Pulmonary artery aneurysm associated with a non-stenotic bicuspid pulmonic valve: a role for genetics?

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Abstract

Background Bicuspid pulmonic valves are quite uncommon, being described in only 0.1% of donor hearts, while pulmonary artery aneurysms are even more rare, having been found in 8 out of 109,571 autopsies. This rarity makes it difficult to characterize the relationship between them. Materials & Methods We describe the case of a 66-year-old female who was found to have a bicuspid pulmonic valve and pulmonary artery aneurysm (5.1cm) on imaging by her cardiologist. Discussion & Conclusion This case raises the question of whether the association between bicuspid semilunar valve disease and vascular wall anomalies are more genetic or hemodynamic. Even on the aortic side, despite the robust association between bicuspid aortic valves and thoracic aortic aneurysms, the mechanism still remains unclear. In our patient there was no significant gradient across the bicuspid pulmonic valve, suggesting that hemodynamics are not the primum mobile of this association.

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Pulmonary artery aneurysm with bicuspid valve

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Background

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Materials & Methods

We describe the case of a 66-year-old female who was found to have a bicuspid pulmonic valve and pulmonary artery aneurysm (5.1cm) on imaging by her cardiologist.

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This case raises the question of whether the association between bicuspid semilunar valve disease and vascular wall anomalies are more genetic or hemodynamic. Even on the aortic side, despite the robust association between bicuspid aortic valves and thoracic aortic aneurysms, the mechanism still remains unclear. In our patient there was no significant gradient across the bicuspid pulmonic valve, suggesting that hemodynamics are not the *primum mobile* of this association.

Introduction

A 66-year-old female (BMI 32.61kg/m^2) with a past medical history of hypertension, hyperlipidemia, type 2 diabetes mellitus, hypothyroidism, anxiety, obstructive sleep apnea, osteoarthritis, mechanical falls, paroxysmal atrial fibrillation/flutter (not on anti-coagulation) status post pulmonary vein isolation (2008) with repeat ablation/isolation (2014, 2016) and left atrial appendage (LAA) occlusion (2017) presented to her cardiologist for a routine visit where she underwent transthoracic echocardiography (TTE) and was found to have a pulmonary artery aneurysm (PAA). Her PAA measured 5.1cm distal to the pulmonic valve, and pulmonary artery (PA) systolic pressure was estimated to be 55mmHg. The aortic valve and root/ascending aorta size were measured as normal.

On exam, our patient lacked pathologic features of a connective tissue disorder, and she noted no family history of such. Her auscultatory exam did not reveal any murmurs/gallops that would suggest valvular disease. She denied angina, dyspnea, orthopnea, paroxysmal nocturnal dyspnea, syncope, and change in exercise tolerance, but endorsed occasional episodes of palpitations and right-sided chest pain (attributed to acid reflux).

Materials and Methods

The PAA was diagnosed by TTE, but it was unclear what the growth pattern and etiology were. We were able to retrospectively follow the growth of the aneurysm because she had multiple computed tomography scans for electrophysiologic procedure planning. International Review Board, consent statement, and clinical trial registration were not applicable to deidentified report of her case.

Results & Discussion

In 2014, the PA was 4.2 cm, in 2016 it was 4.6 cm, and in 2018 it was 4.7 cm, despite the absence of any hemodynamically significant pulmonary stenosis. Transesophageal echocardiogram from 2018 obtained for LAA occlusion device monitoring noted EF 65% with normal biventricular function, a trileaflet aortic valve, and normal Sinus of Valsalva (2.9cm)/ascending aorta (3.1cm). It did not have descriptors of PA pressures/size. Follow-up computed tomography angiography (CTA) reported a bicuspid pulmonic valve without stenosis and PA dilation to 4.7cm (Figures 1, 2).

While the risk of rupture and dissection with PAAs is less than that of aortic aneurysms, it has been reported. Therefore, care should be taken to monitor those with a >2mm increase in size/year, a PA pressure >50mmHg, and a diameter >7.5cm.[1] PAAs that become large enough undetected are eventually

found due to sequelae of compression of nearby structures.[2] Indications to intervene on a bicuspid valve if stenotic include a PA systolic pressure gradient >64mmHg and right ventricular dysfunction.[1]

Although our patient presented without symptoms that could be directly attributed to a bicuspid pulmonic valve and PAA, symptomatic patients can exhibit exertional dyspnea, weakness, cough, and hemoptysis.[2,3] In their case series of 7 patients with isolated bicuspid disease and PAA, Izumida et al found that over half were asymptomatic with diagnosis made on investigation of other diseases.[1]

Because the patient did not meet size criteria for operative intervention and because symptomatology was not thought to be direct sequelae of physiologic/mass effect of the PAA, the decision was made to continue surveillance with imaging.

Conclusions

Causes of PAAs have been divided into those associated with Eisenmenger's physiology, pulmonic valve abnormalities, and connective tissue disorders.[4] Gupta et al also included infectious causes such as tuberculosis (Rasmussen's aneurysm), vasculitis, and endocarditis.[4] This rare entity has been associated with congenital defects in almost half of cases.[2] The pathophysiology of PAAs remains poorly understood, especially in the absence of connective tissue disorders.[2] This case report describes the rare incidence of a normally functioning, non-stenotic bicuspid pulmonic valve and a PAA, suggesting a developmental association versus a hemodynamic one.

