MYOCARDIAL INFARCTION REVEALING NON-COMPACTION OF THE LEFT VENTRICLE: A RARE CASE-BASED DISCUSSION.

Abstract :

Left ventricular non-compaction (LVNC) is a cardiomyopathy due to disturbances in the normal development of the heart muscle leading to the persistence of hypertrabeculation. its incidence is more common in males.

The most common clinical presentations at the time of the first diagnosis are heart failure, thromboembolic events, and various rhythm disorders.

We report the case of a 52-year-old man, with a probable unexplored history of cerebrovascular accident, who consulted in the emergency room for acute chest pain constrictive retrosternal.

The electrocardiogram showed negative T waves in the lateral and inferior septum. The echocardiography showed a trabeculated aspect of the apex of the left ventricle, inter trabecular recesses taking color on color Doppler. Ultra-sensitive troponins were elevated to 351 ng/l.

Coronary angiography was performed returning normally.

Myocardial ischemia with healthy coronaries secondary to non-compaction of the LV remains poorly understood and may be secondary to an anomaly of the microcirculation, coronary embolization of a thrombus formed at the level of the ventricular trabeculations.

The dreadful complications of the discovery of LVNC must draw the cardiologist's attention to this pathology for better subsequent management.

Keys words :

Left ventricular non-compaction, myocardial infarction, case report

Introduction :

Left ventricular non-compaction (LVNC) or previously called embryonic spongy left ventricle is characterized by the presence of excessive trabeculations of the left ventricle, forming deep cavities that fill with blood.

It is due to disturbances in the non-compaction process of multifactorial origin leading to the persistence of hypertrabeculation and LVNC.

The most frequent clinical presentations at the time of the first diagnosis are heart failure, thromboembolic events, and various rhythm disorders.

We report the case of a 52-year-old patient admitted with acute non-ST-segment elevation coronary syndrome in whom LVNC was discovered incidentally on echocardiography. There are a few similar cases in the literature.

Case presentation :

We report the case of a 52-year-old man, a 20-year-old smoker, with no personal or family history of cardiovascular pathology or sudden death. His symptoms date back to two months before his admission with the onset of constrictive retrosternal chest pain at rest, which gave way spontaneously; the patient consulted the emergency room. In addition, the patient reported a transient hemiparesis occurring 3 years before his admission, which was not the subject of a consultation or exploration.

On admission, the patient was conscious, hemodynamically and respiratory stable, eupneic, normocardial at 95 bpm, and normotensive at 110/65 mmHg. The physical examination did not reveal any heart murmurs or signs of cardiac insufficiency with normal pleuropulmonary auscultation. The neurological examination did not reveal any motor or sensory deficit.

The electrocardiogram showed negative T waves in the lateral and inferior septum. The echocardiography showed a trabeculated aspect of the apex of the left ventricle, intertrabecular recesses taking color on color Doppler, an aspect of non-compacted tissue at the apical level with a ratio of compacted to non-compacted tissue at 3.14. (figure 1.2.3) The echocardiography did not reveal any segmental kinetic disorder, nor any individualizable thrombus, with a left ventricular ejection fraction (LVEF) preserved at 55%. The chest X-ray was without abnormality. The coronary angiography performed within 24 hours was normal with no atheromatous plaques or thrombotic lesions. A Holter ECG was performed and found no supra-ventricular rhythm disorder. Cardiac MRI was not performed because it was not available.

In view of the neurological symptoms described by the patient, a cerebral scanner was performed, showing ischemic lesions of a sequential nature.

On biological examination, the ultra-sensitive troponins were elevated to 351 ng/l, microcytic hypochromic anemia at 9.7 g/dL, and a correct renal function. A thrombophilia test did not reveal any abnormality.

Faced with myocardial infarction with non-obstructive coronary arteries on a non-compaction of the left ventricle, the possible diagnoses were myocardial infarction (MI) due to an attack on the microcirculation found in the non-compaction of the LV, or the embolization of a thrombus formed at the apical level of the LV despite its non-visualization on echocardiography and the absence of thrombus at the level of the coronaries on coronary angiography, or a thrombus linked to atrial fibrillation in the context of LVNC. Type 2 MI was also suspected in the presence of iron deficiency anemia. The most likely mechanism in the face of coexisting sequelae of ischemic brain lesions is embolization from a thrombus in the trabeculations not visualized on echocardiography or paroxysmal atrial fibrillation (AF).

