

Recovery from heart failure in a patient with cardiac amyloidosis treated with autologous stem cell transplantation

Amyloidoza serca — wczesne rozpoznanie i leczenie poprawia funkcję serca. Obserwacja dwuletnia

Monika Gawor¹, Łukasz Mazurkiewicz^{1, 2}, Blanka Milanowska¹, Jacek Grzybowski¹

¹Department of Cardiomyopathy, Institute of Cardiology, Warsaw, Poland

²Magnetic Resonance Unit, Institute of Cardiology, Warsaw, Poland

A 40-year-old female, diagnosed by renal and bone-marrow biopsies with light chain amyloidosis, was referred to our institution with suspicion of cardiac amyloidosis, for further evaluation. She suffered from progressive fatigue and exertional dyspnoea (NYHA class II at initial presentation). On physical examination the patient was normotensive, there was mild

oedema of lower extremities, jugular veins were normal, lungs were clear on auscultation, and the liver was not enlarged. The baseline N-terminal pro B-type natriuretic peptide (NT-proBNP) level was 3430 (range 0–125) pg/mL, and troponin T was 16.7 (range 0–14) ng/L. No significant pathology was present on chest X-ray. The standard 12-lead electrocardiogram demonstrated sinus rhythm with low QRS voltage in the limb leads, QS complexes in V1–V3 leads, and nonspecific ST-T wave changes (Fig. 1). Cardiac magnetic resonance (CMR) scan revealed the presence of concentric left ventricular (LV) hypertrophy with a maximal wall thickness of 18 mm and increased myocardial mass indexed for the body surface area (LV mass index [LVMI] 91 g/m², range 48–77). Systolic function of the LV was slightly decreased with an ejection fraction (EF) of 59% and with compromised mitral annular plane systolic excursion (MAPSE 5 mm). The right ventricular function was also impaired with diminished tricuspid annular plane systolic excursion (TAPSE 14 mm). Additionally, small pericardial effusion was found. Late gadolinium enhancement (LGE) was present in the subendocardial part of all segments of LV. Two years after autologous stem cell transplantation (ASCT) preceded by three cycles of bortezomib and dexamethasone, the patient was completely asymptomatic. Physical examination was normal with no peripheral oedema. NT-proBNP level decreased (1620 pg/mL). Repeated CMR scan showed reduction of the maximal LV wall thickness to 16 mm (Fig. 2) and decrease of LVMI (80 g/m²) with improvement in both LV (EF 72%, MAPSE 14 mm) and right ventricle function (TAPSE 20 mm). Moreover, the remote regression of the subendocardial areas of LGE was found (Fig. 3). Prognosis of patients with light chain amyloidosis is poor with a median survival of eight months in the absence of treatment [Muchtart E et al., *Acta Haematologica*, 2016; 135: 172–190]. Survival depends mostly on the severity of organ involvement, particularly the presence of cardiac amyloidosis, and on the haematological response to therapy. This case shows that early diagnosis and intensive treatment including ASCT improve cardiac function and prolong survival.

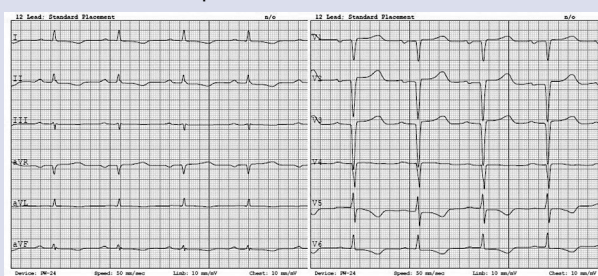


Figure 1. The standard 12-lead electrocardiogram showing sinus rhythm with low QRS voltage in the limb leads, QS complexes in V1–V3 leads, and nonspecific ST-T wave changes in the limb and precordial leads

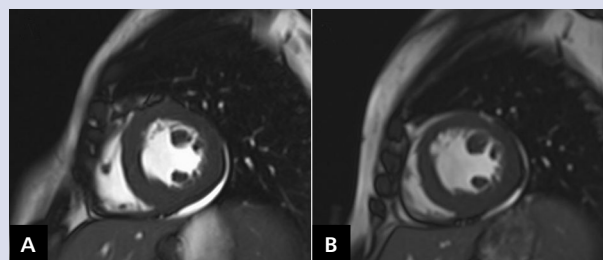


Figure 2. Baseline cardiac magnetic resonance (CMR) scan (A) and two years after autologous stem cell transplantation CMR scan (B) showing reduction of the left ventricular wall thickness

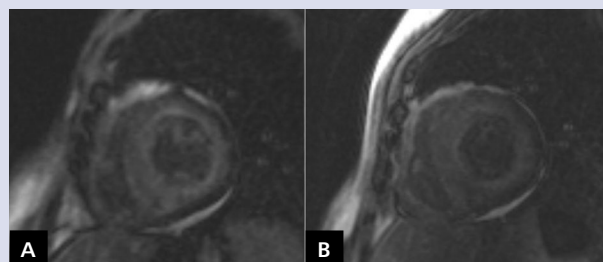


Figure 3. Baseline cardiac magnetic resonance (CMR) scan (A) and two years after autologous stem cell transplantation CMR scan (B) showing remote regression of the subendocardial areas of late-gadolinium-enhancement

Address for correspondence:

Monika Gawor, MD, Department of Cardiomyopathy, Institute of Cardiology, ul. Alpejska 42, 04-628 Warszawa, Poland, e-mail: mgawor@ikard.pl

Conflict of interest: none declared

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