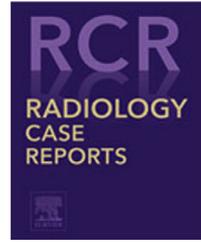




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Case Report

MRI aspects of left ventricular non compaction (LVNC): About 3 cases from Sub-Saharan Africa and review of the literature[☆]

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ABSTRACT

Left ventricular non compaction (LVNC) is a relatively rare variety of cardiomyopathy of genetic origin.

We report three cases of LVNC diagnosed on cardiac magnetic resonance imaging (MRI) in Abidjan in patients aged 42, 46 and 60 years, referred for suspected LVNC on echocardiography.

We used a 1.5 T MRI and performed the following sequences: black blood and white blood, LV minor axis, LV major axis, 4 cavities, and T1 SPIR Gadolinium (early and late enhancement at 10 minutes).

MRI made the diagnosis of LVNC based on a double-layered myocardium, the inner (endocardium) non compacted, fibrillar thickened and the outer (epicardium) compacted thin with a non compacted to compacted myocardium ratio greater than 2.3, making the formal diagnosis.

Cardiac MRI is an excellent diagnostic tool for LVNC. Its recent use in Africa should be common in the management of this cardiomyopathy.

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Introduction

Left ventricular non compaction of myocardium (LVNC) is a very rare condition in adults [1]. It is considered as a disorder of endo-myocardial morphogenesis characterized by the persistence of numerous left ventricular trabeculae and deep inter-trabecular recesses leading to a double myocardial layer appearance: an internal (endocardium) non-compacted and an external (epicardium) thin compacted layer. The first African cases were described in Marseille (France) in 2007 by Paule P. et al [2].

The diagnosis can be made by echocardiography in symptomatic forms called major. The asymptomatic forms called minor or subtle are difficult to diagnose by echocardiography.

The practice of cardiac magnetic resonance imaging (MRI), particularly in the diagnosis of LVNC (left ventricular non compaction), is quite recent in Africa. We emphasize the interest of the practice of cardiac MRI in the diagnosis of this pathology in the African context.

Case 1

A 42-year-old woman consulted for dyspnea, asthenia. Blood pressure was normal at 110/80 mm Hg. The electrocardiogram (ECG) showed a sinus rhythm of 78 beats/min with repolarization disorders.

NCVG (non compaction du ventricule gauche or in english)-type cardiomyopathy was suspected on echocardiography. Cardiac MRI demonstrated:

Functionally, a 35% systolic ejection fraction associated with global hypokinesia; Morphologically, cryptic hypertrophy of the endocardium of the left ventricular apex and dilatation of the left ventricle with right hypoplasia; trabecular appearance of the lateral and inferior wall of the left ventricle more pronounced on the inferior sectors, with an appearance of double myocardial layer (Figs. 1 and 2) :

The inner layer (endocardium) is 22.6 mm thick and not compacted;

The outer layer (epicardium) is compacted to a thickness of 6.46 mm and the inner layer to outer layer ratio of 3.49 in diastole, meeting the diagnostic criteria for LVNC according to the OECHSLIN criteria.

The study of late enhancement did not reveal any late contrast, especially in the subendocardial region. There was no intra-cavity thrombus.

Case 2

A 46-year-old man with cardiac situs inversus presented with left heart dysfunction with major dyspnea during follow-up. Physical examination was normal with a blood pressure of 108/75 mm Hg.

The ECG showed a sinus rhythm disorder at 70 beats/min. Echocardiography suspected LVNC by the demonstration of some trabeculations at the tip of the left ventricle.

