

Valve Sparing Surgery for Papillary Fibroelastoma of the Aortic Valve

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Cardiac papillary fibroelastoma (CPF) is a rare benign tumor that involves the heart valves and may cause thromboembolism or arrhythmia. The case is presented of an asymptomatic, 60-year-old woman in whom a fibroelastoma was localized on the ventricular aspect of the non-coronary leaflet of the aortic valve. The patient had only palpitation of the heart, and the tumor was accidentally found during transthoracic echocardiography. The patient underwent immediate surgical intervention. The tumor was removed without destruction of the valve. The postoperative course was uneventful. Follow-up echocardiography 4 years after the operation showed no recurrence of the tumor. Due to the high incidence of embolism and low risk of recurrence, the tumor must be surgically removed by simple excision immediately after a diagnosis is confirmed.

Key Words: Aortic valve • Papillary fibroelastoma

CASE REPORT

A 60-year-old female visited a local cardiologist due to intermittent palpitation for 1 month. The echocardiography showed a tumor on the aortic valve. She was referred to us for surgical intervention. At our hospital, the 2-D echocardiography revealed a 0.7 cm × 0.7 cm mass on the non-coronary cusp of the aortic valve (Figure 1). To prevent cardiac embolic events, we did an urgent operation.

During the operation, we found a round tumor attaching to the ventricular aspect of the non-coronary aortic valve leaflet. The tumor was 0.7 cm in diameter, and its appearance was semi-parent, pedunculated, and villous-like (Figure 2). Using a number 15 blade, we ex-

cised the tumor without destruction of the aortic valve. The cardiac chambers were not opened since there was no specific finding on the preoperative transthoracic echocardiography and intraoperative transesophageal echocardiography. Microscopically, the specimen showed numerous papillae with fibrinous, hyalinized matrix covered by a single layer of flattened lining endothelial cells (Figure 3). The pathologic diagnosis was cardiac papillary fibroelastoma.

The patient experienced an uneventful postoperative course. After discharge, she has been followed up at our



Figure 1. Echocardiography revealed a round mass on the ventricular aspect of the aortic valve.

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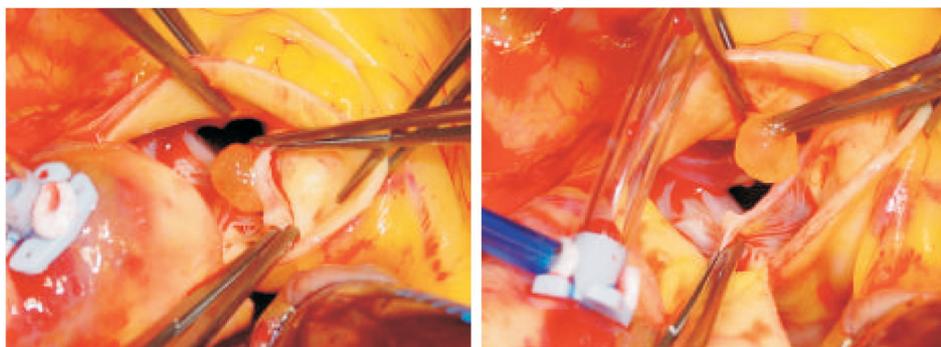


Figure 2. Left: Tumor attached on the ventricular surface of the non-coronary cusp; Right: Tumor was easily excised with a blade, without destruction of the valve tissue.

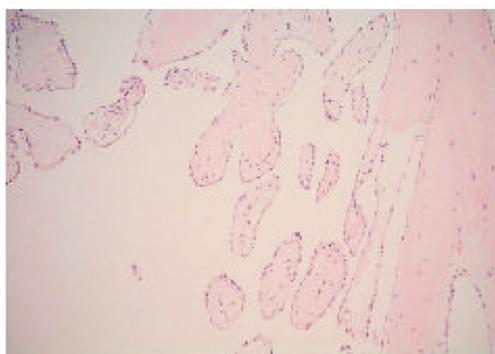


Figure 3. Microscopic findings of the papillary fibroelastoma: numerous papillae with fibrinous, hyalinized matrix covered by a single layer of flattened lining endothelial cells.

outpatient clinic for 4 years. Periodical echocardiographic studies showed normal valve function and no evidence of recurrence of the tumor.

