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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

Belirgin Biventriküler Hipertrofi Gösteren Son Dönem Hipertrofik Kardiyomiyopati Olgusu: Yeni Ekokardiyografik Yöntemlerle Değerlendirme

ÖZET

Hipertrofik kardiyomiyopati en sık rastlanan genetik kökenli kalp hastalığıdır. Gençlerde gözlenen ani kardiyak ölümün en sık sebebi olan bu hastalıkta tipik olarak sol, bazen de sağ ventrikülün idiyopatik hipertrofisi gözlenmektedir. Bu çalışmada, belirgin biventriküler hipertrofi bulguları gösteren son dönem hipertrofik kardiyomiyopatili bir olgu yeni ekokardiyografik modaliteler ile incelenmiştir.
Anahtar Kelimeler: Hipertrofik kardiyomiyopati; belirgin hipertrofi; görüntüleme

INTRODUCTION

Hypertrophic cardiomyopathy due to it's 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.

Parasternal short axis image of two-dimensional echocardiography revealed impressive biventricular hypertrophy; posterior free wall: 37 mm, anterior ventricular septum: 39 mm, anterolateral free wall: 38 mm, posterior ventricular septum: 35 mm, right ventricular (RV) free wall: 15 mm. Mild reduction of LV systolic function was detected by biplane Simpson's method (EF: 50%). There was no dynamic LV outflow tract obstruction by Valsalva maneuver. Conventional and tissue Doppler imaging revealed restrictive type diastolic filling (E: 0.9 m/s, A: 0.3 m/s, DT: 101 ms, Se: 4 cm/s, Sa: 2 cm/s). A significant reduction of LV global longitudinal (-7.53%), radial (8.34%) and circumferential (-10.12%) strain was detected by 2-D speckle tracking echocardiography (Figure 1,2). Significant dyssynchronic motion of the basal posterior, lateral and anterior segments was also detected (Figure 3). LV twist was found to be reduced (2.3°) (Figure 4). RV free wall longitudinal strain (-14.53%) and longitudinal displacement (10.28 mm) were also reduced (Figure 5). For better determination of ventricular cavity and walls 3-D echocardiography was performed.

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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

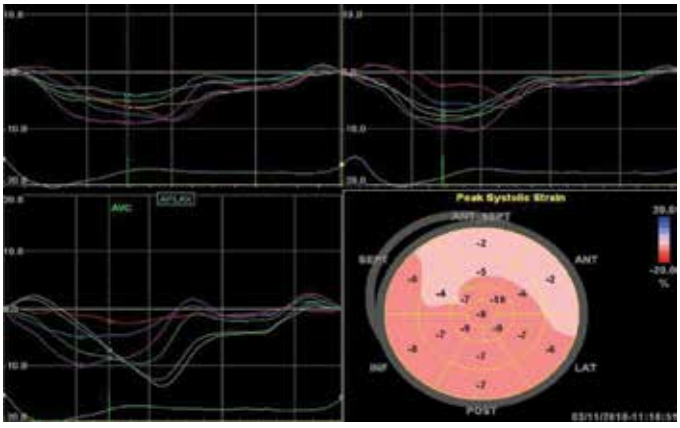

Figure 1. Bull-eye demonstration of reduced left ventricular peak systolic strain value by 2D speckle tracking echocardiography.

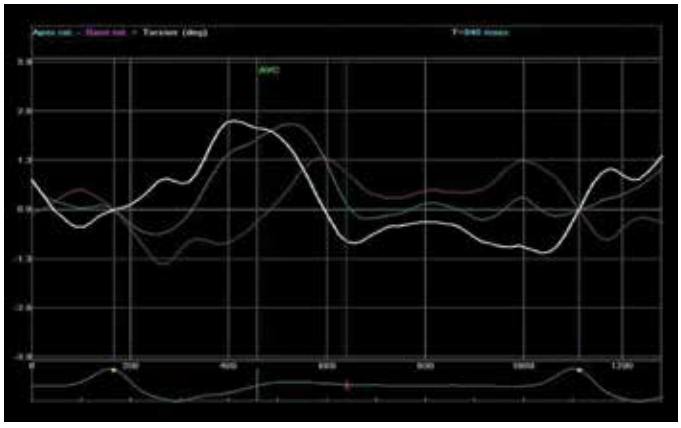

Figure 4. Reduced left ventricular basal and apical rotation and net twist by 2D speckle tracking Echocardiography.