Chest X-ray showed predominantly left-sided cardiomegaly. The cardiac MRI performed demonstrated:

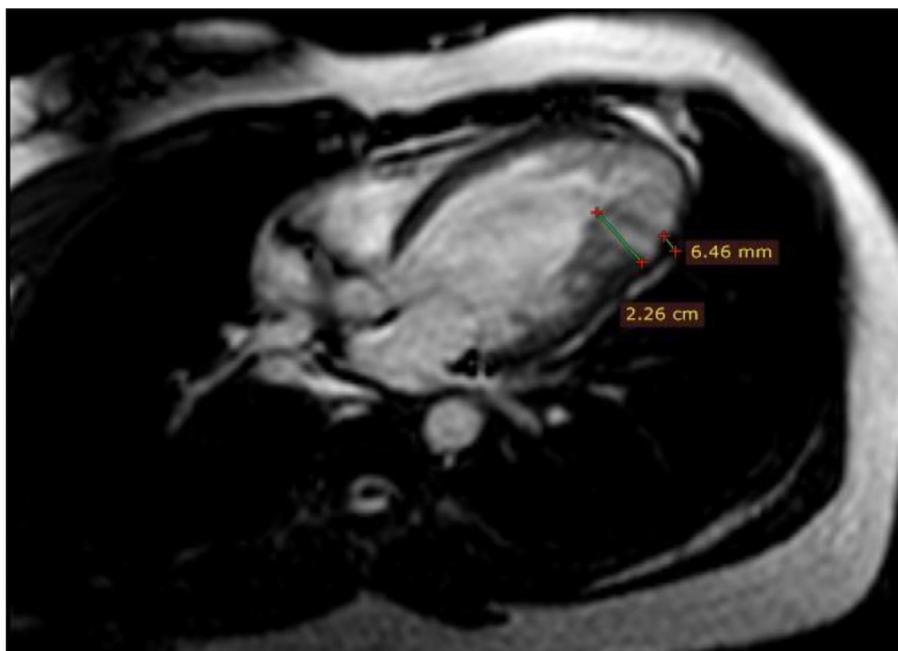


Fig. 1 – Cardiac magnetic resonance imaging (MRI) T2 sequence; four-cavity slice horizontal long axis (HLA); Hypertrophy of the endocardium with the endocardium/epicardium ratio estimated at 3.49.

