CLINICAL IMAGE

Suspicion of myocarditis in a patient with mitral valve prolapse

Mateusz Polak[1], Romuald Wojnicz[2], Jarosław Myszor[1], Jan Szczogiel[1], Katarzyna Mizia-Stec[1]

1 1st Department of Cardiology School of Medicine in Katowice, Medical University of Silesia, Katowice, Poland
2 Department of Histology and Embryology, Medical University of Silesia, Zabrze, Poland

A 23-year-old woman with diagnosed mitral valve prolapse (MVP) was admitted to a cardiac department because of frequent polymorphic ventricular extrasystoles and episodes of nonsustained ventricular tachycardia on Holter monitoring.

On admission, the patient reported palpitations, persistent weakness, and episodes of nonspecific chest pain and arthralgia. Blood count, biochemical parameters, and myocardial necrosis markers were within the reference ranges. Baseline electrocardiogram (ECG) showed sinus rhythm with frequent ventricular extrasystoles and episodes of nonsustained ventricular tachycardia, without any conduction disorder. Flat T waves in the inferior leads (III, aVF) were present (FIGURE 1A). A 24-hour ECG recording showed a sinus rhythm of 62 bpm with frequent episodes of polymorphic ventricular arrhythmia (8760

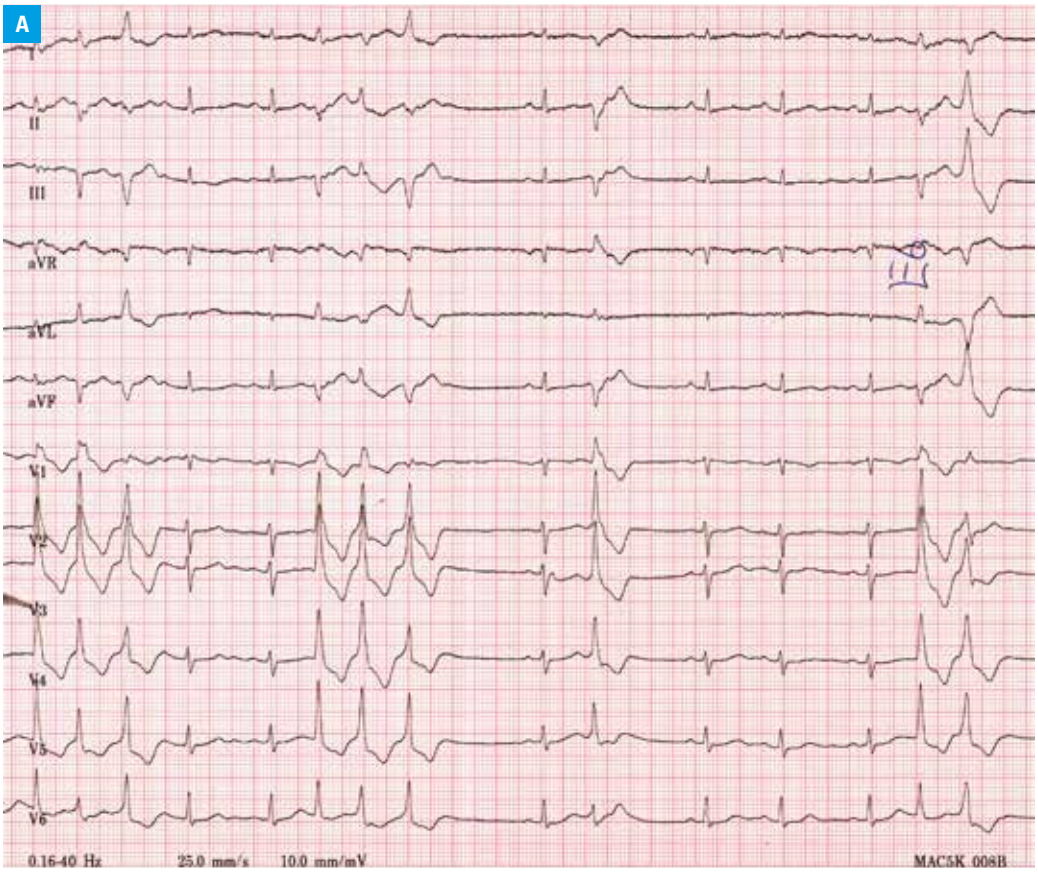

FIGURE 1 A – baseline electrocardiogram: sinus rhythm with ventricular extrasystoles and nonsustained ventricular tachycardia, flat T waves in the inferior leads (III, aVF)

