Case Report

Quadricuspid Pulmonary Valve in an Adult Patient Identified by Transthoracic Echocardiography and Multi-Detector Computed Tomography

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Key words:
Quadricuspid pulmonary valve, congenital heart disease.

Quadricuspid pulmonary valve is a rare congenital heart disease. It is infrequently associated with significant clinical complications and tends to be clinically silent. Because of its benign nature, it has been diagnosed mainly *post mortem*. Its diagnosis by transthoracic echocardiography is very difficult because of the anatomical features. We describe a case of quadricuspid pulmonary valve diagnosed by transthoracic echocardiography and electrocardiography-gated multi-detector row computed tomography.

Manuscript received:
May 1, 2013;
Accepted:
November 25, 2013.

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Quadricuspid pulmonary valve is rare and tends to be clinically silent. Because of its benign nature, it has been diagnosed mainly post mortem. With recent advances in cardiac imaging, such as computed tomography (CT) and magnetic resonance imaging (MRI), more cases have been discovered incidentally in living patients.[1,2]

We report a patient with a quadricuspid pulmonary valve diagnosed by transthoracic echocardiography and ECG-gated multi-detector row CT performed for the evaluation of intermittent chest discomfort.

Case presentation

A 53-year-old male patient visited our clinic with intermittent chest discomfort during exercise. He had no significant past medical or family history. Physical examination revealed no abnormal findings. The resting ECG showed normal sinus rhythm. Transthoracic echocardiography (TTE) in the parasternal short-axis view at the level of the aortic valve showed thickened and systolic doming pulmonary valve leaflets with mild pulmonary regurgitation and a dilated main pulmonary artery (Figure 1A). The diameters of the aorta and main pulmonary artery were 33 mm and 41 mm, respectively. Continuous wave Doppler at the pulmonary valve showed mild pulmonary stenosis, peak flow velocity across the pulmonary valve of 2.0 m/s, and a peak pressure gradient of 16 mmHg. With angulation of the transducer superiorly from the left parasternal window, a short-axis view of the pulmonary valve was obtained, showing a quadricuspid valve with a deficit of central coaptation (Figure 1B). The patient was further evaluated by ECG-gated multidetector row CT. CT angiography revealed 40-50% stenosis in the right coronary artery and a quadricuspid pulmonary valve with valvular thickening and a mildly dilated main pulmonary artery. The pulmonary valve showed 3 equal cusps and 1 smaller cusp on axial view (Figure 2). As the patient showed no limitations on treadmill testing, he was put on regular follow up.