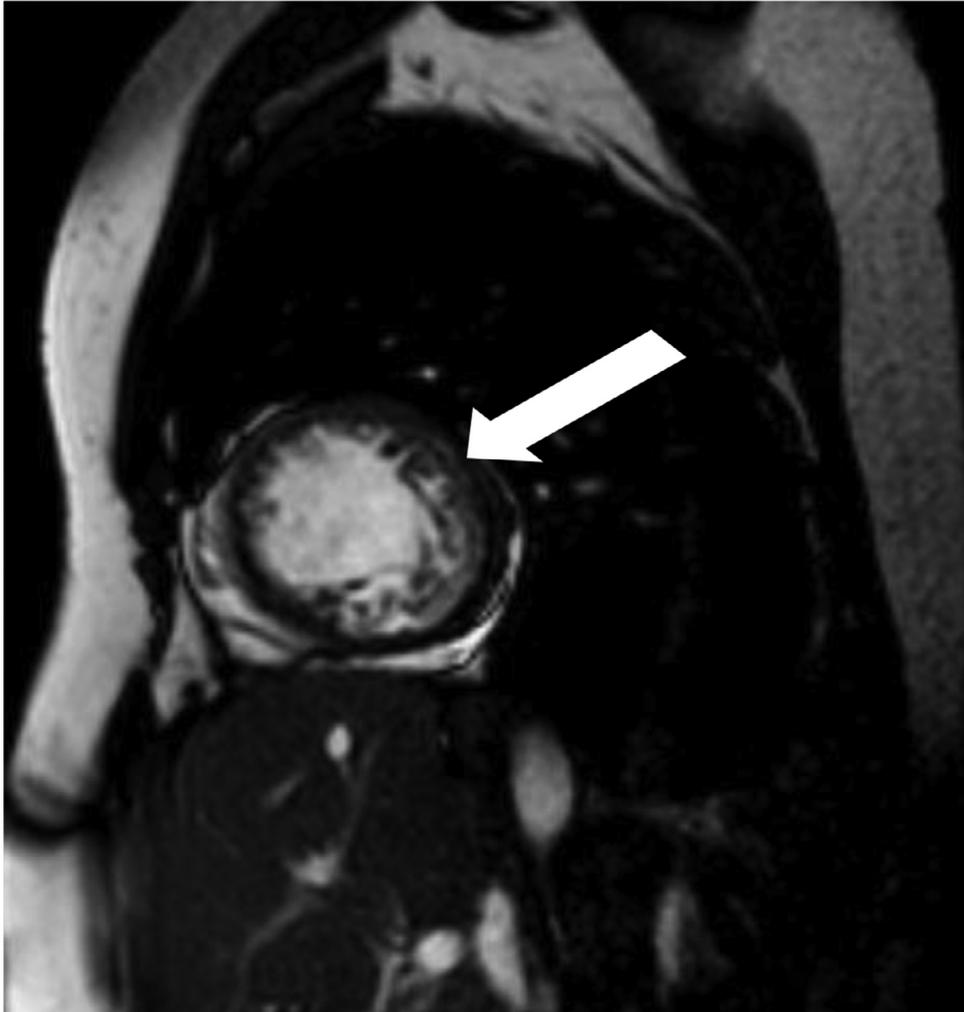


Fig. 2 – Cardiac magnetic resonance imaging (MRI) T2 sequence; short axis section of the heart passing through the apex of the left ventricle. Cryptic hypertrophy of the endocardium (white arrow).

Functionally, left ventricular hypokinesia with a decrease in systolic ejection fraction to 35%;

Morphologically, a cardiac situs inversus with dextrocardia and a double myocardial layer appearance of the left ventricle; an inner layer (endocardium) that was non compact and 22.9 mm thick, with numerous trabeculations at the apex of the left ventricle, and an outer layer (epicardium) that was 7.14 mm thick was compact (Figs. 3 and 4).

The ratio of non compacted to compacted myocardium in diastole was 3.20 consistent with LVNC.

After injection, no late contrast was found, especially in the subendocardial region. There was no intra-cavity thrombus.

Case 3

A 60-year-old man presented with left heart failure with episodes of ventricular tachycardia. On clinical examination, blood pressure was 130/85 mm Hg. The ECG showed a sinus rhythm of 80 beats/min.

Echocardiography suspected LVNC. Cardiac MRI objectified: Functionally, a systolic ejection fraction of 43%;

Morphologically, dilatation of the cardiac chambers without pericardial effusion or valve abnormalities; a double-layered appearance of the lateral and inferior wall of the

