Papillary fibroelastoma of the pulmonary valve: a 14-year follow-up

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Abstract

Primary cardiac tumors are exceedingly rare. Among them, papillary fibroelastomas (PFEs) are the third most common. These tumors are often incidental findings but can also precipitate a myriad of clinical presentations, mainly embolic events. Most common in the left-sided valves, PFEs rarely occur in the right side of the heart. They are usually resected surgically following diagnosis, thwarting assessment of their natural history. We present the case of a woman diagnosed with pulmonary valve PFE following recurrent pulmonary embolism who did not undergo surgery, allowing for an extended follow-up of the condition.

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Abstract

Primary cardiac tumors are exceedingly rare. Among them, papillary fibroelastomas (PFEs) are the third most common. These tumors are often incidental findings but can also precipitate a myriad of clinical presentations, mainly embolic events. Most common in the left-sided valves, PFEs rarely occur in the right side of the heart. They are usually resected surgically following diagnosis, thwarting assessment of their natural history. We present the case of a woman diagnosed with pulmonary valve PFE following recurrent pulmonary embolism who did not undergo surgery, allowing for an extended follow-up of the condition.

Introduction

Primary cardiac tumors are exceedingly rare, with an estimated prevalence between 0.0017% and 0.028%[1-3] in autopsy series and 0.15% in echocardiographic series.[2] Among these tumors, papillary fibroelastomas (PFEs) are the third most common, accounting for less than 10% of them.[4-7] These pedunculated lesions

chiefly grow in cardiac valves, the most common heart tumors in this location.[7] The pulmonary valve is least commonly affected.[8] Although histologically benign and most often found incidentally, PFEs can precipitate life-threatening conditions, usually embolic events, but also heart failure and sudden cardiac death.[2,8] Surgical tumor removal is considered curative, and a plurality of patients undergo tumor excision.[8] This approach, however, has dampened attempts to elucidate their natural history. We present the case of an older woman with recurrent pulmonary emboli attributable to a pulmonary valve papillary fibroelastoma (PVPFE) who did not undergo surgery and had a 14-year follow-up.

Case report

A 74-year-old woman presented to a local hospital in 2008 with shortness of breath and chest pain. Computerized tomography showed filling defects of the pulmonary artery branches and a small pulmonary valve nodule that was not initially recognized as pathological (Fig. 1). The patient was diagnosed with pulmonary embolism (PE) and treated with anticoagulation therapy for six months.

She presented back in 2016 with recurrent symptoms attributable to another pulmonary embolus. A repeat CT confirmed the diagnosis of PE and revealed growth of the pulmonary valve nodule. The nodule measured 8mm and was attached to the pulmonary valve by a stalk (Fig. 2). A presumptive diagnosis of pulmonary valve papillary fibroelastoma was made. Surgical resection of the lesion was considered. However, the patient deferred surgical intervention, partly due to her several comorbidities, including advanced age, frailty, and multiple orthopedic issues such as numerous back and cervical spine surgeries. Instead, the patient was commenced on long-term anticoagulation with warfarin.

Her latest follow-up was in 2022, with a CT scan showing further growth of the pedunculated nodule (Fig. 3). It now measured 20mm, representing a 2mm/year growth rate. Transthoracic and transesophageal echocardiograms were performed, further delineating the PVPFE (Figs. 4 and 5). They showed normal valve function. The patient had not had any other embolic events since 2016 and elected to maintain anticoagulation without surgical intervention.

Discussion

PFEs are rare tumors that can paint a challenging clinical picture. Their infrequent nature combined with variable manifestations demands clinicians to maintain a high suspicion index for them. In our case, even with proper imaging, the tumor was not immediately recognized. Imaging is the mainstay of diagnosis, particularly echocardiograms, which allow for evaluation of the mass and assessment of valve function.[9]

When imaged, PFEs usually present as round, oval, or irregular nodules. Sun et al. have reported that about half of them have stalks, which are associated with increased tumor mobility.[10] These tumors are best evaluated with TTE or TEE, as CT and MRI may fail to detect smaller tumors. However, CT and MRI may still have a role in diagnosing neoplasms in atypical locations and detecting concomitant extracardiac disease.[9] Increasing imaging quality and application of new techniques should improve sensitivity for these exams.[9,11] They can also help evaluate a patient with established PFE for preoperative planning or assessment of alternate diagnoses.

