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Case Reports

Papillary Fibroelastoma Incidentally Found on Left Atrial Wall During Minimally Invasive Aortic Valve Replacement

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A 60-year-old man was about to undergo minimally invasive aortic valve replacement when transesophageal echocardiography revealed an intracardiac mass on the left atrial free wall. Multimodal images from 5 months earlier had shown no mass. We converted the procedure to open surgery. The excised mass resembled a cardiac myxoma but was determined to be a papillary fibroelastoma. This case illustrates that papillary fibroelastomas can form and grow rapidly, warranting alertness for their unexpected discovery before and during cardiac surgical procedures. ***(Tex Heart Inst J 2022;49(4):e217725)***

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Primary cardiac tumors have an incidence of <0.3% in autopsy series,[1] and about 25% are malignant.[2] Of the benign tumors, papillary fibroelastoma (PFE) is second most prevalent among the tumors that we have found in our patients.[3] In our experience, 80% of the PFEs have caused symptoms; approximately 50% have involved the left side of the heart, and 74% have originated from a valve. We recommend surgery to remove left-sided PFEs[3]; the need to excise right-sided PFEs is less obvious, because the risk of embolization and the potential for growth are not well known.[4] We present a case of a rapidly growing left-sided PFE that we found incidentally when planned minimally invasive aortic valve replacement (AVR) was to begin.

Case Report

A 60-year-old man with a history of atrial fibrillation was referred to our hospital for evaluation of severe, symptomatic aortic stenosis. He had begun experiencing dyspnea on exertion 12 months earlier. Five months before his referral, cardiac magnetic resonance (CMR) images and a transthoracic echocardiogram (TTE) had revealed severe aortic stenosis, mildly depressed left and right ventricular ejection fractions, and no intracardiac mass. The patient declined surgery at that time because his symptoms were mild; however, they worsened after another 4 months, and he was referred to our institution for possible AVR.

At the current presentation, coronary angiograms showed an occluded right coronary artery (RCA) that could not be engaged. Coronary computed tomographic angiograms revealed a heavily calcified proximal RCA with poor distal vessels and severe stenosis, and a heavily calcified proximal ascending aorta. Our cardiac surgical team recommended AVR through a small right anterior thoracotomy without intervening in the right-sided coronary system, which was supplied by collateral vessels from the left coronary artery. The RCA occlusion was not thought to be causing the patient's symptoms.

As the AVR procedure began, transesophageal echocardiography (TEE) revealed a mean gradient of 39 mmHg across the severely calcified aortic annulus. The TEE also showed a mobile, pedunculated, 0.75 × 0.8-cm mass that originated from the left atrial free wall between the left atrial appendage and the anterior mitral valve annulus (Fig. 1).

The AVR procedure was paused to enable reviews of the CMR and TTE images from 5 months before, and these again showed no mass.

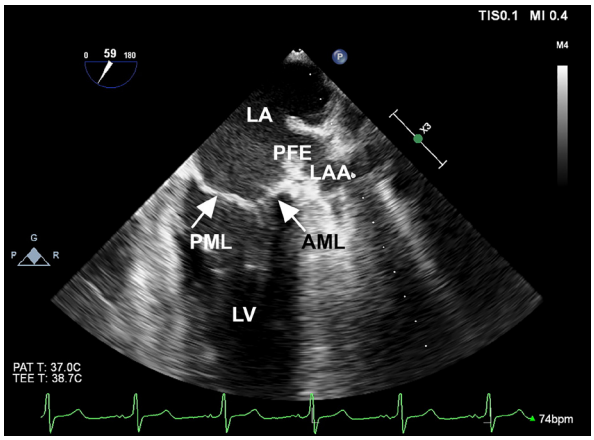

Fig. 1 *Transesophageal echocardiogram shows a papillary fibroelastoma (PFE) attached to the left atrial (LA) free wall between the LA appendage (LAA) and the anterior mitral valve annulus.*

AML = anterior mitral leaflet; LV = left ventricle; PML = posterior mitral leaflet

Supplemental motion image is available for Figure 1.

The mass posed embolic risk, so the planned thoracotomy was converted to a standard median sternotomy. Cardiopulmonary bypass was initiated through the ascending aorta with bicaval cannulation. We cross-clamped the aorta, administered cardioplegic solution, and performed a left atriotomy through the interatrial groove. The mass had a gelatinous, friable consistency and was initially thought to be a myxoma (Fig. 2). We resected it for pathologic examination, then proceeded with AVR. The aortic root was heavily calcified all the

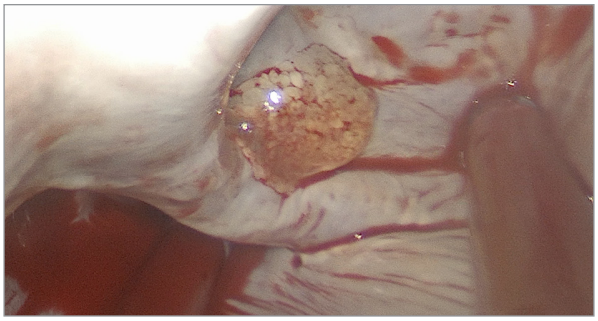

Fig. 2 *Intraoperative photograph shows the mass attached to the left atrial wall.*

