Truth will out: a rare case of amyloid cardiomyopathy

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DESCRIPTION

A 65-year-old patient with no medical history of notice was referred to our cardiology outpatient clinic for the evaluation of exertional dyspnoea. After a thorough work-up, the suspicion of an amyloid cardiomyopathy was raised by the cardiac magnetic resonance (CMR) findings (figure 1A–C), but laboratory tests, abdominal fat pad and bone marrow biopsies failed to prove the diagnosis. Eventually endomyocardial biopsies confirmed the diagnosis of secondary (AA) amyloidosis. Furthermore, the CMR scout images unveiled a large abdominal mass (figure 1D), with subsequent CT being suggestive of a pancreatic neuroendocrine tumour with multiple hepatic metastases.

Clinically apparent heart disease or cardiac amyloidosis is uncommon in secondary amyloidosis. Secondary amyloidosis may complicate chronic inflammatory diseases and is rarely associated with other causes, for example, neoplasms. In the diagnosis of cardiac amyloidosis, the characteristic echocardiographic finding of an increased echogenicity of the myocardium has limited

Figure 1 Cardiac magnetic resonance images showing diffuse subendocardial late gadolinium enhancement of the left ventricle, right ventricle and atria. (A) Four-chamber image; (B) two-chamber image; (C) midventricular, short-axis image and (D) balanced steady-state free precession (SSFP) scout image showing a large abdominal mass.

specificity and sensitivity.² In contrast, CMR imaging with a distinctive late gadolinium enhancement pattern can provide evidence strongly suggestive of cardiac amyloid deposition and can be used to guide myocardial biopsies, which have a higher sensitivity compared with subcutaneous fat or rectal biopsies.^{2 3}

In our patient, the characteristic CMR findings proved to be pivotal for the diagnosis of the secondary or AA amyloidosis with cardiac manifestation. Furthermore, the CMR scout images unveiled an important incidental finding, that is, a large abdominal mass, possibly associated with the amyloidosis in this patient. ¹

Learning points

- Cardiac magnetic resonance (CMR) has a vital role in the work-up of suspected (cardiac) storage diseases.
- ► Endomyocardial biopsy has a high sensitivity for the diagnosis of cardiac amyloidosis.
- Use all CMR images, including the scout images.

Contributors JJ is the first author and wrote the manuscript. SG helped with the interpretation of the CMR and supplied the images. MG supervised the CMR reporting and supervised the clinical course of the patient as well as the manuscript.

Competing interests None declared.

Patient consent Obtained from next of kin.

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