

[ CASE REPORT ]

## Pulmonary Artery Aneurysm Associated with Bicuspid Pulmonary Valve

Seiya Izumida, Hiroaki Kawano, Akira Tsuneto, Yoshiyuki Doi and Koji Maemura

### Abstract:

A 73-year-old Japanese man was admitted with an asymptomatic pulmonary artery aneurysm. However, chest X-ray and contrast-enhanced thoracic computed tomography revealed a protrusion at the second left branch that in fact was a pulmonary artery aneurysm with a diameter of 50 mm. Transesophageal echocardiography showed a bicuspid pulmonary valve, and cardiac catheterization revealed pulmonary stenosis with a pressure gradient of 45 mmHg, but no other heart diseases were noted. An extremely rare pulmonary artery aneurysm associated with an isolated bicuspid pulmonary valve was diagnosed.

**Key words:** anomaly, congenital heart disease, systolic murmur

(Intern Med 59: 1867-1871, 2020)

(DOI: 10.2169/internalmedicine.4415-20)

### Introduction

Pulmonary artery aneurysms (PAAs) are rare, having been identified in only 8 of 109,571 autopsies. They generally occur in younger persons compared with other types of aneurysms with an equal sex incidence (1, 2).

Several pathologies are related to PAAs, and pulmonary valve stenosis is one cause of PAAs due to dilation after stenosis (3). Although pulmonary valve stenosis is not unusual in terms of congenital heart diseases, isolated bicuspid pulmonary valves without other congenital abnormalities are rare (4).

We herein report an extremely rare PAA with an isolated bicuspid pulmonary valve.

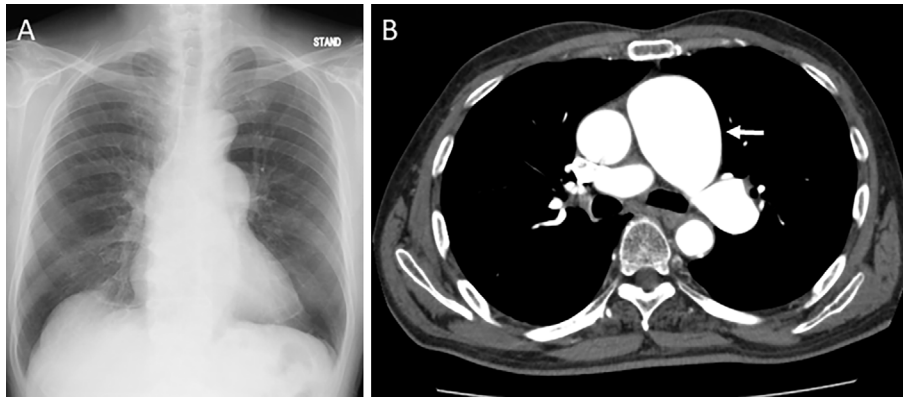
### Case Report

A 73-year-old Japanese man was admitted with a PAA. Pulmonary emphysema associated with smoking (cigarette 30/day for 35 years) had been pointed out one year before. His family history showed no remarkable disease, including cardiovascular diseases, infectious diseases, and autoimmune diseases. Although a heart murmur had been identified at a medical checkup several years previously, it had not been further assessed. Chest X-ray during a routine medical

