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Three-Dimensional Echocardiography: A Promising Tool for the Diagnosis of Quadricuspid Pulmonary Valve and Pulmonary Artery Aneurysm

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A 64-year-old male patient presented to the emergency room with a history of dyspnea persisting for the last two weeks, clinically classified as NYHA class II equivalent. The patient, who quit smoking 22 years ago, had no other cardiac risk factors or concomitant diseases.

The clinical examination revealed minimal bilateral lower leg edema, a blood pressure of 113/72 mmHg, normal lung sounds, and a fixed split S2 sound. Blood tests demonstrated normal complete blood count, kidney and liver function tests, as well as troponin and NT-pro-BNP values. The ECG showed sinus rhythm with a mild first-degree atrioventricular block (PR time: 205 min), left axis deviation, and persistent S wave up to V6.

The patient was scheduled for transthoracic echocardiography (TTE) and spiroergometry. Three-dimensional TTE (3D TTE) revealed a quadricuspid pulmonary valve (PV) (Figure 1, Video 1) and an aneurysmatic pulmonary artery (55 mm) within a normal heart. Cardiac computed tomography (CCT) (Figure 2, Video 2) and cardiac magnetic resonance imaging (CMRI) (Figure 3, Video 3) were performed for further cardiac assessment. The findings of both these procedures were consistent with the 3D TTE findings. Spiroergometry revealed no cardiac, respiratory, or metabolic exhaustion.

Neither CCT nor CMRI indicated an additional pulmonary pathology. Behçet's disease was ruled out through clinical evaluation, following the International Criteria for Behçet's Disease. To address dyspnea, we performed left and right cardiac catheterization, ruling out coronary heart disease and pulmonary hypertension. The dyspnea was non-progressive; hence, we recommended an initial short-term follow-up and a cardiopulmonary exercise program. The patient was regularly followed-up for 3 years. Dyspnea resolved after regular exercise initiation, and the aneurysm showed no progression.

A comprehensive anatomical and hemodynamic analysis is necessary for accurately diagnosing and treating valvular heart disease. Quadricuspid PV is an uncommon and usually incidental occurrence in clinical practice.[1] Its incidence has been reported to be 0.2% in a European autopsy study.[2] The standard TTE exam may be relatively difficult to visualize, potentially leading to underdiagnosis. The clinical implications of quadricuspid PV or pulmonary aneurysm remain unclear.

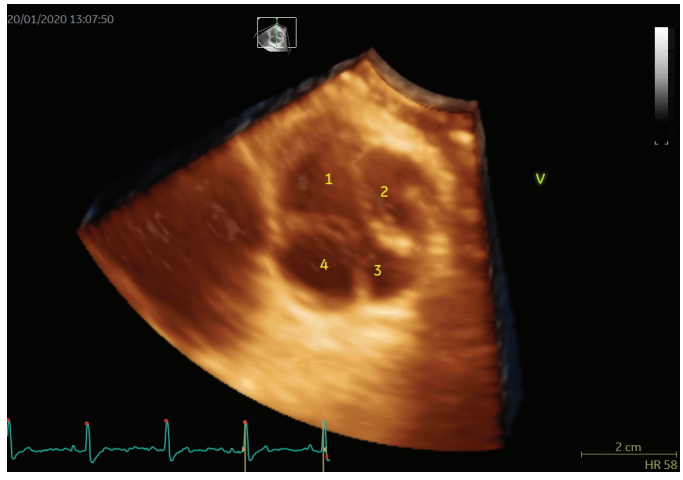

FIG. 1 and Video 1. Three-dimensional transthoracic echocardiography images showing the short-axis view of the quadricuspid pulmonary valve.

Video 1: 10.4274/balkanmedj.galenos.2024.2023-10-5.video1

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3DE is a promising new tool for evaluating quadricuspid PV and all PV lesions.[3,4] Before progressing to additional cardiovascular imaging modalities, it is advisable to use 3DE as a supplementary tool in PV evaluation following two-dimensional echocardiography.[5] 3DE has been utilized for diagnosing pulmonary artery aneurysms only in a few patients.[6,7] To the best of our knowledge, we present the first documented case of a patient with quadricuspid PV and an aneurysmal pulmonary artery, identified through 3D TTE and subsequently verified by CCT and CMRI.