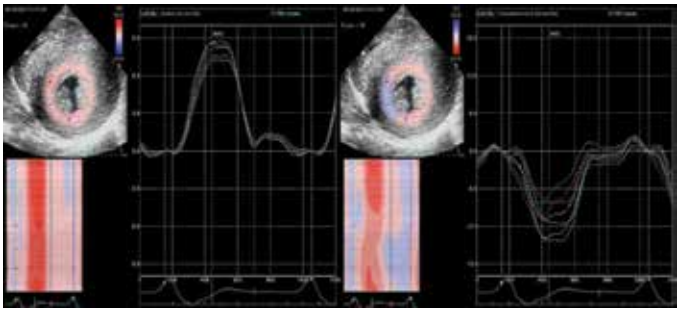

Figure 2. Reduced radial and circumferential strain values of the mid ventricular region by 2D speckle tracking echocardiography.

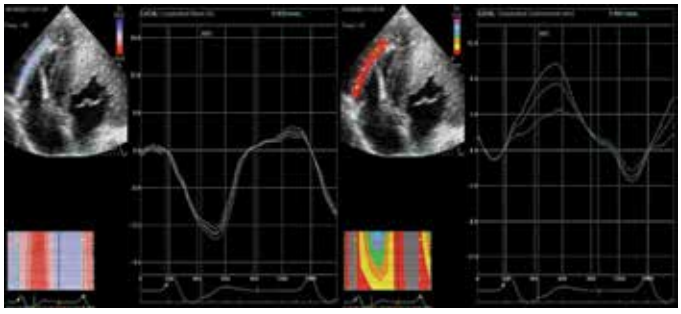

Figure 5. Right ventricular free wall longitudinal strain and displacement by 2D speckle tracking echocardiograph.

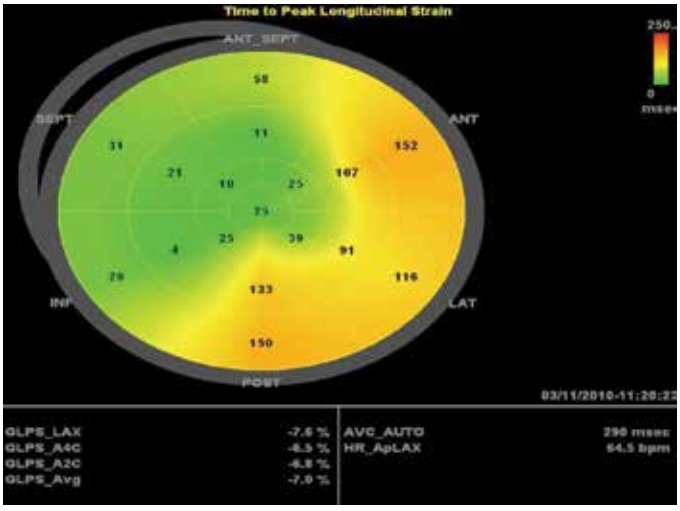

Figure 3. Bull-eye demonstration indicating significant intraventricular systolic dyssynchrony.

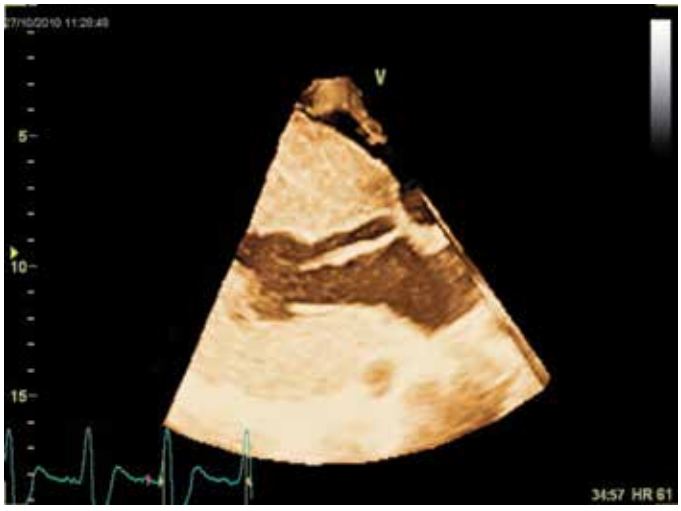

Figure 6. 3D Echocardiographic view demonstrating impressive thickening of septum and posterior left ventricular wall.

