CLINICAL VIGNETTE

Implantable cardioverter-defibrillators in cardiac amyloidosis: a grey zone requiring an individual approach

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A 66-year-old Caucasian woman was admitted to the cardiology department because of fainting episodes in order to determine further treatment. Nine months earlier, she was diagnosed with stage 2 immunoglobulin G kappa–type multiple myeloma (MM) and underwent chemotherapy with bortezomib, thalidomide, and dexamethasone (VTD). Autologous peripheral blood stem cell transplant was planned as further treatment. On hospital admission, the patient presented signs and symptoms of decompensated heart failure (New York Heart Association functional class III). She reported recurrent syncopal episodes, the course of which suggested that orthostatic hypotension was the underlying cause, but arrhythmias could not be clearly ruled out. Blood tests revealed an increased serum brain natriuretic peptide level of 218 pg/ml (reference range <132 pg/ml) and a high-sensitivity cardiac troponin I concentration of 0.07 ng/ml (reference range <0.03 ng/ml). Twenty-four-hour Holter monitoring revealed first-degree atrioventricular block, nonsustained ventricular tachycardia (VT; 115 bpm, 7 beats), and paroxysmal atrial tachycardia (100 bpm, lasting almost 7 hours) (FIGURE 1A). Transthoracic echocardiography (TTE) showed left atrial enlargement, biventricular wall thickening with sparkling myocardial appearance, an apical "sparing" pattern in the left ventricular (LV) global longitudinal strain assessment, preserved 3-dimensional LV ejection fraction (60%), and LV diastolic dysfunction (FIGURE 1B).[1] Magnetic resonance imaging confirmed TTE findings and additionally revealed an increased extracellular volume fraction, a significant increase in T_1 and T_2 relaxation times, global late gadolinium enhancement within the right and left ventricles, and an abnormal pattern of myocardial nulling in the inversion scout sequence, which are typical features of cardiac amyloidosis (CA) (FIGURE 1C–1E).[2] Clinical, laboratory, and imaging findings provided a definitive diagnosis of monoclonal immunoglobulin light-chain amyloidosis (AL amyloidosis) as a complication of MM with dominant cardiac involvement. A single-lead proMRI implantable cardiac device (ICD) was inserted (Intica 5 VR-T DX, Biotronik, Berlin, Germany). Three months later, high-dose melphalan-based chemotherapy was administered and autologous peripheral blood stem cell transplant was performed, resulting in complete MM remission. Six months after implantation, 3 episodes of nonsustained VT (most severe: 188 bpm, 18 beats; FIGURE 1F) were revealed at routine ICD follow-up, and TTE showed no changes compared with the previous examination (Supplementary material, *Video S1–S4*).

There are currently no European guidelines on ICD treatment in patients with CA. It has been suggested that, in the case of cardiac involvement, the prognosis is poor and, besides, bradyarrhythmias or pulseless electrical activity can cause sudden death.[3] However, thanks to modern therapies, the prognosis in AL amyloidosis has significantly improved.[4] Lin et al[5] showed that the rate of appropriate ICD shocks in patients with CA was 32% in the first year after implantation, but no translation into overall survival benefit was seen, which may suggest that eligibility evaluation for ICD treatment was imprecise.