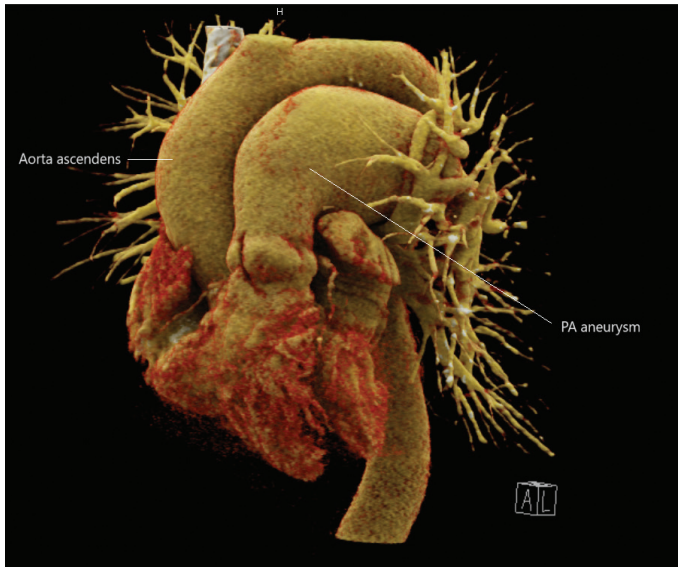

FIG. 2 and Video 2. CT pulmonary angiogram 3D reconstruction reveals an aneurysmal main pulmonary artery. PV indicates pulmonary artery.

CT, computed tomography, PV, pulmonary valve.

Video 2: 10.4274/balkanmedj.galenos.2024.2023-10-5.video2

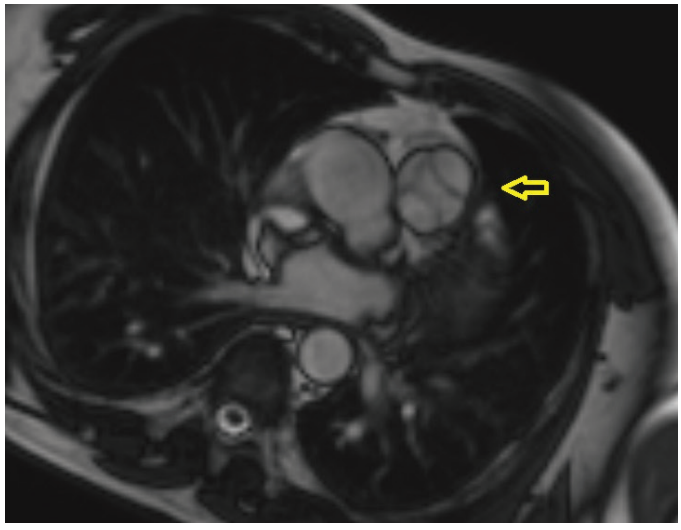

FIG. 3 and Video 3. Cardiac magnetic resonance imaging showing the quadricuspid pulmonary valve (arrow).

Video 3: 10.4274/balkanmedj.galenos.2024.2023-10-5.video3

While routine follow-up is sufficient for most patients with this anomaly, some may have coexisting congenital heart disease. Although rare, extreme cases with an abnormally functioning valve (e.g., stenosis) may necessitate intervention.[8] In our patient, the presence of quadricuspid PV and aneurysmatic pulmonary artery findings may be incidental or related to dyspnea, as there was no association between dyspnea and any other heart disease. Guidelines lack consensus on the follow-up or treatment of these cases because most practitioners determine their strategy based on the associated presentation, such as pulmonary stenosis. Addressing the need for routine prophylaxis against infective endocarditis poses another challenge.

Informed Consent: We obtained written informed consent from the patient for this study.

Authorship Contributions: Conception- T.K.Ö.; Design- T.K.Ö.; Supervision- T.K.Ö.; Data Collection and/or Processing- V.K., U.A.; Analysis and/or Interpretation- V.K., U.A., T.K.Ö.; Literature Search- V.K., U.A., T.K.Ö.; Writing- V.K., U.A., T.K.Ö.

Conflict of Interest: No conflict of interest was declared by the authors.

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