Case Reports

Autologous Pericardial Patch Repair for Papillary Fibroelastoma on an Aortic Valve Leaflet

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A 50-year-old man with no history of cardiovascular disease was referred to our hospital because of an abnormal electrocardiogram. Echocardiograms and computed tomograms revealed a 9-mm mass on the underside of an aortic valve leaflet. We chose surgical treatment, to prevent embolic events. The tumor's appearance and intraoperative frozen section were consistent with myxoma. We resected the tumor and its attachment, including the free margin of the aortic valve leaflet, and repaired the defect with use of a glutaraldehyde-treated autologous pericardial patch. The postoperative histopathologic diagnosis was papillary fibroelastoma. Six months later, echocardiograms showed mild aortic regurgitation and no recurrence of the aortic valve mass.

Papillary fibroelastoma and myxoma can be difficult to distinguish intraoperatively, yet the diagnosis has considerable influence on the surgical strategy, including whether valve-sparing excision is an option. Therefore, it is necessary to at least suspect both entities if the tumor characteristics are unusual. This case is instructive for surgeons and pathologists. ***(Tex Heart Inst J 2017;44(2):144-6)***

Papillary fibroelastoma (PFE), a benign endocardial papilloma, is the second most prevalent primary cardiac tumor and the most prevalent cardiac valvular tumor.[1] A PFE contains many papillary fragments, has frond-like projections, and is attached to the endocardium. It can resemble myxoma, making it difficult to distinguish one from the other intraoperatively. We report our experience with an aortic valve PFE that was diagnosed as myxoma intraoperatively. After excision, aortic valve repair was performed. We discuss lessons for surgeons and pathologists that can be learned from this case.

Key words: *Aortic valve/pathology; diagnosis, differential; fibroma/diagnosis/surgery; heart neoplasms/diagnosis/surgery; heart valve diseases/diagnostic imaging; treatment outcome*

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Case Report

In March 2014, a 50-year-old asymptomatic man with no relevant medical history was referred to our hospital after inverted T waves were seen on his routine electrocardiogram. Echocardiograms and coronary computed tomograms (CT) revealed a 9-mm-diameter mobile mass on the underside of the aortic valve, apparently on the right coronary cusp. A coronary CT revealed no stenosis. We could not determine whether the mass was thrombus or tumor; in either case, there was risk of embolization. Therefore, surgical treatment was planned. Transthoracic echocardiograms showed no asynergy, morphologic abnormalities, or valvular disease, including aortic regurgitation. Laboratory data revealed no abnormalities.

To perform mass resection, we established cardiopulmonary bypass between ascending aorta cannulation and right atrial drainage. After aortic cross-clamping, cardiac arrest was obtained with use of antegrade cardioplegia. The aortic valve tumor was stalkless and attached to the underside of the right coronary cusp. Its surface was smooth, indicative of myxoma (Fig. 1). Intraoperative frozen-section examination yielded spindle cells in edematous interstitial tissue, also consistent with myxoma.

We decided to resect the tumor and its attachment, with sufficient margins. Resection created a large trapezoidal defect, including the free margin of the aortic valve leaflet. We repaired the defect with use of a glutaraldehyde-treated autologous pericardial patch and 6-0 Prolene running sutures (Fig. 2). To test for regurgitation, we filled the aortic root with water and confirmed that the water level did not drop. In-

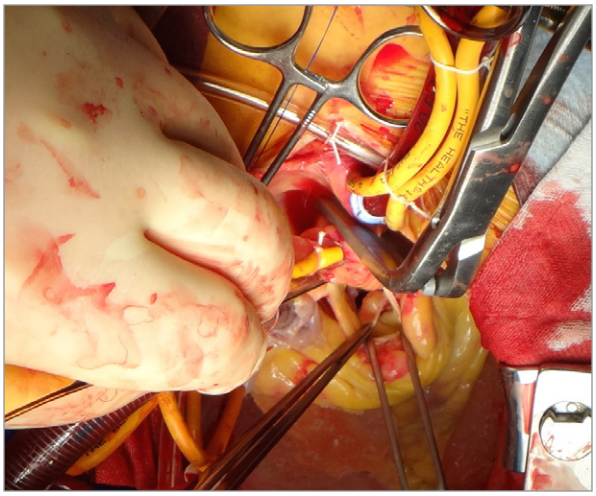

Fig. 1 *Intraoperative photograph shows a tumor with a smooth surface on the underside of the right coronary cusp.*

