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Mid-ventricular obstructive hypertrophic cardiomyopathy with apical aneurysm and bidirectional gradient in a patient with multivessel coronary disease

Short title: MVO HCM with apical aneurysm in a patient with coronary artery disease

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A 69-year old female was referred to the hospital due to syncope and newly diagnosed atrial fibrillation (AF). The patient had a family history of sudden cardiac death, mitral valve prolapse, ventricular arrhythmia and premature coronary artery disease. She was treated for hypertension, type 2 diabetes and hyperuricemia. Electrocardiogram (ECG) showed atrial fibrillation with average QRS rate of 90 bpm (Figure 1A). The echocardiography revealed enlarged left ventricle (LV end-diastolic diameter 58 mm) and left atrium (LA volume index 80 ml/m^2), massive LV hypertrophy — up to 21 mm in the basal anteroseptal and all mid

segments, including papillary muscles, creating mid ventricular obstruction with peak systolic pressure of 25 mm Hg (Figure 1B). The apex was dilated and dyskinetic — aneurysmal area of 16 cm^2. Color Doppler imaging showed instantaneous, paradoxical, bidirectional systolic flow at the mid-cavity level reflected by double proximal isovelocity surface area (Figure 1C). In continuous wave Doppler we observed physiological systolic blood flow to the base of the heart followed by paradoxical diastolic flow, as the midventricular obstruction was relieved (Figure 1D). The results met criteria for mid-ventricular obstructive hypertrophic cardiomyopathy (MVO HCM) [1, 2]. Left ventricle ejection fraction (EF) measured by 3D method was 37% (Figure 1E). NT-proBNP concentration was elevated: 1906 pg/ml. In 24-hour ECG monitoring 10747 premature ventricular complexes were recorded, including 725 pairs and 44 non-sustained ventricular tachycardia (nsVT) episodes of maximum 193 bpm, the longest consisting of 37 beats. Cardiovascular magnetic resonance (CMR) examination confirmed the diagnosis of HCM and demonstrated EF of 39%, confirming estimation from the echo exam. Transmural late gadolinium enhancement was present in all apical segments, which suggested previous myocardial infarction [1, 2] (Figure 1F). Coronary arteriography showed severe stenosis of the right coronary artery (RCA) and diffuse atherosclerotic lesions in the remaining vessels — multivessel disease (MVD). Percutaneous angioplasty of RCA was performed. The HCM Risk-SCD (risk for sudden cardiac death) was estimated at 9.07%. In the further course elective catheter ablation of AF was conducted.

We present various echocardiographic manifestations of MVO HCM phenotype including an extremely rare (<1%) bidirectional gradient accompanied by a diastolic flow. Mid-ventricular obstructive hypertrophic cardiomyopathy with apical aneurysms occurs in 2%–5% of patients with HCM and apical aneurysm is observed in 20% of patients with MVO HCM. It is an independent risk factor for sudden cardiac arrest (SCA) in the VT/ventricular fibrillation mechanism. However, in this case the origin of apical aneurysm was difficult to be established due to coexisting MVD and CMR results suggesting post myocardial infarction scar. In the available guidelines, left ventricular aneurysm in the course of HCM is an indication for implantable cardioverter-defibrillator implantation. For this reason, regardless of the etiology, in the presence of HCM and high 5-year risk of sudden cardiac death, the patient had ICD implanted [1, 5]. Surgical removal of an aneurysm should be considered in severely symptomatic patients with HCM to reduce the overall risk of adverse events, especially ventricular arrhythmias and embolic stroke [4].

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal.