Bicuspid pulmonic valves are quite uncommon, being described in only 0.1% of donor hearts, while PAAs are even more rare, having been found in 8 out of 109,571 autopsies.[1,5-7] This rarity makes it difficult to characterize relationships. Even on the aortic side, despite the robust association between bicuspid aortic valves and thoracic aortic aneurysms, the mechanism still remains unclear. It has been postulated that with a bicuspid valve, there is an increase in matrix metalloproteinase 2 activity and a decrease in fibrillin-1 that leads to dilation.[8] Fedak et al found this decrease in fibrillin-1 to also be present in PAs of patients with bicuspid aortic valves independent of valvular function.[8] Abnormal neural crest cell migration could also be a culprit since neural crest cells are involved in both semilunar valve and vascular wall formation.[1]

Previously noted relationships between bicuspid valves and PAAs has been attributed to post-stenotic dilation.[3] However, in our patient there was no significant gradient across the bicuspid pulmonic valve, suggesting that hemodynamics are not the *primum mobile* of this association and that progression can occur in the absence of significant pulmonic stenosis. Furthermore, the association between PAAs and bicuspid *aortic* valves adds additional weight to a common developmental abnormality, along with the more recognized relationship between bicuspid aortic valves and aortopathies.[1,9]Further studies are needed to determine if the incidental finding of a bicuspid pulmonic valve warrants routine imaging for the development of a PAA, but given the continued enlargement of the PAA in our patient, we will perform surveillance imaging.

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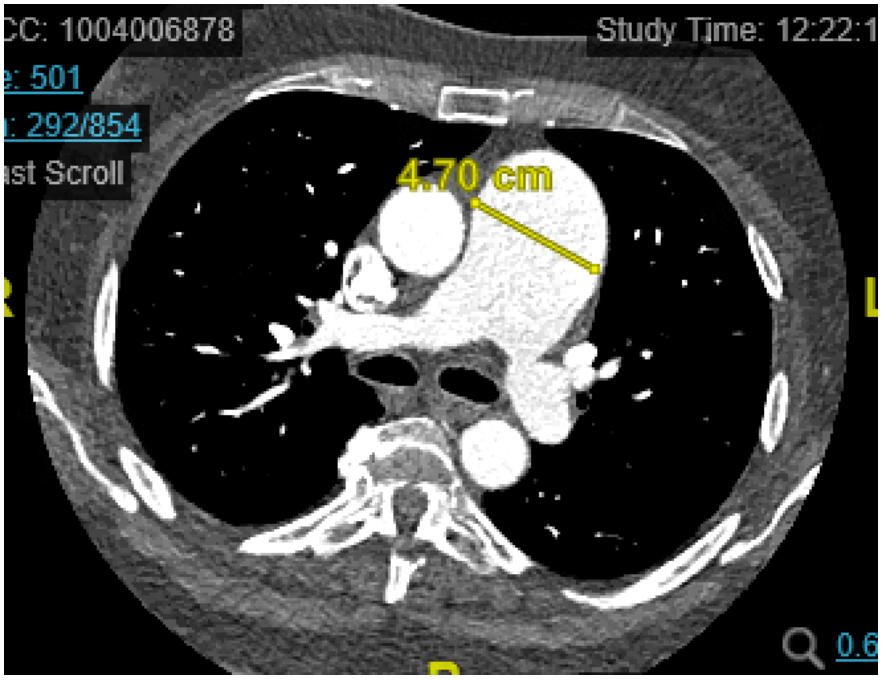
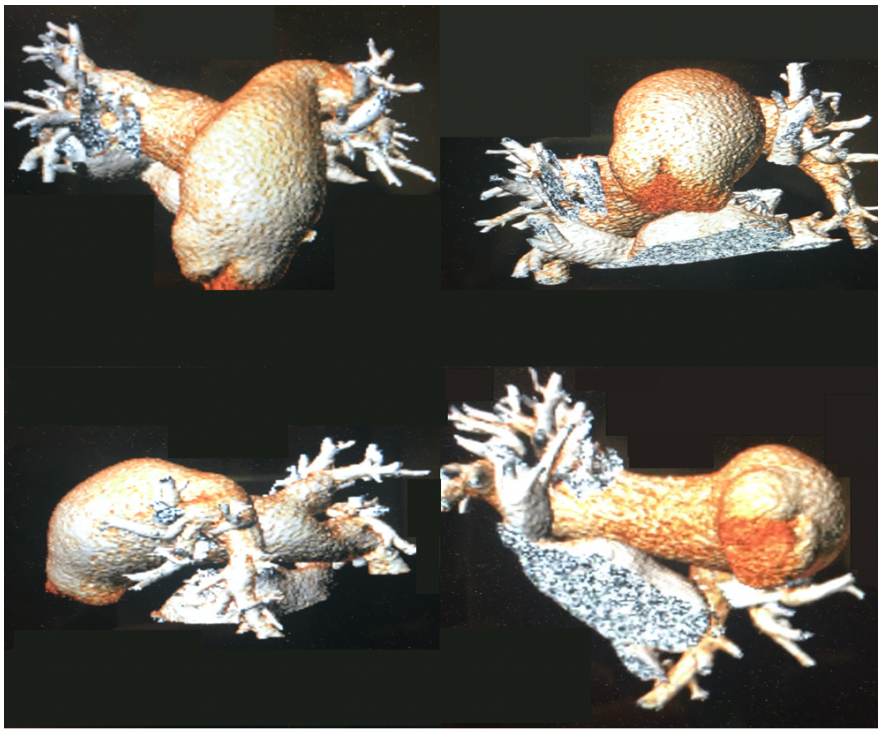
Figures

Figure 1. Axial CTA



Axial CTA demonstrating the main pulmonary artery aneurysm (PAA) at 4.7cm, which is 0.5cm growth from 4 years prior.

Figure 2. Pulmonary Artery Reconstruction



Reconstruction demonstrating the bicuspid pulmonic valve and main pulmonary artery aneurysm (PAA).