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Case Reports

Papillary Fibroelastoma in Differential Diagnosis of Left Atrial Appendage Masses

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Papillary fibroelastomas are benign tumors that usually originate from cardiac valves but may have other endocardial origins. We report the cases of 2 patients in whom left atrial appendage masses were initially diagnosed as thrombus. They were treated for embolic stroke and their symptoms resolved; however, their left atrial appendage masses did not regress. After surgery, histologic analysis of the resected masses revealed papillary fibroelastoma in both cases. We discuss the diagnostic and therapeutic dilemmas encountered in patients with papillary fibroelastomas and cardiac masses other than thrombus. ***(Tex Heart Inst J 2021;48(1):e197088)***

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Papillary fibroelastoma (PFE) is a benign heart tumor often diagnosed incidentally or during evaluation for cardioembolic events.[1] These tumors can be asymptomatic, but left-sided PFEs tend to embolize. The aortic valve is most often affected, followed by the mitral, tricuspid, and pulmonary valves. The left ventricle is the chief nonvalvular location.[1]

We describe the cases of 2 patients whom we treated conservatively for stroke when masses were identified in their left atrial appendage (LAA). Initially presumed to be thrombus, the masses did not respond to therapy. As a result, we had to establish the correct diagnosis of PFE rapidly and determine the best treatment. Our experience emphasizes the need for differential diagnosis of all cardiac masses.

Case Reports

Patient 1

A 75-year-old man was admitted for stroke that affected the left middle cerebral artery and caused right-sided hemiparesis. Intravenous thrombolysis substantially resolved his symptoms. A transesophageal echocardiogram (TEE) revealed a 13.5 × 12.7-mm mass attached to the ridge in the left atrium (LA) between the LAA and left upper pulmonary vein (Fig. 1). The presumptive diagnosis was thrombus. We placed the patient on intravenous unfractionated heparin (target partial thromboplastin time, 50 s) for 23 days; however, TEEs at 1 and 2 weeks after therapy had begun showed no shrinkage of the mass. We suspected myxoma or another growth, so surgery was indicated. During surgical excision, a second mass (size, 5 mm) was discovered and excised (Fig. 2A). Because occult atrial fibrillation was possible, we occluded the LAA. Histopathologic analysis of both masses revealed PFE (Fig. 2B–C). After an uneventful 8-day hospital stay, the patient was discharged with instructions to take warfarin for 3 months. Through 4 years of postoperative follow-up, he had no stroke symptoms.

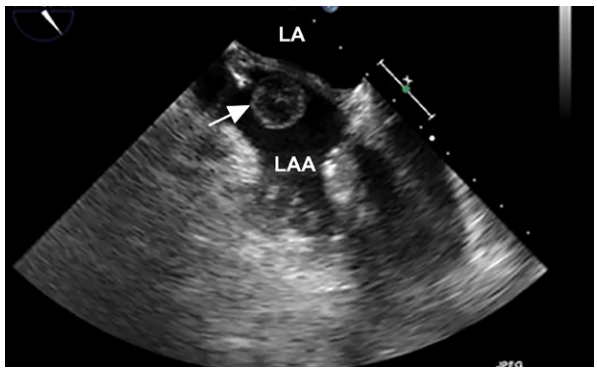

Fig. 1 *Patient 1. Transesophageal echocardiogram shows a 13.5 × 12.7-mm mass (arrow) between the left atrial appendage (LAA) and left upper pulmonary vein.*

LA = left atrium

Patient 2

A 58-year-old man presented at our stroke unit 10 hours after the gradual onset of right-sided hemiplegia and dysarthria. Computed tomograms revealed ischemic stroke caused by occlusion of the left middle cerebral artery. The patient's medical history included hypertension, type 2 diabetes, and hyperlipoproteinemia. His late presentation after symptom onset was outside the therapeutic window for thrombolysis or endovascular intervention, so he was prescribed 100 mg/d of aspirin. Electrocardiography at admission and 6-day adhesive-patch electrocardiography revealed continuous sinus rhythm. A TEE showed an 8 × 9-mm free-floating mass in the LAA (Fig. 3A). The presumptive diagnosis was LAA thrombus. The patient was discharged from the hospital with instructions to take warfarin (target international normalized ratio, 2–3) and undergo serial TEE for 6 months. When TEE showed no regression of the mass after one month, cardiac magnetic resonance (CMR) was performed to determine whether the mass was thrombus or a tumor. However, definite diagnosis was not possible because it was too small (10 × 5 mm) (Fig. 3B), so surgery was indicated. The mass was surgically excised from the LAA through a right anterolateral thoracotomy. Histopathologic analysis revealed PFE (Fig. 4). The patient recovered uneventfully and was discharged from the hospital 7 days postoperatively. All anticoagulation was discontinued. One year later, he had no residual neurologic symptoms.

