Clinical vignette

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CMR scarring in a patient with hypertrophic cardiomyopathy correlates well with histological findings of fibrosis

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A 38-year-old female patient with hypertrophic cardiomyopathy and congestive heart failure underwent late gadolinium enhancement cardiovascular magnetic resonance (CMR). Cine CMR (Panel A) revealed normal dimensions of the left ventricle (left ventricular end-diastolic diameter 45 mm) with moderately reduced left ventricular systolic function (ejection fraction 40%). The maximum wall thickness was 16 mm and there was severe dilation of both atria. Using a T1-weighted inversion-recovery sequence 15 min after the application of 0.2 mmol/kg body weight gadolinium-DTPA, the anteroseptal, inferoseptal, and the anterior walls (Panel B) of the left ventricle showed extensive regions of myocardial enhancement. During her hospital course, the patient developed malignant ventricular arrhythmias and underwent urgent heart transplantation 30 days after the CMR scan. The explanted heart was examined histologically for the extent of fibrosis and disarray. The macroscopic examination of the explanted heart showed massive scarring. Microscopy showed areas of myofibril disarray and scars. These findings were diagnostic of HCM. Regions of gadolinium enhancement correlate well with regions of macroscopic pale myocardium (Panel C) and regions of fibrosis (Panel D).

Panel A. Pre-contrast diastolic long-axis cine image in vivo obtained with cine CMR.

Panel B. Post-contrast image demonstrating myocardial hyperenhancement in vivo. There is a large area of mid-myocardial late gadolinium enhancement of the anterior wall (arrows).

Panel C. Corresponding gross specimen from the explanted heart with well-defined regions of macroscopic mid-myocardial pale myocardium similar to the distribution of late gadolinium enhancement in Panel B.

Panel D. Histological section (haematoxylin and eosin stain) shows a magnification of the box in Panel C with pronounced fibrosis (sheets of collagen stained pink). There is a concordance between regions of gadolinium enhancement in vivo and regions of fibrotic tissue.