A stone heart: fatal cardiac microcalcification

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Marked cardiac uptake of 99mTc-methyl-diphosphonate was noted on a bone scan performed in a patient with end-stage renal failure, on long-term dialysis. Amyloidosis was considered a possible explanation for clinical features that included severe left ventricular hypertrophy, low-voltage ECG, atrial and ventricular arrhythmia. Cardiac magnetic resonance (CMR) imaging demonstrated an unusual pattern of delayed contrast hyperenhancement of the entire left ventricular myocardium, suggesting diffuse myocardial fibrosis. This pattern was not typical for amyloidosis. Non-contrast CT demonstrated diffusely increased Hounsfield units in the myocardium and unconventional windowing demonstrates patchy calcification in the myocardium.

The patient died of intractable ventricular arrhythmia. At autopsy, there were diffuse microcalcification and interstitial fibrosis of the left ventricular myocardium. Congo red staining for amyloid protein was negative and von Kossa stain for calcium was positive.

CMR and radionuclide pyrophosphate imaging may be abnormal in the setting of either cardiac amyloidosis or cardiac interstitial fibrosis owing to microcalcification. The risk of nephrogenic systemic fibrosis now precludes the use of gadolinium in this setting from cardiac amyloidosis.

End-stage renal failure and dialysis may be associated with ectopic calcium deposition owing to elevated serum phosphorous and calcium-phosphate (Ca × P) product and elevated parathyroid hormone. Calcium-based phosphate binding therapy may contribute to hypercalcaemia. Diffuse cardiac microcalcification may cause intractable heart failure and malignant arrhythmia, but is most often diagnosed at autopsy.

The pre-contrast black blood (Panel A) and bright blood (Panel B) CMR demonstrate increased diastolic wall thickness of the left ventricle in the four-chamber view. The corresponding post-contrast-delayed hyperenhancement pattern (Panel C) is unusual with diffusely increased signal intensity in the left ventricular myocardium and normal null (dark appearance) of the right ventricular myocardium. The CT shows diffusely increased left ventricular Hounsfield units (Panel D), and when windowed unconventionally (Panel F), a speckled appearance of the myocardium can be appreciated. Abnormal visualization of soft tissue (left ventricular) uptake of 99mTc-methyl-diphosphonate is seen on the bone scan (Panel E). Haematoxylin and eosin staining of the left ventricle shows the extensive interstitial fibrosis with microcalcifications (Panels G and H), and the calcifications are evident as brown-black granules in intracellular and extracellular locations, von Kossa stain (Panel I).

Clinical vignette

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