**Case
Report**

Simultaneous Surgery for Patent Ductus Arteriosus Associated with Papillary Fibroelastoma

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We describe a case of patent ductus arteriosus (PDA) in a 76-year-old woman with a history of stroke, atrial fibrillation, and chronic obstructive pulmonary disease. Cranial diffusion-weighted imaging (DWI) performed for preoperative assessment showed a hyperintense lesion in the left cerebellum. Preoperative transesophageal echocardiography (TEE) demonstrated two highly mobile masses approximately 5 mm in diameter adherent to the left and non-coronary cusps of the aortic valve. We performed transpulmonary patch closure of PDA under hypothermic circulatory arrest. Subsequently, two frond-like masses were completely shaved off the cusps, preserving the native aortic leaflets. Pathological examination confirmed the diagnosis of papillary fibroelastoma (PFE). To our knowledge, this is the first report of PDA associated with PFE. Perioperative use of TEE is an effective tool for management of cardiovascular patients with suspected cardiogenic embolism.

Keywords: patent ductus arteriosus, papillary fibroelastoma, simultaneous surgery, transesophageal echocardiography

Introduction

Patent ductus arteriosus (PDA) usually shows more severe calcification and fragility around the ductus in the elderly than in the younger patients. Several techniques have been reported to perform PDA closure safely. As the use of transesophageal echocardiography (TEE) has become widespread, abnormal structures such as papillary fibroelastoma (PFE) have been found frequently and incidentally. These lesions carry a high risk of

systemic embolization. We describe the surgical case of a 76-year-old woman with a PDA who underwent a transpulmonary patch closure under hypothermic circulatory arrest, with a simultaneous excision of PFEs, without a valve repair.

Case

A 76-year-old woman was admitted to our hospital complaining of dyspnea on effort. She had been treated

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Fig. 1 Enhanced computed tomography demonstrated a large patent ductus arteriosus (PDA) with severe calcification in circumference and no aneurysmal change of the distal arch.

for hypertension, persistent atrial fibrillation, chronic obstructive pulmonary disease, and cerebral infarction for several years. The anticoagulation therapy with warfarin was shown to be adequate by a prothrombin time-international normalized ratio (PT-INR) of 1.92. A continuous murmur was heard near the left sternal border. The patient was febrile, and a chest radiograph showed severe cardiomegaly with a cardiothoracic ratio of 68.6% and an increase in size of the pulmonary trunk. An electrocardiogram showed atrial fibrillation with a low voltage of the f wave. Transthoracic echocardiography revealed an L-R shunt toward the pulmonary trunk, severe pulmonary regurgitation, and severe tricuspid regurgitation, however detected no intracardiac mass. Cardiac catheterization revealed a pulmonary/systemic flow ratio of 2.5, pulmonary artery systolic pressure of 83 mm Hg, mean pulmonary artery pressure of 53 mm Hg, pulmonary capillary wedge pressure of 23 mm Hg, and pulmonary artery resistance of 984 dynes · sec/cm⁵. No significant coronary stenosis was observed by coronary angiography. Enhanced computed tomography demonstrated a large PDA with severe calcification in circumference and no aneurysmal change of the distal arch (**Fig.1**). TEE demonstrated 2 highly mobile masses approximately 5 mm in diameter adherent to the left and non-coronary cusps of the aortic valve, with neither aortic regurgitation nor left atrial thrombus. Cranial diffusion-weighted imaging (DWI) performed as a preoperative assessment showed a hyperintense lesion in the left cerebellum. The patient was asymptomatic

Fig. 2 Two frond-like masses were resected. One was attached widely to the edge of the left coronary cusp (**a**) and the other to the ventricular side of the non-coronary cusp (**b**).

neurologically, and the relationship between the brain infarction and the aortic valve tumor was unknown. We decided to perform surgical treatment for both PDA and aortic valve tumors simultaneously.

A median sternotomy was performed, and a cardiopulmonary bypass (CPB) was instituted in the standard fashion of ascending aortic and bicaval cannulation. While the patient was being cooled after inserting an LV vent, we controlled backflow from the aorta by putting pressure on the pulmonary artery corresponding to the opening of the PDA. Then, we performed tricuspid ring-annuloplasty and resected the left atrial appendage. When the bladder temperature was lowered to 20°C, the ascending aorta was cross-clamped. Under hypothermic circulatory arrest, the pulmonary trunk was longitudinally opened. Because the pulmonary opening of the PDA was 10 mm in diameter, patch closure was performed with 6 pledgeted 4-0 polypropylene mattress sutures. Subsequently, the ascending aorta was opened. Two frond-like masses measuring approximately 5 mm × 5 mm were observed on the 2 coronary cusps. One was attached widely to the edge of the left coronary cusp and the other to the ventricular side of the non-coronary cusp. No thrombi were observed on the surface of the tumor (**Fig. 2a and 2b**). Both the lesions were completely shaved off the cusps, preserving the native aortic leaflets. Postoperative TEE showed no residual mass, normal aortic valve function, and no residual shunt through the PDA. Postoperative course was uneventful. During pathological examination, the tumor was found to exhibit a papillary structure covered with a layer of endothelial cells. It consisted of multiple papillae with an elastic core and myxoid material. These findings confirmed the diagnosis of PFE.