The patient was treated medically and put on anticoagulation with vitamin K antagonist (VKA).

DISCUSSION

Left ventricular non-compaction describes a phenotypic phenomenon characterized by the presence of excessive trabeculations of the left ventricle, forming deep cavities that fill with blood. The first autopsy report describing an "embryonic spongy pattern of the left ventricle" in newborns was published in 1926 [1]. "The first case of an isolated sinusoidal anomaly of the left ventricle diagnosed by echocardiography in a 33-year-old woman was published in 1984". [2].

Its classification as cardiomyopathy differs between the European Society of Cardiology and the American Heart Association. The former defines it as an "unclassified cardiomyopathy", and the latter as a "genetic cardiomyopathy" [3 , 4].

"The prevalence is 4.1-5%, and men are almost three times more affected" [5].

"Disturbances in the normal development of the heart muscle leading to the persistence of hypertrabeculation. Trabeculations appear at the end of the 4th week of gestation. The thin subepicardial part of the muscle forms the "compacted myocardium" while the trabecular part forms the "non-compacted" part. In the following weeks, the trabeculae thicken, increasing the volume of the compacted layer, while the intertrabecular recesses compress and form capillaries" [6]. Disruptions in this process, for whatever reason, lead to the persistence of hypertrabeculation and LVNC. Normally, the process of compaction progresses from the epicardium to the endocardium, from the septum to the free wall, and from the base to the apex in the LV. Moreover, it is more often observed in the LV than in the right ventricle. Therefore, apical non-compaction is the most common type.

"Furthermore, the presence of many cases of familial LVNC argues for a genetic origin of the disease and about 66 genes have been considered to be associated with LVNC, 82% of which code for sarcomeric proteins" [7]. In patients with LVNC, various concomitant congenital heart diseases and neuromuscular disorders have been reported.

"Furthermore, hypertrabeculation has not always been pathological and may be reversible. It has been observed as an adaptive phenomenon to increased cardiac output in athletes, pregnant women, and patients with haemoglobinopathy" [8]. The trabeculated ventricle can function more efficiently by generating the same systolic ejection volume at lower strain and wall stress.

The most common clinical presentations at the time of the first diagnosis are heart failure, thromboembolic events, and various rhythm disorders.

Ventricular dysfunction is maybe systolic or diastolic. Although the mechanism of heart failure associated with LVNC is not clear, it has been suggested that myocardial ischemia may play an important role. This is supported by subendocardial perfusion deficits that have been observed on cardiac MRI and PET scans that will lead to subendocardial fibrosis and necrosis and these perfusion deficits unrelated to atherosclerosis are explained by an overly

thick subendocardial uncompacted layer, preventing the endothelial myocardium from contacting the epicardial coronary arterial system, thereby causing ischemia.

"The incidence of atrial fibrillation ranges from 15 to 23% and may lead to systemic embolization. It has been observed more frequently in patients with reduced left ventricular systolic function" [9]. "Ventricular tachyarrhythmias are more common and occur in 47% of cases" [10]. "Ventricular fibrillation is a common cause of sudden death in patients with LVNC. Thromboembolic complications can occur in up to 38%" [11]. "Excessive trabeculation and atrial fibrillation of the left ventricle have been shown to be a risk factor for the thromboembolic event". [12].

"Echocardiography is the most widely used diagnostic tool. Many diagnostic criteria have been proposed and include the presence of at least three trabeculations and the presence of blood flow between the trabeculations. In addition, the ratio of compacted to non-compacted myocardium must be>2 at the end diastole, and without secondary causes. Finally, the absence of a "golden standard" makes diagnosis very difficult. Cardiac magnetic resonance has become the method of choice for diagnosing LVNC" [13].

"LVNC has no specific therapy to date. Treatment should be applied according to the clinical situation. In patients with decreased LVEF, treatment of heart failure should be applied according to guidelines" [14].

In the prevention of embolic events, anti-vitamin K and new oral anticoagulants (NOAC) can be considered. In patients with LVNC and AF or a history of embolic events, NOAC is the first choice. In patients with left ventricular thrombus, AVK is the drug of choice.

