Imaging: Myocardial Disease

Aetiology-discriminative multimodality imaging of hypertrophic cardiomyopathy: deformation patterns relate to synchrotron-based assessment of microstructural tissue remodelling

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Background: The aetiology of left ventricular hypertrophy (LVH) is a relevant clinical challenge with consequences for patient management. Phenotypes resulting from hypertensive remodelling and sarcomere mutation often overlap. Synchrotron X-ray phase-contrast imaging (X-PCI) is a technique that can provide 3-dimensional detailed information on myocardial micro-structure non-destructively. The aim is to relate macrostructural/functional, non-invasive, imaging phenotypes of hypertrophic cardiomyopathy (HCM) to the underlying myocardial micro-structure assessed with X-PCI.

Methods: Myocardial tissue samples were obtained from three patients (P1-3) with obstructive myocardial hypertrophy undergoing septal myectomy. Medical history and the 5-year HCM risk scores were evaluated. The patients were imaged with magnetic resonance imaging and echocardiography prior to procedure. Myocardial structure was assessed with wall thickness, late gadolinium enhancement (LGE), whereas function with speckle-tracking deformation (STE) and tissue Doppler imaging (TDI). Myectomy tissue was imaged with X-PCI in the TOMCAT beamline, using a multiscale propagation-based protocol combining a low-resolution (LR) and a high-resolution (HR) setup (5.8 and 0.7 um pixel size, respectively).

Results: The clinical and imaging data are shown in Fig 1. On initial assessment, wall thickness, LGE distribution, global longitudinal strain and septal TDI demonstrated a similar macrostructural and functional phenotype of P1 and P2, whereas P3 stood out with more severe hypertrophy, scarring and dysfunction. Additional regional deformation analysis with STE revealed reduced deformation in the basal and mid septum in P1, paired with a hypertensive pattern of post-systolic shortening (PSS) (yellow arrows). In comparison, in P2 and P3, deformation was more heterogeneous regionally, with regions of almost complete absence of deformation (orange arrows). Upon further exploration with TDI, areas with abnormal deformation were identified on the transition from basal to mid septum in both P2 and P3, whereas deformation was normal, but reduced in P1, and paired with PSS. LR X-PCI defined regions of interest to scan with HR (yellow frame), where HR revealed extensive interstitial fibrosis (orange arrow) with normal myocyte size and organisation in P1, compatible with severe hypertensive remodelling. However, in P2 and P3, patches of fibrosis (yellow arrow) paired with enlarged myocytes organized in visible disarray, considerably more prominent in P3, were both compatible with sarcomere-mutation HCM.

Conclusion: The results demonstrate multiscale phenotyping of HCM - relating micro- and macrostructural findings to function, and integrating multimodality data. In-depth regional deformation analysis, validated by synchrotron-based microstructural analysis, showed potential to identify distinct imaging phenotypes in HCM, distinguishing between overlapping presentations in different aetiologies.

Abstract Figure 1