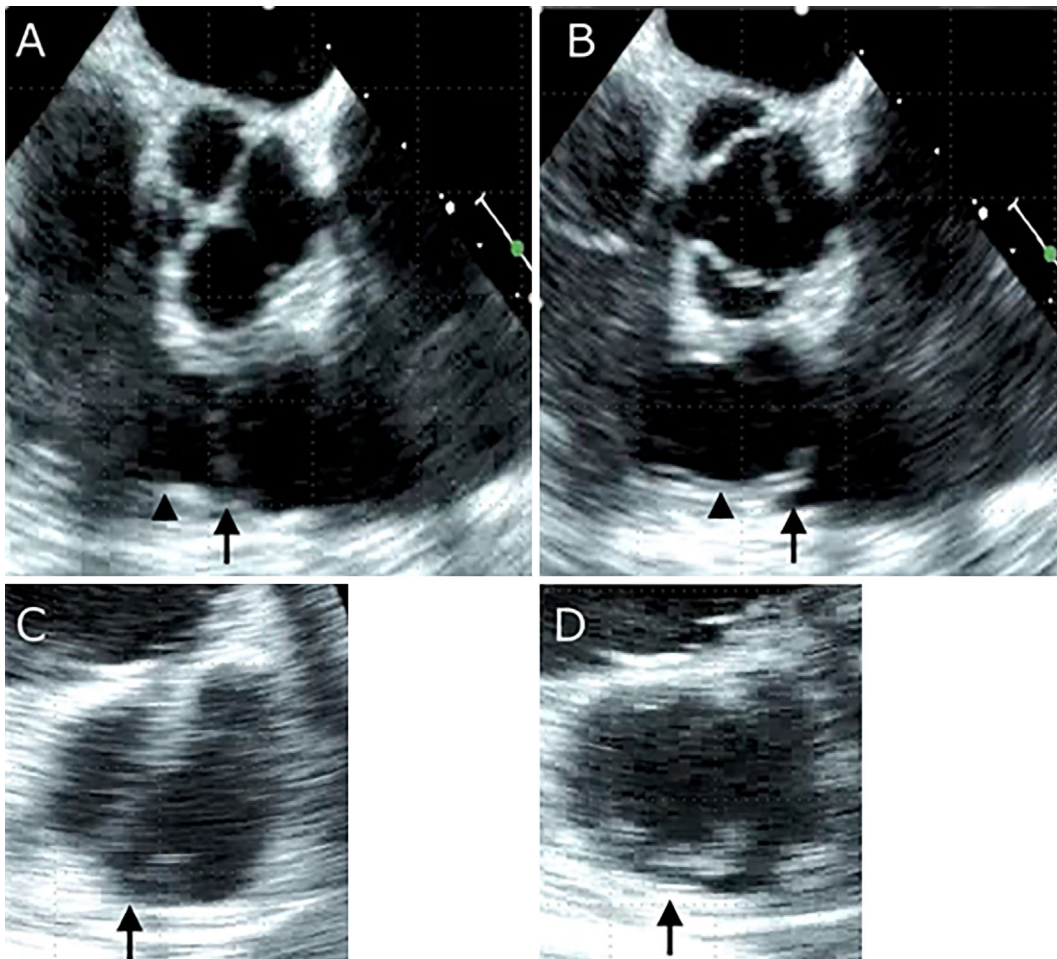
checkup at 72 years old had shown a round mass in the left middle lung field (Fig. 1A). Contrast-enhanced chest computed tomography (CT) 6 months ago revealed a PAA with a diameter of 50 mm (Fig. 1B). He had had no symptoms, such as dyspnea or chest pain, when he worked as a fisherman.

A physical examination upon admission showed height 155 cm, body weight 56.6 kg, blood pressure 138/75 mmHg, heart rate 74/min, and oxygen saturation 95% on room air. A Levine grade 3 systolic murmur was heard with normal respiratory sounds, and he had no hepatosplenomegaly and no leg edema. He also had no skin lesions, aphtha, or arachnodactyly. We consulted with an ophthalmologist, who reported no abnormal findings in his eyes, including lens dislocation and uveitis.

Laboratory data were normal, including N-terminal pro brain natriuretic peptide (NT-pro BNP) 45.8 pg/mL, C-reactive protein 0.09 mg/dL, anti-nuclear antibody <20,  $\beta$ -D-glucan 5.4 pg/mL (normal <20), and the T-SPOT test (-). Electrocardiography findings were normal without right heart overload. Transthoracic echocardiography (TTE) showed a normal left ventricular systolic function, no right-sided cardiac overload, pulmonary valve stenosis with mosaic flow in the pulmonary valve, and peak systolic velocity of transpulmonary flow of 3.4 m/s, indicating a mean systolic pressure gradient in the transpulmonary valve of 45



