Systemic wild-type transthyretin amyloidosis combined with valvular and ischemic cardiomyopathy

Tereza Knaflec[1]*, Siniša Roginić[1], Ivan Jakšić[2], Martina Roginić[1], Marija Čajko[1], Nikolina Mijač Mikačić[1], Domagoj Futivić[1]

[1]General Hospital Zabok and Hospital of Croatian Veterans, Zabok, Croatia
[2]University Hospital Centre Sestre Milosrdnice, Zagreb, Croatia

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***ADDRESS FOR CORRESPONDENCE**: Tereza Knaflec, Opća bolnica Zabok i bolnica hrvatskih veteran, Bračak 8, HR-49210 Zabok, Croatia. / Phone: +385-98-318-813 / E-mail: tknaflec@gmail.com

ORCID: Tereza Knaflec, https://orcid.org/0000-0002-4915-3935 • Siniša Roginić, https://orcid.org/0000-0002-0384-8088 Ivan Jakšić, https://orcid.org/0000-0001-8776-0560 • Martina Roginić, https://orcid.org/0000-0001-5463-5392 Marija Čajko, https://orcid.org/0000-0001-7084-707X • Nikolina Mijač Mikačić, https://orcid.org/0000-0002-0933-6577 Domagoj Futivić, https://orcid.org/0000-0003-4363-1008

Aim: To show a case of systemic wild-type transthyretin amyloidosis (wtATTR) combined with valvular and ischemic cardiomyopathy.

Case report: 78-year-old man presented with worsening of congestive chronic heart failure. Medical history includes arterial hypertension, stage 3b chronic kidney disease, coronary artery disease, hypothyroidism, syndrome Raynoud (**Figure 1**) and implantation of ICD in secondary prevention. The electrocardiogram showed atrial fibrillation and right bundle branch block. Transthoracic echocardiography (**Figure 2, Figure 3**) showed reduced left ventricle ejection fraction, biventricular wall thickening[1], global and segmental hypokinesia of inferior wall and basal inferoseptum, decreased global longitudinal systolic function; moderate calcified aortic stenosis[2]; biatrial enlargement, moderate mitral and severe secondary tricuspid regurgitation. Based on the absence of monoclonal protein, cardiac scintigraphy was performed with injection of technetium-based compound which confirmed the diagnosis of amyloid transthyretin cardiomyopathy[3] (**Figure 4 A and B**). Furthermore, recoronarography excluded progression of coronary artery disease. Peripheral polyneuropathy consistent

FIGURE 1. Raynaud's syndrome; pallor 5th finger of the left hand and cyanotic hands.

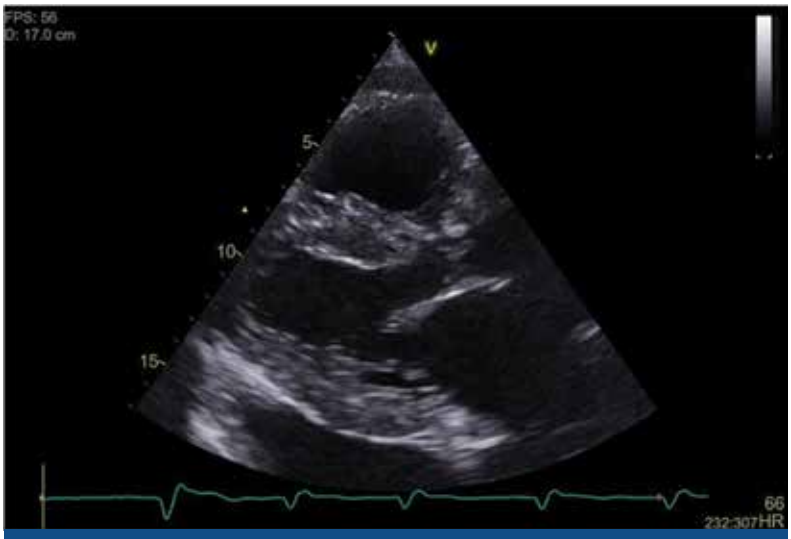

FIGURE 2. Parasternal long axis view: small left ventricle cavity, left atrial enlargement, biventricular wall thickening.