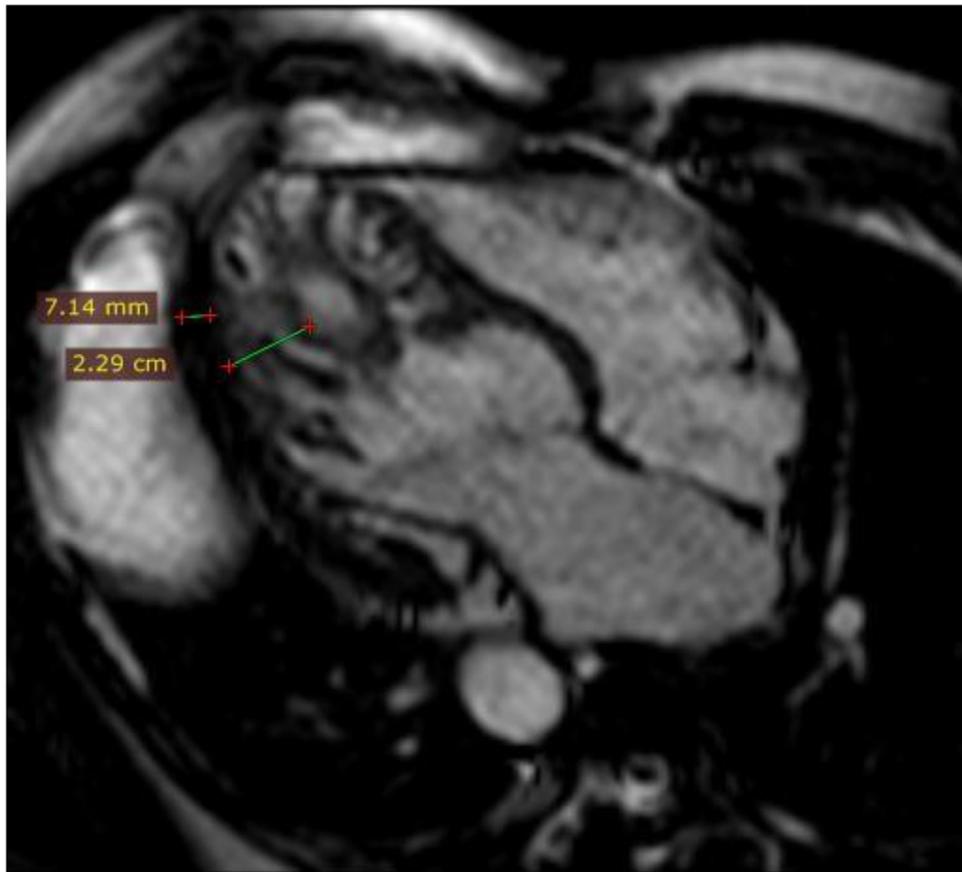


Fig. 3 – Cardiac magnetic resonance imaging (MRI) T2 sequence, Four-cavity section (HLA), Situs inversus with dextrocardia; The endocardium/epicardium ratio at 3.20.

left ventricle; a 20.2-mm non compacted inner layer (endocardium) with a trabecular appearance and a 5.31-mm thinned compacted outer layer (epicardium); the ratio of non compacted myocardium to compacted myocardium was reported to be 3.80 (Figs. 5 and 6).

Elsewhere, no abnormalities of segmental kinetics, late enhancement or intra-cavity thrombus were noted.

Discussion

Left ventricular non-compaction (LVNC) is a rare congenital cardiomyopathy resulting from the arrest of normal myocardial embryogenesis [2]. It is characterized by a trabecular subendocardial layer and a relative thinning of the compacted outer myocardium (Fig. 7) [3], which distinguishes it morphologically from other cardiomyopathies. The interest of

this pathology lies in its variable anatomical characteristics, which are the cause of uncertain diagnostic criteria in its difficult diagnosis, relying mainly on echography and cardiac Magnetic Resonance Imaging (MRI).

LVNC has recently been described as a genetic cardiomyopathy caused by abnormal embryogenesis of the endocardium and myocardium in utero. It is characterized by a double-layered myocardium with a non-compacted inner layer (endocardium), composed of prominent ventricular trabeculations with deep inter-tabular recesses and a compacted outer layer (epicardium) [4].

It can be isolated or associated with various cardiac anomalies such as right or left ventricular outflow tract obstruction, complex cyanotic congenital heart disease and coronary anomalies. According to the literature, it can be associated with cardiac malformations in 3 to 5% of cases such as transposition of the great vessels and atrio-ventricular discordance, persistence of the right aortic arch in 80% of cases



Fig. 4 – Cardiac magnetic resonance imaging (MRI) T2 sequence; short axis section of the left ventricular apex on situs inversus with evidence of large trabeculations (white arrow).

and the presence in almost all cases of situs inversus with le-
vocardia.

This was the case of the 46-year-old patient (case 2) who
presented with a situs invertus.

Genetically, two forms can be distinguished [4]:

A familial form more often diagnosed in childhood related
to a mutation of the G4.5 gene located on chromosomal region
Xq28

A sporadic form encountered in adults of autosomal dom-
inant transmission corresponding well to our observations
with an age range from 42 to 60 years. (Our patients are 42,
46 and 60 years old).

The age of our patients was identical to that of Diop's study
in Senegal of two sporadic cases aged 40 and 61 years [5].

Clinically, it can be completely asymptomatic and discov-
ered by chance during a routine examination (echocardiogra-
phy) in children or adults, carried out for a different reason.

It may be symptomatic and present with a wide range of
manifestations. Diagnosis may be delayed due to limited
knowledge or even misunderstanding of imaging and clinical
features [6].