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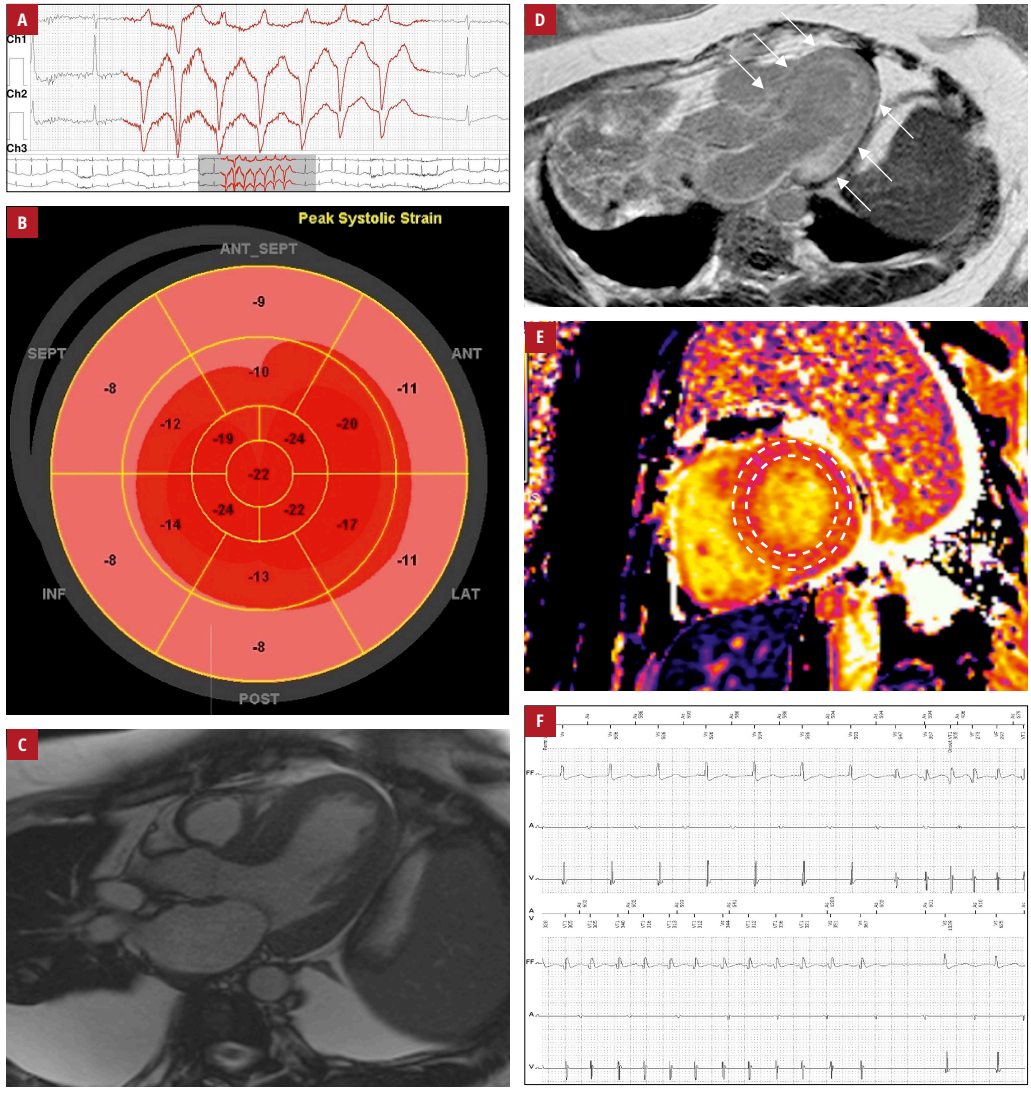

FIGURE 1 **A** – 24-hour Holter monitoring: nonsustained ventricular tachycardia at 115 bpm; **B** – transthoracic echocardiography (Vivid S70, GE Healthcare, Chicago, Illinois, United States): the apical "sparing" pattern in the left ventricular global longitudinal strain assessment; **C** – magnetic resonance imaging (MRI; Siemens Aera 1.5 T, Erlangen, Germany): a long-axis still frame from routine, balanced steady-state free precession cine sequence, showing marked myocardial thickening and a large amount of fluid in the pleural spaces; **D** – MRI (Siemens Aera 1.5 T), phase-sensitive inversion recovery sequence, showing the extent and distribution of late gadolinium enhancement: massive diffuse subendocardial and intramural late gadolinium enhancement involving the entire left ventricle (arrows); **E** – MRI (Siemens Aera 1.5 T), modified Look–Locker inversion recovery sequence: markedly increased myocardial native T_1 relaxation time (mean global region of interest T_1, 1231 ms; reference range, 993 ±21 ms). The extracellular volume fraction calculated based on native and postcontrast (not shown) T_1 values was also markedly increased at 45% (reference range, 26% ±2%); **F** – an intracardiac electrocardiogram showing nonsustained ventricular tachycardia recorded by an implantable cardioverter-defibrillator

In conclusion, the final decision to implant an ICD in a patient with CA should be taken on a case-by-case basis, probably with a lower threshold for implantation in patients with syncope of unknown origin. This was the case in the presented patient, who had only relative indications for ICD placement, but ICD monitoring confirmed the proarrhythmogenic state and recurrent nonsustained VT already at 6 months of follow-up. There is a need to identify risk factors for ventricular tachyarrhythmias and sudden cardiac death in this group of patients.

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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