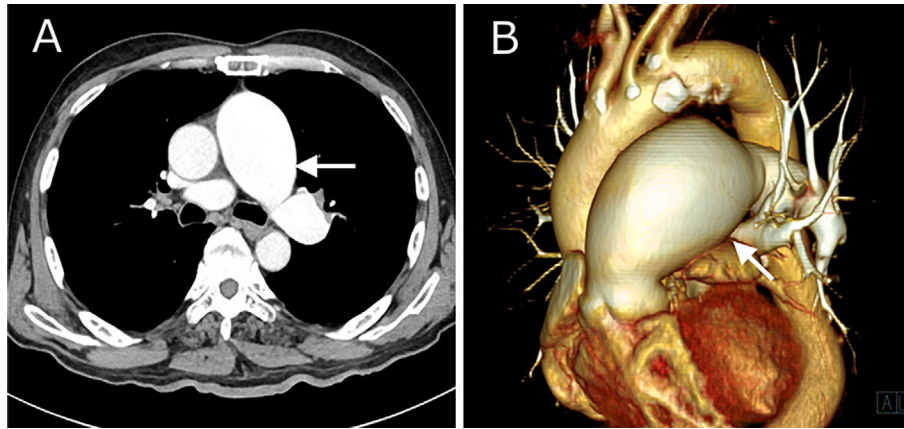
**Figure 1.** Chest radiography findings one year ago and contrast-enhanced chest computed tomography findings six months ago. (A) Chest radiography shows protrusion of the left second branch. (B) Contrast-enhanced chest computed tomography shows pulmonary artery aneurysm with a diameter of 50 mm (arrow).



**Figure 2.** Pulmonary valve views and transesophageal echocardiography findings. The long-axis view of the pulmonary valve in the diastolic (A) and systolic (B) phases. The short-axis view of the pulmonary valve in the diastolic (C) and systolic (D) phases. Transesophageal echocardiography shows systolic doming of pulmonary valve (arrow) in the long-axis view (B), no pulmonary valve calcification, bicuspid pulmonary valve in short-axis views (C and D), and no stenosis of the right ventricular outflow (B, arrowhead).

mmHg. Transesophageal echocardiography (TEE) confirmed these findings and revealed systolic doming of the pulmo-

nary valve in the long-axis view, no pulmonary valve calcification, and a bicuspid pulmonary valve in the short axis



**Figure 3.** Contrast-enhanced chest computed tomography findings on admission. (A) Axial section. (B) Three-dimensional image shows pulmonary artery aneurysm with diameter of 50 mm (arrow).



**Figure 4.** Right ventriculography findings. Image shows doming of systolic pulmonary valve and jet flow to the pulmonary artery.

(Fig. 2). Planimetry showed that the area of the pulmonary valve was  $1.7 \text{ cm}^2$ , but cardiac shunt diseases were undetectable.

The PAA findings on contrast-enhanced CT were the same as those from six months earlier (Fig. 3). Pulmonary ventilation perfusion scintigraphy did not reveal a pulmonary artery embolism. Cardiac catheterization showed normal systolic, diastolic, and mean pulmonary artery pressures of 31, 11, and 18 mmHg, respectively, and the peak-to-peak pressure gradient from the pulmonary artery to the right ventricle was 45 mmHg. Right ventriculography showed systolic pulmonary valve doming and jet flow to the pulmonary artery with the PAA (Fig. 4). The blood gas sampling test showed no  $\text{O}_2$  step-up in the heart. Other data of cardiac catheterization were as follows: mean pulmonary capillary wedge pressure, 4 mmHg, right atrial pressure (a wave/v wave/mean), 4/0/0 mmHg, right ventricular pressure (systolic/diastolic/end-diastolic), 66/0/5 mmHg, left ventricular pressure (systolic/diastolic/end-diastolic), 117/0/3 mmHg, cardiac index,  $2.74 \text{ L/min/m}^2$ , and the pulmonary valve area was  $1.1 \text{ cm}^2$ . He was finally diagnosed with PAA due to

congenital bicuspid pulmonary valve.

We decided to monitor the patient without intervention, such as surgical repair or transcatheter pulmonary valve dilation, because he was asymptomatic, his pulmonary pressure was normal, the pressure gradient of the pulmonary valve was not severe, and the PAA had not expanded over time.

## Discussion

PAA is rare, and its pathogenesis remains unclear. Congenital heart diseases, such as patent ductus arteriosus, atrial septal defect, ventricular septal defect and pulmonary valvular anomalies, cause about 60% of PAAs (5). Other causes are acquired pulmonary hypertension; infectious diseases, such as tuberculosis; fungal diseases and syphilis, connective tissue diseases, such as Marfan and Loeys-Dietz syndromes; inflammatory diseases, such as Behçet disease; trauma, including iatrogenic injury; and idiopathic sources (6).