Patients with a final diagnosis of LVNC are often referred
to a cardiologist for unexplained heart failure, palpitations,
or certain echocardiographic findings, as was the case with

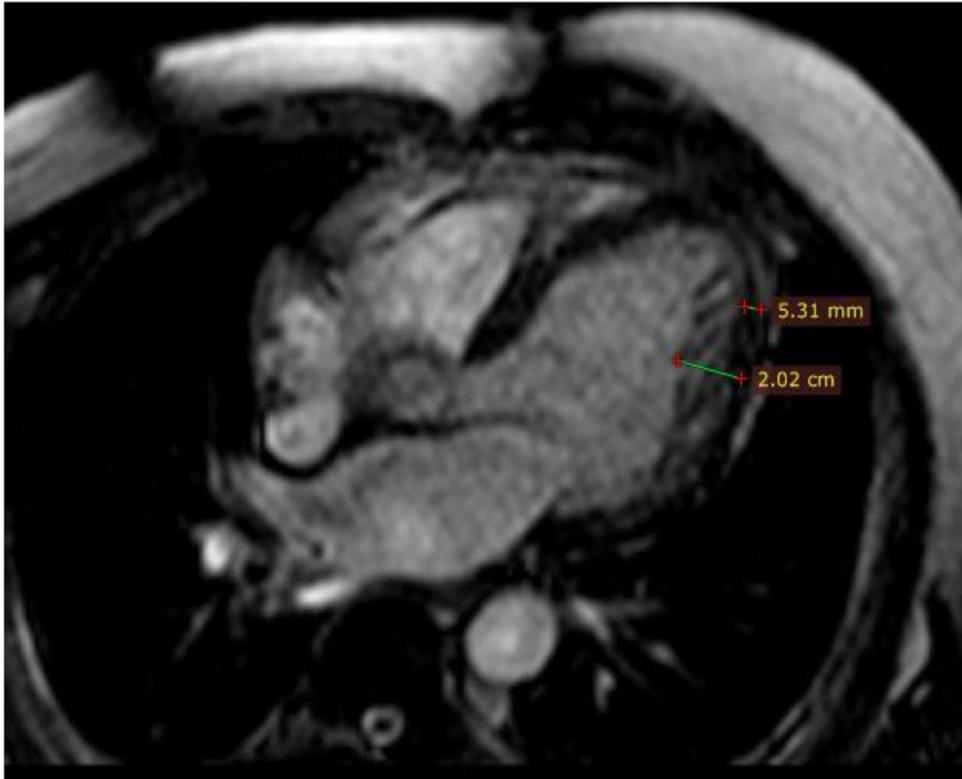


Fig. 5 – Cardiac magnetic resonance imaging (MRI) T2 sequence; Four-cavity section (HLA); The endocardium/epicardium ratio is of more 3.80 than 2.3.

the three reported cases. Traditionally, the diagnosis of LVNC is based on 2D echocardiography characterized by the presence of numerous prominent trabeculations with deep recesses in hypertrophied, often hypokinetic segments of the left ventricle (LV). The most frequently affected areas are the LV apex, lateral and inferior walls [4]. The most commonly used echocardiographic criteria for the diagnosis of LVNC in adults are consistent with the Oechslin proposal [5,7].

Cardiac MRI has established itself as the gold standard in the diagnosis of left ventricular non-compaction. It has not only a diagnostic but also a prognostic role.

Because of its higher spatial resolution, it allows the detection of subtle forms of left ventricular non-compaction as in our first patient who was followed for another anomaly (situs inversus).

It assesses prognosis, as in the three patients, by providing data on cardiac function, myocardial perfusion and degree of myocardial fibrosis (study after injection), identification of vascular thrombi and search for alternative diagnoses.

During an MRI examination in the context of a LVNC the following elements are objectified:

Numerous excessive trabeculations in the left ventricle with predominant involvement of the apical and middle segments of the lateral and inferior walls [4]

Thinning of the left ventricular wall during diastole

The presence of a myocardium structured in two layers with, in the diastolic phase, a myocardial ratio between the compacted surface and the non-compacted surface greater than 2.3 [8]. In our study the ratios vary from 3.20 to 3.80 values well above normal.

Similarly, the calculation of left ventricular mass with trabeculations by the following formula remains the best diagnostic criterion for left ventricular non-compaction [9]:

$$\frac{\text{Global left ventricular mass} - \text{the compacted left ventricular mass}}{\text{Global left ventricular mass}}$$

The measurements are made at the console.