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with amyloidosis was also diagnosed. Guideline based heart failure management resulted in rapid recovery and after discharge patient was ambulatory (NYHA III). He was adherent to therapy without side-effects typical for cardiac amyloidosis (hypotension, etc.) probably due to combined aetiology of cardiomyopathy. Tafamidis[4] is the only medication approved for the treatment of wtATTR cardiomyopathy, slowing the dissociation of transthyretin and further progression of the disease, reducing all-cause mortality and cardiovascular-related hospitalizations compared to placebo. Unfortunately it is not indicated in advanced heart failure present in our patient.

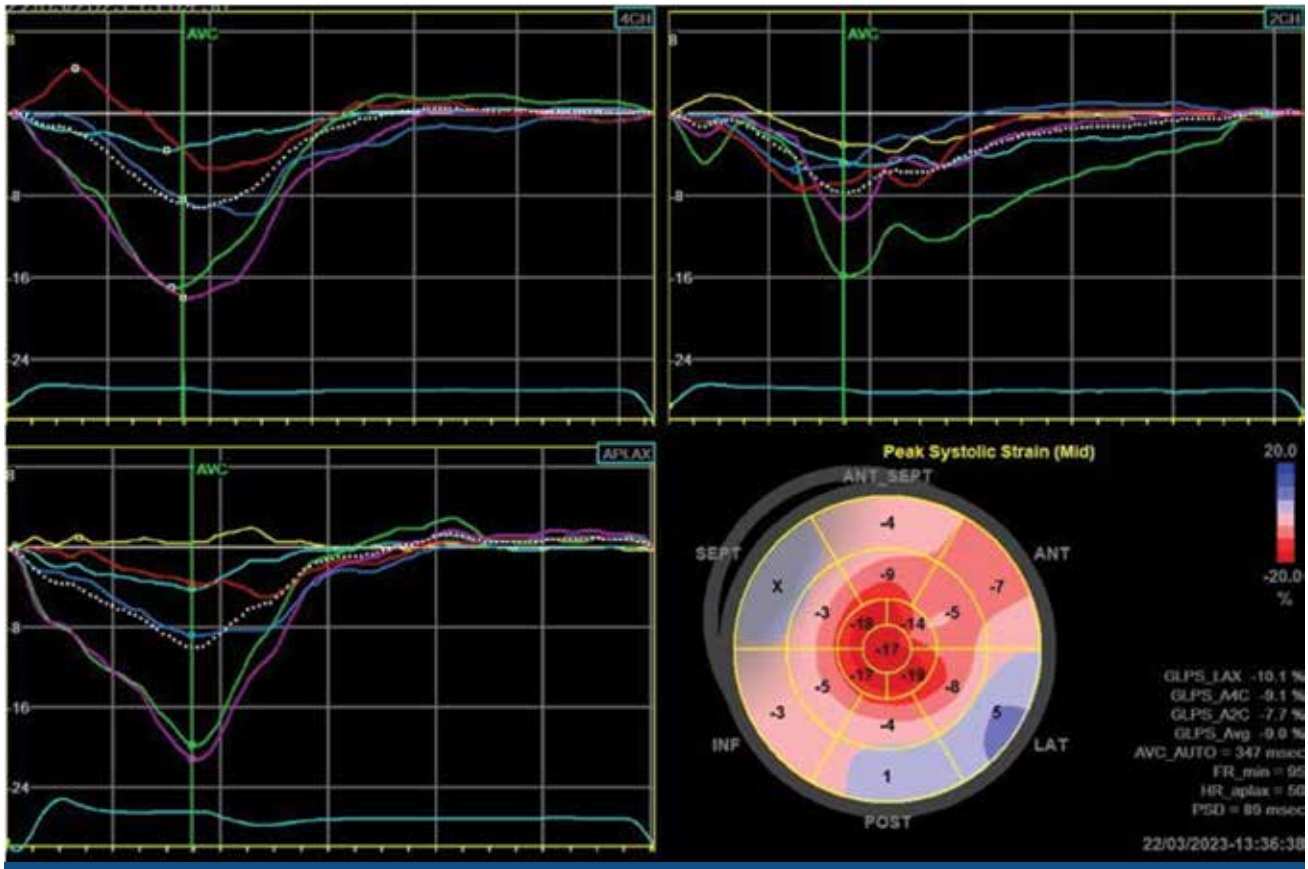

FIGURE 3. Speckle tracking echocardiography: severely reduced global longitudinal strain (-9.0%) with an "apical sparing"/"cherry on the top" pattern.

