

## A Rare Case of Aortic Valve Myxoma: Easy to Confuse With Papillary Fibroelastoma

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Myxoma of the aortic valve is an exceedingly uncommon condition. In this article, we report the case of a 72-year-old man with myxoma arising from the aortic valve. We extirpated the mass and repaired the aortic valve with the patient under cardiopulmonary bypass. The post-operative course was uneventful. Histological examination confirmed that the mass was a myxoma. (**Korean Circ J 2012;42:281-283**)

**KEY WORDS:** Myxoma; Aorta valve.

### Introduction

Cardiac myxoma usually develops in the atria and presents in the left atrium in more than 75% of patients and in the right atrium in only 15-20% of patients.<sup>1-9</sup> Only 3% to 4% of myxomas are detected in the left and right ventricle each.<sup>10-12</sup> Myxomas of the heart valves are rare,<sup>12</sup> and primary cardiac valve myxoma is unusual, especially aortic valve myxoma.<sup>2</sup>

### Case

A 72-year-old man was referred to our cardiovascular center with previous history of hypertension for 5 years and a 2-month history of worsening dyspnea on exertion that motivated him to seek medical attention. Atrial fibrillation was detected during a recent medical check up at a local clinic, and the patient was referred to our hos-

pital and admitted for closer examination. The laboratory data was unremarkable. His electrocardiogram revealed new-onset atrial fibrillation compared with previous data. A chest X-ray film revealed no particular anomalies. Two-dimensional transthoracic echocardiography (2D TTE) showed 1.04×0.87 cm nodular mass attached to the aortic valve. The echogenic mass originated from the ventricular side of the non-coronary cusp of the aortic valve (Fig. 1). Based on these findings, a papillary fibroelastoma of the aortic valve was suspected. Because of his old age and atrial fibrillation, scheduled surgery was performed to avoid systemic embolization. After aortic cross clamp, the aorta was opened. Inspection of the aortic root revealed a normal structure. A mass was found on the ventricular side of the non-coronary cusp. The mass was successfully excised from the aortic valve (Fig. 2). The tumor measured 1.5 cm and had a smooth surface with a spherical shape. The aortic valve was repaired rather than replaced after tumor resection. Intra-operative transesophageal echocardiography showed normal left ventricular outflow without regurgitation and no residual mass.

Histopathological examination of the tumor revealed a benign soft tissue tumor mass. The margin of the mass was irregular but was easily distinguishable from the normal valve tissue. The tumor consisted of plump gray purplish myxoid stroma and slender and long spindle stromal cells. The stromal cells had eosinophilic scanty cytoplasm and dark small nuclei. Fibrocollagenous bands of aortic valve tissue were also noted in the myxoid stroma (Hematoxylin and eosin; ×200) (Fig. 3). The final pathologic diagnosis was a myxoma of the aortic valve. The patient's postoperative convalescence was uncomplicated and he has discharged on aspirin with regularly sch-

