Photo Clinic

A Case of Isolated Bicuspid Pulmonic Valve and Pulmonary Artery Aneurysm

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A 33-year-old woman with a history of thyroid surgery for thyroid cancer and radioactive iodine therapy was referred for echocardiography due to dyspnea on exertion.

Transthoracic echocardiography showed normal left ventricular size and function (the ejection fraction = 55%), a prolapsing mitral valve with redundant chordae, mild mitral regurgitation, a tricuspid aortic valve, mild aortic insufficiency, and mild tricuspid regurgitation. The most remarkable echocardiographic findings were moderate right ventricular dilation with mild systolic dysfunction, moderate right atrial dilation, an aneurysmal pulmonary artery (the main pulmonary artery = 47 mm), mild pulmonary stenosis (the peak gradient = 22 mmHg), and severe pulmonary regurgitation (the vena contracta = 6-7 mm and the pressure half time = 105 ms).

Transesophageal echocardiography with the use of 3D modalities demonstrated a bicuspid pulmonic valve with doming and poor coaptation of the pulmonic valve leaflets (Figure 1). Additionally, a large patent foramen ovale was visualized in color Doppler (the flap separation = 2 mm and the tunnel length = 11 mm) with bubble passage in agitated saline injection.

Bicuspid pulmonic valves constitute a rare finding, and they are most often associated with other congenital heart diseases. Isolated bicuspid pulmonic valves are extremely rare, with an incidence rate of about 0.1% in clinical practice.¹ Pulmonary artery aneurysms also comprise a rare abnormality, with an incidence rate of approximately 1 in 14 000 cases in most studies.² The association between bicuspid pulmonic valves and pulmonary artery aneurysms has been reported, and the pathophysiologic causes of this association include hemodynamic alterations due to bicuspid pulmonic valves and most likely the abnormal migration of neural crest cells.³

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Figure 1. Transesophageal echocardiography with 3D imaging in (A) the multiplanar reconstruction mode demonstrates (B) the bicuspid pulmonic valve en face with doming and the poor coaptation of the PV leaflets (the arrow). AO, Aorta; AV, Aortic valve; PA, Pulmonary artery; PV, Pulmonary valve

The diagnosis of a bicuspid pulmonic valve by 2D imaging is challenging and sometimes impossible. Using 3D echocardiography and reconstruction confers a better assessment of the pulmonic valve morphology and identification of bicuspid pulmonic valves.

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