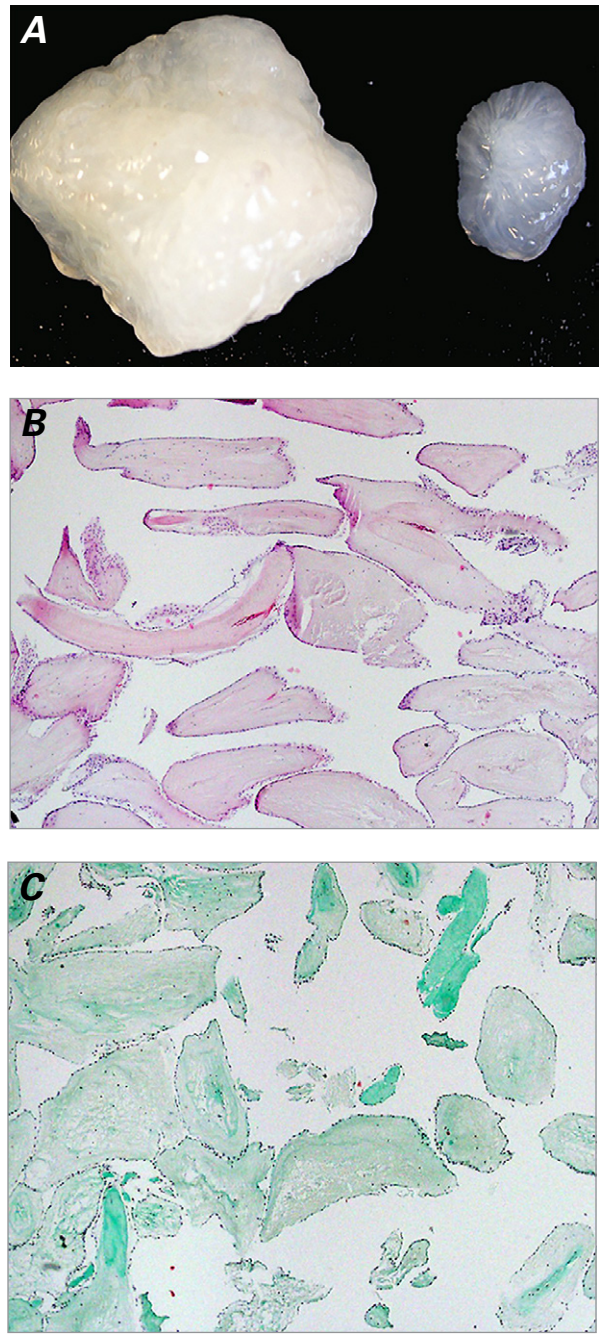

Fig. 2 *Patient 1.* ***A****) Photograph shows the larger mass seen on the echocardiogram and the smaller mass discovered during surgery. Photomicrographs of specimens from the larger tumor show* ***B****) delicate papillary protrusions covered by endothelium (H & E, orig. ×5) and* ***C****) papillary stalks containing various amounts of collagenous fibrous tissue (Masson-Goldner trichrome stain, orig. ×5), confirming papillary fibroelastoma. The smaller mass was similar.*

Discussion

Papillary fibroelastomas constitute 5% to 10% of all cardiac tumors. Approximately 80% of PFEs are found on cardiac valves; presentation in the cardiac chambers is unusual.[1] The incidence of embolization associated with PFE is as high as 69% in symptomatic patients. When myxomas are identified, surgical excision is recommended to reduce the risk of obstruction, embolization, or sudden cardiac death,[2] but no similar guidance for treating PFEs has been provided beyond the observations presented in a 14-patient case series.[3] The practice of watchful waiting in cases of asymptomatic or small, nonmobile PFEs has been challenged.[3,4]