Discussion

Quadricuspid pulmonary valve is a rare

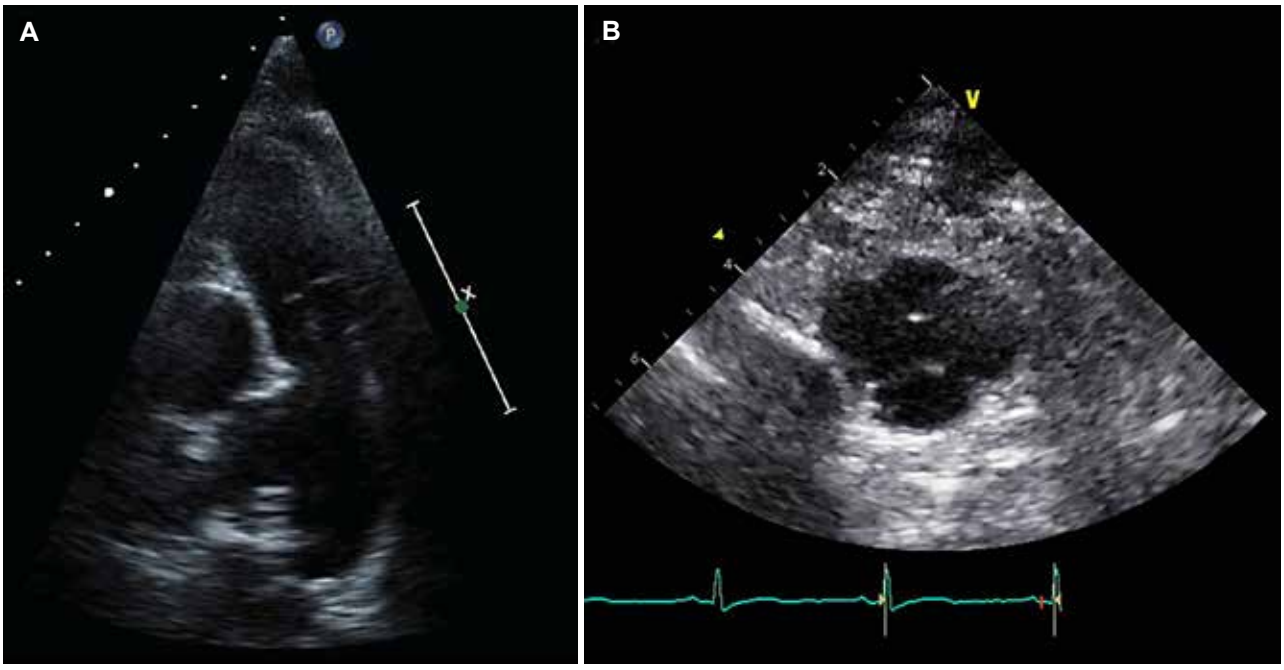

Figure 1. Two-dimensional echocardiography, showing a thickened pulmonary valve with systolic doming, a dilated main pulmonary artery (A) and a quadricuspid pulmonary valve in the diastolic frame (B).

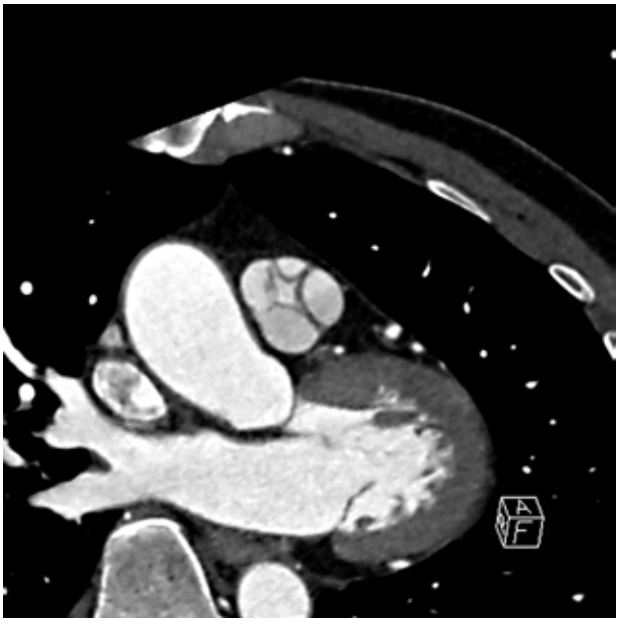

Figure 2. ECG-gated multi-detector row CT axial image, showing a quadricuspid pulmonary valve with incomplete coaptation during diastole.

congenital anomaly. It affects predominately male patients, with a ratio of 2:1 according to the data of Hurwitz and Roberts.[3] It has been found in from one in 400 to one in 2000 autopsies.[3,4]

Quadricuspid pulmonary valve has been commonly associated with aortic valve malformations, because the semilunar valves arise from mesenchymal swellings on the common vascular trunk.[1] It is sometimes associated with other congenital heart conditions, including atrioventricular defect, atrial septal defect, ventricular septal defect, patent *ductus arteriosus*, and coarctation of the aorta.[3,5] Quadricuspid pulmonary valve has anatomical variations, with the most common consisting of three equal-sized cusps and one smaller cusp.[3] No correlation has been found between anatomical variation and functional status.

The condition tends to be without serious clinical complications and has an asymptomatic course, even in combination with the other cardiac anomalies.[3,5] Diagnosis of quadricuspid pulmonary valve by TTE is very difficult, because of the anatomical disposition of the valve with respect to the thoracic wall. For this reason, it has almost always been diagnosed at autopsy. In our case, we were able to image the short axis of the pulmonary valve by TTE, because the pulmonary valve orifice had shifted anteriorly as a result of the pulmonary artery dilation.

Recent advances in cardiac CT and MRI permit improved visualisation of morphological valvular features, with any associated congenital and acquired structural deformities, and provide functional infor-

mation.[2] More cases have been discovered incidentally in living patients.[1,2] In the future, with newer techniques, three-dimensional TTE, CT, and MRI will play an important role in the diagnosis of quadricuspid pulmonary valve.

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