Nuclear medicine imaging with 99mTc-Pyrophosphate scintigraphy in patients with suspected cardiac amyloidosis

Marina Prpić*, **Danijel Planinc**, **Maja Franceschi**

University Hospital Centre "Sestre milosrdnice", Zagreb, Croatia

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***ADDRESS FOR CORRESPONDENCE:** Marina Prpić, Klinički bolnički centar Sestre milosrdnice, Vinogradska 29, HR-10000 Zagreb, Croatia. / Phone: +385-1-3787-111 / E-mail: marina_prpic@yahoo.com

ORCID: Marina Prpić, http://orcid.org/0000-0002-0635-3806 • Danijel Planinc, http://orcid.org/0000-0002-1799-8554 Maja Franceschi, http://orcid.org/0000-0002-7427-3875

Introduction: Amyloidosis is an infiltrative disease characterized by extracellular deposition of insoluble fibrillary protein. There are two major types: light-chain (AL) amyloidosis and transthyretin-related cardiac amyloidosis (ATTR, mutant and wild type). Cardiac involvement can lead to restrictive cardiomyopathy (CMP) and heart failure. 99mTc-Pyrophosphate (99mTc-PPY) has high affinity for TTR amyloid, allowing differential diagnosis with AL and other nonamyloidotic CMP with hypertrophic phenotype, in which 99mTc-Pyrophosphate (PYP) is low or absent.[1-4]

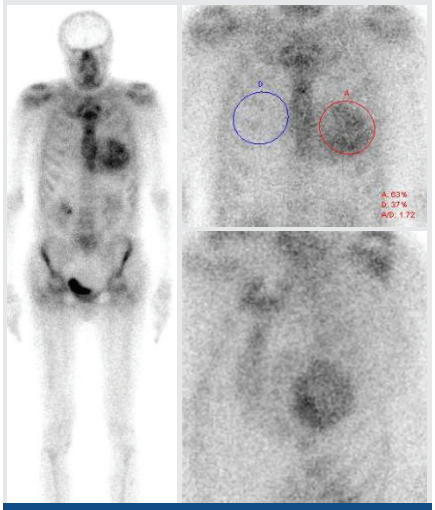

FIGURE 1. Transthyretin-related cardiac amyloidosis. The planar 99mTc-Pyrophosphate scintigraphy (whole body, anterior and left oblique images) showing diffusely intense myocardial activity, greater than in ribs and sternum; regions of interest drawn over the myocardium and the contralateral side show heart/contralateral uptake ratio of 1.72.

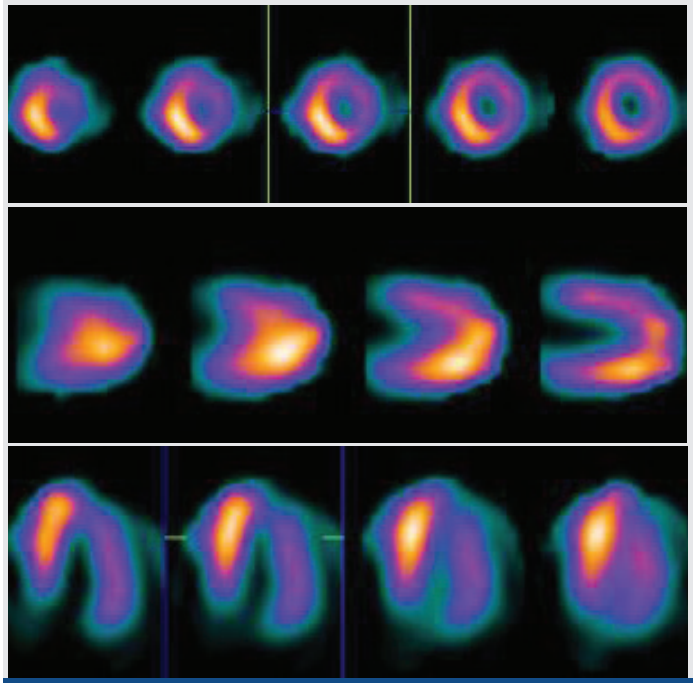

FIGURE 2. Reconstruction of single photon emission CT images in short and long axes show uniform and intense myocardial uptake of 99mTc-Pyrophosphate, most prominent in septum and inferior wall.

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Patiens and Methods: We are presenting four patients with confirmed diagnosis of cardiac amyloidosis, as follows: 54-year-old lady hospitalized for evaluation of progressive dyspnea and chest pain; 69-year-old lady, with history of hypertension, hospitalized for congestive heart failure; 50 and 64 old gentlemen with progressive dyspnea and intolerance of exertion. Whole body, anterior and lateral static images and SPECT/CT with heart centered in the field of view, were performed 1 and 3 hours after intravenous injections of 740MBq (20mCi) 99mTc-PYP.

