

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

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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 Raynaud (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 thickening¹, global and segmental hypokinesia of inferior wall and basal inferoseptum, decreased global longitudinal systolic function; moderate calcified aortic stenosis²; 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 cardiomyopathy³ (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. Tafamidis⁴ 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.

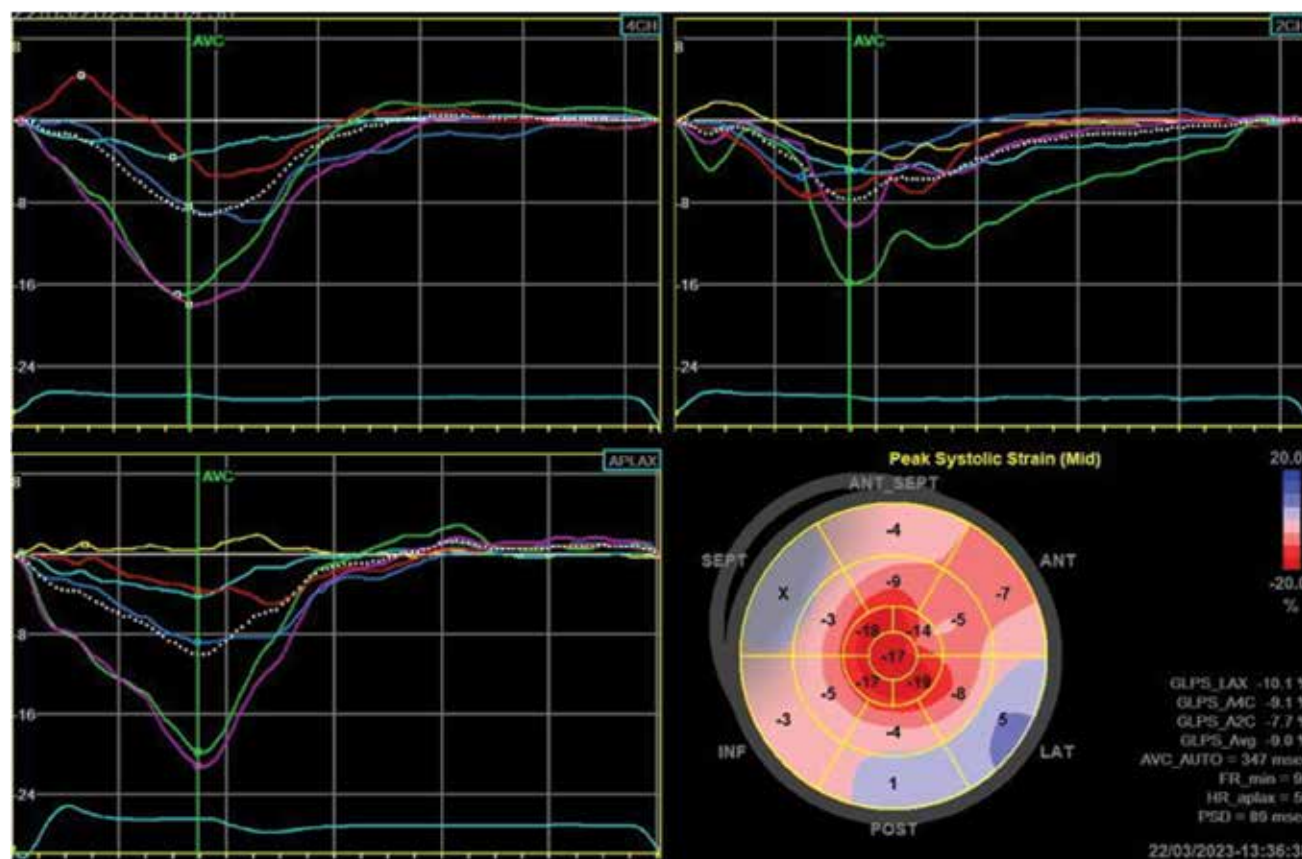


FIGURE 3. Speckle tracking echocardiography: severely reduced global longitudinal strain (-9.0%) with an “apical sparing”/“cherry on the top” pattern.

