WHICH IS YOUR DIAGNOSIS?

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A male, 38-year old patient, weighting 82 kg, 1.78 m in height, with non-sustained ventricular tachycardia, has been referred to the Service of Radiology and Diagnostic Imaging at Hospital Procardíaco to be submitted to magnetic resonance image (MRI) of the heart.

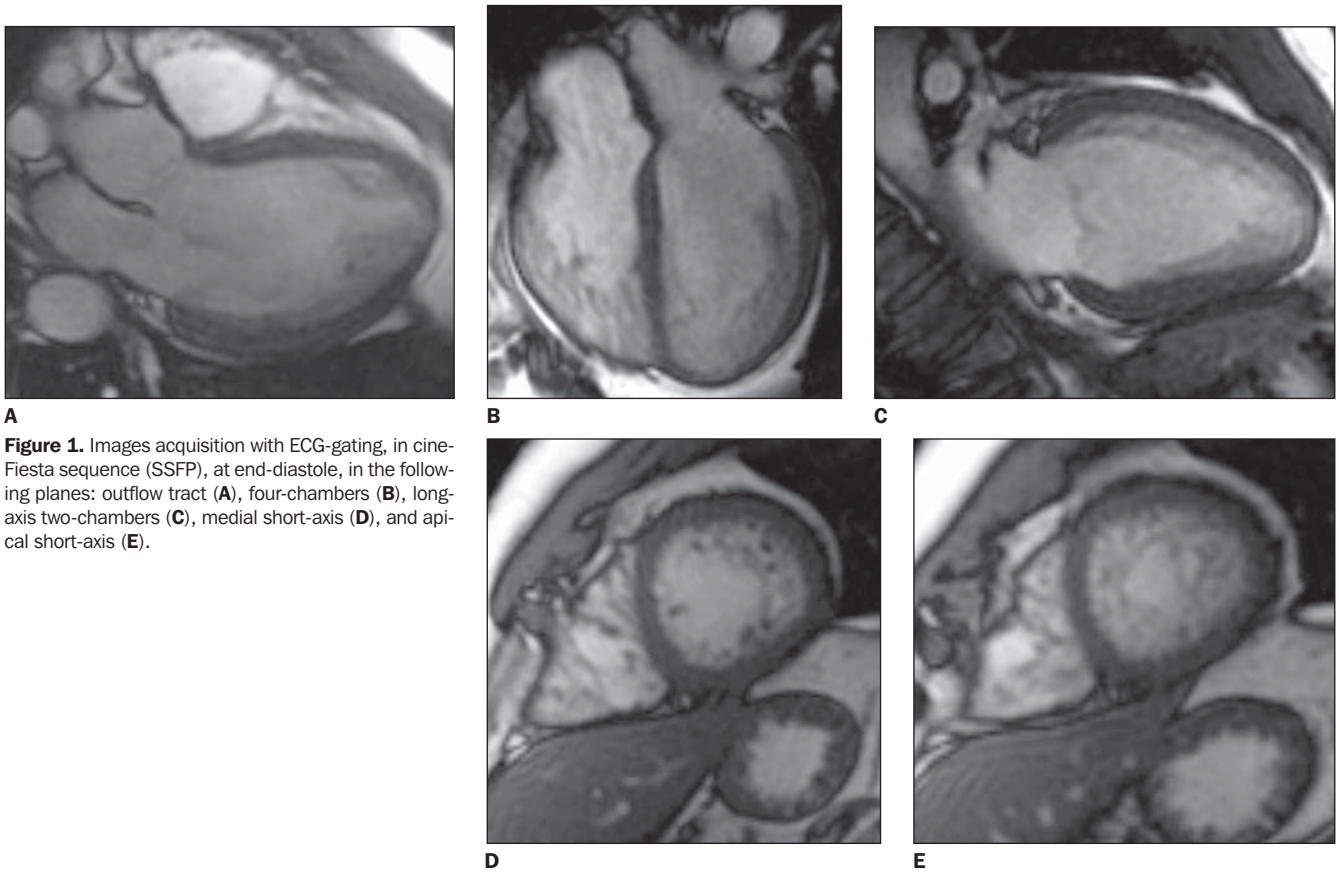

Figure 1. Images acquisition with ECG-gating, in cine-Fiesta sequence (SSFP), at end-diastole, in the following planes: outflow tract (**A**), four-chambers (**B**), long-axis two-chambers (**C**), medial short-axis (**D**), and apical short-axis (**E**).