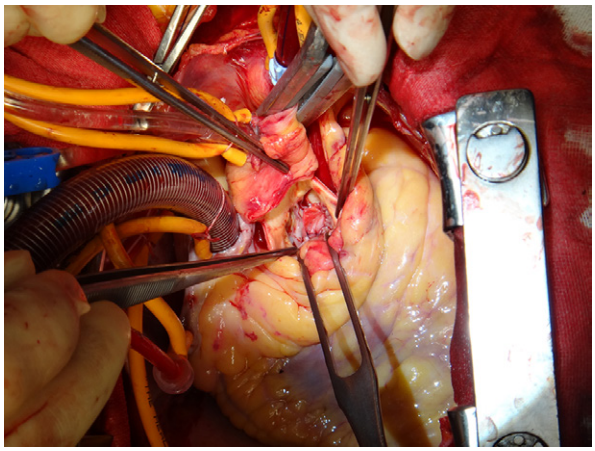

Fig. 2 *Intraoperative photograph shows the right coronary cusp leaflet after repair with a glutaraldehyde-treated autologous pericardial patch and 6-0 Prolene running suture.*

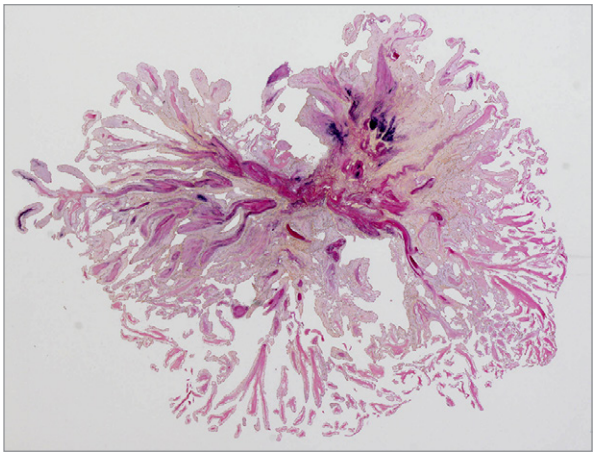

Fig. 3 *Photomicrograph shows papillary fronds containing a central fibroelastic core surrounded by a layer of endothelial cells (Elastica van Gieson stain, orig. ×50).*

traoperative transesophageal echocardiograms showed trivial aortic regurgitation and no residual mass. Postoperative histopathologic results revealed that the tumor had papillary fronds containing a central fibroelastic core surrounded by a layer of endothelial cells (Fig. 3). Thus, the tumor was finally diagnosed as a PFE. Six-month follow-up echocardiograms showed mild aortic regurgitation, no recurrence of the aortic valve mass, and a normal left ventricular ejection fraction of 0.67. Twelve months postoperatively, the patient had no signs of heart failure.

Discussion

Papillary fibroelastoma constitutes 4.4% to 8% of primary cardiac tumors, making it the second most prevalent after myxoma; PFE is the most prevalent cardiac valvular tumor.[1,2] It is widely considered to be a hamartoma or organized thrombus. Some authors have speculated that PFE is a virus-induced local growth[3] or that it might be caused by prior cardiac surgery.[4,5]

Although PFE is a slow-growing, benign neoplasm, there is a risk of embolization leading to myocardial or cerebral infarction. There are no guidelines on PFE management beyond a seeming consensus that symptomatic patients should be treated surgically. Ngaage and colleagues[6] further suggested that asymptomatic patients with a mobile PFE >10 mm in the left-sided heart chambers should undergo cardiac surgery, to prevent thromboembolic events or sudden death. Gowda and co-authors[7] reported that 12 of 25 medically treated patients died of tumor-related causes. Therefore, even though our patient's tumor was only 9 mm in diameter, we chose surgical treatment.

The typical treatment for PFE is simple shave excision. In one study of 725 patients with PFE, 425 were treated surgically: 81% underwent simple shave excision; 10%, valve replacement; and 9%, valve repair.[7] Had our patient's tumor been definitively diagnosed as PFE, we would have performed valve-sparing surgery; however, the gross appearance and the intraoperative frozen section were consistent with myxoma, not PFE. Resection of myxoma with inadequate margins leads to recurrence in approximately 5% of patients,[8] so patients with myxoma need valve repair or replacement to enable complete resection of the tumor and its attachment. We suspected myxoma and therefore trimmed the leaflet of the right coronary cusp with sufficient margins, then repaired the resultant large defect with a patch.

Because it can be difficult to distinguish PFE from myxoma on preoperative echocardiograms and CT, surgeons typically must reach a diagnosis intraoperatively. However, myxoma can be solid and have an appearance like that of PFE. A chief factor in the differential diagnosis is the location of the tumor, but rare cases of aortic

valve myxoma have been reported.[9,10] Accordingly, it is prudent to suspect both PFE and myxoma if a tumor's characteristics are atypical.

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