An older man with a rare cause of heart failure

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DESCRIPTION

A previously healthy 74-year-old man presented with symptoms of heart failure over the last 10 months. Initial tests revealed raised levels of brain natriuretic peptide (617 pg/mL) and a low-voltage ECG. An echocardiogram showed symmetric left ventricular (LV) hypertrophy, systolic dysfunction (ejection fraction 35%; figure 1A, video 1), restrictive filling pattern and decreased global longitudinal strain (figure 1B). As the cardiomyopathy aetiology was unclear, he underwent further testing. A cardiac MRI demonstrated diffuse subendocardial delayed myocardial enhancement after gadolinium injection (figure 1C) and amyloid deposits were found on endomyocardial biopsy (figure 1D). The serum κ/λ free-light chains (FLC) ratio was 2.4 (increased) and a bone marrow biopsy ruled out multiple myeloma. The final diagnosis was cardiac amyloid light-chain (AL) amyloidosis and he was started on cyclophosphamide, thalidomide and dexamethasone. His congestive symptoms improved

Video 1 Transthoracic echocardiography showing reduced left ventricular ejection fraction, marked left ventricular wall thickening, and increased echogenicity of the myocardium.

with diuretics. At 15 months of follow-up, he is in New York Heart Association class II.

Figure 1 (A) Transthoracic echocardiography showing a non-dilated left ventricular cavity with increase of septal and posterior wall thickness. Myocardial 'granular sparkling' is also noticeable. (B) Bulls-eye view quantitative data for global and segmental wall motion of the left ventricle. Apical deformation is preserved (normal $-18\pm 2\%$) while the remaining segments are completely abnormal. (C) Cardiac MRI demonstrating diffuse myocardial delayed enhancement (red arrows) after gadolinium injection. (D) Endomyocardial biopsy stained with Congo red and viewed with polarising microscopy. Amyloid deposits exhibit apple-green birefringence. LV: left ventricle, RV: right ventricle.

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Only 5% of patients with unexplained cardiomyopathy are found to have amyloidosis.¹ Amyloid infiltration of the heart results in increased LV thickening seen on echocardiography (associated with reduced ECG voltage).² As this finding is indistinguishable from other causes of LV hypertrophy, the use of more advanced imaging techniques such as speckle-tracking echocardiography and cardiac MRI can provide non-invasive evidence suggestive of amyloid cardiomyopathy.³ However, the diagnosis of cardiac amyloidosis is only confirmed when the biopsy specimen stains with Congo red. In AL amyloidosis, λ or κ FLC is also detectable in serum or urine. Finally, the treatment involves the use of chemotherapy and/or autologous stem cell transplantation.

Learning points

- ▶ Cardiac amyloidosis should always be considered in patients without hypertension with left ventricle hypertrophy and low-voltage ECG.
- ▶ Cardiac MRI provides strongly suggestive evidence of amyloid cardiomyopathy.

Contributors All authors were equally involved in the planning, execution, and reporting of the work.

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