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Magnetic Resonance Imaging / Formation image de résonance magnétique

Magnetic Resonance Imaging of Hypertrophic Cardiomyopathy: Beyond Left Ventricular Wall Thickness

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Abstract

During the past decade, cardiac magnetic resonance has gained increasing popularity in the diagnosis of hypertrophic cardiomyopathy because of its greater accuracy and better characterization of cardiac morphology compared with other imaging modalities. In this pictorial essay, a global clinical portrait of hypertrophic cardiomyopathy will be drawn. The various radiologic findings associated with each variant of hypertrophic cardiomyopathy, and the clinical edge offered by cardiac magnetic resonance will be discussed.

Résumé

La résonance magnétique cardiaque connaît depuis une dizaine d'années un essor important dans le diagnostic de la cardiomyopathie hypertrophique, offrant une meilleure précision ainsi qu'une meilleure description de la morphologie myocardique lorsque comparée à d'autres modalités. Dans cet article, nous dressons un portrait clinique de la cardiomyopathie hypertrophique, nous abordons les trouvailles radiologiques qui sont associées aux différents types de cardiomyopathie hypertrophique et nous discutons des avantages cliniques qu'offre la résonance magnétique cardiaque.

Key Words: hypertrophic cardiomyopathy; Cardiomyopathy; Adult heart disease; Magnetic resonance cardiac imaging; Late gadolinium enhancement

Cardiovascular magnetic resonance (CMR) has proved to be a useful tool for assessing a hypertrophic heart, especially when echocardiography is inconclusive or suboptimal. In this article, we review the pathology of hypertrophic cardiomyopathy (HCM), including etiology; clinical manifestation; physiopathology; different phenotypes and implication; management; diagnostic imaging workup; the advantages and disadvantages of ultrasound and CMR; specific information provided by CMR for clinical management; and CMR differentiation of HCM from other cardiomyopathies.

What Is HCM?

Definition

The generally accepted clinical definition of HCM is a left ventricular (LV) hypertrophy associated with a nondilated ventricular chamber that cannot be explained by a cardiac or a systemic disease, including abnormal loading conditions [1,2]. HCM is a relatively common genetic cardiac disease, with a prevalence in the general population estimated at approximately 1:500 [3]. In young people, including children and athletes, it is the most common cause of sudden death [4,5]. In adults of all ages, HCM can present as refractory dyspnea, angina, syncope, palpitation and heart failure. Nonetheless, annual mortality rates are approximately 1%, and almost 25% of these patients have preserved longevity [1,2,6].

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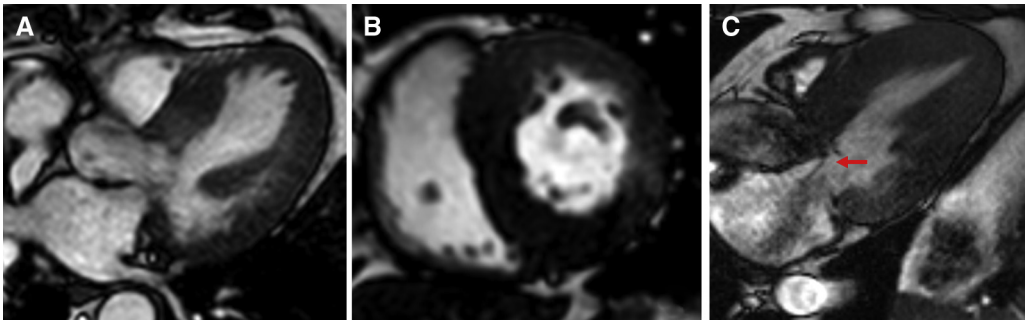

Figure 1. Asymmetric hypertrophic cardiomyopathy septal myocardial thickening on (A) the long-axis view and (B) the short-axis view. (C) Mitral valve regurgitation secondary to systolic anterior movement of the mitral leaflet (red arrow).

Etiology

Hypertrophic cardiomyopathy is caused by mutations in genes that encode cardiac sarcomere proteins [7]. It is inherited in an autosomal-dominant fashion, with variable expression and penetrance. A thick LV wall is the general morphologic phenotype. However, because of variable gene mutations and variable transmission, cardiac involvement can vary and rarely presents with a normal LV thickness or a right side involvement. The wide range of genetic expression can also affect the age of presentation. Although it usually presents in the pediatric population, some phenotypes can present in adulthood [2,7].

Physiopathology and Clinical Consequences

The physiopathologic consequences, depending on the distribution and the degree of myocardial thickening, may be obstruction, arrhythmia predisposition, cardiac dysfunction, aneurysmal formation, or embolic disease [2,5]. Obstructive diseases are divided into subaortic or mid ventricular [7]. When the thickening involves the interventricular septum near the LV outflow tract (LVOT), it will create turbulent flow. Consequently, the anterior leaflet of the mitral valve may have a systolic anterior motion that causes contact with the septum, which contributes to outflow obstruction (Figure 1) [2,5,7,8]. Hypermobility of the papillary muscles or an unusual orientation of papillary muscles (anteroapical displacement and bifid muscle) are other notable causes of subaortic obstruction in HCM and can account for obstruction even in patients with near-normal maximal wall thickness (MWT) [9]. LVOT obstruction produces typical symptoms of dyspnea, angina, or syncope on exertion. To determine whether or not the LVOT thickening is obstructive, a pressure gradient can be calculated with the modified Bernoulli equation: pressure gradient $= 4 \times \mathrm{Vmax}^2$. The cutoff value of 30 mm Hg is now generally accepted [9,10]. Patients with an LVOT gradient higher than 30 mm Hg show a higher risk of heart failure, atrial fibrillation, stroke, and sudden death [6,9,10]. Patients may have a normal LVOT gradient at rest, and the obstruction is often detected by using provocative maneuvers, such as exercise, Valsalva maneuver, or pharmacologic provocation [2,5,10]. The other form of obstruction is mid ventricular, which occurs when there is apposition of a hypertrophic portion of the wall to another part of the LV chamber during systole. In this context, an apical aneurysm may develop [11].

Aside from obstruction, symptoms of cardiac insufficiency may occur secondary to diastolic dysfunction of the hypocontractile muscle [2,5,10]. Over time, with scarring, systolic dysfunction may also develop. When there is an obstruction, diastolic dysfunction, or atrial enlargement, blood stagnation and clot formation may lead to cardioembolic stroke [5,10]. With myocardial disarray and interstitial fibrosis, regions of hypertrophied cardiac muscle fail to conduct electricity in a normal way. Heart block or a re-entrant circuit that leads to sustained or nonsustained ventricular arrhythmia can be created [2].

Different Phenotypes

Although there is no consensual classification of the various phenotypes of HCM, a largely used nomenclature distinguishes among asymmetric septal, apical, mid ventricular, masslike, and symmetric (concentric) hypertrophy. A last phenotype seen in end-stage disease, the ‘‘burned-out phase,’’ also is described [7,9,11].

Asymmetric septal HCM

Asymmetric septal hypertrophy is by far the most common phenotype, with an estimated incidence of 80% [7]. The anteroseptal wall is usually involved, with a localized thickening of the septum. LVOT obstruction is detected in up to 30% of these patients [9]. Patients may have a normal LVOT gradient at rest, and the obstruction often is detected by using provocative maneuvers, such as exercise or the Valsalva maneuver, or by using pharmacologic provocation. Typical symptoms of dyspnea, angina, or syncope on exertion will orient the clinician towards an obstructive disease. The mitral valve is frequently abnormal in these patients. Elongation of the anterior leaflet or both leaflets and secondary mitral regurgitation are common. Mitral