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

1. Cheng T. Hypertrophic cardiomyopathy is a biventricular disease. Int J Cardiol 200816;129:3-4.
2. Maron MS, Hauser TH, Duborow E, Horst TA, Kissinger KV, Udelson JE et al. Right ventricular involvement in hypertrophic cardiomyopathy. Am J Cardiol 2007;100:1293-8.
3. Maron BJ, Piccininno M, Casey SA, Bernabò P, Spirito P. Relation of extreme left ventricular hypertrophy to age in hypertrophic cardiomyopathy. Am J Cardiol 20031;91:626-8.
4. Maron BJ. Hypertrophic cardiomyopathy: a systematic review. JAMA 2002;287:1308-20.
5. Olivotto I, Cecchi F, Gistri R, Lorenzoni R, Chiriatti G, Girolami F, et al. Relevance of coronary microvascular flow impairment to long-term remodeling and systolic dysfunction in hypertrophic cardiomyopathy. J Am Coll Cardiol 2006;47:1043-8.
6. Harris KM, Spirito P, Maron MS, Zenovich AG, Formisano F, Lesser JR, et al. Prevalence, clinical profile, and significance of left ventricular remodeling in the end-stage phase of hypertrophic cardiomyopathy. Circulation 2006;114:216-25.
7. Melacini P, Basso C, Angelini A, Calore C, Bobbo F, Tokajuk B, et al. Clinicopathological profiles of progressive heart failure in hypertrophic cardiomyopathy. Eur Heart J 2010;31:2111-23.
8. Spirito P, Bellone P, Harris KM, Bernabo P, Bruzzi P, Maron BJ. Magnitude of left ventricular hypertrophy and risk of sudden death in hypertrophic cardiomyopathy. N Engl J Med 2000;342:1778-85.
9. Nagueh SF, Bachinski LL, Meyer D, Hill R, Zoghbi WA, Tam JW, et al. Tissue Doppler imaging consistently detects myocardial abnormalities in patients with hypertrophic cardiomyopathy and provides a novel means for an early diagnosis before and independently of hypertrophy. Circulation 2001;104:128-30.
10. D'Andrea A, Caso P, Severino S, Cuomo S, Capozzi G, Calabrò P, et al. Prognostic value of intra-left ventricular electromechanical asynchrony in patients with hypertrophic cardiomyopathy. Eur Heart J 2006;27:1311-8.
11. Serri K, Reant P, Lafitte M, Berhouet M, Le Bouffos V, Roudaut R, et al. Global and regional myocardial function quantification by two-dimensional strain: application in hypertrophic cardiomyopathy. J Am Coll Cardiol 2006;47:1175-81.
12. Popović ZB, Kwon DH, Mishra M, Buakhamsri A, Greenberg NL, Thamilarasan M, et al. Association between regional ventricular function and myocardial fibrosis in hypertrophic cardiomyopathy assessed by speckle tracking echocardiography and delayed hyper enhancement magnetic resonance imaging. J Am Soc Echocardiogr 2008;21:1299-305.

13. Sun JP, Stewart WJ, Yang XS, Donnell RO, Leon AR, Felner JM, et al. Differentiation of hypertrophic cardiomyopathy and cardiac amyloidosis from other causes of ventricular wall thickening by two-dimensional strain imaging echocardiography. Am J Cardiol 2009;103:411-5.
14. Chang SA, Kim HK, Kim DH, Kim JC, Kim YJ, Kim HC, et al. Left ventricular twist mechanics in patients with apical hypertrophic cardiomyopathy: assessment with 2D speckle tracking echocardiography. Heart 2010;96:49-55.
15. Abozguia K, Nallur-Shivu G, Phan TT, Ahmed I, Kalra R, Weaver RA, et al. Left ventricular strain and untwist in hypertrophic cardiomyopathy: relation to exercise capacity. Am Heart J 2010;159:825-32.
16. Salustri A, Kofflard MJ, Roelandt JR, Nosir Y, Trocino G, Keane D, et al. Assessment of left ventricular outflow in hypertrophic cardiomyopathy using anyplane and paraplane analysis of three-dimensional echocardiography. Am J Cardiol 1996;78:462-8.
17. Oe H, Hozumi T, Arai K, Matsumura Y, Negishi K, Sugioka K, et al. Comparison of accurate measurement of left ventricular mass in patients with hypertrophied hearts by real-time three-dimensional echocardiography versus magnetic resonance imaging. Am J Cardiol 2005;95:1263-7.
18. De Gregorio C, Recupero A, Grimaldi P, Coglitore S. Can transthoracic live 3-dimensional echocardiography improve the recognition of midventricular obliteration in hypertrophic obstructive cardiomyopathy? J Am Soc Echocardiogr 2006;19:1190.1-4.