Unusual form of ‘obstructive hypertrophic cardiomyopathy’: unique contribution of comprehensive cardiovascular magnetic resonance

Thomas Hovasse, Thierry Unterseeh, and Jérôme Garot*

Department of Cardiovascular MR, Institut Hospitalier Jacques Cartier-ICPS, 6 Av du Noyer Lambert, 91 300 Massy, France

*Corresponding author. Tel: +33 1 60 13 46 02, Fax: +33 1 60 13 46 03, Email: j.garot@icps.com.fr

A 50-year-old male was referred for dyspnoea on exertion and systolic murmur. The 12-lead ECG was normal. Transthoracic echocardiography revealed an obstructive hypertrophic cardiomyopathy (OHCM) with a 23 mm thick basal septum, systolic anterior motion of the anterior mitral leaflet, moderate mitral regurgitation, and 70 mmHg dynamic intraventricular gradient in the basal conditions. The patient underwent cardiovascular magnetic resonance (CMR) (1.5 T Siemens Symphony®, Erlangen, Germany) for the assessment of fibrosis and risk stratification.

Steady-state free precession cine-CMR (Supplementary material online, Movie 1), pre-contrast T1 (not shown) and T2-weighted black blood spin echo images (Panel A), and echocardiography (Panel B) revealed typical patterns of OHCM. Late contrast-enhanced CMR showed intramyocardial patchy enhancement within the hypertrophied septum, concordant with the presence of fibrosis in OHCM (Panels C and D). Most strikingly, dynamic first-pass perfusion imaging revealed rapid enhancement of a well limited lobular mass within the basal part of the interventricular septum that persisted over the minute of acquisition (Panels E and F; Supplementary material online, Movie 2). The diagnosis of benign myocardial tumour was made, most likely a rhabdomyoma (same T1 and T2 signals as normal myocardium). The full surgical resection of the mass was deemed hazardous at this location, and the patient was treated on beta-blocker with subsequent decrease of the intraventricular gradient (20 mmHg) and good clinical outcome. Eighteen month follow-up CMR demonstrated perfect stability of the mass.

This case highlights the crucial importance of acquiring and carefully analysing first-pass dynamic perfusion CMR images in the setting of OHCM to rule out the presence of a septal mass. In this case, CMR provided unique diagnostic accuracy above and beyond that of echocardiography. The vascularized nature of the tumour rises the question of the potential efficacy of alcoholic septal ablation in this setting.

Supplementary material is available at European Heart Journal online.

Pre-contrast T2-weighted (Panel A) black blood turbo spin echo images and B-mode echo in the long-axis parasternal view (Panel B) showing typical patterns of OHCM. Note the same T1 (not shown) and T2 signals in the septum as in the remote non-hypertrophied myocardium and the homogeneous echo signal of the interventricular septum. Late-enhanced inversion-recovery CMR images (Panels C and D) acquired 10 min after gadolinium injection (0.1 mM Dotarem®, Guerbet, Aulnay, France) show intramyocardial and patchy hyperenhancement in the septum (arrows), concordant with the presence of fibrosis in hypertrophic cardiomyopathy. Diastolic still frames extracted from ultrafast dynamic perfusion CMR at 10 s (Panel E) and 30 s (Panel F) after gadolinium injection demonstrate rapid enhancement of a lobular septal mass (arrows, 24 mm maximal diameter), persisting over the minute of acquisition. This pattern is very different from the passive and slow diffusion of gadolinium into the interstitial space in case of fibrosis in OHCM.