Article information

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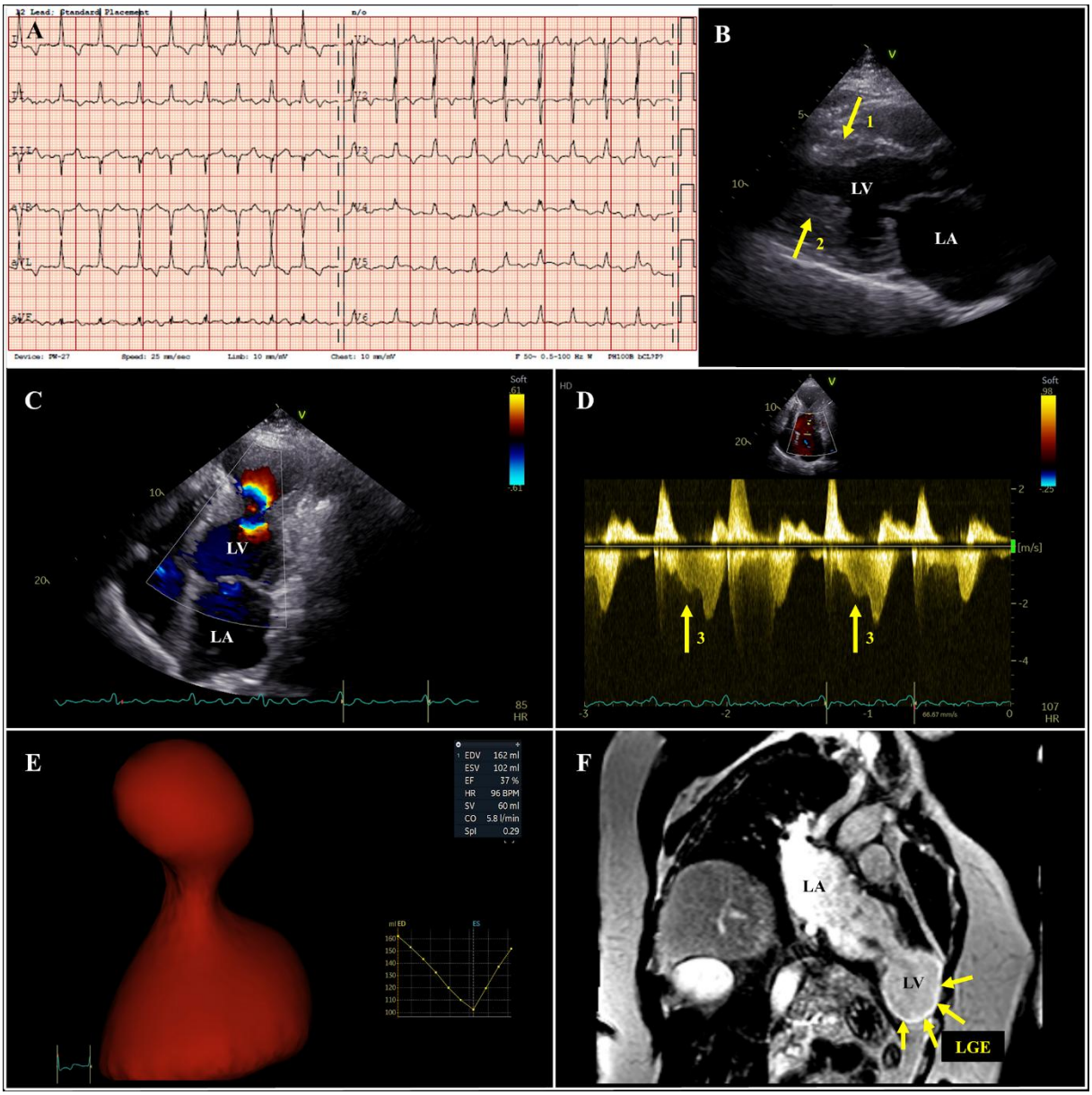

Figure 1. A. Electrocardiogram showing early R/S transition in precordial leads, "dagger-like" Q-wave in lead III, negative T-waves in the leads I, II, aVL, V2–V6; **B.** hypertrophy of basal anteroseptal and all mid segments (arrow 1), including papillary muscles (arrow 2); **C.** Color Doppler, mid-cavity obstruction caused pressure build up and blood trapping in the apical cavity; it resulted in bidirectional systolic flow at the mid-cavity level reflected by double proximal isovelocity surface area at systole; **D.** Continuous wave Doppler, paradoxical diastolic flow across the aortic valve (arrow 3); in this case, physiological systolic blood flow to the base was followed by paradoxical diastolic flow as the midventricular obstruction was relieved and trapped blood left the apical cavity; **E.** 3D model used to estimate ejection fraction; **F.** cardiovascular magnetic resonance, hyperdense late gadolinium enhancement (LGE) in all apical segments indicating regions of cardiomyocyte necrosis or myocardial tissue fibrosis