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Cardiac amyloidosis: the role of non-invasive imaging

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Topic(s):
Cross-Modality and Multi-Modality Imaging Topics

Citation:

Introduction: Cardiac Amyloidosis is a potentially fatal condition and early diagnosis is essential to improve outcome. Although endomyocardial biopsy remains as gold standard for Cardiac Amyloidosis, non-invasive imaging has emerged as an important tool in this context. Case report: A 81-year-old-male with history of hypertension, chronic obstructive pulmonary disease and chronic kidney disease presented with progressive exertional dyspnea that had persisted for the previous month. A previous history of chronic artery disease and some episodes of syncope were known. Tilt test and 24 hour holter had confirmed carotid sinus hypersensitivity and paroxysmal atrial fibrillation, respectively. Patient was admitted to the cardiac care unit for acute heart failure (HF) in a Heart Association, class IV. Initial laboratory data presented high levels of NT-proBNP. Electrocardiogram demonstrated first-degree atrioventricular block and right bundle branch block. Transthoracic echocardiogram revealed enlarged left atrium and left ventricular volumes, mild concentric left ventricular hypertrophy, overall moderate systolic dysfunction (EF=41%) and diffuse hypokinesis. Impaired relaxation and elevated filling pressures with restrictive mitral inflow pattern were consistent with diastolic dysfunction grade II (pseudonormal). Mild mitral regurgitation was also noticed. Coronary angiography demonstrated non-obstructive coronary artery disease ruling out ischemic etiology. A Cardiac MRI was pursued and depicted severe biventricular systolic dysfunction and linear meso epicardium late gadolinium enhancement encompassing the interventricular septum consistent with non-ischemic fibrosis. A diagnostic of cardiac amyloidosis was suspected and 99m-Tc-Technetium-Pyrophosphate (99mTc-PYP) scintigraphy was performed. Planar images showed abnormally increased radiotracer activity in the heart with a calculated heart-to-contralateral ratio (H/CL) of 1,54 (normal < 1,5) and SPECT/CT images demonstrated abnormally increased radiotracer activity throughout the myocardium, greatest in the interventricular septum and right ventricle and less at left ventricle lateral wall consistent with transthyretin amyloidosis (ATTR). 24-hour-urine protein electrophoresis was normal. Conclusions: The authors wish to emphasize the importance of non-invasive imaging in the diagnosis, prognosis and therapeutic management of Cardiac Amyloidosis.