Massive atrial thrombus in sinus rhythm cardiac amyloidosis is not a wild goose chase?

Piotr Gościniak1, 2, Bartłomiej Baumert3, 4, Justyna Rajewska-Tabor 5, Joanna Jędrzychowska-Baraniak1, Małgorzata Pyda6, Bogusław Machaliński3, 4

1Laboratory for Non-Invasive Cardiac Imaging, Tomasz Sokolowski Public Hospital, Pomeranian Medical University, Szczecin, Poland
2Department of Endocrinology and Internal Diseases, Pomeranian Medical University, Szczecin, Poland
3Department of Pathology, Pomeranian Medical University, Szczecin, Poland
4Department of Hematology and Transplantology, Pomeranian Medical University, Szczecin, Poland
51st Department of Cardiology, Poznań University of Medical Sciences, Poznań, Poland

The recent diagnostic guidelines and therapeutic recommendations [1] in patients with cardiac amyloidosis (CA) have changed the approach to modern cardiological diagnosis of the causes of myocardial hypertrophy while facilitating hematological evaluation [2].

We present probably the first case in the Polish population of a massive thrombus of the left atrium appendage in a patient with IgG lambda multiple myeloma with concomitant CA in the presence of sinus rhythm.

In a 54-year-old woman, a significant deterioration in exercise tolerance (New York Heart Association [NYHA] class III), hypotension, and weight loss were observed for about a year. Echocardiography revealed concentric myocardial hypertrophy, good left ventricular systolic function, and impaired grade-II diastolic function. After five months, due to the persistence of symptoms, coronary angiography was performed, which showed no narrowing of the coronary arteries. Due to the lack of clinical improvement, the patient was referred to the cardiac magnetic resonance unit. T1 time mapping during stress perfusion was carried out before and after a gadobutrol injection. Cine images showed a decreased ejection fraction (41%) and concentric hypertrophy of the myocardium. Native T1 sequences and extracellular volume (ECV) maps showed diffused fibrosis with high native T1 values (mean, 1095 ms; normal values, 953–981 ms) and extended ECV (mean 51%) (Figure 1A, B). The late gadolinium enhancement images revealed subendocardial and transmural fibrosis.

The diagnostics were extended to a histopathological examination of the adipose tissue, in which amyloid deposits were found, and the bone marrow examination, which showed 15% infiltration of clonal plasma cells. Specific tests showed: protein M 1.17 g/dl, free light chain (FLC) λ 330 mg/l, FLC ratio κ/λ 0.03, monoclonal protein IgG λ and trace λ band in serum immunofixation, troponin T 31.78 pg/ml, N-terminal pro-brain natriuretic peptide (NT-proBNP) 2544 pg/ml, IgG 19.1 g/l, and urine protein 16.9 mg/dl. The treatment included four cycles of cyclophosphamide, bortezomib, and dexamethasone (CyBorD). Hematological response at the PR level was achieved, with a reduction in the NT-proBNP level from 2653 to 2037 pg/ml in the cardiac response.

The resting electrocardiography showed a sinus rhythm and a low voltage of the QRS complexes. The observation showed no atrial fibrillation. Echocardiography revealed a granular structure of the myocardium, left and right ventricular hypertrophy, a thickened interatrial septum, pericardial fluid, an apical sparing in global longitudinal strain (Figure 1C), and impaired diastolic function of the LV. The peak longitudinal left atrial strain was reduced (Figure 1D). Moreover, an additional echo in the left atrial appendage (LAA) was observed.

For this reason, 3D transesophageal echocardiography was performed, revealing a massive thrombus modeling the entire LAA (Figure 1E, F). Apixaban treatment was initiated and autologous hematopoietic stem
cell transplantation (aHSCT) was temporarily postponed. On control transesophageal echocardiography at 8 weeks, only self-contrast in the blood was observed in the LAA. Nowadays the patient is undergoing aHSCT with melphalan 200 mg/m² conditioning and low molecular weight heparin prophylaxis.

The most common form of amyloidosis is light chain amyloidosis, which accounts for about 70%–80% of all forms of the disease [3]. In untreated light-chain cardiac amyloidosis, survival is estimated to be less than six months from diagnosis. The prognosis may also be influenced by the presence of a thrombus in the LAA despite the presence of a sinus rhythm [4, 5]. Therefore, detailed and modern cardiac imaging should become the standard approach.

**Article information**
**Conflict of interest:** None declared.
**Open access:** This article is available in open access under Creative Common Attribution-Non-Commercial-No Derivatives 4.0 International (CC BY-NC-ND 4.0) license, allowing to download articles and share them with others as long as they credit the authors and the publisher, but without permission to change them in any way or use them commercially. For commercial use, please contact the journal office at kardiologiapolska@ptkardio.pl.

**REFERENCES**


**Figure 1.** A. Diffusely increased T1 values on CMR native T1 map (1095 ms), “pericardial effusion, short axis view. B. Increased ECV values (51%) in extracellular volume map, short axis view. C. TTE global longitudinal strain — “apical sparing”. D. TTE reduced longitudinal left atrial strain. E. TEE — an additional echo in the LAA (the red arrow). F. 3D TEE — a massive thrombus (the red arrow) modeling the LAA

Abbreviations: CMR, cardiac magnetic resonance; ECV, extracellular volume; LAA, left atrial appendage; TEE, transesophageal echocardiography; TTE, transthoracic echocardiography.