Scintigraphic images of transthyretin cardiac amyloidosis progression

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Scintigraphic images of transthyretin cardiac amyloidosis progression

Scintigraphic images of ATTR CA

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In Cardiac amyloidosis (CA) myocardial infiltration of amyloid fibrils causes structural and functional changes that determine heart failure (HF), angina and arrhythmias (atrial fibrillation, affection of the conduction system). [1]

Light chain amyloidosis (AL) and transthyretin amyloidosis ATTR (wild type or hereditary) account for most cases of heart involvement. [2]

There is an increasing interest in using noninvasive cardiac imaging techniques to diagnose CA. Echocardiography can demonstrate a variable range of diastolic abnormalities in the setting of Left Ventricular wall thickening and the apical sparing strain pattern may differentiate CA from other causes of Left Ventricular Hypertrophy (LVH).

In Cardiac Magnetic Resonance (CMR), Late Gadolinium Enhancement (LGE), T1 mapping and Myocardial Extracellular Volume (MEV) are the techniques used for the diagnosis and the prognosis of CA. [4]

Studies demonstrated that (99m) Tc-3,3-diphosphono-1,2propanodicarboxylic acid (99mTc-DPD) and (99m) Tc-pyrophosphate (99mTc-PYP) have high affinity for ATTR uptake and thus can differentiate this form from AL. The mechanism of uptake seems related to the high calcium content in ATTR amyloid deposit. For these reasons, nuclear scintigraphy may ultimately obviate the need of biopsy to diagnose ATTR-CA. [5]

Here we describe an 84-year-old man with a past medical history of diet-controlled diabetes mellitus type 2, hypertension, HF and PCI for Acute Coronary Syndrome (ACS) occurred three years before. In this occasion, Troponin T high-sensitivity assay (hs-TnT) value was 90 ng/l (15 ng/l) and Trans Thoracic Echocardiography (TTE) showed: LV wall thickening (13 mm), preserved Left Ventricular Ejection Fraction (LVEF 55%) and signs of abnormal diastolic relaxation pattern. A previous TTE performed twelve years before, had a similar
pattern of LVH attributed to a long standing Hypertension. 99mTc-DPD scintigraphy did not demonstrate significative cardiac uptake suggestive for ATTR (Figure 1-a).

At the admission to our Institution, he had a painful and swelling right shoulder, lumbar pain and loss of strength of the legs. Blood Cultures were positive for a Methicillin Sensitive Staphylococcus Aureus and Spinal Magnetic Resonance imaging was consistent with L4-5 spondylodiscitis. TTE did not revealed signs of endocarditis, but showed a left ventricular wall thickening (14 mm), LVEF 48 %, left atrial dilatation and moderate Mitral Valve regurgitation. During the hospitalization, he developed an acute cardiogenic pulmonary edema treated with high dose of diuretics and Non-Invasive Mechanical Ventilation (NIMV). A Trans Esophageal Echocardiography (TEE) demonstrated a severe functional mitral regurgitation without signs on valvular endocarditis. After two weeks, the patients progressively improved and the further TTE revealed a significative reduction of mitral regurgitation. Before the discharge, he underwent to 99mTc-DPD scintigraphy that was significative for ATTR (Figure 1-b). The laboratory examination demonstrated: Natriuretic Peptide (NT-proBNP) 18628 ng/l, hs-TnT 188 ng/l. According to the patient’s age, which made the diagnosis of wt-ATTR more likely, we did not performed any genetic investigation.

In ATTR, scintigraphy repetition could identify a greater myocardial amyloid burden that determines a clearer diagnostic imagine. Even if the specific wt-ATTR therapy is actually only supportive, the diagnosis allows a better prognostic stratification and therapeutic strategies.
REFERENCES


Figure 1 A-B. Comparison between two scintigraphic images (99mTc-DPD) made three years apart