Value of cardiovascular magnetic resonance for determining cardiac involvement in systemic amyloidosis

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A 66-year-old woman presented with atypical chest pain, symptoms of heart failure (NYHA class III), and loss of appetite. Her past medical history was notable for previous episode of decompensated heart failure and two transient cerebral ischaemic attacks. Physical examination revealed a raised jugular venous pressure, marked peripheral oedema, hepatomegaly, and bilateral rales. An adjunct S4 heart sound was heard suggesting a stiff left ventricle. Electrocardiogram documented right bundle branch block and Q-waves in the inferior leads. Chest X-ray confirmed bilateral pleural effusions\(^+\). Echocardiogram showed asymmetric left ventricular hypertrophy (LVH), diastolic dysfunction, and pulmonary hypertension. Right and left cardiac catheterization showed unobstructed coronary arteries, pulmonary arterial pressure of 44/15 mmHg, mean 24 mmHg, and a wedge pressure of 20 mmHg.

Cardiovascular magnetic resonance (CMR) was performed to characterize her underlying cardiomyopathy. Her scan showed severely impaired biventricular systolic and diastolic function, concentric LVH (maximal wall thickness 18 mm), a small pericardial effusion, and bilateral pleural effusions (Panels A and B). In the cine images, the presence of a large mobile thrombus was seen in the left atrial appendage (Panel C) and confirmed by the early inversion-recovery imaging following gadolinium-DTPA contrast administration (Panel D). On the delayed enhancement images, the blood pool was characteristically dark and there was circumferential late enhancement of the endocardial and mid-wall layers suggestive of an extensive infiltrative process and in particular cardiac amyloidosis (Panels E and F). Subsequently, a serum amyloid P component (SAP) scan was performed showing amyloid deposition in the spleen and bone marrow (Panel G). Serum blood tests showed an IgG lambda serum paraproteinaemia and serum-free light chain assay confirmed an excess of lambda light chains. Primary systemic AL amyloidosis with severe cardiac involvement was diagnosed.

Cardiac involvement is the cause of death in approximately 50% of patients with AL amyloidosis and is associated with overt congestive heart failure. SAP scintigraphy is an effective non-invasive tool for diagnosis of systemic AL amyloidosis but it is considered inadequate for evaluating the heart. In cardiac amyloidosis, CMR presents a characteristic pattern of circumferential subendocardial late enhancement that is related to the histological distribution of amyloid protein. The potential of CMR in detecting cardiac amyloid load may suggest diagnostic value of this innovative non-invasive imaging technique and may yield an opportunity to assess therapeutic response.