Quadricuspid pulmonic valve and great pulmonary artery aneurysm by multimodality imaging

A 71-year-old man was admitted to our hospital with chest tightness and progressive dyspnea. Physical examination revealed normal left ventricular (LV) impulse, systolic and diastolic murmur at the left sternal border.

Chest X-ray revealed enlarged contour of the vessel below the aortopulmonary window (Fig. 1a). An aneurysmal dilation of the main pulmonary artery and left pulmonary artery was revealed through transthoracic echocardiography, and the right pulmonary artery was seen slightly dilated (Fig. 1b). A short-axis view of pulmonary valve in a higher intercostal window demonstrated a quadricuspid anomalous valve (Fig.1c, Video 1). Moderate pulmonary stenosis (transvalvular peak velocity of 3.3 m/s) and moderate regurgitation (Fig.1d, Video 2) were seen through color Doppler flow imaging at the pulmonary valve. Cardiac computed tomography and three-dimensional volume-rendered computed tomography confirmed a quadricuspid pulmonary valve with four equal-sized cusps, and great aneurysm of the pulmonary artery trunk (73 mm) extended into the left pulmonary artery (43 mm), while the right pulmonary artery was dilated (31 mm) (Fig. 2a–2d). Therefore, the diagnosis of quadricuspid pulmonic valve complicated with pulmonary artery aneurysm was made by multimodal imaging.

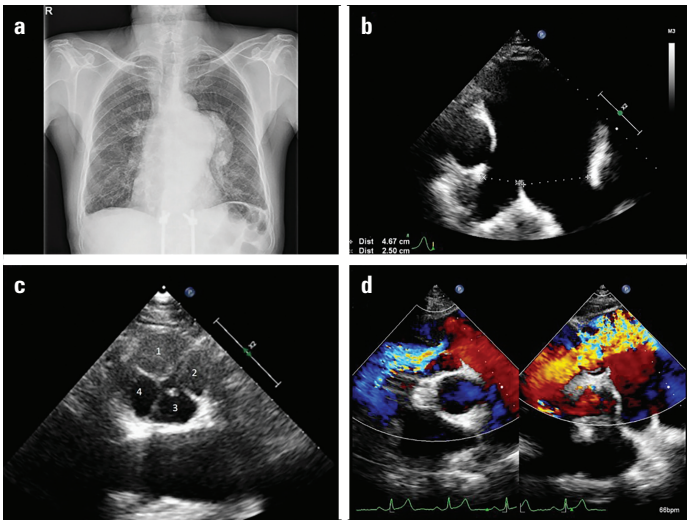

Figure 1. Chest X-ray revealed an enlarged contour of the vessel below the aortopulmonary window (a). Transthoracic echocardiographic image revealed aneurysmal dilation pf pulmonary artery trunk and left pulmonary artery (b). A short view of pulmonary valve obtained by transthoracic echocardiography showed a quadricuspid pulmonary valve (c). Color Doppler flow imaging demonstrated the moderate pulmonary insufficiency and systolic flow acceleration (d)

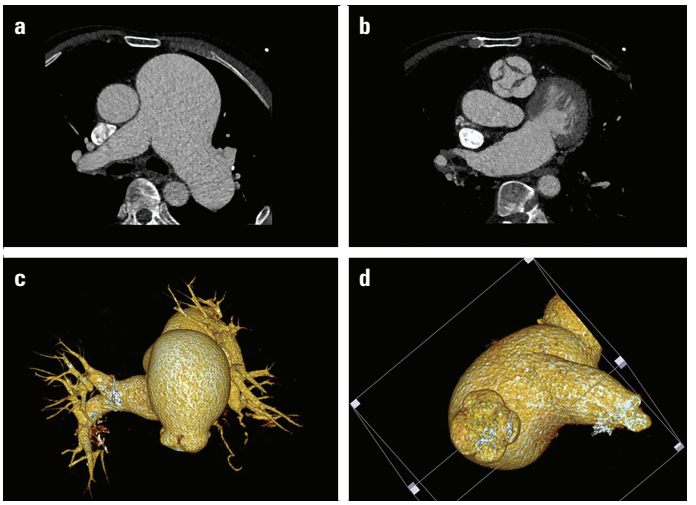

Figure 2. The computed tomography angiography (CTA) and three-dimensional volume-rendered computed tomography (3D VR CT). Axial image of the chest at the level of the bifurcation of the main pulmonary artery showed aneurysmal dilation of the main (73 mm) and left (43 mm) pulmonary arteries, while the right pulmonary artery was seen slightly dilated (31 mm) (a). Reconstructed CTA of the chest at the level of pulmonary valve demonstrated a quadricuspid pulmonary valve with four cusps of similar size (b). 3D VR CT confirmed that quadricuspid pulmonary valve had four cusps of similar size and showed the great aneurysm of the pulmonary artery intuitively (c and d)

The quadricuspid pulmonary valve is a rare congenital anomaly with an incidence reported from 1 in 1000 to 1 in 20000 (1). Pulmonary artery aneurysms are rare and infrequently diagnosed (2). Limited reports are available on the quadricuspid valve in combination with pulmonary artery aneurysm (3, 4). Cardiac computed tomography allowed an adequate characterization of the valve morphology, and echocardiography provided the information about the function of the anomalous valve. This case highlights the crucial role of multimodality imaging evaluation for detailed noninvasive depiction of pulmonary valve disease.

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Video 1. A transthoracic echocardiography demonstrated the quadricuspid pulmonary valve

Video 2. Color Doppler flow imaging indicated the moderate pulmonary valve insufficiency and systolic flow acceleration

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