Table summarizes the seven cases of pulmonary aneurysm complicated with an isolated bicuspid pulmonary valve that have been reported, along with the present patient (7-12). The age of the reported patients ranged from 55-73 years, 5 were women, 2 were men, and 4 patients were asymptomatic. The present patient is the second man and the oldest of these patients. Three of the patients, including our own, had been asymptomatic at the time of the PAA diagnosis. Thus, imaging tests including, chest X-ray, TTE, CT, and magnetic resonance imaging (MRI), are important for the diagnosis of PAA.

The diagnosis of a bicuspid pulmonary valve is even more difficult than that of a PAA. The present case was revealed by TEE, although it was unable to be diagnosed by contrast CT because it was not synchronized on electrocardiograms. MRI is one of the most important imaging modalities in such cases, as bicuspid pulmonary valve was diagnosed by MRI in three of these patients.

The mechanisms underlying PAA formation in patients with congenital heart disease are an increased pulmonary blood flow due to left-right shunt, volume overload due to

**Table. Characteristics of This and Previous Reports.**

Reference	Age	Sex	Opportunity of diagnosis	UCG	PG of PS (mmHg)	PAA diagnosis	PA diameter (mm)	BAV diagnosis	Treatment
This patient	73	M	Medical check	PS BAV in TEE	45	CT	50	TEE	Medical follow up
7	53	F	CT for other disease	Unknown	Unknown	MRI	Unknown	MRI	unknown
8	55	M	SOB	PS	52	CT	55	Surgery	PA retraction PV incision
9	54	F	SOB	RV dilation	23	CT	60	Already known	PA partial resection PVR
10	70	F	CT for other disease	PS	22	CT	48×62	MRI	Unknown
11	73	F	SOB	Unknown	Unknown	UCG	44	CT	Unknown
12	59	F	Unknown	Unknown	Unknown	MRI	Unknown	MRI	Unknown

BAV: bicuspid aortic valve, BPV: bicuspid pulmonary valve, CT: computed tomography, F: female, M: male, MRI: magnetic resonance image, PA: pulmonary artery, PAA: pulmonary artery aneurysm, PG: pressure gradient, PS: pulmonary stenosis, PVR: pulmonary valve replacement, RV: right ventricle, SOB: shortness of breath, TEE: transesophageal echocardiography, UCG: ultrasound cardiography

severe pulmonary valve regurgitation, and dilation after pulmonary valve stenosis (5). A separated post-stenotic flow and increased wall shear stress in a bicuspid pulmonary valve might also play important roles in PAA formation (13). However, PAA develops even when the degree of pulmonary stenosis due to a bicuspid pulmonary valve is mild. Thus, a bicuspid pulmonary valve and PAA might be secondary to changes in the development of normal neural crest cells because the migration of these cells is needed for the normal development of semilunar valves and septation of the outflow tract into the aorta and pulmonary artery as well as remodeling of the aortic arch (12).

The risk of rupture and dissection of a PAA is lower in patients with a low pulmonary pressure and no pulmonary hypertension or shunt disease than in others (14). The factors associated with an increased risk of rupture and dissection are a pulmonary artery pressure >50 mmHg, diameter >75 mm and expansion >2 mm within one year (15).

The histopathology of PAA associated with an isolated bicuspid pulmonary valve remains unknown, although cystic medial necrosis and elastic fragmentation of the pulmonary artery and aortic walls have been identified in some patients with a bicuspid aortic valve (16). The indications for surgical or catheter treatment for pulmonary valve stenosis are a class I maximum systolic pressure gradient >64 mmHg, class IIa maximum systolic pressure gradient <64 mmHg, symptomatic condition, and right ventricular dysfunction with right-to-left shunt caused by other congenital heart diseases (17, 18).

The PAA and bicuspid pulmonary valve were not treated in the present patient because his pulmonary artery pressure was low, the pulmonary artery diameter was 50 mm and had not expanded over time, the pressure gradient of the pulmonary valve was 45 mmHg, he was asymptomatic, and he had no other congenital heart diseases. His symptoms were monitored, and the pressure gradient of the pulmonary valve and the diameter of the pulmonary artery were respectively

assessed by TTE and CT.