The differential diagnosis of PFEs encompasses vegetations, thrombi, and other cardiac tumors - mainly myxomas.[8,11] Compared to PFEs, myxomas are more likely to be located in the right atrium, be larger, and cause symptoms.[9] Thrombi can be differentiated by their laminated appearance, irregular contours, and absence of a pedicle.[8,11] Vegetations can be similar to PFEs in imaging; however, patients usually have clinical signs of endocarditis, valvular dysfunction or destruction, and evolving lesions and clinical course. Other less common differential diagnoses include Libman-Sacks vegetations and Lambl's excrescences.[8]

The pathogenesis of PFEs remains unknown. On the one hand, some believe they are reactive processes to trauma and hemodynamic stress, while on the other hand, others have postulated them to be hamartomas.[9,12] The presence of other tumors in patients with PFEs has been reported, but the meaning of this association is unknown.[12]

Histologically, PFEs are small and highly papillary masses often attached to cardiac tissue through a stalk. This appearance has led some researchers to liken them to sea anemones.[5,10,13] They are also firmly attached to the endocardium, and embolization may occur from its fragile papillary fronds or a thrombus formed on its surface rather than dislodgement of the tumor itself.[8]

PFEs are usually found incidentally during cardiac surgery, imaging, or autopsy. Their management usually involves surgical resection to prevent complications.[8] This has made it hard to assess their natural history and the effectiveness of medical treatment to prevent complications such as embolization. This is especially true for PVPFEs since the pulmonary valve is one of the least common sites for the development of PFEs, representing only 8% of cases.[8]

Our patient, an older woman with multiple comorbidities, elected not to have her tumor excised. The 14-year follow-up she received is, to the best of our knowledge, the longest described follow-up of a patient with PVPFE in the literature. This allowed us to highlight the natural history of PVPFEs and their growth rate. Our findings suggest that PFEs may have a variable rate of growth than the previously described growth rate of 0.5 ± 0.9 mm/year.[14]

Conflicts of interest

All authors declare that they have no conflicts of interest.

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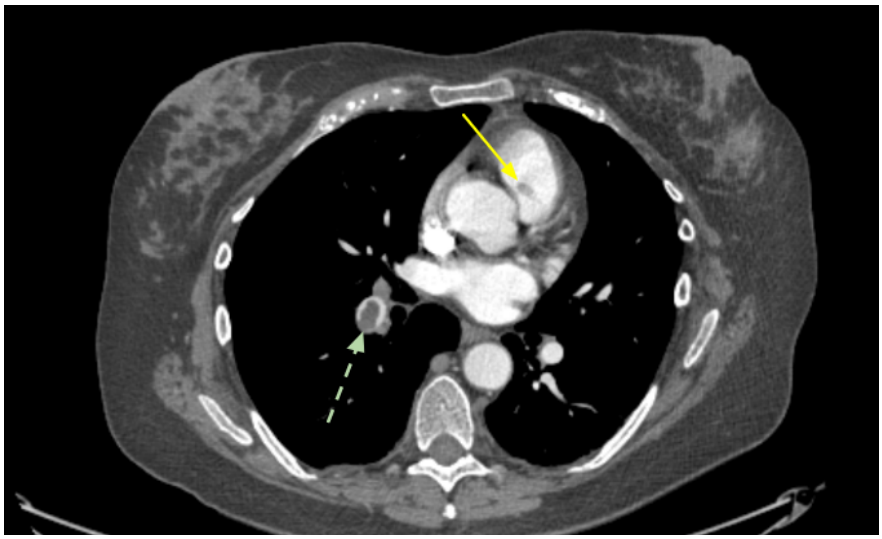

Fig. 1 . 2008 CT scan showing filling defects in the pulmonary artery branches (green dashed arrow), confirming the diagnosis of pulmonary embolism. A small filling defect can be noted in the RV outflow tract, representing a small PVPFE (yellow solid arrow).

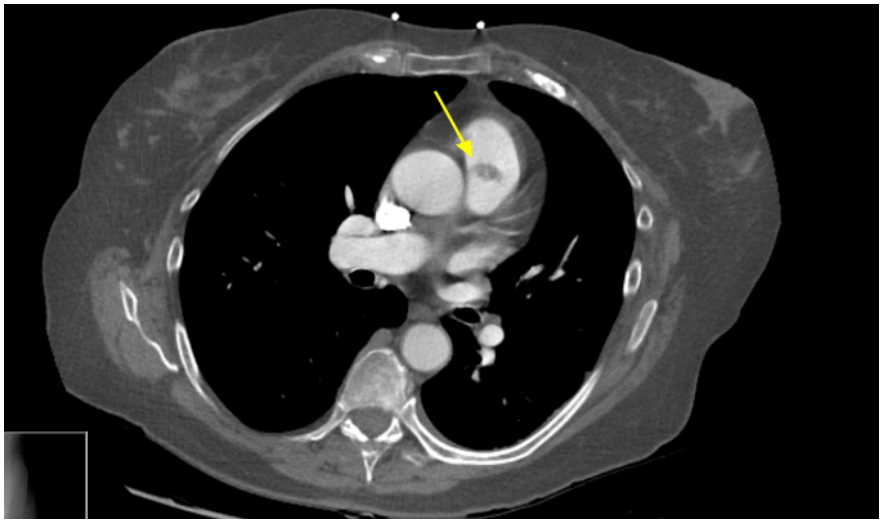

Fig. 2. 2016 CT scan showing an 8mm nodule with a stalk in the RV outflow tract (arrow), consistent with a PVPFE.

