End-Stage Hypertrophic Cardiomyopathy with Impressive Biventricular Hypertrophy: Evaluation with Novel Echocardiographic Modalities

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ABSTRACT

Hypertrophic cardiomyopathy is the most common genetic cardiac disorder. This disease, which is the leading cause of sudden cardiac death in young patients, is typically characterized by idiopathic hypertrophy of the left and sometimes the right ventricle. We present a case of end-stage hypertrophic cardiomyopathy with impressive biventricular hypertrophy which was evaluated with novel echocardiographic modalities.

Key Words: Hypertrophic cardiomyopathy; impressive hypertrophy; imaging

INTRODUCTION

Hypertrophic cardiomyopathy due to its frequency and significant outcomes like sudden cardiac death, is subject to many cardiac imaging techniques in order to establish a definite diagnosis and guide the therapy. Imaging modalities that complement each other help clinicians to decide on the treatment strategy. Especially in patients with obscure indications for a given therapy option, with the help of multimodality imaging techniques a rational solution may be found. We presented an end stage hypertrophic cardiomyopathy patient which was evaluated with novel echocardiographic modalities.

CASE REPORT

A 26 year old man with hypertrophic cardiomyopathy (HCM) was referred for ICD battery exchange. The initial diagnosis of HCM was made at the age of 21 years after a syncope episode. Based on a syncope history and left ventricular (LV) wall thickness greater than 30 mm, a cardioverter-defibrillator had implanted for primary prevention of sudden death after initial diagnosis.
Interventricular septum was measured 51 mm on parasternal image of 3D echo (Figure 6). The patient was diagnosed end stage hypertrophic cardiomyopathy after evaluating with these novel echocardiographic modalities. Appropriate medical treatment for heart failure was initiated for follow up.

**DISCUSSION**

HCM is the most common genetic cardiac disorder, the leading cause of sudden cardiac death in young patients and typically characterized by idiopathic hypertrophy of LV sometimes accompanied by hypertrophy of RV. The distribution
of hypertrophy is also variable; ranging from “classic” septal hypertrophy to isolated apical hypertrophy. In addition the present observations suggest that the genetic process in HCM is more diffuse than previously regarded as a LV disease and the presence of RV hypertrophy was shown recently(1,2). On the other hand, Maron et al. described that extreme degrees of LV wall thickening (≥30 mm) occur relatively frequently in young patients(3). It was suggested that this morphology responsible for increased risk of sudden cardiac death in the absence of severe heart failure symptoms. The present patient has extreme biventricular hypertrophy and syncope history; an ICD had implanted for primary prevention of sudden cardiac death.

The clinical presentation and natural history is particularly heterogeneous, ranging from benign asymptomatic forms to more malignant expressions that may result in sudden cardiac death(4). A small proportion of patients with HCM eventually progress to a stage with reduced systolic function and LV remodeling. Mechanisms responsible for transformation of typical HCM to end stage phase are unresolved but diffuse myocardial ischemia due to micro-vascular dysfunction and extensive myocardial scarring associated with LV chamber remodeling are thought to be important(5,6).

Severe heart failure as end stage phase, with or without LV chamber remodeling has been reported in 3-5% of HCM cohorts(6,7). Although end stage phase is often associated with wall thinning and cavity dilatation it was shown that these patients with and without LV cavity enlargement did not differ with regard to clinical outcome, including cardiac death, transplantation or ICD shocks and only about 50% of patients had evidence of complete remodeling with the triad of LV wall thickness regression, cavity dilatation, and reduced ejection fraction(6). Two-dimensional, M-mode and Doppler echocardiography allow for assessment of the morphology and hemodynamic conditions in HCM, some of which have profound prognostic value(8). The development of 3D echocardiography and speckle tracking echocardiography (STE) has allowed measurement of LV strain (including longitudinal shortening, radial thickening, circumferential shortening), twisting and untwisting rates in determining LV contractility. These novel technologies have facilitated preclinical diagnosis(9) and improved risk stratification of HCM(10).

In a study of Serri et al. LV longitudinal, circumferential and radial strain values were reduced by STE despite preserved ejection fraction(11). In another STE study; all strain values were found to be reduced in patients HCM patients independent from the degree of myocardial fibrosis(12). Besides, reduced LV longitudinal, radial and circumferential strain values, significant deterioration of RV free wall longitudinal strain and displacement values in the present case confirms biventricular systolic dysfunction and end stage phase of HCM.

LV systolic twist patterns have markedly variability due to HCM morphology and extent of hypertrophy(13). It was revealed that LV twist in apical HCM was significantly decreased due to reduction in apical rotation(14). On the other hand the resulting untwisting may represent a useful marker of diastolic function and it was shown that delayed untwisting contribute significantly to exercise limitation in non-obstructive HCM patients(15). LV twist was also reduced in our case supporting the literature.

Real-time 3-dimensional echocardiography allows for better visualization and understanding of the mechanics of systolic anterior motion(16), evaluating accurate estimation of LV ejection fraction as well as LV mass in hypertrophied hearts (comparing with cardiac magnetic resonance imaging(17), facilitating recognition of location and extent of LV cavity obliteration(18). Impressive LV hypertrophy and lack of systolic anterior motion of the mitral anterior leaflet clearly demonstrated in our case by real time 3D echocardiography.

As a conclusion; novel echocardiographic imaging modalities are feasible and beneficial in routine clinical evaluation of patients with HCM especially in the end stage phase of this complex cardiac disorder.

CONFLICT of INTEREST

The authors reported no conflict of interest related to this article.

REFERENCES

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