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Variability of Dilated Cardiomyopathy Presentation in Magnetic Resonance Imaging Observed in a Single Patient

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Abstract

Dilated Cardiomyopathy (DMC) characterized by Left Ventricular (LV) dilation and systolic dysfunction in the absence of coronary artery disease is a common cause of a Heart Failure (HF) in young adults. Being the final response of myocardium to different genetic and environmental factors, DCM is the most frequent indication for heart transplantation in young population. Assessment of LV dimensions and ejection fraction is indispensable in diagnosis, treatment and risk stratification. Although echocardiography is the most frequently used imaging technique in cardiology, Magnetic Resonance Imaging (MRI) remains the gold standard for LV visualization ensuring precise quantification of cardiac chambers' diameters and volumes. The case presents acute heart failure decompensation in patient with DCM confirming that MRI could be used not only in deferential diagnosis and therapy strategies appointment but also as an excellent method of assessing treatment outcomes. In this patient baseline and control MRI showed significant LV volumes and systolic function improvement after 3 months of adequate pharmacotherapy, as well as change in cardiomyopathy phenotype pattern.

Keywords: Cardiomyopathy • Heart failure • Late gadolinium enhancement

Introduction

Both primaries DCM caused by presence of DNA mutations and secondary due to toxic, infectious, metabolic, and autoimmune or arrhythmogenic background could lead to sever myocardial damage. The role of the echocardiography in DCM diagnosis is limited. While the evaluation of DCM deviated from the standard approach to systolic HF, advanced imaging techniques enables excellent LV volumes and function measurements, as well as the assesment of cardiac remodeling etiology. Evaluation of myocardial edema and classification of Late Gadolinium Enhancement (LGE) in MRI are essential in HF differential diagnosis. While, impaired LV systolic function and LV dilation are major determinants of adverse outcomes in DCM, reverse remodeling is an overriding aim of the therapy. Moreover, proper systolic function estimation in DCM patients is pivotal in sudden cardiac death risk stratification and qualification to Cardioverter-Defibrillator (ICD) implantation. While, one-third of DCM patient may experience partial or even complete recovery, adequate treatment depending on DCM etiology should be implemented. Patient evaluation for an ICD must be carried out after at least 3 months of HF treatment and take into account the risk of unnecessary implantation due to further systolic function improvement. MRI with LGE assesment could identify patients with high remodeling potential, in which group time to decision of ICD implantation should be extended [1,2]. This case presents importance of MRI in proper DCM diagnosis and management.

Case Presentation

A 50-year old patient without previous medical history of cardiac disease was admitted to the cardiology unit with severe dyspnea. At admission the patient showed respiratory effort, expressed crepitations, tachycardia (145 bpm) and hypotension (90/60 mmHg). Electrocardiography revealed an atrial flutter with 2:1 conduction, left axis deviation and left bundle branch block (Figure 1A). LV enlargement, severe mitral and tricuspid regurgitation, as well as reduced ejection fraction (LV EF 10%) was observed in transthoracic echocardiography. Pericardial effusion and distended inferior vena cava with diminished inspiratory collapsibility was noted (Figure 1B). Because of the presence of cardiogenic shock, the patient underwent electrical cardioversion and hemodynamically efficient sinus rhythm was restored. The decision was made to intubate and mechanically ventilate the patient due to persistent respiratory failure. Pharmacotherapy including intravenous pressors and diuretics infusion was introduced with satisfactory effects. Coronary angiography showed no significant coronary arteries' stenosis. After stabilization of the patient's state extubation and MRI imaging were implemented. MRI revealed extensive akinesia of dilated LV (309 ml) with severe reduction of ejection fraction (LV EF 16%), pericardial and pleural effusion and slight Intraventricular Septum (IVS) edema. Rich trabeculation of LV without thrombus presence was noticed (Figure 1C). LGE showed intramuscular changes including 90% of IVS thickness without a vascular scar pattern (Figure 1D). Overall view allowed diagnosing chronic DCM.

After 3 months of adequate heart failure pharmacotherapy the patient with subjective symptoms resolution was re-admitted to the cardiology unit to assess indications for Cardioverter-Defibrillator (ICD) implantation (Figures 1E and 1F). Control MRI revealed LV volume reduction (130 ml *vs*. >300 ml), systolic function improvement (LV EF 38%) and pericardial and pleural effusion regression. Mitral and tricuspid valve regurgitations significantly improved (mild *vs*. severe grade). Substantial IVS edema and overgrowth (18.5 mm) without LV outflow tract obstruction was noticed [Figure 1G]. Intramuscular enhancement of IVS in LGE was observed as before [Figures 1H and 1I]. The observed IVS overgrowth was suspected to be due to the relative increase in scar mass and continuing edema, and finally the diagnosis of DCM was sustained. However, differential diagnosis including Hypertrophic Cardiomyopathy (HCM) and LV no compaction should be carried out with necessity of further MRI verification in 6-12 months.

