THE ANATOLIAN JOURNAL OF CARDIOLOGY

Official journal of the TURKISH SOCIETY OF CARDIOLOGY

A Challenging and Rare Morphology in a Patient with Hypertrophic Cardiomyopathy: Quadricuspid Mitral Valve

A 60-year-old woman, previously diagnosed with hypertrophic cardiomyopathy (HCM) since 2019, hypothyroidism since 2018, and paroxysmal atrial fibrillation, with a known history of intracardiac defibrillator (ICD) implantation in 2019, presented to the emergency department with an ICD shock. Further tests revealed shock due to transient rapid atrial rate. Thus, radiofrequency AF ablation was planned as a treatment. To exclude intra-atrial thrombi, transesophageal

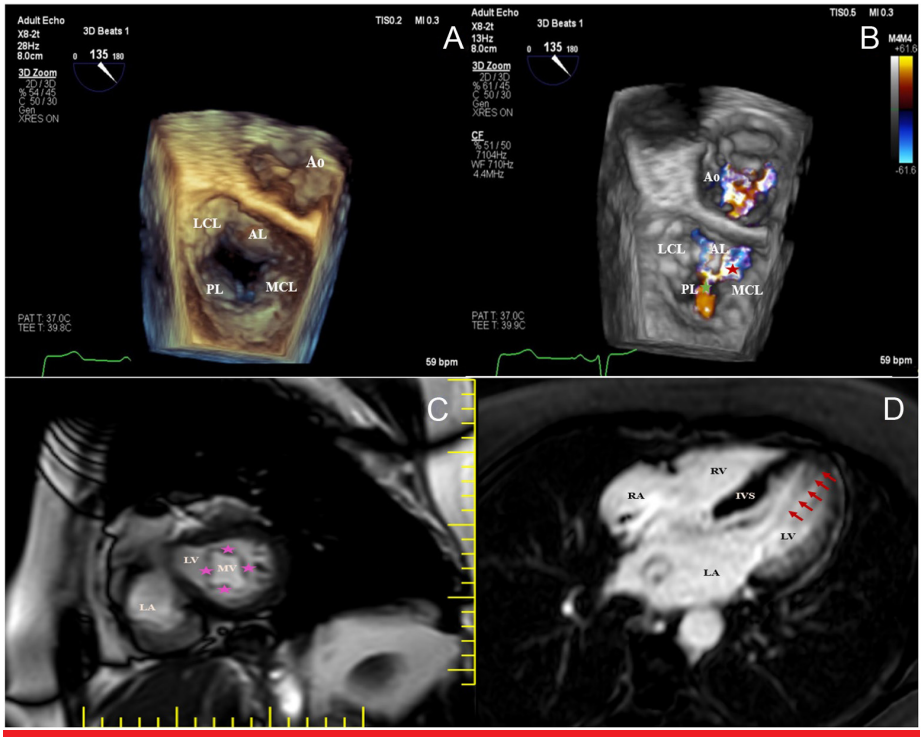

Figure 1. 3D en face TEE imaging of MV and cardiac MRI imaging of MV and papillary muscles. A) A quadricuspid MV structure with a quadrangular mitral annulus geometry with larger anterior and posterior leaflets and smaller commissural leaflets medially and laterally (Live 3D imaging). B) Double orifice mitral regurgitation jet originating between the medially located commissural mitral leaflet and the anterior (red star) and posterior (green star) mitral leaflets (3D color imaging). C) Cine short-axis imaging showed the quadrangular morphology of the MV annulus (pink stars). D) Late contrast imaging (Phase Sensitive Inversion Recovery sequence) showed an abnormally located accessory papillary muscle extending from the apex to the mitral annulus (red arrows). AL, anterior leaflet; Ao, aortic valve; IVS, interventricular septum; LA, left atrium; LCL, lateral commissural leaflet; LV, left ventricle; MCL, medial commissural leaflet; MV, mitral valve; PL, posterior leaflet; RA, right atrium; RV, right ventricle.

E-PAGE ORIGINAL IMAGE

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Cite this article as: İnan D, Tekin M, Pamuk FÖ, et al. A challenging and rare morphology in a patient with hypertrophic cardiomyopathy: Quadricuspid mitral valve. *Anatol J Cardiol.* 2024;28(11):E-33-E-34.

