

Tricuspid Papillary Fibroelastoma Mimicking Tricuspid Vegetation in a Patient with Severe Neutropenia

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We report a 72-year-old male with known myelodysplastic syndrome who presented to the emergency department with a 7-day history of fever and dyspnea. Echocardiography revealed a round echogenic mass 13×16 mm in size attached to the atrial side of the tricuspid valve. Considering the high risk of infective endocarditis in the patient with a low absolute neutrophil count ($130/\text{mm}^3$), emergency surgery was performed. Intraoperatively, a single gelatinous neoplasm was resected, and subsequent reconstruction of the involved leaflet was accomplished using autologous pericardium. The tumor was pathologically confirmed as papillary fibroelastoma with no evidence of infective endocarditis. Papillary fibroelastoma is a rare cardiac neoplasm that occurs in either the mitral or aortic valves. Interestingly, a few cases of tricuspid valve papillary fibroelastoma have been reported so far. Similar echocardiographic findings between vegetation and tricuspid valve neoplasm make it difficult to distinguish these two disease entities.

Key words: 1. Heart neoplasms
2. Fibroelastoma
3. Tricuspid valve

CASE REPORT

A 72-year-old male with known myelodysplastic syndrome presented to Seoul St. Mary's Hospital with a 7-day history of fever and dyspnea. His planned chemotherapy for myelodysplastic syndrome had been stopped a month before due to fungal infection of the orbit. The initial absolute neutrophil count was only $130/\text{mm}^3$, and antibiotic therapy was initiated under the possibility of pneumonia or aggravation of orbital infection. Even after 7 days of antibiotic therapy, the fever persisted and no evidence of infection was found. We

performed transthoracic echocardiography under suspicion of infective endocarditis; a 13 mm echogenic mass was found on the atrial side of the tricuspid valve (Fig. 1). Similar findings were also revealed by transesophageal echocardiography. Fortunately, no functional abnormalities of the valve were seen on echocardiography.

Based on the clinical symptoms and the echocardiographic findings, the patient was highly suspected of having infective endocarditis of the tricuspid valve. He was scheduled for emergency surgical removal due to the possibility of fungal endocarditis and the poor clinical course of infective endo-

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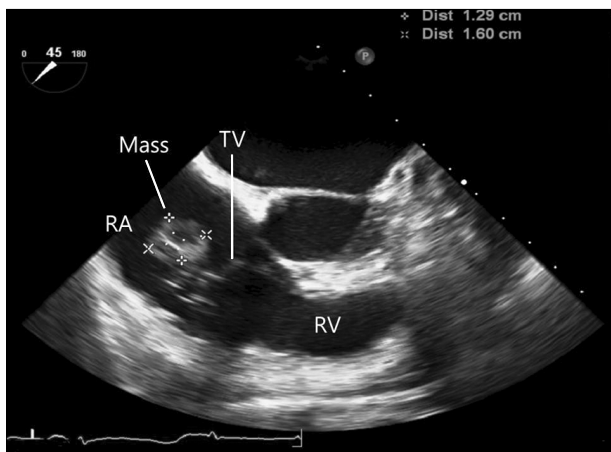


Fig. 1. Preoperative transthoracic echocardiogram shows an oscillating 13×16 mm echogenic mass which was attached on the atrial side of the tricuspid valve. RA, right atrium; RV, right ventricle; TV, tricuspid valve.

carditis in immunocompromised patients. With a cardiopulmonary bypass, the right atrium was opened after aortic cross-clamping and cardioplegic arrest. On the exploration, a solitary polypoid gelatinous mass 13×16 mm in size infiltrating the septal leaflet of the tricuspid valve was found in the surgical field (Fig. 2). Although the tumor had a stalk, simple resection without making a defect in the septal leaflet appeared to be impossible, because the leaflet was too thin. Therefore, the mass was excised with part of the septal leaflet of the tricuspid valve using the quadrangular resection technique, followed by reconstruction with autologous pericardium. After successful weaning from the cardiopulmonary bypass, intraoperative transesophageal echocardiography was performed, showing no residual tumors. The tricuspid valve was found to be competent without any functional deficits after operation. Papillary fibroelastoma was pathologically confirmed, and no evidence of infective endocarditis was found (Fig. 3). Follow-up transthoracic echocardiography showed no residual or recurrent tumor, with uneventful post-operative recovery.

DISCUSSION

Primary cardiac tumor is very rare, and its frequency is approximately 0.02% according to the data from 22 autopsy

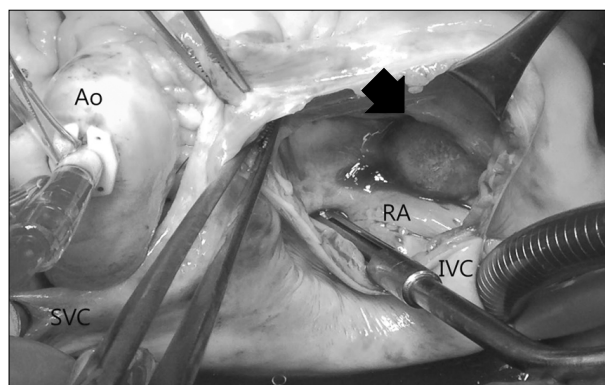


Fig. 2. Intraoperative finding shows the mass (arrow) attached to the septal leaflet of tricuspid valve without stalk. AO, aorta; SVC, superior vena cava; IVC, inferior vena cava.

cases [1]. Cardiac papillary fibroelastoma is the third most common benign primary cardiac neoplasm and the most common cardiac valvular tumor [2].

