

Cardiac Papillary Fibroelastoma Presented as Angina: A Case Report

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Case report

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Abstract

Background: Cardiac papillary fibroelastoma is a rare, benign primary cardiac tumor, which always remains asymptomatic. Severe complications also had been reported in many cases. And the only effective therapy is surgical excision.

Case presentation: Here we report a case of cardiac papillary fibroelastoma with the beginning symptom of chest pain and first diagnosis of non-ST-segment elevation myocardial infarction (NSTEMI). The patient finally underwent tumor excision surgery and recovered well.

Conclusion: For patients with symptomatic cardiac papillary fibroelastoma we provide a series of comprehensive data before, during and after operation. This might be helpful for the further diagnosis and treatment of the tumor.

Background

Cardiac papillary fibroelastoma is classified as a rare, benign primary cardiac tumor, accounting for approximately 10% of all cardiac tumor[1]. Although most patients remain asymptomatic, the tumor might result in a series of severe complications occasionally. Diagnosis of such tumor is usually incidental and surgical excision is the only effective therapy so far. Here we present a patient with non-ST-segment elevation myocardial infarction (NSTEMI) secondary to fibroelastoma of aortic valve, who was successfully treated by surgical intervention.

Case Presentation

A 75-year-old female with chest pain and palpitation was admitted for acute coronary syndrome from emergency department. The initial electrocardiogram showed negative for ischemia, along with a mildly elevation of initial serum troponin T level of 61.5 ng/L. After further episodes of chest pain and ST depression in leads V_{2-6} , I, II, III, aV_F , ST elevation in lead aV_R , the patient was considered as left main trunk (LMT) obstruction and referred for cardiac catheterization.

No coronary stenosis was observed from coronary angiogram (Fig 1). However, during the angiography, this patient developed shortness of breath, with blood pressure decrease. Therefore, atropine and dopamine were administrated to treat the hypotension. When her discomfort resolved, she was transferred to CCU for special care and more examination.

Further blood test showed a peak serum troponin T of 4852.0 ng/L. Repeated electrocardiogram revealed widespread ST-segment depression again. Moreover, the patient felt markedly decreasing in exercise tolerance. Thus, echocardiography and computed tomographic angiography (CTA) were considered to identify whether she had heart failure, Aortic dissection or pulmonary embolism. Unexpectedly, echocardiogram demonstrated a isodense mass on the upper margin of the right coronary cusp, while CTA demonstrated a nodular filling-defect on the aortic valve (Fig 2).

Due to angina, myocardial infarction history and presence of tumor, the patient received surgical intervention. Transoesophageal echocardiography (TEE) was also performed both in and after the operation to reveal the tumor, guide the excision and evaluate the valvular function. Long-axis(at the level of left ventricle) and short-axis(at the level of aortic valve) in operation demonstrated a 18×12mm pedunculated, isodense mass attached to the left coronary cusp margin of the aortic valve, moving along with the opening and closing of aortic valve. There was also a mild regurgitation on the aortic valve (Fig 3). Aortotomy identified an approximately 2×1cm pedunculated mass arising from the left coronary cusp margin of aortic valve near the boundary between the left and right cusp. It was freely mobile with multiple fronds, resembling a sea anemone (Fig 4). There was no thrombus on its surface and it was suspected to intermittently obstruct the left coronary ostia by its movement. During the surgery, the tumor was dissected from the valve and the aortic valve was repaired. Postoperative TEE demonstrated the valvular function was good. Histologic characteristics of the tumor specimen included a centre core of collagen and elastin and the characteristic papillary fronds of a cardiac fibroelastoma (Fig 5). The patient did well postoperatively and was discharged home on the 10th postoperative day (Fig 6).

Discussion And Conclusion

Cardiac papillary fibroelastoma is classified as a rare, benign primary cardiac tumor accounting for approximately 10% of all cardiac tumors[2]. The etiology of this tumor is still unclear. There are many hypotheses of the original mechanism including neoplasms, hamartomas, organized thrombi, and unusual endocardial responses to infection or hemodynamic trauma[3-5]. The histological feature is described as an inner vascular core of connective tissue covered by endothelium that surrounds a layer of acid mucopolysaccharide, which is often with a peduncule or stalk, resembling a sea anemone[6].