Discussion

An adult PDA can be rarely diagnosed based on physical examination finding such as a characteristic heart murmur. Various complications related to PDA include heart failure, pulmonary hypertension, atrial fibrillation, aneurysm, and infectious endocarditis. Relief from these symptoms requires the closing of the left-right shunt, except in cases of Eisenmenger syndrome. PDA in the elderly patients usually shows severe calcification and more fragility around the ductus than in younger patients. Since it is hazardous to divide or ligate the ductus, hypothermic circulatory arrest and balloon occlusion using CPB have been reported as the standard procedures for treatment of adult PDA.¹⁾ In our case, it was safe and effective to perform the transpulmonary patch closure under hypothermic circulatory arrest while securing a good exposure of the PDA. As the PDA was large, the use of a coil and Amplatzer duct occluder was contraindicated. Because we found no aneurysmal change, there was no necessity for invasive procedures such as intra-aortic patch repair, total arch replacement with open-stent grafting, or thoracic endovascular aortic repair (TEVAR).²⁾ Only for PDA, TEVAR concomitant with axilloaxillary bypass would be a useful option. To reduce the likelihood of thromboembolic events due to manipulation of an occlusion balloon through the ductus, the procedure was performed under hypothermic circulatory arrest. Any recanalization or aneurysmal change in the PDA during a long follow-up period must be addressed. If an aneurysmal change progresses in the PDA, TEVAR would be indicated as a less invasive alternative in the future.

PFE is a rare benign endocardial tumor secondary to myxoma. The tumor typically has a small pedicle and is similar to a sea anemone with multiple papillary fronds. PFE occurs more commonly on the aortic valve than on other valves, and sometimes occur in locations such as an interventricular septum, atrial endocardium, papillary muscle, and chordae tendineae. PFEs have been generally thought to be originated from various sources, including hamartomas, inflammation, thrombosis, and neoplasms. Grandmougin and colleagues have suggested the possibility of a virus-induced tumor source based on the immunohistochemical investigations.³⁾ Symptoms thought to be related to PFE include coronary embolism, cerebral embolism, pulmonary embolism, acute valvular dysfunction, ventricular fibrillation, and sudden death.

Embolization may arise from the papillary fronds themselves or from fibrin or thrombi originating from the tumor surface. In our patient, TEE was performed because of a history of brain infarction, in addition to the incidental presence of a new lesion on magnetic resonance imaging. TEE is a very useful modality in identifying cardiac masses in patients with suspected cardiogenic embolism. Therefore, for in patients for whom cardiovascular surgery is planned, TEE should be performed not only intraoperatively but also during preoperative assessment.⁴⁾ On TEE, PFE appeared as a pedunculated, homogenous, and mobile mass. Other pedunculated lesions include vegetations, thrombi, and myxomas. To differentiate among these conditions, it is necessary to evaluate the lesions, valve regurgitation, and clinical findings. Freedberg, et al. suggested that our patient's lesions could have been Lambl's excrescences, which are seen on pathologic examination of the valve strands to be highly mobile, fine, threadlike excrescences less than 1 mm in width.⁵⁾ Distinguishing between PFE and giant Lambl's excrescences is very difficult on echocardiography. The former may have a more sessile or broader stalk than the latter. Although PFE is a benign tumor, surgical removal, including simple excision with or without valve repair and valve replacement, has been recommended on an urgent basis to prevent catastrophic embolic complications.⁶⁾ In our case, we could perform a simple excision without valve repair despite the presence of a sessile tumor.

To our knowledge, this is the first report of PDA associated with PFE. The relationship between PDA and PFE is unknown in this case, but endocarditis due to PDA may be a cause of PFE.

Conclusion

We report the surgical case of an adult PDA patient who underwent transpulmonary patch closure under hypothermic circulatory arrest, with simultaneous excision of PFEs without valve repair. Perioperative use of TEE is an effective tool for management of cardiovascular patients with suspected cardiogenic embolism.

Disclosure Statement

None declared.

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