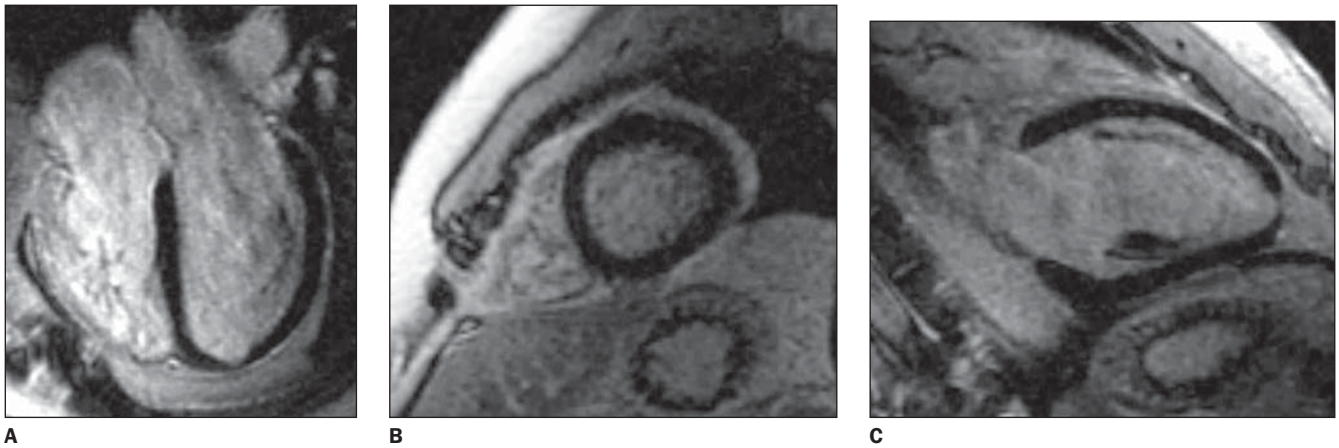

Figure 2. Images acquisition with ECG-gating. Delayed enhancement – four chambers (**A**), short-axis (**B**), and long-axis - two chambers (**C**).

Images description

Figure 1. Images acquisition with ECG-gating, in cine Fiesta sequence (SSFP), at end-diastole, in the following planes: outflow tract (**A**), four-chambers (**B**), long-axis – two-chambers (**C**), medial short-axis (**D**), and apical short-axis (**E**). Observe normal sized atriums, right ventricle with preserved diameters; the right ventricular global and segmental function was preserved. The left ventricle (LV) presents with slightly increased diastolic diameter, with preserved global and segmental function. Note the increase in subendocardial LV trabeculation in medial apical, anterior and inferior segments. The maximum diastolic myocardial ratio of non-compacted (N/C) to compacted (C) thickness was 4 (mean ratio = 2.4).

Figure 2. Images acquisition with ECG-gating. Delayed enhancement – four-chambers (**A**), short-axis (**B**), and long-axis – two-chambers (**C**). Observe the absence of delayed myocardial enhancement, compatible with absence of myocardial necrosis/fibrosis.

Diagnosis: Isolated non-compacted myocardium.

COMMENTS

Non-compacted myocardium is a rare disease, usually diagnosed in the pediatric population, and associated with other structural congenital malformations of the heart, predominating in patients with congenital left or right ventricular outflow tract obstruction[1,2]. Isolate non-compacted myocardium, defined by the absence of other associate structural alteration of the heart, is an even more rare presentation, and has been poorly reported in the literature[2].

Also called LV hypertrabeculation, spongy myocardium or isolated LV abnormal trabeculation, this disease was firstly described in 1932, after necropsy. So far, a few cases have been reported in the Brazilian literature. Its estimated prevalence was 0.05% to 0.24%, but, with the current development of diagnostic imaging methods, especially in the field of MRI, this prevalence tends to increase[1–4].