As a past report presented, cardiac papillary fibroelastoma can be found anywhere inside the heart such as valvular surfaces, papillary muscles, chordae tendineae, ventricular septum or endocardial surface[7]. Frequently, it appears on aortic valve and mitral valve, especially on the aortic valve[7, 8]. The certain location of the tumor may be associated with the occurrence of the complications.

Although most patients are asymptomatic, serious complications have been reported, such as neural symptoms, angina, myocardial ischemia, distal embolic events[9-12]. The mechanism is thought to be intermittent obstruction of the coronary ostia or valve by the mobile tumor mass and embolization of tumor mass or platelet thrombi on the tumor into the coronary arteries or distal arteries[13]. It can also cause a sudden death by occlusion of the coronary artery reported through an autopsy[14]. As a result, the tumor will bring potential risks in to patients who have yet been symptomatic.

The diagnosis in living patients was rare before the introduction of echocardiography[15]. With the development of radiological technology, computed tomography scans(CT) or magnetic resonance imaging(MRI) has been introduced a limited use of diagnosing this tumor[16, 17]. Above all these techniques, echocardiography has been defined as a convenient and noninvasive way to diagnose and should be the first choice of tests to search for cardiac papillary fibroelastoma which can be

demonstrated as a pedunculated, mobile and echodense mass. And the use of transoesophageal echocardiography (TEE) provides a more visualized image to identify the location and guide the excision. CT and MRI can be supplements of echocardiography.

Surgical excision is the only therapy of cardiac papillary fibroelastoma[18].For those symptomatic patients, excision is recommended regardless of the size. For those asymptomatic patients, mobile lesions or lesions in the specific locations like coronary ostia also should be surgically excised because of the higher risk of obstructive and thromboembolic complications. And a valvuloplasty or a valve replacement often needs to be performed after cutting of the tumor. Other patients with nonmobile lesions less than 1 cm in diameter can be managed expectantly or with anticoagulation. In addition, recurrence of PFE is very low[6].

In this case reported here, the patient presented with angina and was admitted by the primary diagnosis of acute coronary syndrome and paroxysmal atrial fibrillation. But the coronary angiogram was normal, while the definite symptom, electrocardiogram, enzymatic test suggested there was a myocardial infarction on this patient. Following tests of echocardiography and computed tomography scans (CT) both revealed a mass on the left coronary cusp margin of the aortic valve. With the evidence of persistent symptom and aortic valvular mass, surgical excision was performed. Intraoperative findings identified the location of this mass. The histological test confirmed the mass was cardiac papillary fibroelastoma. According to other cases reported, embolization of tumor fragments or thrombotic material could not be excluded from the potential mechanism of myocardial infarction, but it is more possible that the tumor may block the coronary ostia causing the coronary ischemia. To our knowledge this might be the most comprehensive case of cardiac papillary fibroelastoma, so we have meticulously recorded the data of primary symptoms, enzymatic test, electrocardiogram, angiogram, echocardiogram, CT and histological test included before and after the operation. It is our belief that these might be more helpful in diagnosis and treatment of the tumor.

Abbreviations

CTA: computed tomographic angiography

PFE: Cardiac papillary fibroelastoma

TTE: Transthoracic echocardiography

Declaration

Acknowledgements

Not applicable.

Authors' contributions

J S participated in the surgery procedure and performed the CT scan, ZF K performed the statistical analysis and drafted the manuscript, M W and YQ G designed the study and revised this manuscript. All authors read and approved the final manuscript.

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none

Availability of data and materials

The data set of this case report is available from the corresponding author on reasonable request.

