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Case Report

Magnetic Resonnance Imaging in the Diagnosis of Predominantly Right Bi Ventricular Non-Compaction: A Case Report

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Abstract

Ventricular non-compaction or excess trabeculations is a rare condition most often affecting the left ventricle. Bi-ventricular or isolated non compaction to the right ventricle has been described. Magnetic resonance imaging is an examination of choice in diagnosis with well-defined and widely used diagnostic criteria.

Keywords: Biventricular Non-Compaction; Excess Trabeculation; MRI

Introduction

Myocardial non-compaction is a rare condition, and it is linked to an alteration of myocardial structure. It is known by several terms: spongy myocardium, fetal myocardium, hypertrabeculation syndrome. The most appropriate term being excessive trabeculation corresponding to a phenotypic aspect, secondary to genetic mutations or integrating into certain cardiomyopathies [1]. The left ventricle is the part most often affected. However, biventricular damage or isolated damage to the right ventricle have been described although they are very rare entities. The magnetic resonance imaging is essential to make the right diagnosis and thereafter guide the care that these patients require. We thus provide an MRI appearance of excess biventricular trabeculations, predominant on the right and with slight left ventricular dysfunction.

Observation

This case is about a young Senegalese woman aged 17, on whom a cardiac ultrasound was performed for suspicion of mitral disease. The examination revealed moderate dilatation of the right ventricle with a trabecular network, well perfused on color Doppler, including deep endocardial spaces. The damage predominated in the apical and mid-ventricular regions, suggesting thereby isolated non-compaction of the right ventricle. A cardiac MRI was additionally performed with a 3 Telsa MAGNETOM Lumia machine from Siemens. The protocol included short-axis, 04-chamber and 02-cavity cine sequences, myocardial perfusion and late enhancement sequences after gadolinium injection. The examination revealed excessive trabeculation of the two ventricles at the apex and mid-ventricular level, predominating on the right. The ratio of uncompacted myocardium to compacted myocardium was 12 for the right ventricle and 5 for the left ventricle (Figure 1). There was no late gadolinium retention in both ventricles or intracardiac thrombus. The LV systolic ejection fraction was

estimated at 44.8%. The diagnosis of biventricular non-compaction predominant on the right with impact on left ventricular function was made.

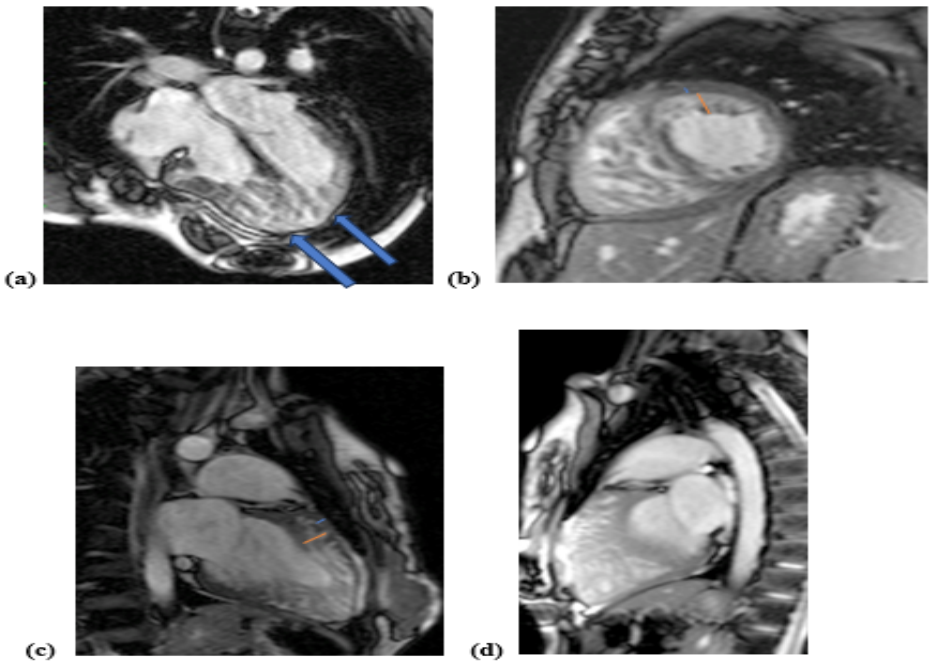

Figure 1: (a) Cine-MRI sections at end of diastole 04 cavities, (b) short axis and (c and d) two right and left cavities. Excessive hypertrabeculations of both ventricles.