Fig. 6 – T2 cardiac magnetic resonance imaging (MRI), short axis slice showing the apex of the left ventricle with numerous trabeculae (white arrow).

The overall mortality of LVNC is low in the asymptomatic form [10], depending on the impairment of systolic function and the presence of associated neurological disease [11]. The symptomatic form has a poor prognosis; its mortality is 40% at 5 years [2].

There is no codified therapeutic management; it is primarily symptomatic. The preventive approach is much debated; anticoagulants are necessary when there is a proven high risk

of thromboembolic events. However, in case of failure of medical treatment, pacemakers or even heart transplants can be considered [4].

Finally, first-degree siblings of any patient diagnosed with left ventricular non compaction should undergo screening echocardiography or even additional MRI in cases of suspected LVNC, which will both confirm the diagnosis and assess cardiac function.

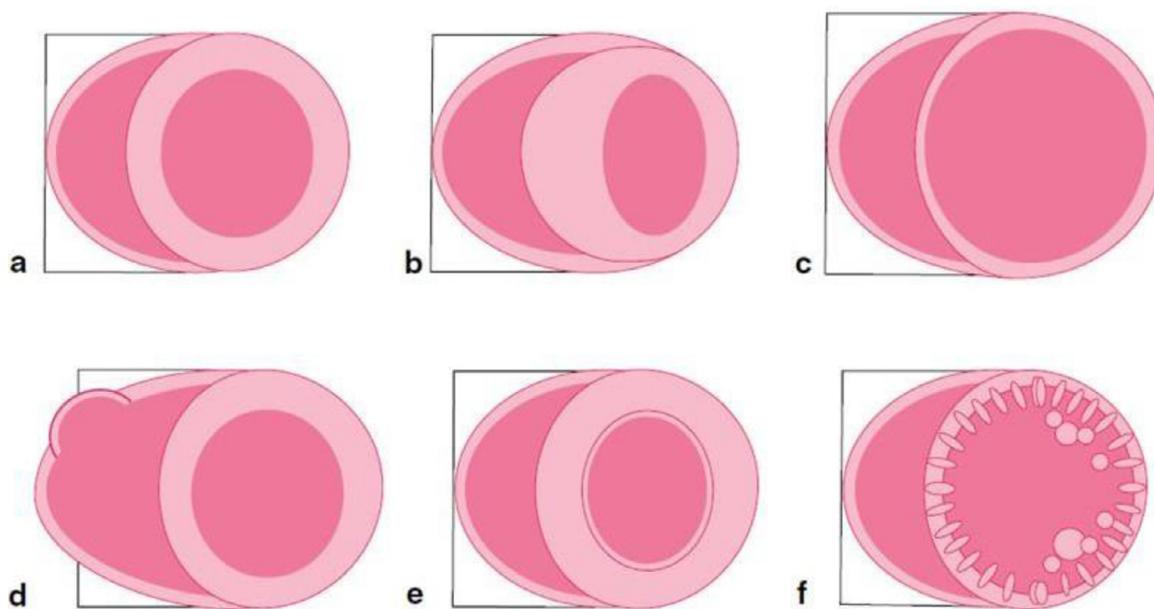


Fig. 7 – Morphological classification of cardiomyopathies. A. Normal heart. B. Hypertrophic cardiomyopathy. C. Dilated cardiomyopathy. D. Arrhythmogenic right ventricular dysplasia. E. Restrictive cardiomyopathy: endocardial lines represent abnormal longitudinal fibers. F. Myocardial noncompaction: subendocardial layer has a trabecular structure with relative thinning of compacted outer myocardium [3].

Conclusion

The two main diagnostic modalities for LVNC in radiology are echocardiography and cardiac MRI.

MRI, the second modality, with its excellent spatial resolution, is the best method because it has not only a diagnostic but also a prognostic role. Any suspicion of ventricular trabeculation should be explored by MRI to rule out LVNC cardiomyopathy. In our African context, this would allow an early diagnosis especially of asymptomatic forms in order to reduce the evolutionary complications and mortality that are often associated with it.

Patient consent

This piece of the submission is being sent via mail.

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