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Case Report

Cardiac Magnetic Resonance Imaging to Diagnose Ebstein's Anomaly Associated with Left Ventricular Non-Compaction

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Abstract

We hereby present the case of a 47-year old female who was admitted to our outpatient clinic to perform cardiac magnetic resonance imaging (CMR) because of unusual texture of the myocardium and tricuspid valve regurgitation in echocardiography. Cardiac magnetic resonance imaging revealed not only non-compaction cardiomyopathy of the left ventricle but also Ebstein´s anomaly in this patient. Isolated non-compaction cardiomyopathy as well as Ebstein´s anomaly is a rare condition and the combination of both is seldom seen. However, both entities have an impact on the patient´s life. This case report in combination with high quality CMR pictures underlines that CMR is a modern imaging technique, which further improves diagnosis of rare cardiac conditions and thereby helps establishing the necessary therapy.

ABBREVIATIONS

CMR: Cardiac Magnetic Resonance Imaging, NC: Non-Compacted, C: Compacted, SSFP: Steady-State Free Precession

INTRODUCTION

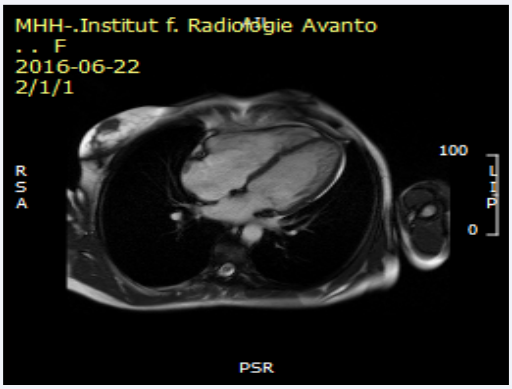
Both isolated left ventricular non-compaction cardiomyopathy and Ebstein's anomaly are rare conditions. We present a case of a 47-year old female in whom cardiac magnetic resonance imaging (CMR) revealed not only non-compaction cardiomyopathy of the left ventricle but also Ebstein´s anomaly.

CASE PRESENTATION

A 47-year-old female was admitted to our clinic to perform cardiac magnetic resonance imaging because of unusual texture of the myocardium and tricuspid valve regurgitation. The initial presentation was for routine cardiac check-up. The patient negated any angina pectoris, dyspnea, dizziness, or syncopes.

The CMR volumetric assessment by left-ventricular short axis SSFP sequences showed a normal function of the left ventricle with a left ventricular ejection fraction of 62%. Non-compacted myocardium was found in antero-septal, anterior, lateral, and inferior segments midventricular to apical with a ratio of > 2.3:1 of non-compacted (NC) to compacted (C) myocardium in diastole (Video 1).

SSFP sequences showed an apical displacement of the septal tricuspid leaflet about 2.5 cm. Furthermore, tricuspid regurgitation could be noted in the SSFP 4-chamber view resulting from failing coaptation of the tricuspid leaflets (Figure). The volumetric assessment of the short axis cine stack of the right ventricle rendered a normal systolic function.



Video 1 SSFP cine imaging of 4-chamber view: non-compacted myocardium is found in midventricular to apical parts of the left ventricle with a ratio of > 2.3:1 of non-compacted (NC) to compacted (C) myocardium in diastole. In the right ventricle, apical displacement of about 2.5cm of the septal tricuspid leaflet is visible with tricuspid regurgitation.

DISCUSSION

Both isolated left ventricular non-compaction cardiomyopathy and Ebstein's anomaly are rare conditions. Isolated left ventricular non-compaction is a rare cardiomyopathy characterized by extensive left ventricular trabeculae and deep intertrabecular recesses in the absence of other cardiac disorders. Ebstein's anomaly denotes a congenital heart disease characterized by the apical displacement of the leaflets of the tricuspid valve and the subsequent "atrialization" of the right ventricle. Prevalence of Ebstein's anomaly accounts to 0.17-0.72/10.000 live births [1, 2]. In rare cases, Ebstein's anomaly can also be associated with non-compacted myocardium [3, 4].

We report a case combining both Ebstein's anomaly and left ventricular non-compaction cardiomyopathy, which was diagnosed through CMR. The ratio of non-compacted to compacted myocardium poses a reliable criterion for the diagnosis of non-compaction cardiomyopathy. A NC/C ratio of >2.3 in diastole identifies non-compaction cardiomyopathy with a sensitivity of 86% and specificity of 86% [5]. Regarding Ebstein's anomaly CMR enables volumetric assessment of the right ventricle and quantification of the right ventricular function. Additionally, CMR is helpful in visualizing the posterior tricuspid valve and to quantify the degree of tricuspid regurgitation [6].

CMR is a modern imaging technique, which further improves diagnosis of rare cardiac conditions such as Ebstein's anomaly accompanied by left ventricular non-compaction.

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