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Case Series

Heart Failure Revealing a Rare Cardiomyopathy Due to Noncompaction of the Left Ventricle: A Series of Three Cases

Insuffisance Cardiaque Révélatrice d'une Cardiomyopathie Rare par non Compaction du Ventricule Gauche : Une Série de Trois Cas

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ABSTRACT

Although rare, the discovery of non-compaction cardiomyopathy of the left ventricle (NCVG) is not exceptional. This is a series of three cases discovered incidentally in patients admitted with symptoms of congestive heart failure. The ultrasound criteria established by Stöllberger and cardiac magnetic resonance imaging were used to diagnose noncompaction in our series. Impairment of the left ventricular ejection fraction was very severe in two of our patients. Ionic disturbance was reported in two patients. Antiarrhythmic treatment combined with an external electric shock allowed the ventricular tachycardia to be resuscitated in the first patient. Coronary angiography would have shed light on a possible association with a coronary anomaly. Diuretic and vasopressor therapy combined with organoprotection with a converting enzyme inhibitor were instituted to relieve the heart failure. This series shows us that an improvement in the technical platform is necessary for adequate management of LVNC in our context.

RÉSUMÉ

Bien que rare, la découverte d'une cardiomyopathie de non-compaction du ventricule gauche (NCVG) n'est pas exceptionnelle. Il s'agit d'une série de trois cas découverts fortuitement chez des patients admis pour des symptômes d'insuffisance cardiaque congestive. Les critères échographiques établis par Stöllberger et l'imagerie par résonance magnétique cardiaque ont été utilisés pour diagnostiquer la non-compaction dans notre série. L'altération de la fraction d'éjection du ventricule gauche était très sévère chez deux de nos patients. Des troubles ioniques ont été rapportés chez deux patients. Un traitement antiarythmique associé à un choc électrique externe a permis de réanimer la tachycardie ventriculaire chez le premier patient. Une coronarographie aurait permis de mettre en évidence une éventuelle association avec une anomalie coronarienne. Un traitement diurétique et vasopresseur associé à une organoprotection par un inhibiteur de l'enzyme de conversion ont été instaurés pour soulager l'insuffisance cardiaque. Cette série nous montre qu'une amélioration du plateau technique est nécessaire pour une prise en charge adéquate de la NCVG dans notre contexte.

INTRODUCTION

Left ventricular noncompaction (LVNC) is a congenital cardiomyopathy characterized by the presence in the left ventricle of fetal myocardium with excessive prominence of a trabecular meshwork and deep intertrabecular cavities [1]. The mechanisms explaining the occurrence of LVNC are still the subject of much speculation. Some authors believe that isolated ventricular non-compaction is caused by the arrest of normal myocardial morphogenesis [2]. The clinical manifestations are very varied, ranging from asymptomatic forms to deterioration in ventricular systolic function, left ventricular dilatation, systemic embolic events and ventricular arrhythmias or sudden cardiac arrest. [3]. Diagnosis of LVNC is based on at least four echocardiographic criteria established by Stöllberger et al, sometimes aided by cardiac magnetic resonance imaging (MRI) [4]. It is a rare primary cardiomyopathy with an estimated prevalence of 0.014% and a male predominance. [5]. We report a series of three cases of noncompaction of the left ventricle discovered

incidentally following symptoms of heart failure in patients of African origin.

CASE PRESENTATION

Clinical Case 1

The patient was a 36-year-old male driver with a sedentary lifestyle as a cardiovascular risk factor. He was admitted with palpitations, exertional dyspnoea, hypersudation and anxiety. Clinical examination revealed arterial hypotension at 90/60 mmHg, tachycardia at 250 beats per minute (bpm), polypnoea at 24 cycles per minute and anxiety. Emergency laboratory tests were normal, apart from hyperuricemia at 562 umol/L. The electrocardiogram (ECG) revealed intolerable ventricular tachycardia (1er episode) at 220 cycles per minute (cpm) (Figure 1). The patient received an external electric shock (EEC) at 200 Joules, followed by amiodarone. The electrocardiogram after EEC and amiodarone showed a regular sinus rhythm at 81 cpm, a normal QRS axis at +50 degrees, symmetrical negative T waves with lateral and inferior peaks, and a corrected QT of 410 ms. Doppler echocardiography showed numerous trabeculations in the left ventricle with a ratio of noncompacted area to compacted area equal to 2.4 in short-axis section and 2.5 in apical 4cavity section. Colour Doppler showed good blood flow within the trabeculations, giving an echocardiographic appearance of non-compaction of the left ventricle (Figure 2). Cardiac MRI (Figure 3) showed a left ventricular ejection fraction of 64% with discrete hypokinesia involving the lateral, medial and apical wall. There was also hypertrabeculation in the lateral, medial and apical walls. The non-compacted myocardium measured 20.5 mm and the compacted myocardium 6 mm, with a ratio of non-compacted area to compacted area of 3.04. The diagnosis was cardiomyopathy due to non-compaction of the LV. The outcome was favourable and the patient was discharged home after one week.

