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MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

BEGINNER

IMAGING VIGNETTE: CLINICAL VIGNETTE

Giant Pulmonary Artery Aneurysm in Bicuspid Pulmonary Valve

Does the Right Side Mimic the Left One?

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ABSTRACT

Bicuspid pulmonary valve is a rare echocardiographic finding, particularly if not associated with other congenital heart diseases. We report the incidental case of a severe giant pulmonary arterial aneurysm associated to bicuspid pulmonary valve in an asymptomatic 79-year-old patient. Multimodality cardiac imaging was important for the correct diagnosis and to exclude any other potential complication. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:1720–2)

Aortic and pulmonary valves are known to stem from the same embryonic arterial trunk, and this could explain the similar presentation of some types of congenital defects in both of the 2 valves. However, due to the limited number reported, more in autoptic analysis, little is known about the effective risk and management of pulmonary abnormalities. Multimodality cardiac imaging is mandatory to better define the valve morphology and to rule out related complications. Overall, surgery remains the cornerstone of therapy, albeit definite surgical thresholds are still lacking.

CASE REPORT

A 79-year-old patient presented to the emergency department for fever and chest pain, described as breathing and cough variable and almost continuous for weeks. No previous cardiovascular history was known, except for arterial hypertension treated with calcium antagonist therapy. A transthoracic echocardiogram performed to investigate a systolic murmur showed a moderately stenotic pulmonary valve and severe pulmonary artery aneurism (PAA) (**Figures 1A and 1B**, Videos 1 and 2). Noninvasive estimation of pulmonary artery pressure resulted in the normal range. The chest pain was explained by a pleuritic inflammation detected at chest x-ray, and a urine test revealed an infection treated with antibiotics. A computed tomography (CT) scan was urgently performed to rule out any acute emergency, although the patient never reported any symptoms related to the PAA compression. The CT and the 3-dimensional CT reconstruction confirmed these data (**Figures 1C and 1D**, Video 3), as far as the magnetic resonance imaging (**Figure 1E**, arrow; Videos 4, 5, 6, and 7).

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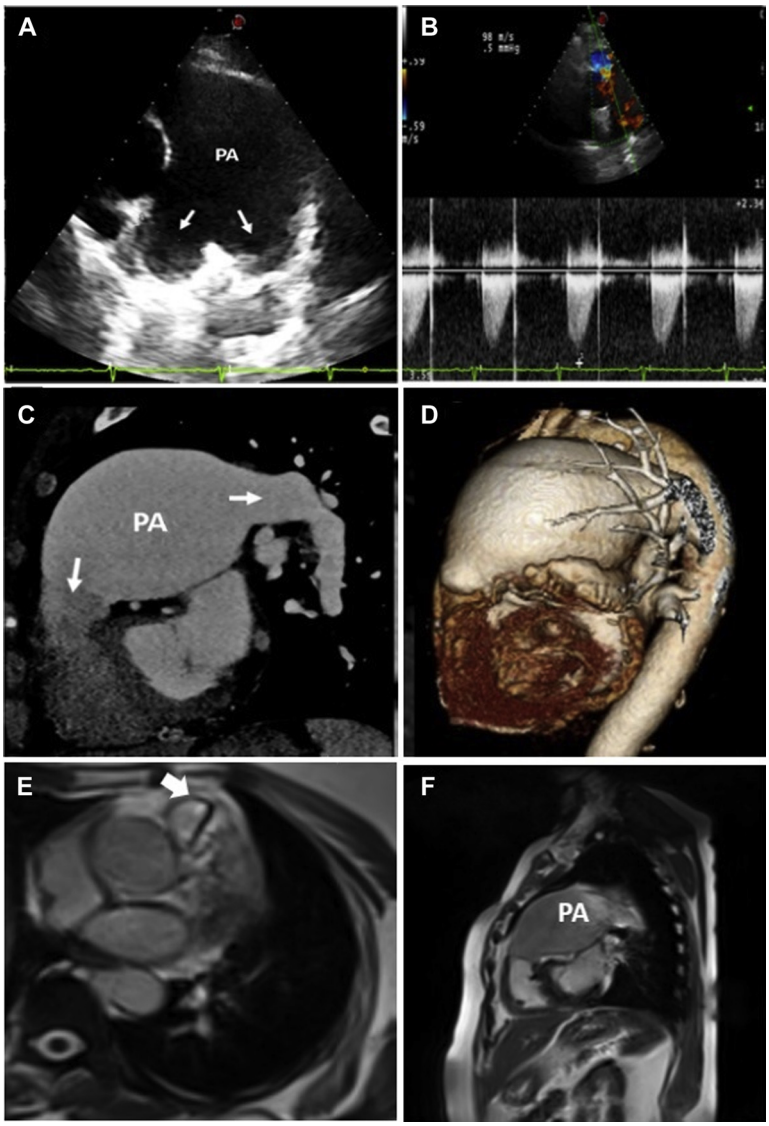
Balancing potential risks and benefits, corrective heart surgery had been proposed to the patient, but was rejected in the absence of symptoms. Currently, after 3 years of follow-up, she is asymptomatic, taking bisoprolol 5 mg/die and routinely transthoracic echocardiogram has been scheduled every 6 months.