Results: In the first patient scintigraphic planar and WB images (**Figure 1**) showed intense accumulation of tracer in myocardium in comparison to bone uptake (ribs and sternum). Heart-to-contralateral (H/CL) ratio=1.72. Myocardial SPECT revealed tracer uptake in entire myocardium, with highest uptake in the apex, septum and inferior wall (**Figure 2**). The finding was highly suggestive for TTR, also confirmed by genetic testing. In other three patients, only mild accumulation of tracer in myocardium was found, similar or lower to bone uptake, with highest uptake in the apex (SPECT/CT images) and

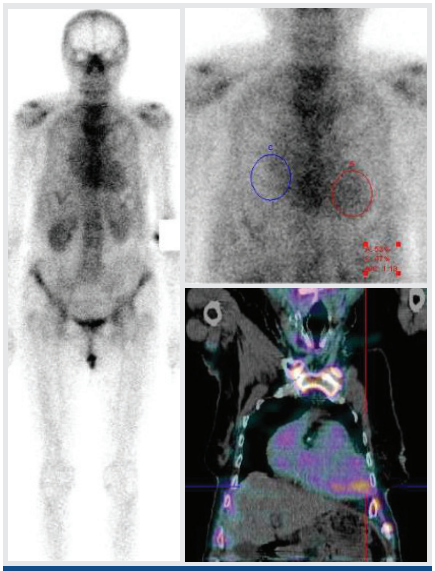

FIGURE 3. Light chain amyloidosis. 99mTc-Pyrophosphate scintigraphy (whole body and anterior) showing moderate uptake in myocardium, similar to bone; heart/contralateral uptake reveals 1.13; SPECT/CT coronal image show area of increased activity in apex.

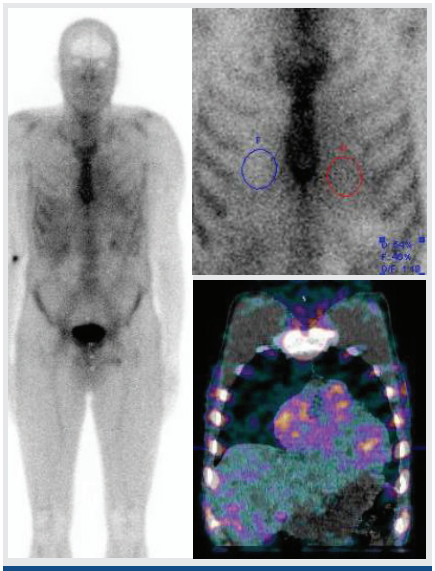

FIGURE 4. Light chain amyloidosis. Whole body and anterior bone 99mTc-Pyrophosphate scintigraphy showing mild uptake in myocardium; heart/contralateral uptake reveals 1.13; c) SPECT/CT coronal image show regional area of increased activity in apex.

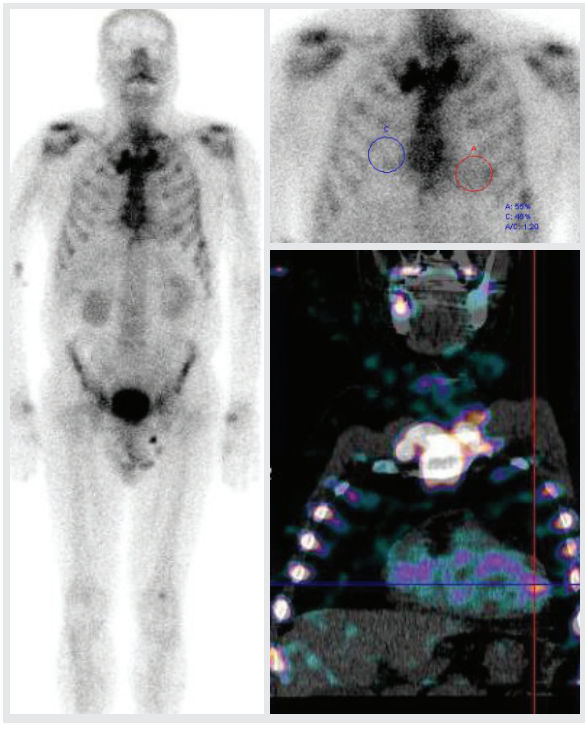

FIGURE 5. Light chain amyloidosis. Planar 99mTc-Pyrophosphate images show diffuse uptake in myocardium; heart/contralateral uptake reveals 1.20; SPECT/CT coronal image show area of increased activity in apex.

consecutive H/CL ratios 1.13, 1.13 and 1.23 (**Figures 3**, **4**, and **5**). The finding was suggestive for AL type. Multiple myeloma (IgG lambda type) was confirmed by bone marrow biopsy with phenotypisation. Endomyocardial biopsy was not performed in all pts.

Conclusion: According to recent recommendations, cardiac 99mTc-PYP scintigraphy using H/CL ratio quantitative score and SPECT/CT is a simple non-invasive imaging technique that can facilitate diagnosis of cardiac amyloidosis and differentiate AL from ATTR, thus avoiding invasive endomyocardial biopsy.

LITERATURE

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