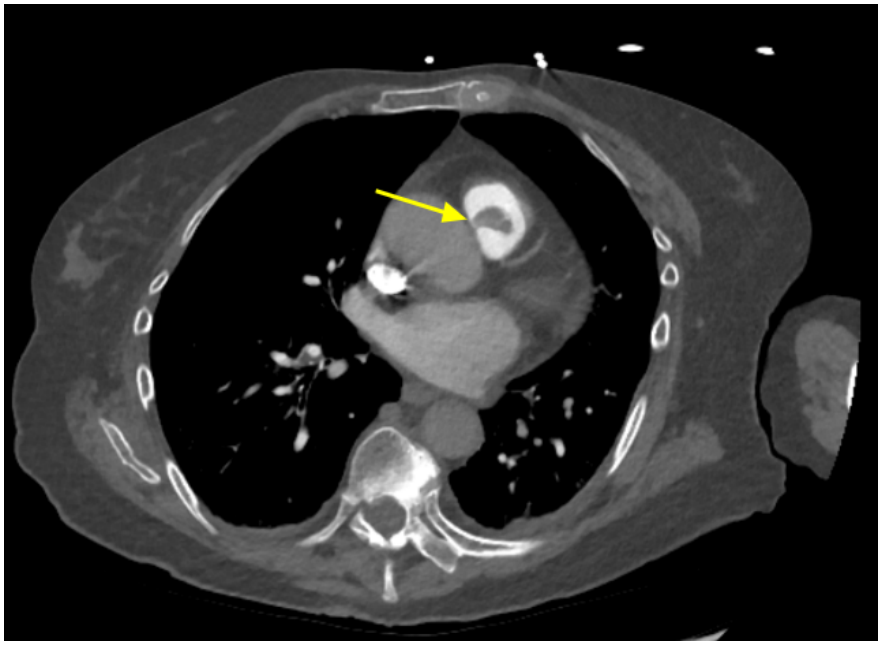

Fig. 3. 2022 CT scan showing further growth of the PVPFE (arrow), now measuring 20mm.

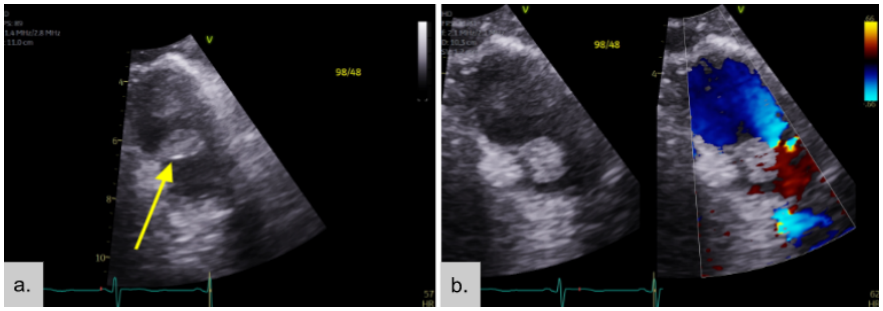

Fig. 4. a. Transthoracic echocardiogram showing the 20mm nodule attached to the pulmonary valve through a stalk (arrow). **b.** 2-D Color Flow Doppler showed normal valve function.

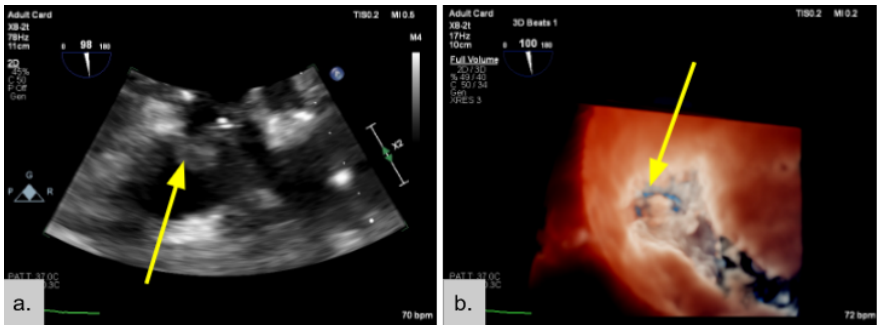

Fig. 5. Transesophageal echocardiogram (TEE) further delineating the PVPFE (arrows) in 2D (**a.**) and 3D (**b.**).