Ethics approval and consent to participate

The hospital ethics committee approval was granted of this case report.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

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Figures

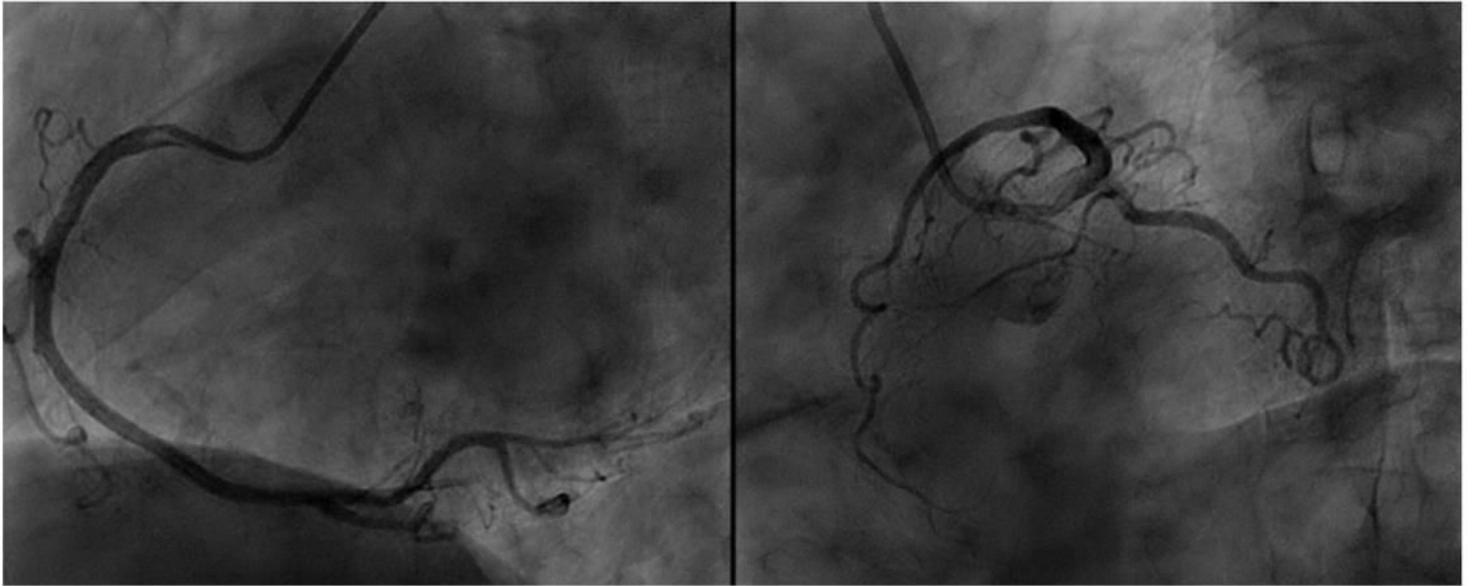


Figure 1

There was no obstruction in both left and right coronary artery.

