

## Papillary Fibroelastoma Developing from the Left Ventricular Septum

Hiroshi KUBO, Atsushi TABUCHI, Sohei HAMANAKA, Yasuhiro YUNOKI,  
Katsuhiko SHIMIZU, Eiichiro INAGAKI, Makio HAYAMA, Yoji KUBO, Takuro YUKAWA,  
Masao NAKATA, Hisao MASAKI, Kazuo TANEMOTO and Kiyoshi YOSHIDA\*

*Division of Thoracic and Cardiovascular Surgery, Department of Surgery, Kawasaki Medical School : 577 Matsushima, Kurashiki, Okayama, 701-0192 Japan*

*\*Division of Cardiology, Kawasaki Medical School : 577 Matsushima, Kurashiki, Okayama, 701-0192 Japan*

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**ABSTRACT.** Case: A 62-year-old male was admitted to our hospital for left side, intermittent claudication and shortness of breath on exertion. An echocardiogram revealed a tumor in the left ventricle and aortic valve stenosis. A coronary artery lesion (one vessel) and left external iliac artery occlusion were also detected by angiography. The left ventricular tumor was resected using the aortic approach. Replacement of the aortic valve was also performed. Based on our pathological observations, we concluded that a papillary fibroelastoma had developed from the ventricular septum. Due to the risk of embolism, it is advised that intracardiac tumors be surgically removed as rapidly as possible.

**Key words** ① papillary fibroelastoma ② left ventricular tumor  
③ cardiac tumor ④ surgery

Papillary fibroelastomas, which are the third most common tumor type, following myxomas and lipomas, account for 10% of benign cardiac tumors. Due to the low frequency of cardiac tumor occurrence, this type of tumor is rarely encountered. The majority of papillary fibroelastoma cases are discovered during cardiectomy or autopsy, but the recent increase in use of echocardiogram analysis has resulted in increased detection of this disease. We report a case involving a papillary fibroelastoma diagnosed by surgical excision of a tumor derived from the left ventricular wall.

### CASE REPORT

Approximately three years ago, a 62-year-old male began experiencing left side, intermittent claudication, as well as shortness of breath, when walking. He had begun to experience intermittent claudication after walking a distance of 200 m four months previously. Consequently, he was referred to our clinic. A systolic ejection murmur (Levin III/VI, dissipated in the cervical region) with peaks at the right

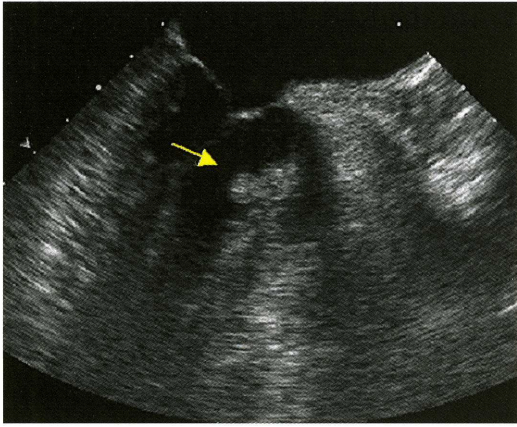


Fig. 1. Transesophageal echocardiogram showing the tumor in the left ventricle.

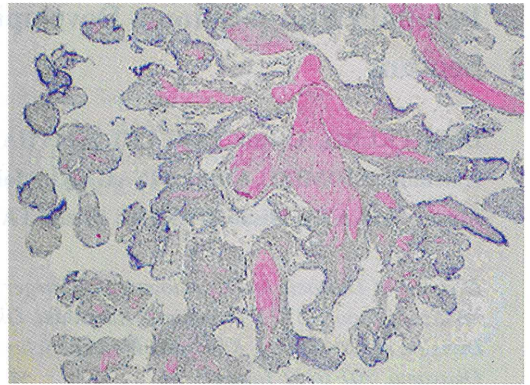


Fig. 2. Histological observations of the surgical specimen [hematoxylin-eosin (H-E) stained; magnification,  $\times 100$ ].

sternum border was detected. The pulse in his left lower limb was very weak [right ankle/brachial index (ABI): 1.14; left ABI: 0.68]. A transesophageal echocardiogram revealed a tumor ( $4.4 \times 5.7$  mm) with clear movement from the left ventricle near the outflow tract over the border region of the frontal wall and septum (Fig. 1). The aortic valve was bicuspid, characterized by a maximum pressure gradient of 77 mmHg and a 57% ejection fraction (EF). An arteriogram of the right coronary artery (seg. 2) revealed 90% stenosis. The left coronary artery was normal. Occlusion left external iliac artery occlusion was detected by arteriography.

In view of the possibility of severe complications such as sudden death due to embolism and brain infarction, surgical excision of the cardiac tumor was performed as a component of the treatment for complications. Surgery was conducted by employing a saphenous vein graft (SVG) for a bypass of the right coronary artery (one vessel). Subsequently, an incision was made in the aorta and the left ventricular tumor was surgically removed via resection of the bicuspid aortic valve, which exhibited severe calcification. The tumor, which was jelly-like, was  $0.8 \times 0.5$  cm in size. It was attached to the inside of the left ventricle, was 1 cm from the aortic valve ring on the apex side, and extended from the cardiac septum as a string stem form. Histological examination revealed papillary tissue characterized by a core consisting of elastic as well as collagenic fiber. The tumor surface was covered with flat cells, which appeared to be endothelial cells, with underlying mucous deposits (Fig. 2). The tumor was diagnosed as a papillary fibroelastoma. Replacement of the aortic valve was performed subsequently with a 23-mm Advancing The Standard (ATS) valve. Finally, a femorofemoral artery crossover bypass was performed with 8-mm expanded polytetrafluoroethylene (ePTFE).

## DISCUSSION

McAllister *et al.*<sup>1)</sup> reported that papillary fibroelastomas comprise 10% of benign cardiac tumors. It is the third most common tumor type, following myxomas and lipomas, with a wide range of onset times (although more than half of these tumors develop after the age of 60) and no gender difference. Papillary fibroelastomas exhibit a sea anemone-like appearance; many papillae developing from a central stem. These

structures layer in soft fiber clusters. The center forms with a collagen core. In addition, these tumors are surrounded by a hyperplastic inner membrane, which is continuous with the inner membrane in the valve<sup>1),2),3),6)</sup>. The majority of papillary fibroelastoma cases are discovered during cardiectomy or autopsy, but the recent increase in the use of echocardiogram analysis has resulted in increased diagnosis of this disease. Of 42 reported cases, 15 tumors were detected in the aortic valve, 9 were observed in the tricuspid valve, 8 occurred in the pulmonary valve, 7 were present in the mitral valve, 3 were discovered in the atria (two cases involving the right atrium and one involving the left atrium), 2 were evident in the right ventricle, and a single example was apparent in the left ventricular septum<sup>1),3),6)</sup>. Three of these cases exhibited multiple tumors. Approximately 90% of the cases displayed development from valves. Furthermore, the majority of cases demonstrated development from the aortic valve.

The present case involved a tumor that had developed from the left ventricular septum. This tumor type is the rarest among all papillary fibroelastoma types. Our single surgical case of papillary fibroelastoma developing from the left ventricular septum was accompanied by complications, including aortic valve stenosis, occlusive arteriosclerosis, and cardiac angina pectoris.

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