Hypertrophic cardiomyopathy mimicking STEMI: The role of cardiac magnetic resonance imaging in the detection of microvascular coronary dysfunction

Keywords: Coronary dysfunction; Cardiac magnetic resonance; Microcirculation

Myocardial fibrosis in hypertrophic cardiomyopathy (HCM) is a progressive phenomenon; it is the result of a potential pressure necrosis occurring in the smaller intramural coronary arteries or arterioles, allowing an early detection of myocardial fibrosis [1].

We report the case of a 20-year-old man with no personal or family history of sudden cardiac death, who presented with chest pain typical of an acute coronary syndrome and significant elevation of cardiac enzymes and Troponin Ic: 0.33 μg/L. The 12 leads ECG showed ST segment elevation in V1–V3 and depressed ST segment in DII, DIII, aVF, V5 and V6 (Fig. 1A). Cardiac catheterization showed normal coronary angiography without significant stenosis (Fig. 1B). Echocardiography revealed left ventricular hypertrophy with predominance in the basal septal segments and left ventricular outflow tract obstruction with systolic anterior motion (SAM). Cine Cardiac Magnetic Resonance (CMR) by steady-state free precession sequence (SSFp) showed a concentric diffuse hypertrophic cardiomyopathy with septal predominance in a very severe form (maximal thickness 35 mm). Left ventricle volumes and ejection fraction were normal (LVEF was 65%) but regional contraction was markedly altered in the most hypertrophic segments (Movie 1 and 2).

First pass gadolinium perfusion detected multiple intramyocardial perfusion abnormalities (Fig. 1C and D). Delayed contrast enhancement images – acquired 10 min after injection of gadolinium – showed several areas of intramural hyperenhancement with preserved subendocardium corresponding to first pass perfusion defects, particularly in the interventricular septum and anterior walls (Fig. 1E and G). These perfusion anomalies were consistent with an ischemic origin involving the microcirculation [2], as acute myocardial infarction due to epicardial coronary artery occlusion, which was ruled out by angiography and there were few arguments in favour of myocarditis (no clinical context and no biological signs of inflammation: CRP: 10 mg/L, fibrinogen: 2.1 g/L, procalcitonin: 0.1 ng/mL). No adenosine stress testing was performed to reveal the microvascular dysfunction.

A similar episode of acute coronary syndrome with ST elevation occurred two years later with the same clinical presentation. An emergency CMR allowed to avoid performing another coronary angiography and confirmed the hypothesis of microvascular coronary dysfunction in this severe form of hypertrophic cardiomyopathy. CMR showed a clear progression of the necrosis area with extension of existing hyperenhancement areas and new focal intramural hyperenhancement territories in favour of new lesions, especially in the apex and basal anterior septum (Fig. 1F and H), fibrosis quantification with full width at half maximum (FWHM) technique showed a progression of the cardiac scar from 33% of the LV to 43% (Fig. 1I and J). CMR also demonstrated altered LVEF compared to the previous exam with 56% ejection fraction of the left ventricle (Movie 3). Implantation of an implantable cardiac defibrillator was proposed to our patient on the grounds of myocardial damage extension and progression in CMR (late gadolinium enhancement), and the detection of ventricular tachycardia bursts.

Hypertrophic cardiomyopathy is the most common genetic disease of the heart, and the most frequent cause of sudden death in the young [3]. It is also a major cause of severe heart failure. We report a rare case of obstructive hypertrophic cardiomyopathy with small intramural coronary arteriole dysplasia (SICAD) presenting as repeated acute myocardial infarction with ST elevation. Microvascular dysfunction is a recognized feature of hypertrophic cardiomyopathy [4], its degree is a strong predictor of clinical deterioration and death [5]. Severe microvascular dysfunction is often present in hypertrophic cardiomyopathy with mild or no symptoms and may precede clinical deterioration by years. This case highlights the role of CMR as an important tool to identify this entity by demonstrating serial episodes of intramyocardial necrosis and subsequent scar sparing the endocardium, with potential implications in patient management and prognosis, since myocardial scarring is often associated with ventricular tachycardia and sudden death.

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Figure 1. A. Standard 12-lead ECG showing ST segment elevation in V1–V3 and depressed ST segment in DI, DIII, aVF, VS and V6. B. Normal coronary angiography. C. CMR images, first episode: first pass gadolinium perfusion, detecting multiple intramyocardial perfusion abnormalities in the basal septum. D. CMR images, second episode: first pass gadolinium perfusion, detecting multiple intramyocardial perfusion abnormalities in the basal septum. E and G. CMR images: delayed contrast-enhanced first episode: in horizontal short axis (E) and vertical long axis (G) showing areas of hyperenhancement. F and H. CMR images: delayed contrast-enhanced second episode: in short axis (F) and vertical long axis (H) showing extension of areas of hyperenhancement and thus, fibrosis territories. I and J. Fibrosis quantification with full width at half maximum (FWHM) technique in the first and second episode.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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The authors of this manuscript have certified that they comply with the Principles of Ethical Publishing.

Appendix A. Supplementary data


References


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