Grossly, cardiac papillary fibroelastoma is a gelatinous mass with multiple narrow papillary fronds, and it attaches to the endocardium via a short pedicle. Fibroelastoma is best visualized when immersed in water or saline. When the tumor is put in saline immediately after excision, it typically has a sea anemone-like appearance. The microscopic appearance is an avascular finger-like process with a core of elastin surrounded by collagen, which is covered by a single layer of endothelium [2,3]. The size of a cardiac papillary fibroelastoma varies from 2 to 70 mm, but the majority of tumors are approximately 10 mm in diameter [3]. The tumor is usually solitary, but rare cases of multiple cardiac papillary fibroelastoma have been reported [4]. Most of the tumors are located on the valvular surface, especially near or on the aortic valve (44%) and mitral valve (35%). Less frequently, the tumor infiltrates the pulmonary valve (8%) or the leaflets of the tricuspid valve (15%) [3].

The clinical presentation of a cardiac papillary fibroelastoma varies from no symptoms to severe thromboembolic complications, including transient ischemic attack, stroke, and myocardial infarction. Other complications include pulmonary embolism, congestive heart failure, near syncope, and sudden death. Cardiac papillary fibroelastoma occasionally manifests with fever, which abates after removal of the tumor [3]. Our patient with cardiac papillary fibroelastoma had a fever,

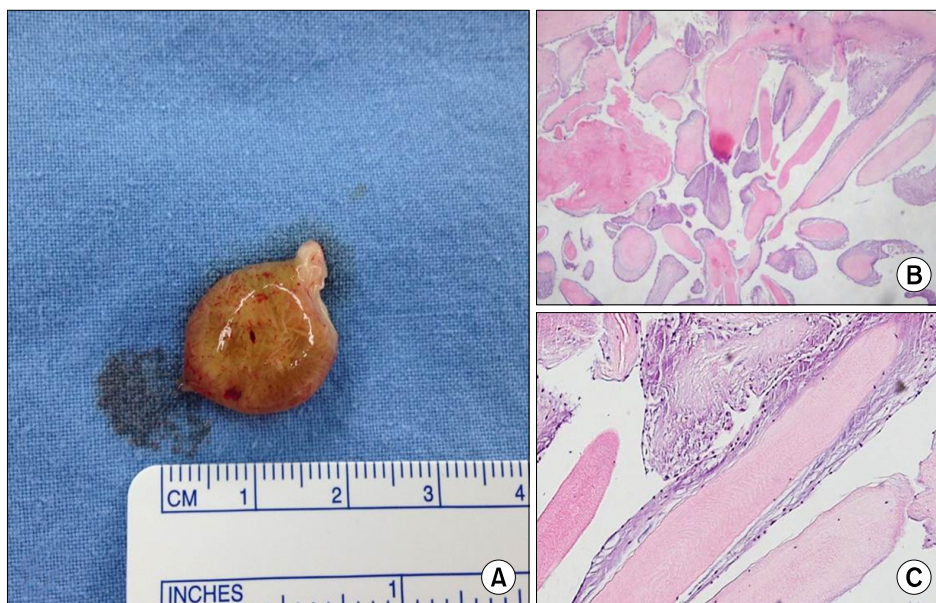


Fig. 3. (A) Flower like appearance with multiple papillary fronds (gross). (B) Distinctive cluster of hair-like projections (H&E, $\times 40$). (C) A central core of collagen surrounded by loose connective tissue, and covered by the endothelial cells (H&E, $\times 200$).

which did not subside even after several days of antibiotic therapy. However, after surgical removal of the tumor, the fever disappeared during the hospital stay.

Transthoracic echocardiography is useful for initial evaluation of a suspected cardiac papillary fibroelastoma. Occasionally, a small cardiac papillary fibroelastoma may not be detected with transthoracic echocardiography; therefore, transesophageal echocardiography is required to further assess such a small tumor. Computed tomography and magnetic resonance imaging are also helpful in detecting cardiac papillary fibroelastoma, but their diagnostic accuracy is relatively low [5]. The mobility and location of the papillary fibroelastoma that has developed on the valvular surface can be misdiagnosed as infective vegetation. In comparison with cardiac papillary fibroelastoma, infective vegetation is usually associated with clinical signs of endocarditis and valvular destruction, and may resolve or change over a short period after treatment [3].

It is obvious that surgical excision is necessary in symptomatic patients with intracardiac tumors proven by echocardiography, regardless of the type and the location of tumors. Management of asymptomatic patients with cardiac papillary fibroelastoma is still controversial. However, when the tumor is a left-sided lesion, surgical excision is recommended due to high-risk thromboembolic events. Controversially,

right-sided lesions are observed and surgically removed only in case they become symptomatic [6].

The risk of cardiac surgery in patients with hematologic malignancies remains to be established. Fecher et al. [7] demonstrated, in a study of 24 patients with hematologic malignancies undergoing cardiac surgery, that there was 1 in-hospital patient death (4.1%), 12 patients (50%) with a minor or major complication, and 7 patients requiring reoperations within 30 days due to bleeding, infection, and other clinical conditions. Chan et al. [8] reported, in a study of 26 patients with hematologic malignancies, that although there was no significant difference in mortality between patients with and without hematologic malignancies, there were significantly higher rates of transfusion, pneumonia, and sepsis in patients with hematologic malignancies than in patients without. The patient in this study improved after removal of the cardiac papillary fibroelastoma without any complications, and he is still alive at the 6-month follow-up.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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