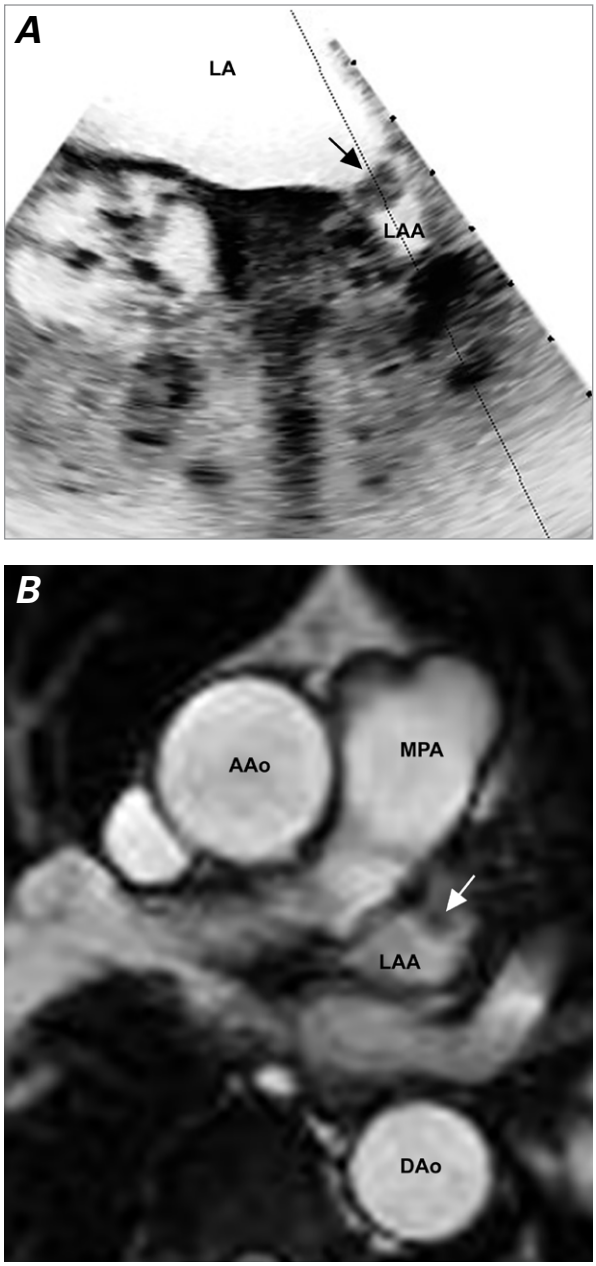

Fig. 3 *Patient 2.* ***A****) Transesophageal echocardiogram shows an 8 × 9-mm mass (arrow) floating freely in the left atrial appendage (LAA).* ***B****) Cardiac magnetic resonance (steady-state free precession sequence in the transverse plane) shows a faint mass (arrow) in the LAA.*

AAo = ascending aorta; DAo = descending aorta; LA = left atrium; MPA = main pulmonary artery

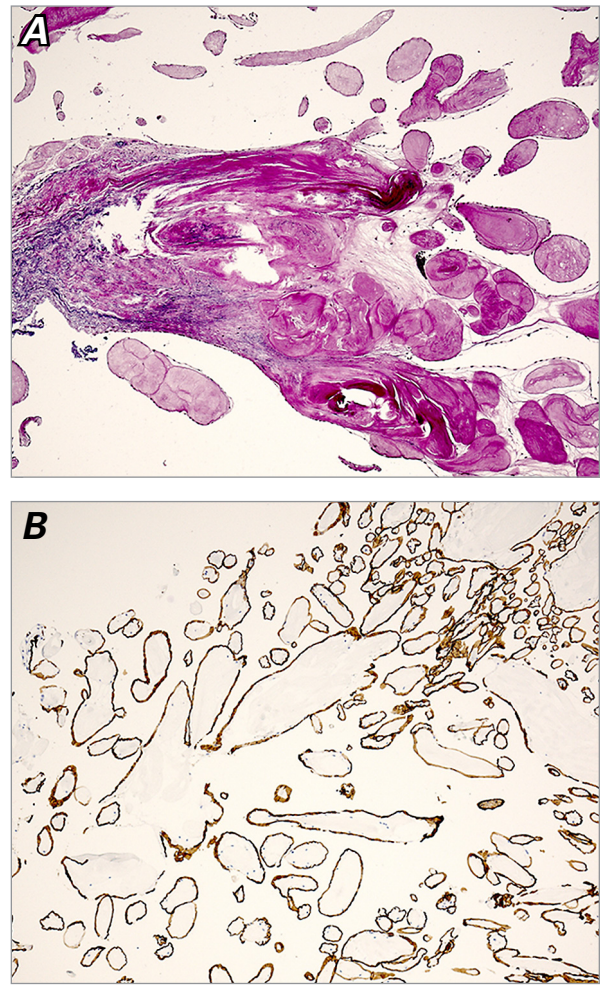

Fig. 4 *Patient 2. Photomicrographs show* ***A****) elastic fibers within the tumor stalk and peripheral papillary protrusions containing smooth collagenous fibrous tissue (Elastica-van Gieson stain, orig. ×5), and* ***B****) a superficial endothelial layer (CD34 immunohistochemical stain, orig. ×2.5).*