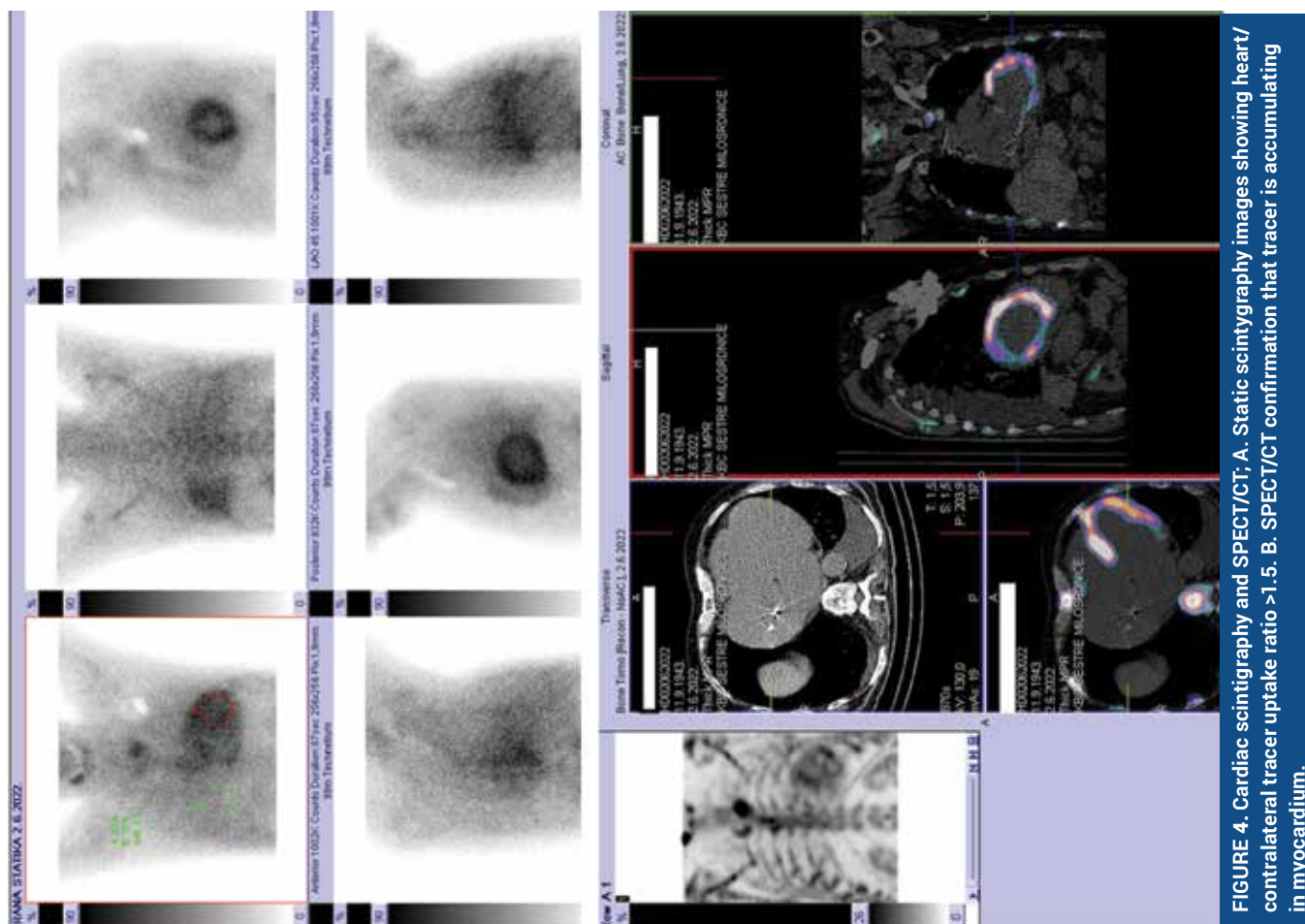


FIGURE 4. Cardiac scintigraphy and SPECT/CT; A. Static scintigraphy images showing heart/contralateral tracer uptake ratio >1.5. B. SPECT/CT confirmation that tracer is accumulating in myocardium.

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