Clinical Image

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Papillary fibroelastoma: an unexpected finding on the aortic valve

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A 50-year-old man presented in May of 2020 with a chief complaint of chest pain. His past medical history was significant for tobacco use, hypertension, and angioplasty to D1 in 2011, along with a drug-eluting stent to the left circumflex coronary artery in 2014. The patient was not taking any medications. He was hypertensive to 210/120 mmHg and was admitted for hypertensive emergency with myocardial ischemia. His pain resolved with lowering of his blood pressure.

An electrocardiogram revealed inferolateral T wave inversions. Troponin T peaked at 1.67 ng/mL. Echocardiography revealed an ejection fraction of 40–45% with apical hypokinesis, mildly-aneurysmal changes of the basal inferior wall, and prominent trabeculation apically and inferolaterally. A spherical, shimmering, echodensity on the aortic valve measuring 1×0.8 cm was also seen. Differential diagnosis included a bacterial vegetation, thrombus, cardiac tumors such as a papillary fibroelastoma (PFE) or myxoma, and noninfectious etiologies such as Libman–Sacks endocarditis. However, given the patient's lack of autoimmune disease history, a primary cardiac tumor or infectious vegetation were favored. Blood cultures were negative, and the patient did not have any stigmata of endocarditis.

A decision was made to pursue computed tomography angiography (CTA) of the coronary arteries as the initial study in the evaluation of the patient's myocardial ischemia, due to the risk of potential catheter-associated embolization of the echodense structure on the aortic valve. The CTA was significant for triple-vessel coronary artery disease, after which a left heart catheterization was performed. A preoperative transesophageal echocardiogram (TEE) showed, again, the echodensity on the ventricular side of the aortic valve (Image A). There was no evidence of aortic regurgitation on color Doppler interrogation. The patient then underwent coronary artery bypass graft surgery, and the aortic valve mass was excised. Pathology revealed a papillary fibroelastoma (Image B). The patient tolerated the surgery well and no complications were seen at his postoperative office visit two weeks later. He has since been lost to follow up.

Cardiac tumors are rare, with prevalence of 0.02% [1]. The vast majority of these, estimated to be up to 75% of cardiac tumors, are benign and primarily of the myxoma subtype [1]. Papillary fibroelastomas comprise 5% of these benign tumors [1, 2]. A previous echocardiographic study [3], however, favored papillary fibroelastomas as more common than myxomas. Discrepancy in these findings may be due to how the tumors were diagnosed. The prevalence estimate given by McAllister and Fenoglio [2] was based on autopsy findings, whereas Tamin et al. [2] showed results from many patients who were diagnosed on echocardiogram [2, 3]. The findings of Hoffmeier et al. [1] also favored higher prevalence of myxoma compared to papillary fibroelastoma; those tumors were diagnosed in living patients as well.

Clinical manifestations occur due to embolism and include entities such as cerebrovascular accident, myocardial ischemia, and pulmonary embolism if a patent foramen ovale is present. Evaluation of a potential PFE should be initiated with a transthoracic echocardiogram (TTE), but TEE may be considered. Papillary fibroelastomas typically grow on the valves, lending higher potential yield from TEE [4]. They can also cause ventricular obstruction leading to manifestations of left or right heart failure [5]. Papillary fibroelastomas are typically detected on echocardiogram, on which they are characterized as frond-like lesions with speckled appearance. Histologically, they are comprised of endothelial cells around a core of connective tissue [6]. When investigating a possible PFE, testing for bacterial endocarditis, lupus, and antiphospholipid syndrome should be done [3].

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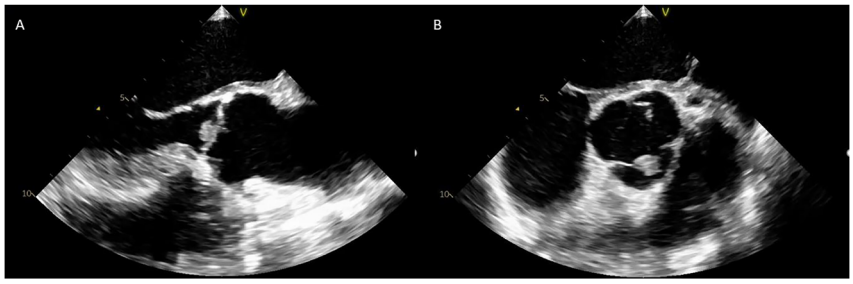

Image A: The patient's transesophageal echocardiogram from long (A) and short (B) axis views showed a spherical, shimmering, echodensity on the aortic valve.