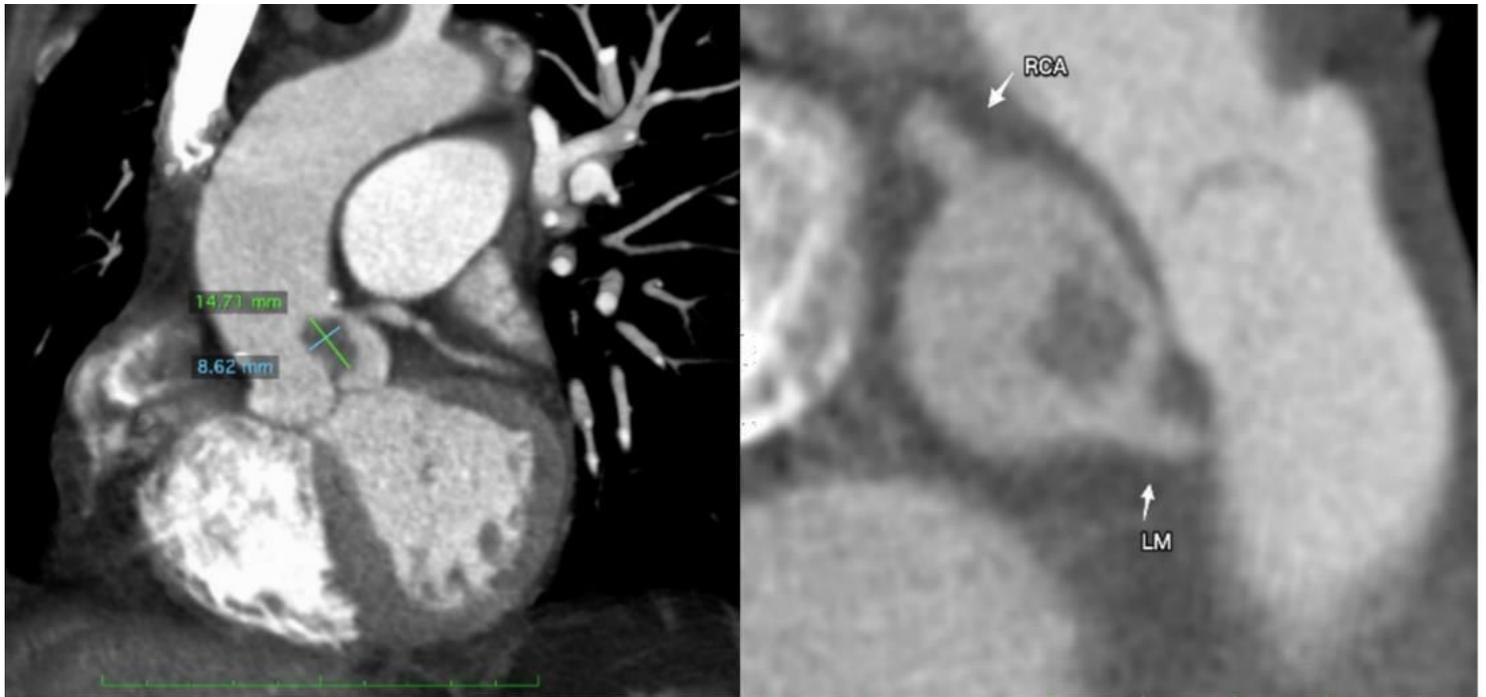


Figure 2

A isodense mass on the upper margin of the right coronary cusp, CTA demonstrated a nodular filling-defect on the aortic valve approaching to the left coronary ostia.