Correspondence to:
Mateusz Polak, MD, I Oddział Kardiologii, Samodzielny Publiczny Szpital Kliniczny nr 7 Śląskiego Uniwersytetu Medycznego w Katowicach, Górnośląskie Centrum Medyczne im. prof. Leszka Gieca, ul. Ziołowa 45-47, Katowice, Poland, phone: +48 32 359 80 00, e-mail: mateuszpolak@mp.pl
Received: March 26, 2017.
Revision accepted: May 15, 2017.
Published online:
Conflict of interests: none declared.
Pol Arch Intern Med. 2017; 127 (6): 452-454
doi:

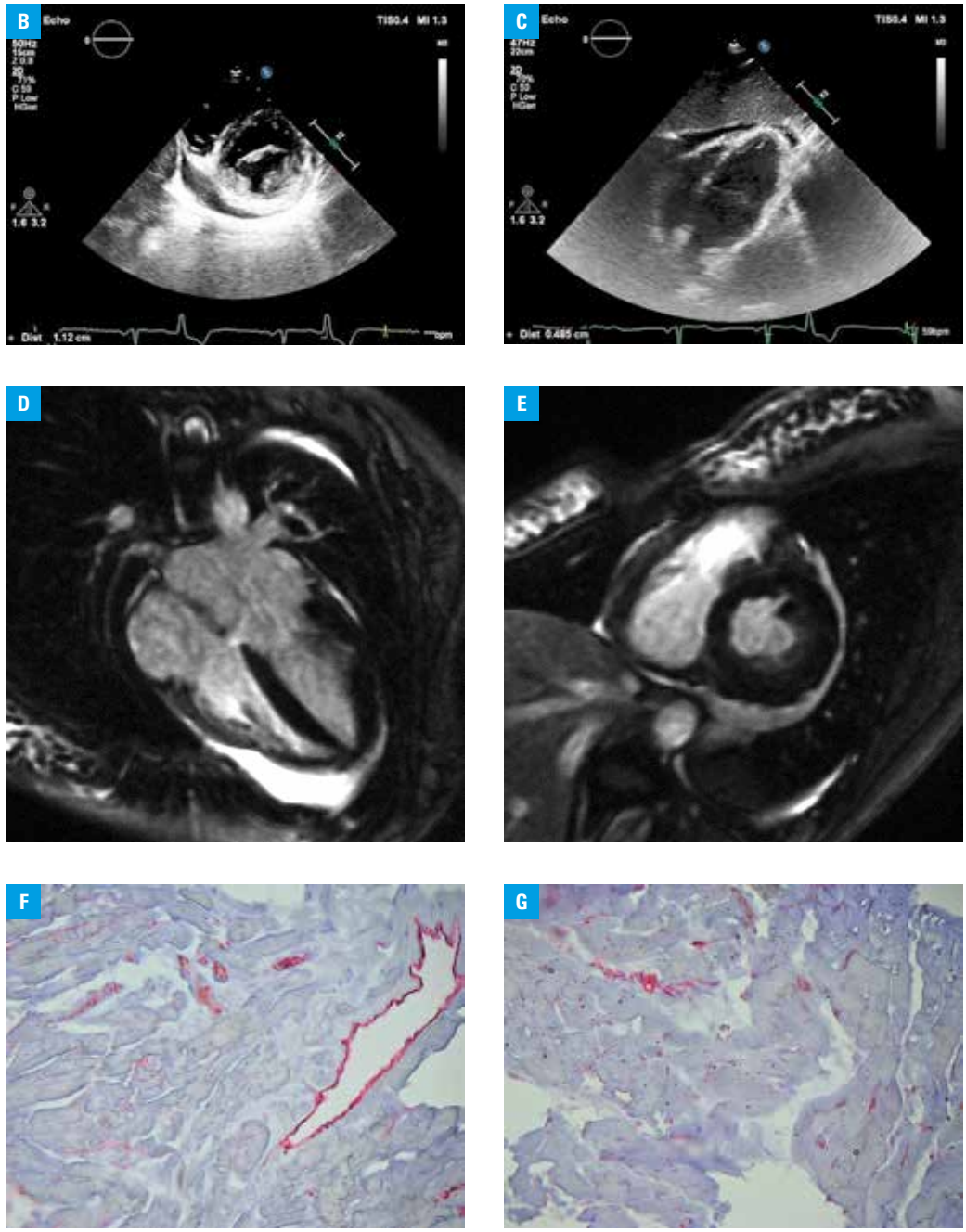

FIGURE 1 B, C – echocardiography: pericardial effusion behind the posterior wall (B), in front of the right ventricle and in the apical area (C); D, E – cardiac magnetic resonance imaging; long-axis view (D) and short-axis view (E); small bands of subepicardial and midmural layer enhancement in the septal and lateral basal segments; F, G – frozen endomyocardial biopsy sections: immunostaining for class I (A, B, C) (F) and class II (DR) (G) human leukocyte antigens on microvascular endothelium (red staining); magnification, × 200.

premature ventricular complexes [PVCs], 40 episodes of nonsustained ventricular tachycardia with 3 to 6 ventricular complexes at a rate of 170 bpm) originating from a single focus with right bundle branch block and left axis deviation. Echocardiography showed a normal size of the cardiac chambers, preserved ejection fraction, and MVP with increased leaflet thickness and without significant mitral regurgitation; pericardial effusion was also present (FIGURE 1BC). Arrhythmia persisted despite treatment with antiarrhythmic drugs (β-blockers, sotalol, ranolazine, propafenone), and administration of β-blockers resulted in symptomatic sinus bradycardia of up to 35 bpm.

Cardiac magnetic resonance (CMR) showed MVP with bileaflet involvement and signs of myocardial edema, myocarditis, and late gadolinium enhancement in the septal and lateral left ventricular (LV) segments (FIGURE 1DE). There was no fibrosis in the papillary muscles or the LV inferobasal wall. LV endomyocardial biopsy was performed to confirm the suspected myocarditis. The biopsy specimens revealed a significant upregulation of class I and II human leukocyte antigens (FIGURE 1FG), accompanied by local thrombogenicity (de novo endothelial expression of tissue factor). Glucocorticoid treatment (first intravenous, then oral) was introduced because of an active inflammatory

process,[1] and nadroparin was administered to improve endothelial function.[2]