The LAA is the most frequent site of thrombus formation in the LA, especially in patients with atrial fibrillation.[5,6] Atrial cardiomyopathy, which indicates LA dysfunction, can cause cryptogenic stroke,[7] and various combinations of structural, architectural, contractile, and electrophysical changes that affect the atria can manifest themselves clinically.[8] Given our patients' presentations and comorbidities, we thought it possible that both had atrial cardiomyopathy and that PFE placed them at risk of aggravation—even without atrial fibrillation, an arrhythmia that is not always easy to rule out entirely.

The diagnostic and therapeutic dilemmas that we encountered, which are not unusual, emphasize the importance of accurate case-by-case diagnosis. Treatment options need to be weighed against the risks associated with watchful waiting and surgical intervention. The decision to perform surgery, and in which patients, should be guided by multimodal imaging of LAA masses. When echocardiographic diagnosis does not correlate with the patient's response to conservative anticoagulation, CMR is indicated. In comparison with cardiac-gated computed tomography, CMR enables more accurate tissue characterization and spatial resolution in differentiating tumor from thrombus and does not necessitate exposing the patient to radiation. Moreover, CMR can distinguish between benign and

malignant masses. If the diagnosis remains unclear after CMR, as was the case in Patient 2, surgery (unless otherwise contraindicated) should follow. Minimally invasive thoracotomy by means of a small incision anterolaterally should be preferable in patients who cannot undergo a sternotomy. Finally, clinicians should remember that prolonging anticoagulation, especially when it has had no effect on reducing the LAA mass, exposes patients to possible bleeding or repeat embolization.[5]

Consequent to our experience with these 2 patients, we recommend performing CMR as soon as cardiac masses have been identified during echocardiographic screening. In the absence of definitive diagnosis on multimodal images, appropriate patients should undergo surgery to resect the LAA mass. Our experience shows that resecting left-sided PFEs can lead to good postoperative outcomes, and we have seen no recurrence of PFE in these 2 patients or others since.

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Conflict of interest disclosure: Dr. Öner has received honoraria from Medtronic Inc.

References

1. Gowda RM, Khan IA, Nair CK, Mehta NJ, Vasavada BC, Sacchi TJ. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. Am Heart J 2003;146(3):404-10.
2. Nehaj F, Sokol J, Mokan M, Jankovicova V, Kovar F, Kubaskova M, et al. Outcomes of patients with newly diagnosed cardiac myxoma: a retrospective multicentric study. Biomed Res Int 2018;2018:8320793.
3. Abu Saleh WK, Al Jabbari O, Ramlawi B, Reardon MJ. Cardiac papillary fibroelastoma: single-institution experience with 14 surgical patients. Tex Heart Inst J 2016;43(2):148-51.
4. Tamin SS, Maleszewski JJ, Scott CG, Khan SK, Edwards WD, Bruce CJ, et al. Prognostic and bioepidemiologic implications of papillary fibroelastomas. J Am Coll Cardiol 2015;65(22):2420-9.
5. Jaber WA, Prior DL, Thamilarasan M, Grimm RA, Thomas JD, Klein AL, Asher CR. Efficacy of anticoagulation in resolving left atrial and left atrial appendage thrombi: a transesophageal echocardiographic study. Am Heart J 2000;140(1):150-6.
6. Brickner ME, Friedman DB, Cigarroa CG, Grayburn PA. Relation of thrombus in the left atrial appendage by transesophageal echocardiography to clinical risk factors for thrombus formation. Am J Cardiol 1994;74(4):391-3.
7. Yaghi S, Kamel H, Elkind MSV. Atrial cardiopathy: a mechanism of cryptogenic stroke. Expert Rev Cardiovasc Ther 2017;15(8):591-9.
8. Goette A, Kalman JM, Aguinaga L, Akar J, Cabrera JA, Chen SA, et al. EHRA/HRS/APHRS/SOLAECE expert consensus on atrial cardiomyopathies: definition, characterisation, and clinical implication. J Arrhythm 2016;32(4):247-78.