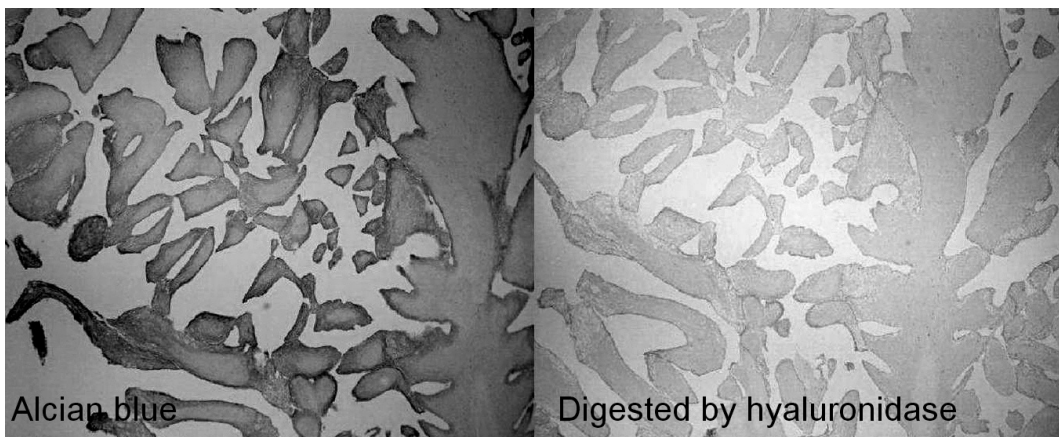


Fig. 7 By alcian blue staining and digestion processing by hyaluronidase of tumor. They confirmed deposition of acid mucopolysaccharide in mucous blanket of a tumor.

We therefore diagnosed it as papillary fibroelastoma.

DISCUSSION

A primary tumor of the heart is rare, ranging from 0.002% to 0.028% among autopsy cases [1]. The 70% of them are benign tumors. Papillary fibroelastoma is extremely rare with 7-9% of benign primary tumor of the heart. It usually derives from aortic or mitral valve

[2]. It sometimes derives from any endocardial surface of heart including left ventricle or atrial septum [3-6]. There is also a report of a multiple tumors [7]. There is no report of the risk factor for papillary fibroelastoma development [8]. Most of the papillary fibroelastoma is incidentally discovered by echocardiography or autopsy. It is sometimes discovered by systemic embolism such as cerebral infarction, myocardial infarction, pulmo-

nary embolism. There was a case of papillary fibroelastoma which was discovered during medical work up for palpitation and examination general malaise. And the symptom disappeared after tumor excision [6]. However, in that case, the tumor did not have papillary structure. The relationship between the pathologic findings and the symptom was not clear.

The tumor is benign, but there is a problem to cause embolism. Embolism may be caused by either thrombus on the tumor or tumor itself [9]. Anticoagulant therapy by heparin administration is recommended for thrombosis prevention, but its efficacy has not been established. There is also a case report of repeated cerebral infarction ever under anticoagulant therapy by Warfarin [10]. Therefore, when we discovered heart tumor even if it is benign tumor, surgical resection of the tumor is the first-line therapy.

A problem on surgical therapy is the range of resection area. Papillary fibroelastoma is benign tumor, but the pathological characteristic is still unidentified. If we restrict the resecting area, we may keep the native valve function. It was reported that native valve function was maintained at 86% of the cases who had papillary fibroelastoma at their cardiac valve [2].

Further epidemiological and pathological studies are necessary to determine the extent of surgical excision in associated with characteristics of papillary fibroelastoma.

CONCLUSION

We experienced a case of papillary fibroelastoma which attached to chorda tendineae of mitral valve who happened to be discovered during medical work up

for angina pectoris. Surgical excision of the tumor was recommended because of danger of embolism. Further pathological studies are necessary to define appropriate range of surgical resection of the tumor.

REFERENCES

- 1) McAllister, HA, Jr and Fenoglio, JJ, Jr: Tumors of the cardiovascular system. Atlas of Tumor Pathology, AFIP, Washington, DC, 1978, pp 20-25.
- 2) Grinda, J M, Couetil, J P, Chauvaud, S *et al.*: Cardiac valve papillary fibroelastoma : Surgical excision for revealed or potential embolization. J. Thorac. Cardiovasc. Surg. 117: 106-110, 1999.
- 3) Yoichi S, Hitoshi Y, Hirono S, *et al.*: A report of Surgical Case of Papillary Fibroelastoma in Left Ventricular Outflow Tract. Ann Thorac Cardiovasc surg 9 : 270-273, 2003.
- 4) Jeffery W Schoondyke, Jonathan W Burrress, Bahaeddin Shabaneh, *et al.*: Papillary Fibroelastoma Involving the Left Ventricular Wall. Reviews IN CARDIOVASCULAR MEDICINE 4 : 184-187, 2003.
- 5) Ramesh M Gowda, Ijaz A Khan, Nirav J Mehta., *et al.*: Cardiac papillary fibroelastoma Originating from Pulmonary Vein Angiology 53 : 745-748, 2002.
- 6) Yoko M, Tatsumi S, Kazuhiro H, *et al.*: Left Ventricular Papillary Fibroelastoma Associated with Chief Complaints of Palpitation, General Fatigue, and Feverish. Kyobugeka 56: 793-796, 2003.
- 7) Fatima Eslami-Varzaneh, Erwin A Burn, Pamela Sears-Rogan: An unusual case of multiple papillary fibroelastoma, review of literature. Cardiovascular Pathology 12: 170-173, 2003.
- 8) Ramesh M Gowda, Ijaz A Khan, Chandra K: Cardiac papillary fibroelastoma: A comprehensive analysis of 725 cases. American Heart Journal 146: 404-410, 2003.
- 9) Gorton, M E, Soltanzadeh, H: Mitral valve papillary fibroelastoma. Ann Thorac Surg 47: 605-607, 1989.
- 10) Kazuma T, Akio I, Yutaka T, *et al.*: A case of Papillary Fibroelastoma of Mitral Valve. Jpn J Cardiovasc Surg 31: 150-152, 2002.