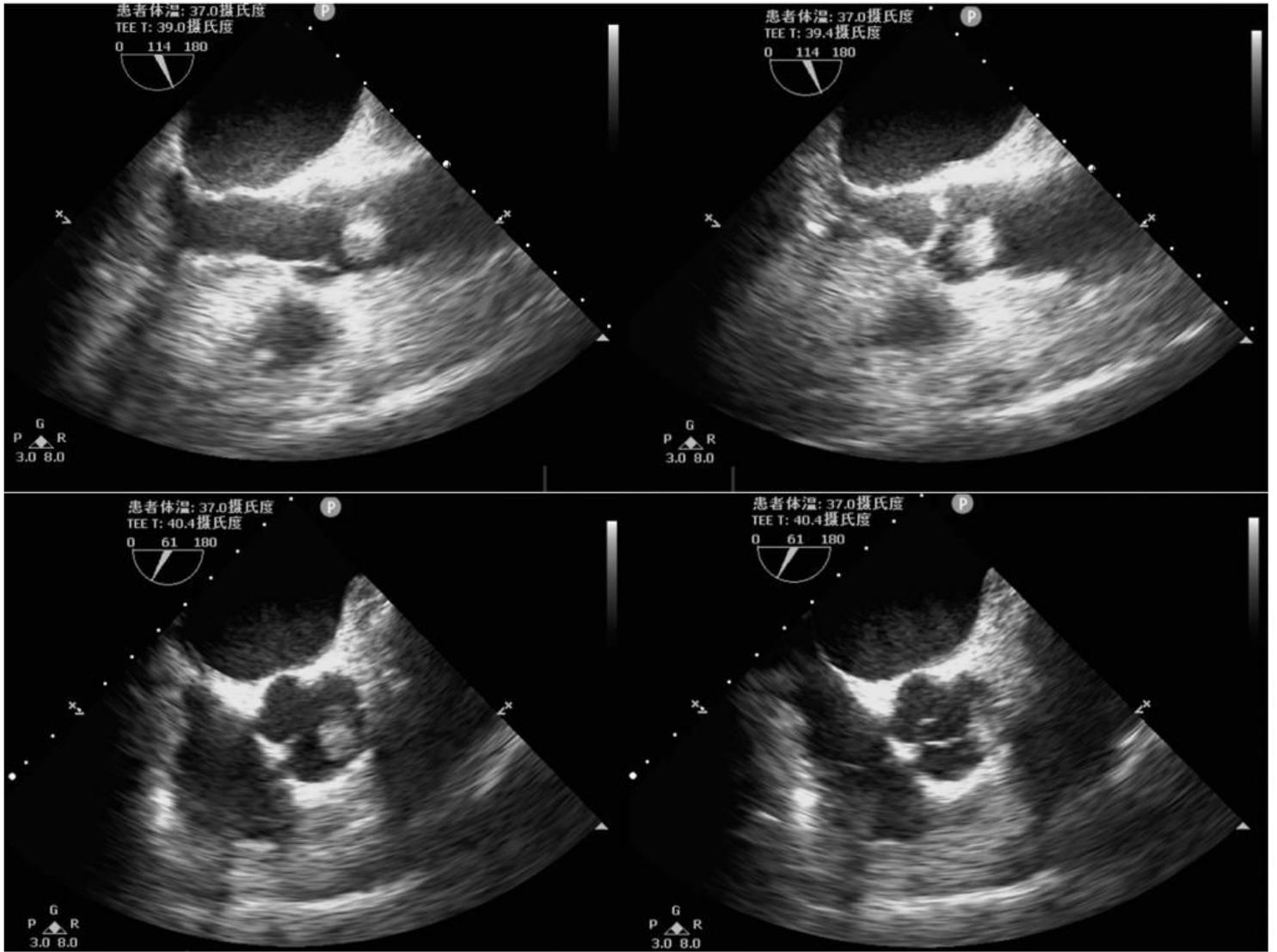


Figure 3

Intraoperative echocardiography showed a 18×12mm pedunculated, isodense mass attached to the left coronary cusp margin of the aortic valve, moving along with the opening and closing of aortic valve.