ABBREVIATIONS AND ACRONYMS

CT = computed tomography scan

PAA = pulmonary artery aneurysm

FIGURE 1 Patient's TTE, CT, and CMR



(A and B) Patient's transthoracic echocardiogram (TTE). The pulmonary trunk was severely dilated (maximum diameter of 6.39 cm), with severe dilatation of the pulmonary arteries (2.97 and 2.8 cm, respectively). The pulmonary cusps appeared myxomatous and calcific. The maximum velocity recorded was 2.98 m/s with a maximum gradient >35 mm Hg. **(C and D)** Patient's computed tomography scan (CT). The CT confirms the presence of a severe dilation of the PA trunk (6.7 × 5.9 cm), right pulmonary artery (3.8 × 3.5 cm), and left pulmonary artery (3.6 × 3.4 cm) with no sign of dissection of the wall. The pulmonary valve appeared morphologically bicuspid. The 3-dimensional CT reconstruction confirmed the severe dilatation of the pulmonary trunk and pulmonary arteries. **(E and F)** Patient's cardiac magnetic resonance imaging (CMR). The pulmonary valve appeared morphologically bicuspid, and the presence of a severe dilation of the PA trunk and pulmonary arteries was confirmed.

DISCUSSION

Bicuspid pulmonary valve is an exceedingly rare finding usually associated with PAA that could result as the consequence of chronic stenosis of the valve. Few sporadic cases of PAA have been reported until now, especially concerning autoptic analysis, but such a giant PAA with bicuspid pulmonary valve in an asymptomatic woman has never been described. Moreover, as demonstrated here, a multimodality imaging approach was crucial to better define the morphology of the pulmonary valve, to exclude any other congenital defects, such as conotruncal anomalies, and to rule out any associated complications. Different cutoffs for PAA have been proposed, from a range of 26.9 mm in women (1) to 29 mm (2) in the general population, and an absolute diameter ≥55 mm has been designated for surgical intervention (3); so our case, where PAA was 64 mm, was clearly enclosed in these ranges. Overall, surgery remains the cornerstone of these pulmonary abnormalities; however, there are no clear guidelines yet on the optimal management of these patients (4). In our case, the absence of complications and symptoms has leaded the patient to give up the proposed corrective surgery, so we have planned a follow-up every 6 months. Moreover, albeit not clearly specified, a pharmacological treatment with beta-blockers has been introduced to reduce vascular wall stress, as is usually also done for the aneurisms of the aortic tract.

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KEY WORDS bicuspid pulmonary valve, congenital disease, giant aneurism, pulmonary artery

APPENDIX For supplemental videos, please see the online version of this paper.