"There is no agreement on treating patients with left ventricular dysfunction without a history of atrial fibrillation with anticoagulants or aspirin. In patients with normal LVEF, the decision can be made taking into account the CHADS 2 score. Chimenti et al. suggested that patients with a CHADS 2 score >2 should be started on NOACs" [15].

"Ischaemia in LVNC can be explained by several mechanisms. The association of LVNC with coronary artery disease is considered rare and may be explained by a common genetic predisposition to LVNC and MI. There is evidence of an association between MI and the activity of cytochrome P450 2C9 and sarcomere-related proteins such as the calcium-binding protein S100A". [17] "When LVNC is diagnosed and the genes for one or more of these proteins are mutated, an increased risk of MI should be suspected. A coronary angiography study in LVNC subjects showed that 29% of patients had significant coronary artery disease" [18]. "In addition, microcirculatory dysfunction has also been proposed as a mechanism of myocardial ischemia in LVNC. This was supported by post-mortem analyses of LVNC hearts, which showed ischaemic subendocardial lesions". [19] A previous study concluded that "coronary reserve decreases in LVNC patients in both compacted and non-compacted heart muscle, including the normal epicardial coronary system" [18]. In addition, LVNC may affect the progression of remodeling in patients with MI and may worsen their prognosis. "Associated with these mechanisms is the embolic mechanism secondary to thrombus in the trabeculations of the LV or secondary to atrial fibrillation common in the LVNC". [20]

Although the relationship between LVNC and coronary heart disease is not well understood, a number of papers have reported an association between NCC and acute myocardial infarction.

"A few cases have been reported including a 73-year-old patient with left ventricular non-compaction (LVNC) who was diagnosed with acute myocardial infarction (MI), with atheromatous involvement of all three coronary arteries and intraventricular thrombus" [21].

Arun Gopi reported a 19-year-old man with signs of heart failure. [22] TTE revealed "LVNC with severe LV dysfunction. He was put on medical treatment including oral anticoagulants. Despite this, 6 months later he presented with an anterior wall MI. The coronary angiography revealed a thrombotic".

Two other patients were reported, aged 20 years, admitted separately for ST-segment elevation ACS, in whom coronary angiography showed normal coronaries. And in whom the TTE found non-compaction of the LV. [23] These latter cases are similar to our patient who was admitted for non-ST-segment elevation ACS, with echocardiography showing LVNC without thrombus and coronary angiography showing normal arteries.

Conclusion :

LVNC is a rare condition but one that should be kept in mind by the cardiologist as it mimics several cardiac conditions.

LVNC is almost always discovered following complications such as heart failure, thromboembolic events, or following a myocardial infarction as our case illustrates.

The limited but growing number of reported cases provides evidence of an association between NCVG and MI. And because of this, patients with NCVG are considered at high risk for MI. Further studies are needed for a better comprehension of the mechanisms incriminated and codification of the management of this pathology.

Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

Consent

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

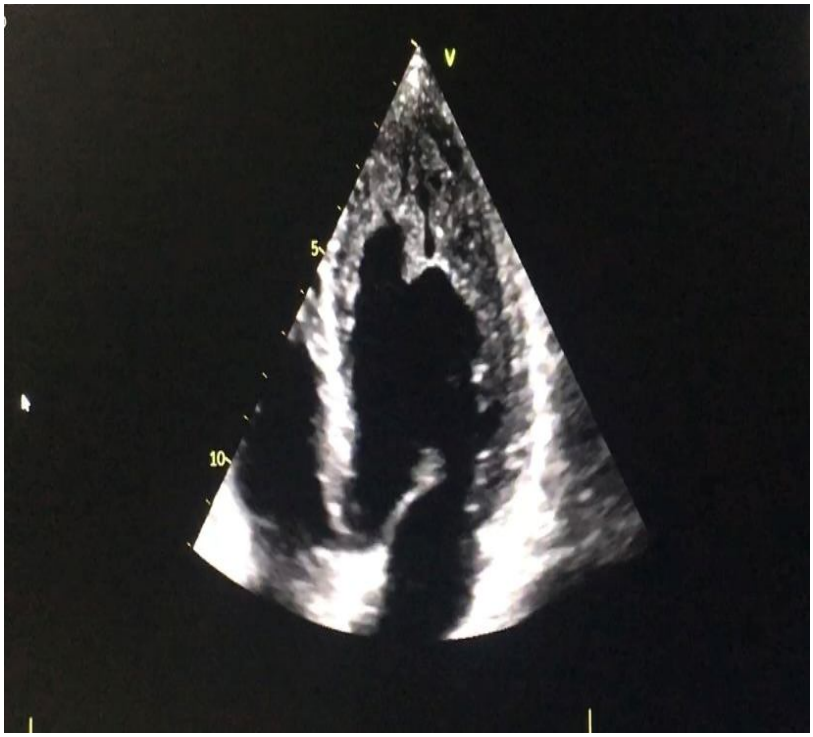

Figure 1 : apical 4-chamber echocardiographic section showing a trabeculated aspect of the apex of the left ventricle

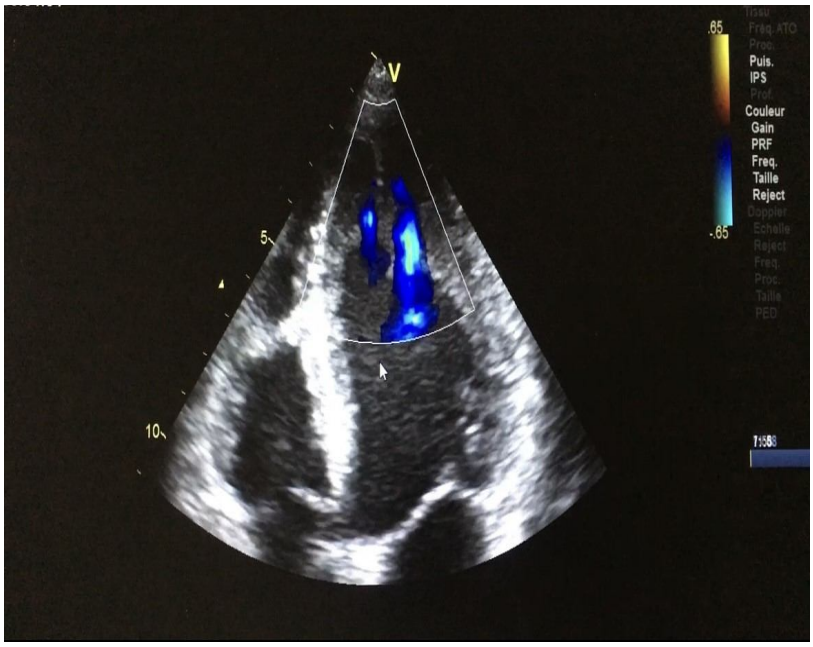

Figure 2 : apical 4-chamber echocardiographic section showing inter trabecular recesses taking colour on colour doppler

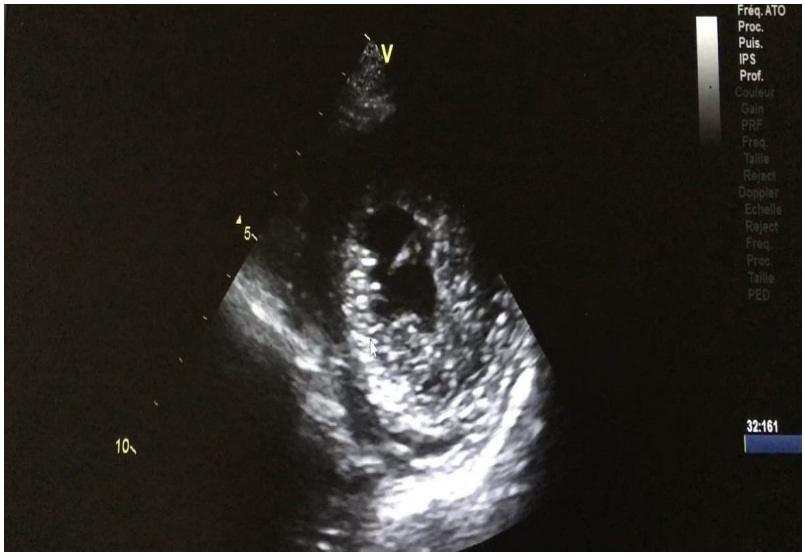

Figure 3 : Parasternal short-axis echocardiographic section showing an aspect of non-compacted tissue at the apical level with a ratio of compacted to non-compacted tissue of 3.14

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