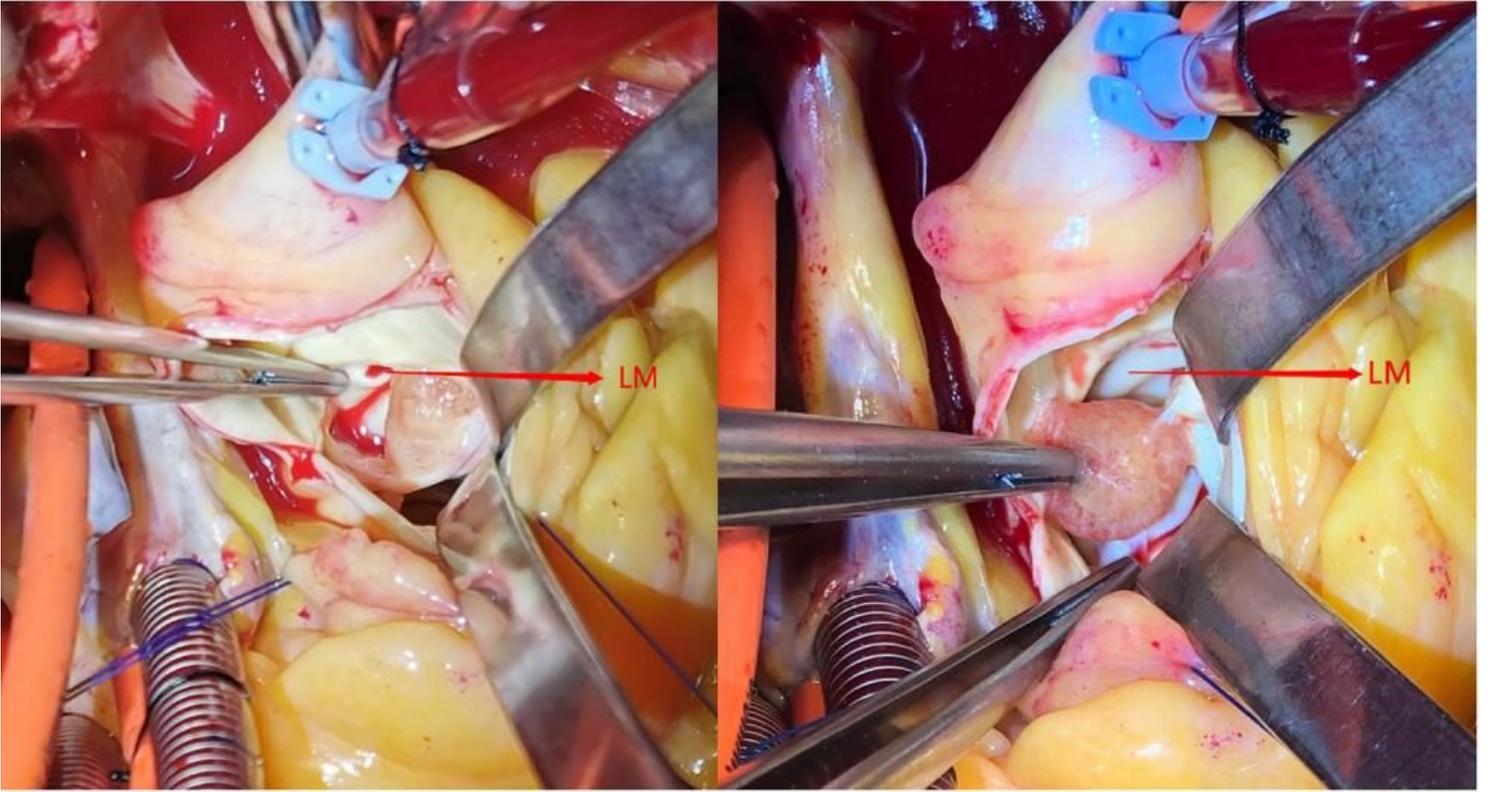


Figure 4

An approximately 2×1cm pedunculated mass resembling a sea anemone arises from the left coronary cusp margin of aortic valve near the boundary between the left and right cusp.