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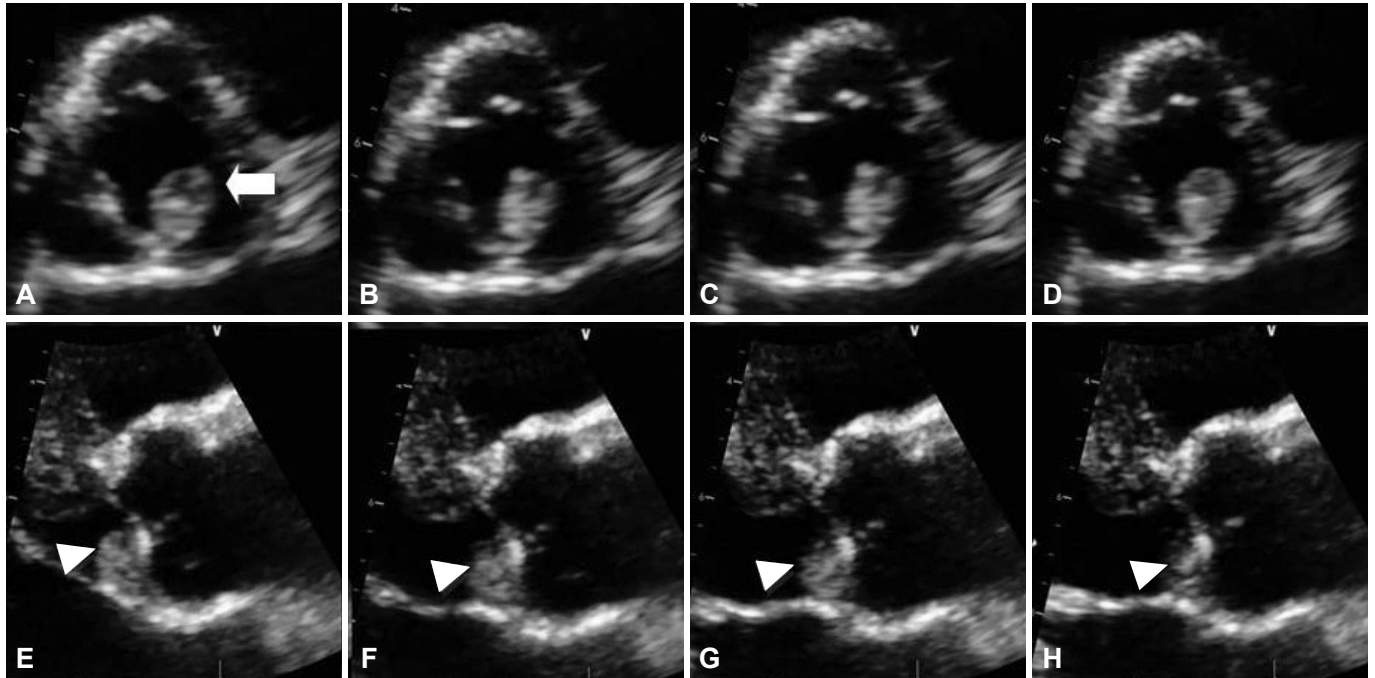
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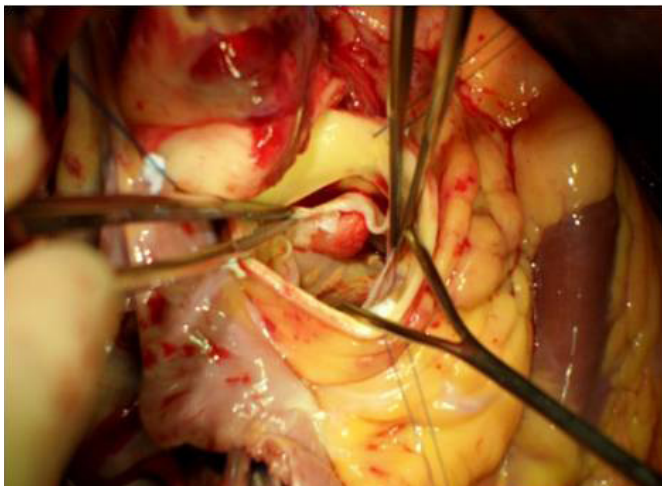
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**Fig. 1.** Echogenic mass in two-dimensional transthoracic echocardiography. This is serial images of cardiac tumor during the cardiac cycle. A-D were systole phase and E-H were diastole phase. A myxoma is seen on the aortic valve. A small spherical mass which shows relatively round surface with central necrosis was attached to the cusp of non coronary (A, arrow) and was highly mobile without pedicle. This tumor originated from the ventricular surface of the semi-lunar valve. It is attached to the leaflet of non coronary cusp (E-H, arrow head).

