



Triple Trouble of the Mitral Valve in a Patient with Hypertrophic Cardiomyopathy

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A 64-year-old male patient with a diagnosis of HCM and no other comorbidities that had been followed up for the last 5 years, who gave informed consent for this publication under the condition of anonymity, was admitted to the cardiovascular imaging department of our hospital to undergo cardiac magnetic resonance imaging (CMRI) with the aim of risk stratification for sudden cardiac death (SCD). The family history of the patient revealed SCD of a 38-year-old male cousin on father's side. The presenting symptoms of the patient consisted of nothing more than occasional palpitations without accompanying presyncope or syncope.

The patient was hemodynamically stable with an arterial blood pressure of 120/80 mmHg, a heart rate of 80 bpm, and a room air O₂ saturation of 99%. Pulmonary auscultation revealed normal lung sounds on cardiac evaluation, with S1, S2, and S4 sounds being heard and S3 being absent, as well as 3/6 systolic murmur at the apical region. No peripheral edema or skin lesions were observed. The remainder of the physical examination was unremarkable.

On transthoracic echocardiography (TTE) performed prior to CMRI, ejection fraction was found to be 65%. Asymmetric septal hypertrophy (maximum thickness of septum: 25 mm; thickness of basal posterior wall: 12 mm) is shown, with no pressure gradient in the left ventricular outflow tract even with the Valsalva maneuver. Moderate thickening of the anterior leaflet of the mitral valve with a hockeystick appearance and doming motion consistent with rheumatic valve disease, as well as chordal systolic anterior motion (SAM) related to asymmetric septal hypertrophy and prolapse of the posterior leaflet were observed altogether (Video 1).

CMRI showed similar findings of the mitral valve as those on TTE, and an acceleration jet in the LVOT was additionally detected.

Moderate mitral regurgitation was evaluated both quantitatively and qualitatively with a regurgitation fraction of 28%. Phase-sensitive inversion-recovery images obtained post gadolinium demonstrated midmyocardial late gadolinium enhancement (LGE) in hypertrophic areas with an LGE extension of 12% (6.36 grams) (Video 2).

On short axis cine imaging, maximum thickness of the left ventricle was measured as 24 mm at the basal anteroseptal wall, and no apical aneurysm was observed. The LVEF was calculated to be 68%. The implantation of implantable cardioverter-defibrillator was not recommended due to lack of signs indicating SCD on clinical examination, history, and cardiac imaging. The patient was followed up with medical treatment (bisoprolol 5 mg once per day) (Figure 1).

The aforementioned case report appears to exhibit a pioneering role in the literature, owing to the extraordinary combination of three types of valve diseases, namely, chordal SAM (Carpentier type 4), rheumatic mitral valve (Carpentier type 3a) disease, and mitral valve prolapse (Carpentier type 2); the third of which is classified as "class 5" according to the modified MRI classification. This case is echocardiographically compatible with group 5 with hybrid pathologies in the modified Carpentier classification suggested by Shah et al.^{1,2} We posit that the rheumatic nature of the anterior leaflet protected the patient from SAM and left ventricular outflow tract obstruction (LVOTO). The intriguing aspect of the present case is that the rheumatic valve disease that developed on top of mitral valve prolapse acted to protect the posterior leaflet from immobility. Although a clinically relevant result on the clinical status of this particular patient, we believe that this is the first case in the literature where these three types of mitral valve pathologies are seen to co-occur.



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FIG. 1. Cardiac MRI figures of the case. (a) Four-chamber phase-sensitive inversion-recovery (PSIR) image: midmyocardial LGE at the base of the septum and multiple crypts at the basal midsection of the anterolateral wall. (b) Short axis PSIR image: LGE at the inferior insertion point. (c) Two-chamber PSIR image: midmyocardial LGE at the base of the inferior wall.

MRI, magnetic resonance imaging; LGE, late gadolinium enhancement.

LVOTO has two fundamental mechanisms: (1) LVOTO narrowing caused by septal hypertrophy, dynamically moving the mitral valve leaflets forward in systole, and leading to abnormal blood flow vectors; (2) anatomical changes, including longer leaflets as well as anterior displacement of the papillary muscles and mitral valve apparatus that make the valve more susceptible to abnormal flow vectors.^{3,4}

In rheumatic mitral valve disease, valve motion is restricted due to fibrosis of the valve.⁵ In this case, the anterior motion of the mitral leaflet in systole, which is one of the pathophysiologic mechanisms underlying LVOTO, is decreased due to the restricted motion of the anterior leaflet of the mitral valve. The key reason behind minimized pressure gradient even on maximal Valsalva maneuver was purported to be in relation to the doming motion of thickened anterior mitral leaflet restricting SAM. Thereby, we have demonstrated the protective effect conferred by one valve disease (rheumatic mitral valve) on another (SAM). Although LVOTO is a risk factor for SCD, rheumatic valve disease may protect this patient from sudden cardiac events caused by hemodynamic collapse.⁶

Informed Consent: The patient has given informed consent for this publication under the condition of staying anonymous.

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Video 1: Echocardiographic images of the case.

<https://www.doi.org/10.4274/balkanmedj.galenos.2022.2022-7-109.video1>

Video 2: Cardiac MRI images of the case. CINE 3 and 4-chamber images.

Important points: Septum hypertrophy, mitral valve anterior leaflet doming, posterior leaflet prolapse, and systolic anterior motion.

MRI, magnetic resonance imaging.

<https://www.doi.org/10.4274/balkanmedj.galenos.2022.2022-7-109.video2>