DISCUSSION

Primary cardiac tumors are rare, with an incidence of about 0.02% of overall autopsies.¹ Although only 7.9% of primary cardiac tumors are cardiac papillary fibroelastoma (CPF),² they are the second most common primary cardiac tumor following myxoma. CPF are the most common primary cardiac valvular tumor and account for 73% of these tumors.³

Grossly, CPF are gelatinous masses with a characteristic “sea anemone” appearance with multiple papillary fronds. They are solitary lesions that measure 1 cm or less in diameter and attach to the endocardial surface by a pedicle.⁴ Microscopically, CPF are avascular papil-

lomas with a single layer of endocardial cells covering the papillary surface.⁵ The matrix consists of proteoglycans, elastic fibers, and rarely spindle cells. The fibrinous matrix is the hallmark of this tumor.

Clinically, most CPF are found incidentally at the time of cardiac investigation for unrelated problem, or at autopsy.³ The most common clinical presentations in symptomatic patients are embolism to the cerebral, systemic or coronary artery circulations, followed by congestive heart failure and syncope.⁶

The treatment of the papillary fibroelastoma is surgical excision with possible valve repair or replacement.⁷ Recurrence after surgical excision has not been reported.⁸ Furthermore, incomplete excision with no regrowth has been reported.⁹

Controversy about surgical indication for the patients with papillary fibroelastoma may exist. Sun et al. concluded that symptomatic patients, patients undergoing cardiac surgery for other lesions, and those with highly mobile and large CPFs should be considered for surgical excision.¹⁰ However, based on the potentially lethal complications of cerebral or coronary embolism, Howard et al. recommended surgical treatment regardless of size and symptoms, since surgical excision is seemingly curative, safe and well tolerated.¹¹ Grinda et al. suggested the potential for life-threatening complications of CPFs is the indication for surgical excision even in asymptomatic patients with tumor as small as 3 mm in diameter, and prophylactic anticoagulant should be initiated at diagnosis.¹²

Cardiac papillary fibroelastomas are rare benign tumors with the risk of embolic events. They are curable by simply surgical excision. Recurrence after excision

almost never happens even after incomplete excision. Therefore, aggressive resection with valve repair or replacement may not be necessary. In this case report, the tumor was excised by simple excision. We conclude that CPF of the aortic valve should be removed by simple excision to prevent embolism soon after the diagnosis is established.

REFERENCES

1. Reynen K. Frequency of primary tumors of the heart. *Am J Cardiol* 1996;77:107.
2. Gowda RM, Khan IA, Nair CK, et al. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. *Am Heart J* 2003;146:404-10.
3. Edwards FH, Hale D, Cohen A, et al. Primary cardiac valve tumors. *Ann Thorac Surg* 1991;52:1127-31.
4. Grebenc ML, Rosado de Christenson ML, Burke AP, et al. Primary cardiac and pericardial neoplasms: radiologic-pathologic correlation. *RadioGraphics* 2000;20:1073-103.
5. Burke A, Virmani R. Papillary fibroelastoma: tumors of the heart and great vessels. In: *Atlas of Tumor Pathology. 3rd series. Washington DC: Armed Forces Institute of Pathology; 1996. Fascicle 16. p. 47-54.*
6. Klarich KW, Enriquez-Sarano M, Gura GM, et al. Papillary fibroelastoma: echocardiographic characteristics for diagnosis and pathologic correlation. *J Am Coll Cardiol* 1997;30:784-90.
7. Ngaage DL, Mullany CJ, Daly RC, et al. Surgical treatment of cardiac papillary fibroelastoma: a single center experience with eighty-eight patients. *Ann Thorac Surg* 2005;80:1712-8.
8. Shahian DM, Labib SB, Chang G. Cardiac papillary fibroelastoma. *Ann Thorac Surg* 1995;59:538-41.
9. Sumino S, Paterson HS. No regrowth after incomplete papillary fibroelastoma excision. *Ann Thorac Surg* 2005;79:e3-4.
10. Sun JP, Asher CR, Yang XS, et al. Clinical and echocardiographic characteristics of papillary fibroelastomas. *Circulation* 2001;103:2687-93.
11. Howard RA, Aldea GS, Shapira OM, et al. Papillary fibroelastoma: increasing recognition of a surgical disease. *Ann Thorac Surg* 1999;68:1881-5.
12. Grinda JM, Couetil JP, Chauvaud S, et al. Cardiac valve papillary fibroelastoma: surgical excision for revealed or potential embolization. *J Thorac Cardiovasc Surg* 1999;117:106-10.