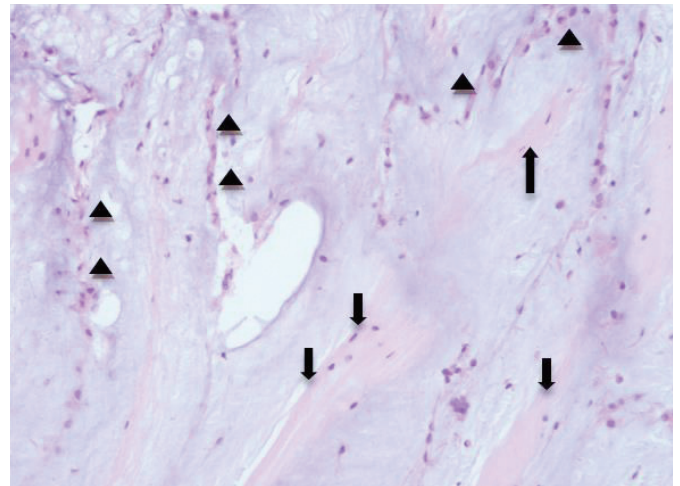


**Fig. 2.** Intra-operation view. After aortic valve opening, photograph shows an ovoid mass attached below non coronary cusp.

eduled follow-up. The post operative echocardiogram showed normal aortic valve function. No residual tumor was evident on the postoperative TTE 7 months after surgery.

## Discussion

Myxomas are one of the most common cardiac tumors but usually confined to the atria, more commonly the left atrium, or the ventricles. Myxomas of the cardiac valves are rarely if ever found. Aor-



**Fig. 3.** Histologic image, H-E stain (×200). Tumor consisted of gray purplish myxoid stroma with slender long spindle cells (arrow heads). Fibrocollagenous band portion of aortic valve is noted in the myxoma stroma (arrow).

tic valve tumors are rare and predominantly papillary fibroelastomas.<sup>13</sup> The differential diagnosis of aortic valve myxoma includes vegetation, tumor, and degenerative changes (Lambli's excrescence).<sup>14</sup> 2D echocardiography may help differentiate thrombus, vegetation, and myxoma, because thrombus typically produces a layered appearance and in some myxomas an area of echolucency may be seen within the tumor. However, pathologic differentiation of cardiac tumor by image is very difficult. Therefore, the size, shape,

location, mobility, and attachment site of a cardiac mass as well as the clinical presentation usually can differentiate these masses. The most important clue to the diagnosis is their location. By far, the most common cardiac tumor of the heart is the myxoma, which is usually single and occurs in the atria. Among tumors that affect the valves (tumors arising from the heart valves are rare), papillary fibroelastomas are by far the most common. This tumor appears as a small mass attached to the mitral or aortic valve with motion independent from the normal valve and often locates on the downstream side of the valve by a small pedicle and are irregularly shaped with delicate frond-like surfaces. In case of our experience, a small spherical mass that showed a relatively round surface with central necrosis was attached to the cusp of non-coronary and was highly mobile without pedicle. Unlike a typical fibroelastoma, according to tumor site, we speculated that the tumor was closer to fibroelastoma than other types of tumors. Although aortic valve myxomas are extremely rare in clinical practice, we should at least suspect them in patients with unusual tumor characteristics.<sup>15)</sup> Nevertheless, the clinical diagnosis of cardiac myxoma is often challenging, and distinguishing myxoma from a vegetation or thrombus by echocardiography is often difficult.

Our patient had an extremely rare aortic valve tumor as most aortic valve tumors are papillary fibroelastomas. A patient with atrial fibrillation and an aortic valve tumor is at an increased risk of systemic embolic events from a cardiac source. Aortic valve myxomas are rare benign cardiac tumors, often discovered after embolization has occurred.

Although long-term results from surgical treatment of cardiac myxomas are not completely understood due to its rarity, the rate of recurrence of aortic valve myxoma appears very low.<sup>16)</sup> Some reports have suggested that recurrence may be due to incomplete removal.<sup>16)17)</sup> Therefore, complete resection of the tumor was performed in this patient, with regular follow-up by echocardiography to monitor the remote chance of recurrence.

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