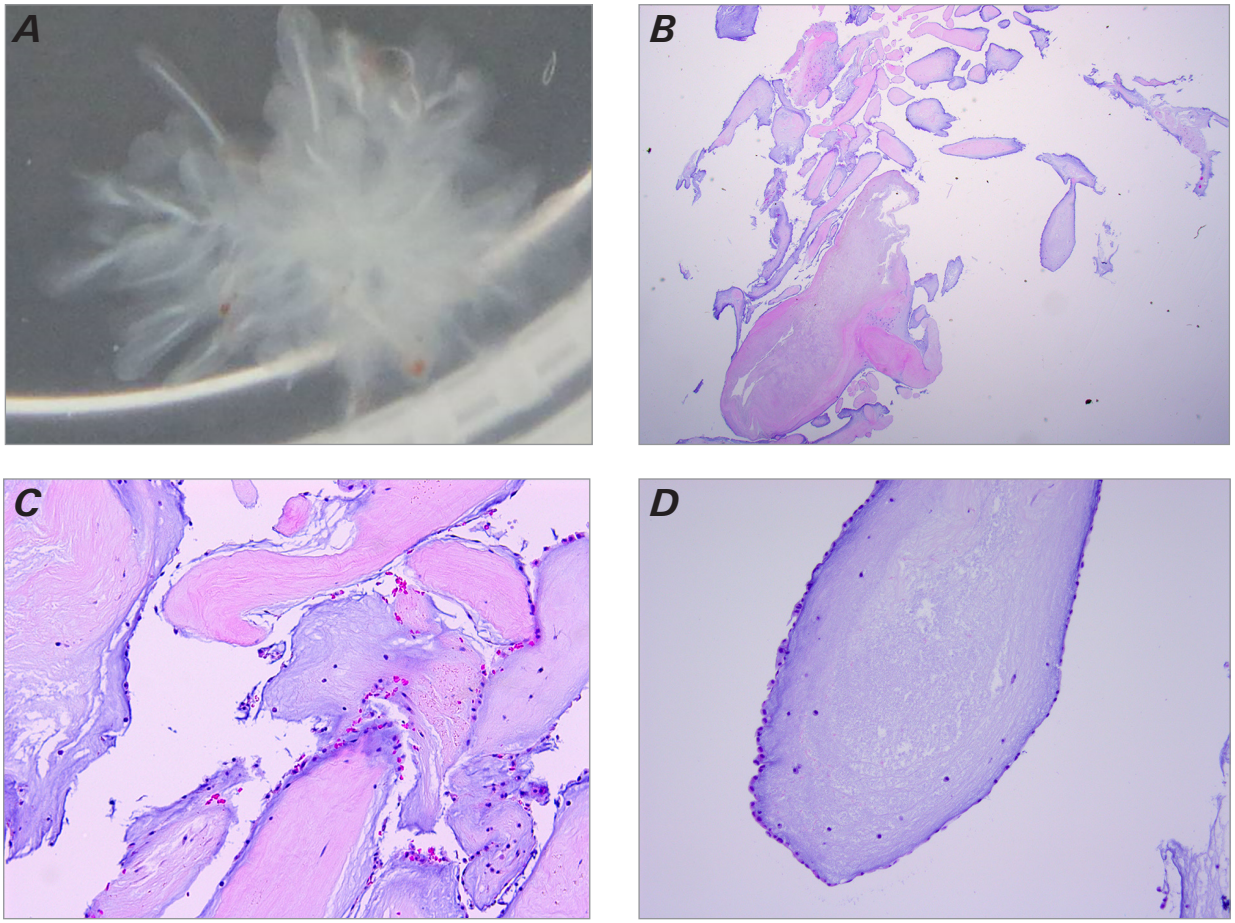

Fig. 3 ***A****) The excised mass placed in water shows the "sea anemone" appearance typically described in these lesions. Photomicrographs show* ***B****) the multiple papillary projections of the mass (H & E, orig. ×4),* ***C****) the papillary projections admixed with fibrin (H & E, orig. ×20), and* ***D****) the papillary fronds composed of a core of collagen and fibrous tissue with an endothelial lining (H & E, orig. ×40).*

way to the anterior mitral valve leaflet, which made placement of annular sutures challenging. We placed a medium-sized bioprosthetic Perceval sutureless aortic valve (LivaNova PLC). Postoperative TEE confirmed the absence of the mass, and the bioprosthetic valve was well seated without paravalvular leak and had good leaflet movement. The mean gradient across the valve was 5 mmHg.

The patient recovered uneventfully, and he was discharged from the hospital 7 days postoperatively. He was well at 6-month follow-up.

Histopathologic analysis of the excised specimen revealed papillary fronds comprising collagen and elastic tissue with endothelial lining, confirming PFE (Fig. 3).

Discussion

Untreated left-sided cardiac PFE is associated with an increased risk of stroke that neither warfarin nor antiplatelet therapy will reduce.[2] Investigators at the Mayo Clinic retrospectively evaluated the presurgical growth of pathologically verified PFEs in 67 patients by reviewing serial TEE studies conducted >30 days apart.[5] The overall mean PFE growth rate in those patients (0.5 ± 0.9 mm/yr) was exceeded by that in our patient (0.75 × 0.8 cm over 5 mo).

Although PFE has been thought to recur only infrequently after surgical excision, investigators at a single center determined recurrence in 12 of 98 patients (12.2%).[6] When left-sided PFE is seen before other cardiac surgery or is found incidentally on intraoperative TEE, resection should be considered. In our previous experience with excision of PFE,[3] which enabled our surgical team to approach this patient's tumor without problems, we reported no deaths and found no need to perform valve replacement later.

Conclusion

Just before performing minimally invasive AVR surgery with use of TEE, we incidentally discovered a PFE that had formed and rapidly grown over 5 months. We recommend that cardiac surgeons be alert to such masses when undertaking other surgical procedures.

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