In conclusion, the timing of intervention is difficult to determine when PAAs are complicated with a bicuspid pulmonary valve because the condition is very rare.

**The authors state that they have no Conflict of Interest (COI).**

## References

- Deterling RA, Clagett OT. Aneurysm of the pulmonary artery: review of the literature and report of a case. *Am Heart J* **34**: 471-499, 1947.
- Groedel FM. Aneurysm of the pulmonary artery. *Radiology* **33**: 219-223, 1939.
- Matsuo S, Sato Y, Higashida R, et al. A giant main pulmonary artery aneurysm associated with infundibular pulmonary stenosis. *Cardiovasc Revascularization Med* **9**: 188-189, 2008.
- Jashari R, Van Hoeck B, Goffin Y, Vanderkelen A. The incidence of congenital bicuspid or bileaflet and quadricuspid or quadrileaflet arterial valves in 3,861 donor hearts in the European Homograft Bank. *J Heart Valve Dis* **18**: 337-344, 2009.
- Bartter T, Irwin RS, Nash G. Aneurysms of the pulmonary arteries. *Chest* **94**: 1065-1075, 1988.
- Robbins RC, Bernstein D, Berry GJ, VanMeurs KP, Frankel LR, Reitz BA. Cardiac transplantation for hypertrophic cardiomyopathy associated with sengers' syndrome. *Ann Thorac Surg* **60**: 1425-1427, 1995.
- Vedanthan R, Sanz J, Halperin J. Bicuspid pulmonic valve. *J Am Coll Cardiol* **54**: e5, 2009.
- Watanabe M, Hoshino S, Iwaya F, et al. Pulmonary artery aneurysm-like associated with pulmonary stenosis. *Kyobu Geka (J Jpn J Thorac Surg)* **42**: 222-225, 1989 (in Japanese, Abstract in English).
- Yamashita K, Akuzawa S, Terada H, Washiyama N, Ohkura K, Kazui T. Surgical treatment of pulmonary valve disease associated with pulmonary arterial dilatation in the adult: reports of two cases. *Japanese J Cardiovasc Surg* **37**: 100-103, 2008.
- Magnoni M, Turri C, Roghi A, Merlanti B, Maseri A. An Inverted location of the bicuspid valve disease. *Circulation* **124**: e513-e515, 2011.
- Krauss T, Berchem L, Blanke P, Zeh W, Pache G. 4D-cine CT imaging of a bicuspid pulmonary valve. *J Cardiovasc Comput To-*

- mogr **8**: 170-171, 2014.
12. Jamis-Dow CA, Barbier GH, Watkins MP, Lanza GM, Caruthers SD, Wickline SA. Bicuspid pulmonic valve and pulmonary artery aneurysm. *Cardiol Res* **5**: 83-84, 2014.
  13. Fenster BE, Schroeder JD, Hertzberg JR, Chung JH. 4-Dimensional cardiac magnetic resonance in a patient with bicuspid pulmonic valve: characterization of post-stenotic flow. *J Am Coll Cardiol* **59**: e49, 2012.
  14. Veldtman GR, Dearani JA, Warnes CA. Low pressure giant pulmonary artery aneurysms in the adult: natural history and management strategies. *Heart* **89**: 1067-1070, 2003.
  15. Duijnhouwer AL, Navarese EP, Van Dijk APJ, Loeys B, Roos-Hesselink JW, De Boer MJ. Aneurysm of the pulmonary artery, a systematic review and critical analysis of current literature. *Congenit Heart Dis* **11**: 102-109, 2016.
  16. de Sa M, Moshkovitz Y, Butany J, David TE. Histologic abnormalities of the ascending aorta and pulmonary trunk in patients with bicuspid aortic valve disease: clinical relevance to the Ross procedure. *J Thorac Cardiovasc Surg* **118**: 588-596, 1999.
  17. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: Executive Summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines for the management of adults with congenital heart disease). *Circulation* **118**: 2395-2451, 2008.
  18. Baumgartner H, Bonhoeffer P, De Groot NMS, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J* **31**: 2915-2957, 2010.

The Internal Medicine is an Open Access journal distributed under the Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view the details of this license, please visit (<https://creativecommons.org/licenses/by-nc-nd/4.0/>).