The etiology of non-compacted myocardium is still to be defined, but heterogeneous genetic factors seem to be closely connected with this disease. During the initial phase of embryonic development, the heart is a trabecular net with a spongy myocardium. The intertrabecular spaces communicate with the cardiac chambers. As the heart develops, the myocardium condenses and the intertrabecular recesses are reduced to capillaries. Non-compacted myocardium is defined as an anomaly of endomyocardial morphogenesis, and it is believed to be an arrest in the compaction of the myocardial fibers, which meet forming an interwoven loose net during intrauterine life. Persistence of non-compacted myocardium is a rare entity, usually diagnosed in the pediatric population and associated with other structural congenital malformations of the heart. It predominates in patients with congenital obstruction of the right or left ventricle outflow tract. The isolate non-compacted myocardium can be detected from the infancy to adulthood. Both sexes are affected and familial recurrence may occur. Familial stratification by cardiac MRI should be considered in relatives of patients with diagnosis of isolated non-compacted myocardium. The present case is in agreement with the literature, where cases with a good myocardial function and absence of constant arrhythmia may present a good prognosis. There is evidence of association with heart failure, severe arrhythmias and embolic events[3–5].

In the majority of reports in the literature, the ventricular non-compaction is associated with other congenital cardiopathies, with predominance of pulmonary atresia and left ventricular outflow tract obstruction combined with an intact interventricular septum. Non-compacted myocardium also has been identified in association with abnormalities in the origin of the left coronary artery from the pulmonary artery trunk. In the case of isolated non-compacted myocardium (Figure 3), its cause remains unknown, and no factor justifying the arrest of ventricular myocardial compaction has been identified. The diagnosis of isolated non-compacted myocardium would be based on a MRI study showing numerous and excessively prominent trabeculations and deep intertrabecular recesses in the absence of coexistent cardiac abnormalities. Contrast-enhanced multidetector computed tomography and MRI studies are complementary and useful for diagnostic confirmation enabling the differentiation between compacted and non-compacted tissues[1,5–7].

Clinical findings may vary from asymptomatic patients to patients with progressive left ventricular dysfunction with arrhythmias and systemic and pulmonary embolic phenomena. In dilated cardiomyopathy, some degree of inferoapical trabeculation associated with the intertra-

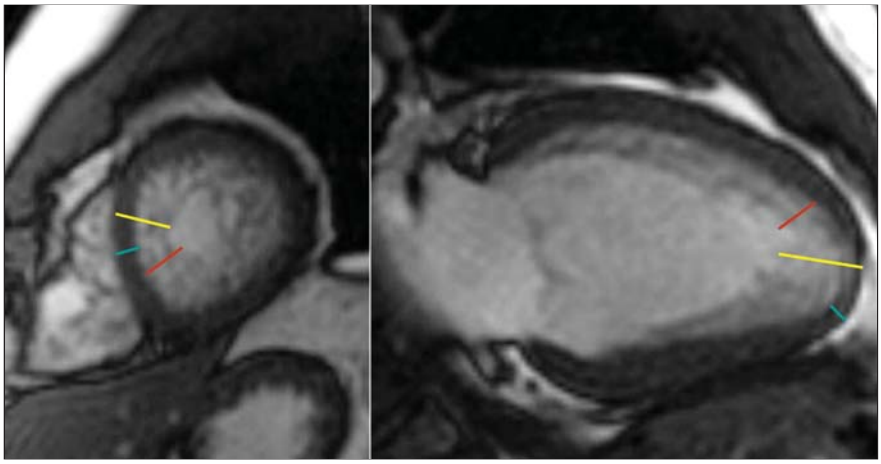

Figure 3. Images acquisition with ECG-gating, in cine Fiesta sequence (SSFP), at end-diastole, in apical short-axis and LV long axis planes. Note the whole wall thickness (yellow tracing), the increase in subendocardial LV trabeculation in medial apical, anterior and inferior segments. The maximum diastolic myocardial ratio of non-compacted (red tracing) to compacted (green tracing) thickness was 4 (mean ratio = 2.4).

becular spaces may be visualized; therefore, a distinction between these two disorders, at least from the morphological point of view, is not always clear. In spite of the fact that demonstration on perfusion MRI of deep perfused intertrabecular recesses is one of the markers for the diagnosis of isolated non-compacted myocardium, transitional variations between the isolated non-compacted myocardium and dilated cardiomyopathy may exist[6–10]. In addition, more discreet cases of isolated non-compacted myocardium without diagnostic confirmation may exist, in the absence of excessive trabeculation in the inferoapical region, hypertrophy, and marked intertrabecular recesses. The high incidence of thromboembolic phenomena in the isolated non-compacted myocardium could result in formation of local thrombi in the deep intertrabecular recesses in addition to ventricular dysfunction[4,9–11].

MRI represents an extremely important method not only for diagnosis, but also for following-up the clinical evolution of these patients.

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