Discussion

Ventricular non-compaction is a cardiomyopathy whose specificity was first mentioned in 1926 by Grant et al [1]. It was recognized as a full-fledged cardiomyopathy in 2006 by the American Heart Association, and in 2013 by the World Heart Federation [2-3]. It is known by several terms: spongy myocardium, fetal myocardium, hypertrabeculation syndrome. On the anatomo-pathological level, excess trabeculations are characterized by a phenotypic appearance of double-layered myocardium with an internal trabeculated endocardial layer and a relatively thin external epicardial layer. A ratio of 2 between the trabeculated layer and the compacted layer is usually set as a diagnostic criterion in anatomical pathology. The trabeculations are separated by recesses which communicate with the cardiac cavity. It is within these recesses that thrombi at risk of distal embolism can form, due to a reduction in flow at this level [1].

Excess trabeculations most often affect the left ventricle, but very rarely does it affect the right ventricle in isolation or being biventricular. A few cases of biventricular damage have been described in the literature [3-5] and Okan et al. [6] in their literature review noted 2 cases of isolated damage to the right ventricle.

The non-compaction is not distributed homogeneously at the level of the endocardium. The latero-apical, infero-apical and latero-medial segments are most often affected. The least affected segments are the basal portions of the left ventricle and the septal segments [1]. The damage in our patient was indeed predominant in the lateral and anterior segments of the apex and mid-ventricles.

Embryologically, non-compaction could be due to a cessation of the compaction process or to an inhibition of the regression of trabeculated embryonic structures, at different stages of embryogenesis. A genetic mutation coding for sarcomeric proteins could be involved and the mutation in the MYH17 gene, coding for myosin, appears to be the most frequently encountered mutation [1]. The genetic origin is undergirded by the existence of familial form and form associated with other genetic diseases [1].

However, acquired forms of excess trabeculation have been noted in adults in situations that cannot be explained by embryological mechanisms. In high-level athletes, in women at the end of pregnancy, in patients with dilated cardiomyopathy, hypertrophic cardiomyopathy, rhythm disorders and in asymptomatic subjects. This fact calls into question the notion of cardiomyopathy attributed to ventricular non-compaction and

some authors prefer to use the term excess trabeculations [1]. Moreover, the latest recommendations from the European Society of Cardiology no longer classify ventricular non-compaction in the group of cardiomyopathies but they rather identify it as a phenotypic trait falling into the category of the different cardiomyopathies [7].

It is a rare condition with variations in prevalence ranging from 0.05% to 3.7%, linked to multiple diagnostic criteria [8]. A predominance among African-American subjects has been described.

Cardiac MRI is the technique of choice deployed for the diagnosis of excess trabeculations. It allows a better assessment of the extent of the damage compared with ultrasound which presents difficulties in identifying pathological trabeculations.

In 2005, Petersen proposed MRI diagnostic criteria based on the ratio of thickness of non-compacted myocardium to compacted myocardium greater than 2.3 measured at the end of diastole and at the level of the maximum thickness of the trabeculations, perpendicular to the non-trabeculated myocardium [9]. This is the simplest and most widespread technique. Jacquier et al proposed, on the other hand, that a trabeculated mass greater than 20% of the overall myocardial mass is a sign of excess trabeculations [10].

MRI allows also the detection of fibrosis resulting in late retention of gadolinium on non-compacted or compacted layers. However, a link between the presence of fibrosis in excess trabeculations and a poor prognosis has not been proven.

Conclusion

Non-compaction or excess myocardial trabeculations is rare and often concerns the left ventricle. Isolated damage to the right ventricle or biventricular damage is possible. The MRI provides key elements for diagnosis with good anatomical resolution.

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