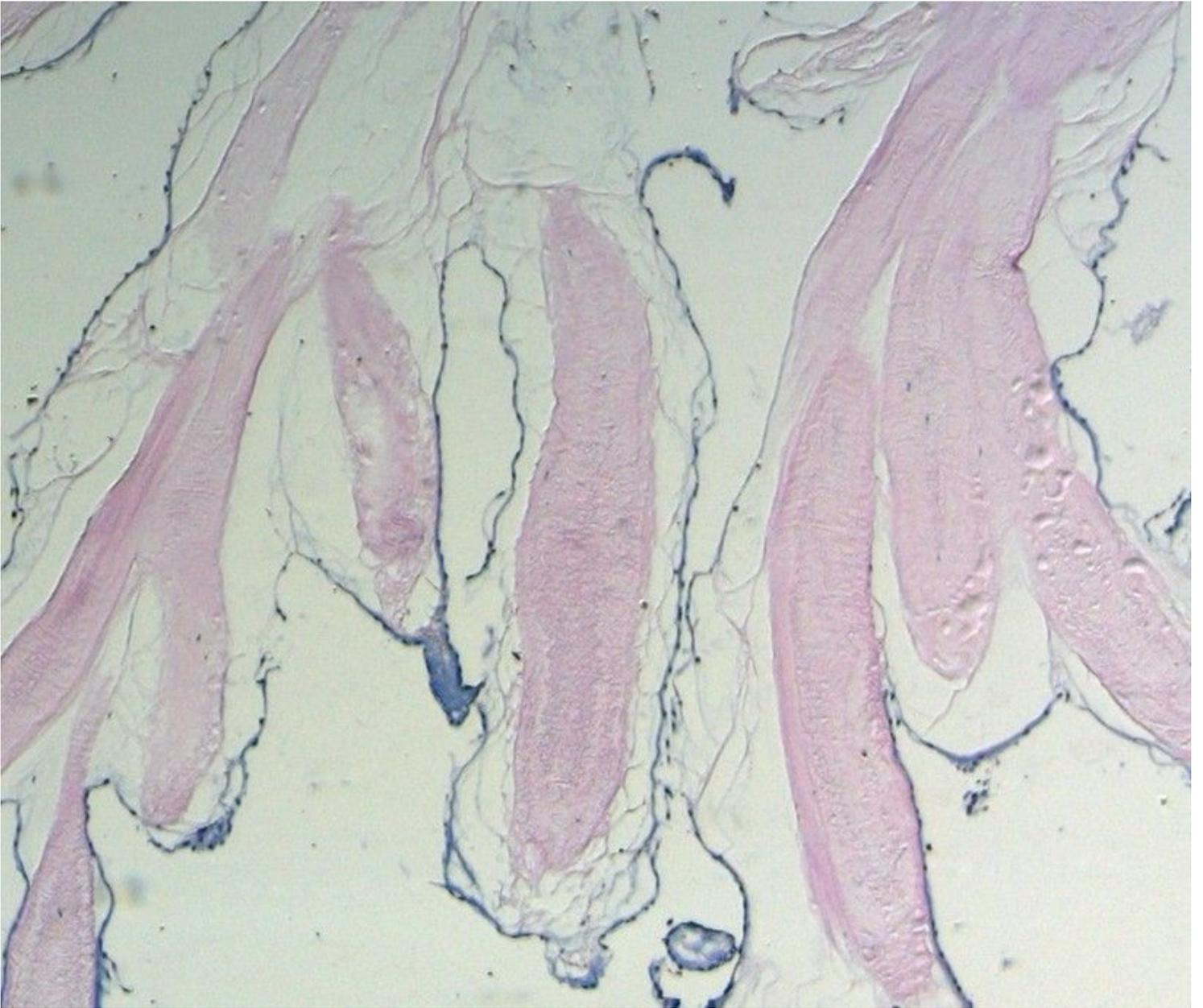


Figure 5

Microscopic appearance of the " tumor" showing a mass of villous projections of myxoid connective tissue with endothelial cell proliferation.

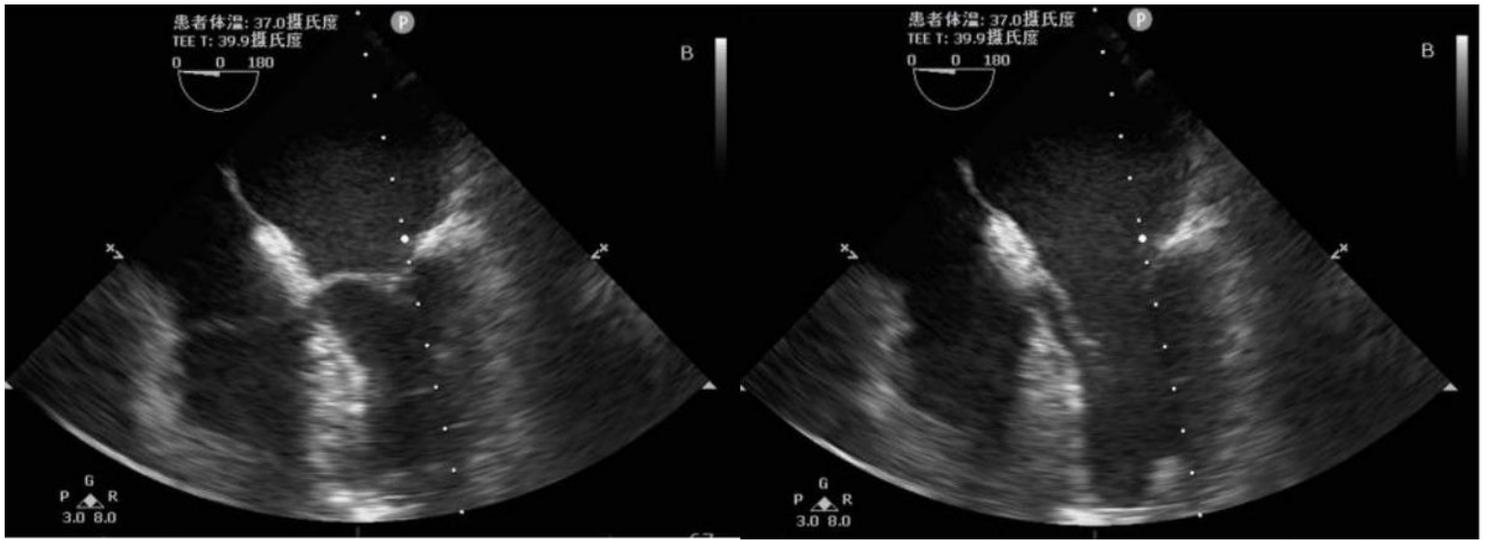


Figure 6

The echocardiogram after operation shows the function of aortic valve was normal.