DOI:10.14744/AnatolJCardiol.2024.4741

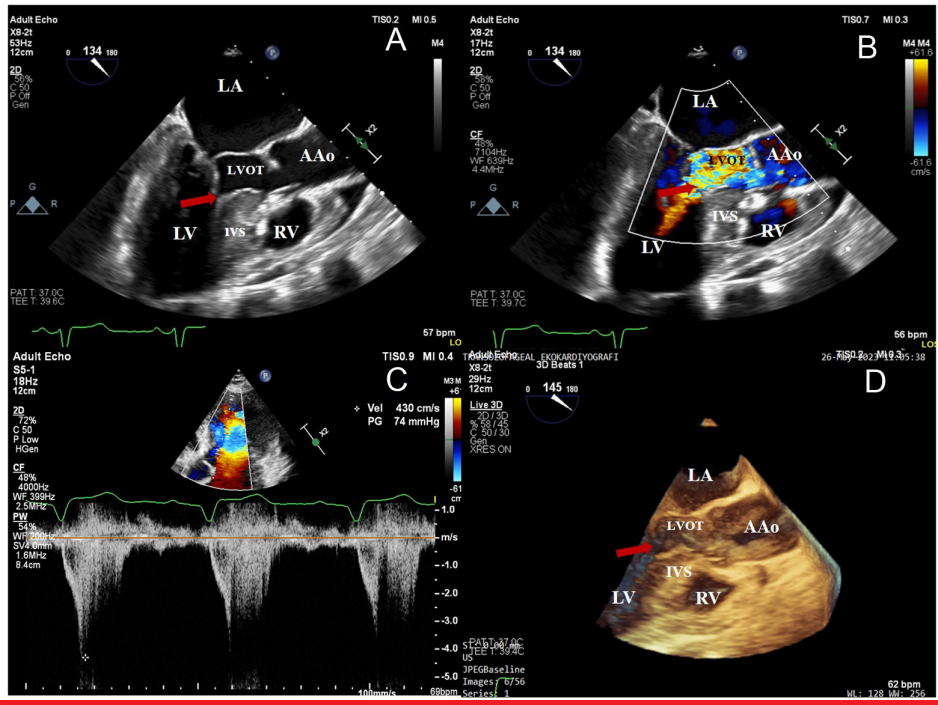

Figure 2. LVOT obstruction associated with SAM and accessory papillary muscle. A) Overt mitral SAM on 2D TEE imaging (red arrow). B) Marked systolic turbulent flow in the LVOT on TEE color imaging (red arrow). C) PW Doppler evaluation on apical 5-chamber TTE imaging, 74 mmHg gradient in the LVOT at rest. D) Overt mitral SAM on 3D TEE imaging (red arrow). AAo, ascending aorta; IVS, interventricular septum; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; MV, mitral valve; PW, pulsed wave; RA, right atrium; RV, right ventricle; SAM, systolic anterior motion; TEE, transesophageal echocardiography; TTE, transthoracic echocardiography.

echocardiography (TEE) was planned. Three-dimensional (3D) TEE evaluation showed a quadricuspid mitral valve (MV) structure with a quadrangular mitral annulus geometry with larger anterior and posterior leaflets and smaller commissural leaflets medially and laterally. Here are some cardiac magnetic resonance imaging (MRI) and 3D TEE images supporting this (Figure 1A-1C). The patient also had obstructive hypertrophy, an accessory papillary muscle extending from the apex to the mitral annulus, and grade 3 systolic anterior motion of the anterior mitral leaflet with a peak systolic gradient of 74 mm Hg, moderate mitral regurgitation, mild aortic and tricuspid regurgitation, and preserved ejection fraction (Figure 1D, Figure 2A-2D). After the ablation procedures, the patient was discharged after adjusting the dose of beta-blocker and adding disopramide treatment for obstruction. Furthermore, the results of HCM genetic testing with a 42-gene panel were negative.

Hypertrophic cardiomyopathy is the manifestation of a complex spectrum that includes not only increased left ventricular wall thickness but also the mitral apparatus and other cardiac structures. We present the third case report of quadricuspid MV supported with significantly rare TEE and MRI findings.

Informed Consent: Detailed information was given to the patient regarding the possible contribution of the case report to the literature. The patient gave written and verbal consent for the publication of the case report.

Declaration of Interests: The authors have no conflict of interest to declare.

Funding: The authors declared that this study has received no financial support.

Video 1: Three-dimensional transoesophageal echocardiography from the atrial view showed a square-like opening in diastole and a cross shape of the commissures during systolic closure, with larger anterior and posterior leaflets and smaller commissural leaflets medially and laterally, and 4 commissures.

Video 2: Three-dimensional color transesophageal echocardiography from the atrial view reveals a moderate, double-orifice mitral regurgitation jet with a total vena contracta 3D vena contracta area of 0.28 cm^2, originating between the medially located commissural mitral leaflet and the anterior and posterior mitral leaflets.

Video 3: Bi-plane transesophageal echocardiographic images illustrated apically located accessory papillary muscle and overt systolic anterior motion of the mitral anterior leaflet.