Cardiac Computed Tomography for the detection of cardiac amyloidosis

Short title: Cardiac CT for detection of cardiac amyloidosis

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Conflicts of interest

None

A 67-year-old Caucasian man presented with atrial flutter on routine 12-lead electrocardiography (ECG). Following appropriate anticoagulation, he successfully underwent atrial flutter ablation. Echocardiography during the admission showed concentric left ventricular (LV) hypertrophy with severely impaired LV systolic function, particularly longitudinal (Figure 1A, video 1), and biatrial dilatation. Due to episodes of non-sustained ventricular tachycardia, he was fitted with an implantable cardioverter-defibrillator (ICD). The ECG QRS complexes on peripheral leads were small compared to the structural wall thickening on echocardiography (Figure 1B) raising the suspicion of cardiac amyloidosis. Preliminary evaluation excluded a plasma cell dyscrasia (negative serum free light chains changes and urinary Bence-Jones protein). Serum amyloid P component (SAP) scintigraphy was negative for visceral organ uptake and bone tracer scintigraphy (Technetium-DPD) was positive with Perugini Grade 2-3 uptake (Figure 1C), consistent with a diagnosis of cardiac transthyretin amyloidosis (ATTR). Cardiac magnetic resonance (CMR) was contra-indicated due to the non-conditional ICD (Figure 1D). After giving fully informed written consent, cardiac computed tomography (CCT) was performed as part of a research study, using a three step protocol (Figure 1E) as previously described¹, allowing calculation of the extracellular volume fraction (ECV) from pre- and 5-minutes post-contrast images. The septal ECV, calculated from a co-registered, segmented myocardial mask displaying pixel-by-pixel ECV values, was in the amyloid spectrum elevated at 63% (Figure 1F), Gene sequencing revealed a heterozygous V122I mutation, confirming the diagnosis of familial amyloid cardiomyopathy (ATTR).

Discussion

Quantification of the myocardial ECV using two-phase cardiac CT can discriminate between cardiac amyloidosis and other disease with myocardial hypertrophy. The ECV by CMR and CT strongly correlated, making CCT a valid alternative to CMR¹, which itself is known to

have good sensitivity and specificity for the diagnosis, prognostication and monitoring of cardiac amyloidosis². Here, CCT added value as there was a MR non-conditional ICD in-situ.

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References

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- 2. Fontana M, Pica S, Reant P, et al. Prognostic Value of Late Gadolinium Enhancement Cardiovascular Magnetic Resonance in Cardiac Amyloidosis. *Circulation*. 2015;132:1570-1579.