After 3 months of therapy, the patient reported none of the previous complaints. Laboratory parameters were within the reference ranges and were comparable with the baseline values. Echocardiography showed a reduction of pericardial effusion. The Holter monitoring revealed a significant reduction of ventricular arrhythmia (only 99 single PVCs). An exercise treadmill test with a maximum load of 10.6 metabolic equivalents was performed and yielded negative clinical and electrocardiographic results, without provocation of any arrhythmia. The control CMR showed a reduction of LV end-diastolic diameter (100 ml vs 136 ml at baseline) and improved cardiac output (4.3 l/min vs 3 l/min at baseline). Myocardial edema was still observed in the lateral segments. Because of resolution of the arrhythmia and improvement of clinical condition, glucocorticoid therapy at a reduced dose (prednisone, 5 mg/d) for the next 3 months was recommended. The follow-up was scheduled to monitor the patient for MVP and residual myocarditis.

MVP is a cause of sudden cardiac death in young adult women. These patients often have ventricular arrhythmias of LV origin, bileaflet involvement, and frequent repolarization abnormalities in the inferior leads. The hallmark of arrhythmic MVP is fibrosis of the papillary muscles and LV inferobasal wall.[3] Myocarditis can also lead to severe arrhythmias or even sudden cardiac death and fibrosis on CMR.[4] These 2 independent pathologies, MVP and myocarditis, may coexist and require a careful differential diagnosis as well as optimal treatment.

REFERENCES

1 Wojnicz R, Nowalany-Kozielska E, Wojciechowska C, et al. Randomized, placebo-controlled study for immunosuppressive treatment of inflammatory dilated cardiomyopathy. Circulation. 2001; 104: 39-45.

2 Wojnicz R, Nowak J, Szyguła-Jurkiewicz B, et al. Adjunctive therapy with low molecular-weight heparin in patients with chronic heart failure secondary to dilated cardiomyopathy: one-year follow-up results of the randomized trial. Am Heart J. 2006; 154: 713.e12 713.e7.

3 Basso C, Perazzolo Marra M, Rizzo S, et al. Arrhythmic mitral valve prolapse and sudden cardiac death. Circulation. 2015; 132: 556-566.

4 Caforio AL, Pankuweit S, Arbustini E, et al. Current state of knowledge on aetiology, diagnosis, management, and therapy of myocarditis: a position statement of the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases. Eur Heart J. 2013; 34: 2636-2648.