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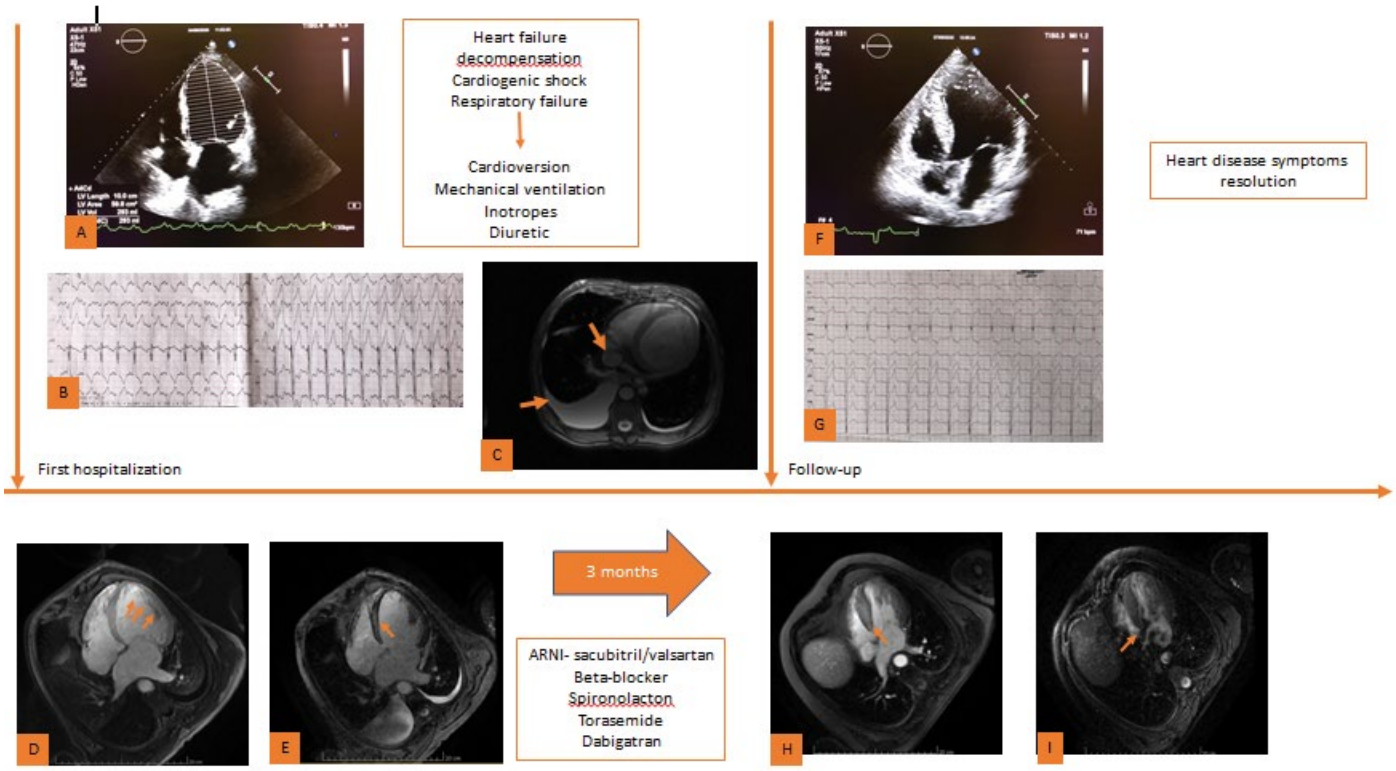

Figure 1(A-I): A: Initial transthoracic echocardiography: LV dilation in apical four-chamber view. B: Initial ECG: Atrial flutter with left bundle branch block. C: Initial MRI: Distended vena cava and pleural effusion. D: Initial MRI: Trabeculations of dilated LV. E: Initial MRI: LGE-MRI showing myocardial enhancement in IVS. F: Follow-up transthoracic echocardiography: IVS overgrowth in apical four-chamber view. G: Follow-up ECG: Sinus rythm with persistent intraventricular conduction block. H: Follow-up MRI: T2-weighted Short- T1 Inversion Recovery (STIR) revealing edema of overgrowth IVS. I: Follow-up MRI: LGE-MRI showing myocardial enhancement in IVS corresponding to STIR.

Discussion

The presented case shows acute decompensation of a cardiomyopathy patient. In that patient's medical history MRI remained useful in every step of the management, from HF decompensation diagnosis, through etiology recognition, differential diagnosis and risk stratification to the evaluation of the treatment results. Baseline severe LV dilatation and dysfunction were significantly and unexpectedly improved. Probably, tachyarrhythmia should probably be regarded as a potent reversible cause of decompensation. Thus, both rhythm conversion and adequate pharmacotherapy were of importance for the clinical success. Comparison of the baseline and control MRI revealed not only a unique reversibility of LV dilatation and dysfunction but also change of LV phenotype pattern from severe LV dilatation with thin walls to enlargement of LV thickness. We used the high spatial resolution of MRI combined with exact delineation of the blood- myocardium interface that enables excellent LV systolic function assesment, which is valid in evaluation of the treatment results. Classification of LGE distribution in MRI and ability to detect myocardial edema could aid etiologic evaluation of cardiac enlargement [3]. In the presented case a differential diagnosis between DCM with cardiac muscle overgrowth due to scar and persistent edema as well as HCM in dilatation stadium were taken under consideration. MRI favors to initial DCM diagnosis. The final diagnosis should be postponed. It is particularly important for sudden cardiac death risk evaluation and ICD implantation.

Conclusion

Although echocardiography is the most commonly used imaging technique in cardiology, the MRI remains the gold standard for left ventricle assesment ensuring precise quantification of cardiac chamber diameters and volumes. In this case MRI was used not only in heart failure deferential diagnosis but also as a tool of the treatment outcome assessment. Moreover, MRI revealed change of cardiomyopathy pattern, which is pivotal in prediction of sudden cardiac death risk.

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