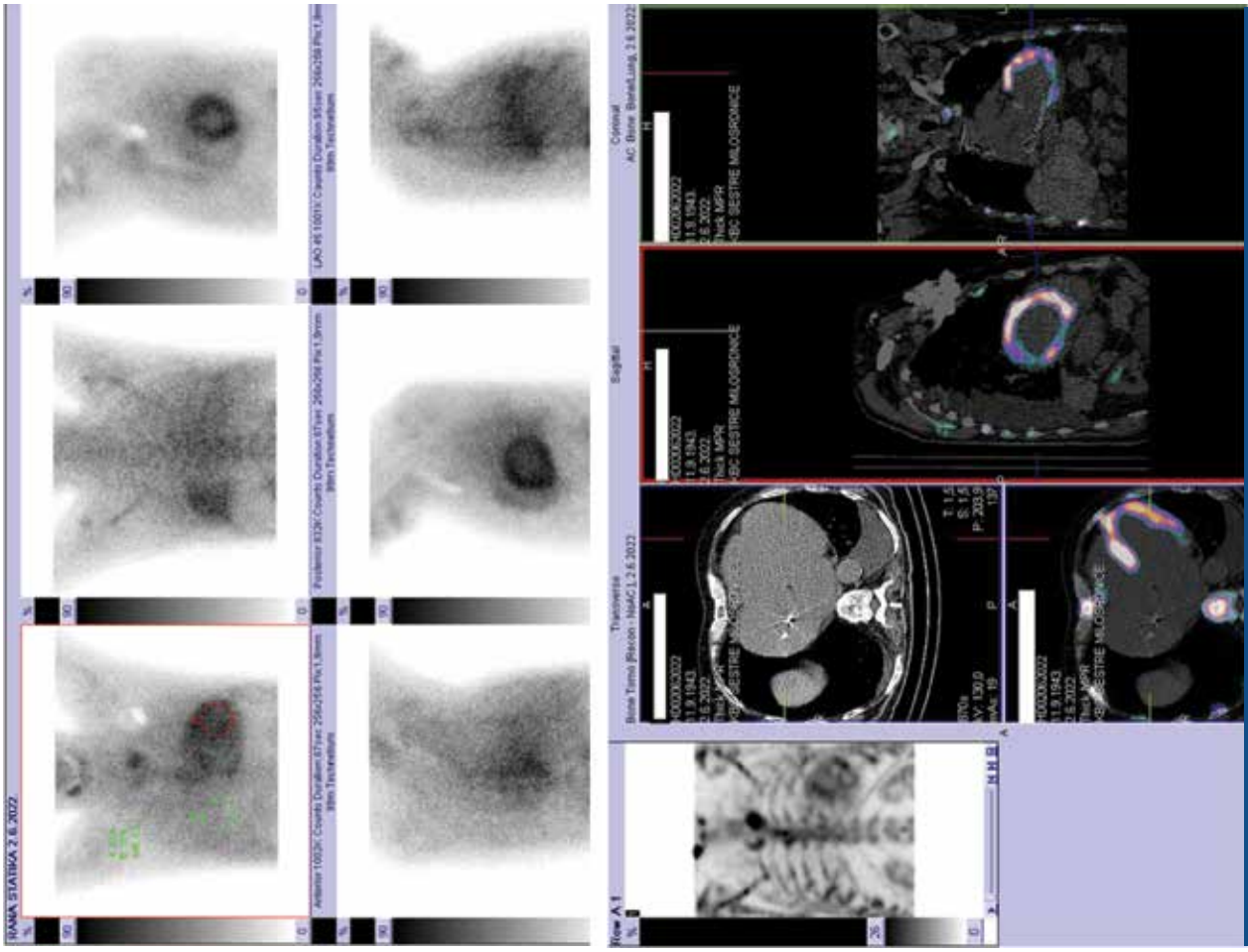

FIGURE 4. Cardiac scintigraphy and SPECT/CT; A. Static scintygraphy images showing heart/contralateral tracer uptake ratio >1.5. B. SPECT/CT confirmation that tracer is accumulating in myocardium.

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