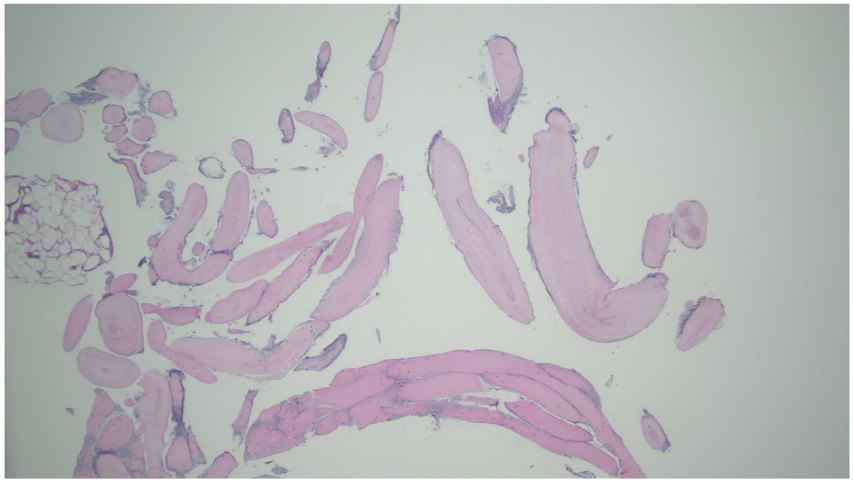

Image B: Pathology from the patient's aortic valve mass showed papillary fronds with central core of fibroconnective tissue surrounded by endothelial cells, consistent with a papillary fibroelastoma.

Recommendations for the management of PFE vary. Even PFE found incidentally can go on to cause significant morbidity due to their embolic risk. There is little controversy in managing symptomatic patients with surgical excision, weighing the surgical candidacy and other indications for surgery in each patient. For asymptomatic patients, ≥1 cm has been used as a threshold for surgical excision, particularly if the mass is mobile due to increased embolic risk. Tumor mobility has been shown to be an independent risk factor for nonfatal embolization and death [5, 6].

The exact follow-up protocol for nonoperative management is unclear. Routine follow up for small (<1 cm), left-sided nonmobile lesions may be performed until either the patient develops symptoms or the mass meets aforementioned size criteria [6, 7]. With this strategy, echocardiography would be the most useful modality for following asymptomatic patients. However, the ideal frequency of imaging is unclear and there are no guidelines for whether TTE or TEE should be used. Transesophageal echocardiography may be considered when a suspected PFE is not well-visualized, specifically to improve measurement and mobility. Anticoagulation has been suggested for poor surgical candidates though there are no guidelines and limited data to support this [7].

Another approach suggested by Tamin et al. [3] allows for a lower threshold for excision in good surgical candidates. With this strategy, all left-sided PFE, irrespective of size or mobility, can be excised [3]. The lack of data from randomized, controlled studies comparing the conservative and invasive management of asymptomatic PFE presents a dilemma. Available data argues that surgical excision has good outcomes, including a low rate valve compromise [3, 8]. The guidelines from the American Society of Echocardiography [4] described a more conservative strategy of close follow up for small, sessile, tumors. However, the authors cautioned against strictly following these recommendations [4]. The decision to proceed with surgical excision vs. close follow up should be made on a case-by-case basis. Each individual patient's surgical candidacy, risk for embolization, willingness to proceed with surgical excision, and other indications for cardiac surgery should be considered.

In summary, papillary fibroelastomas are uncommon, benign, cardiac tumors that carry a risk of thromboembolic complications. There are no established guidelines for the management of PFE, particularly in asymptomatic patients. Surgical excision has shown a favorable outcome. It should be considered even in asymptomatic patients who have a relatively low risk of embolism based on size and mobility of the lesion, weighing both the patient's surgical candidacy and the provider's or institution's expertise level. Further studies are needed to compare outcomes of a conservative surveillance strategy vs. early surgical excision.

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