He was readmitted after 04 months for intolerable ventricular tachycardia (2nd episode) following therapeutic non-compliance. Regression of this last episode was obtained after a new CEE of 300 Joules and initiation of amiodarone.

Clinical case 2

The patient was a 39-year-old male driver whose cardiovascular risk factors included a
sedentary lifestyle and active smoking (10 packs/year). He had no specific pathological history. He was presented with a hacking cough with New York Heath Association stage III dyspnoea and lower limb oedema. Clinical examination revealed congestive heart failure. The ECG showed regular sinus tachycardia at 116 cycles/min. Doppler echocardiography revealed hypokinetic dilated cardiomyopathy with impaired systolic function of both ventricles (left ventricular ejection fraction 15%). There were also significant intraventricular trabeculations on the left, with a ratio of compacted to noncompacted myocardium of 2.33, leading to the conclusion that the left ventricle was not compacted (Figure 4). The laboratory work-up revealed hypokalaemia (K^+ =3mmol/L), hypomagnesaemia (Mg^+ =0.58 mmol/L) and normocytic normochromic anaemia at 11g/dL. The diagnosis of noncompaction of the left ventricle with severely impaired left ventricular ejection fraction decompensated into congestive heart failure complicated by ionic disorders was accepted. The patient was treated with diuretics, a conversion enzyme inhibitor and potassium and magnesium supplements. Progress was favourable and the patient was discharged after two weeks.

Clinical case 3

A 14-year-old girl from secondary school with no cardiovascular risk factors or specific pathological history was admitted with New York Association Stage IV dyspnoea, with dizziness and diffuse oedema over the whole body. Physical examination revealed normal consciousness, arterial hypotension of 85/50, tachycardia of 125 bpm, and congestive heart failure syndrome with anasarca. The ECG showed regular sinus tachycardia at 125 cycles per minute. Doppler cardiac ultrasound showed dilated cardiomyopathy with a reduced left ventricular ejection fraction (LVEF) of 25%. There were also right intraventricular microthrombi and two mobile apical thrombi in the left ventricle. The left ventricular cavity was highly trabeculated with disharmonious flow on colour Doppler (Figure 5). Biologically, the ionogram, renal and liver tests were normal, but there was a normochromic microcytic anaemia at 10g/dL. The diagnosis of noncompaction of the left ventricle with impaired LVEF (25%) decompensated into congestive heart failure complicated by cardiovascular collapse and intracavitary thrombi was accepted. Treatment with a diuretic (Furosemide) and vasopressor drugs (dobutamine and dopamine) was initiated using an electric syringe. The course was marked by persistent collapse and congestive signs despite high doses of diuretics. The patient died on the 10th day of hospitalisation from cardiogenic shock.

DISCUSSION

There is a wide variety of circumstances in which LVNC is discovered, as the symptoms are highly polymorphic. Sometimes asymptomatic, NCVG may be revealed by symptoms of cardiac or even neurological insufficiency. Indeed, Fettouhi et al reported a case of a patient admitted for convulsions in whom the diagnosis of LVNC and inferior ACS were made. [6].

In addition to ventricular arrhythmias, coronary artery disease has been reported in a number of series. **Toufan et al,** reported more than 07 cases of MI in patients with LVNC [7]. Panduranga et al. reported a case of ACS without dyslipidaemia and raised the hypothesis that the

same gene responsible for LVNC could lead to a predisposition to coronary heart disease. [8]. Martini et al reported a series of cases of NCVG associated with familial dyslipidaemia and coronary artery disease [9]. However, this explanation is not unanimously accepted by authors, and the pathophysiology of the association between coronary artery disease and non-compaction of the LV is still under investigation. [10]. Coronary angiography would have shed light on a possible association with a coronary anomaly in our series.

LVNC cardiomyopathy is characterised by electrical abnormalities, including areas of low voltage and scarring, primarily related to the presence and extent of myocardial fibrosis rather than uncompacted myocardium [11]. In this genetically diverse phenotype, the development of fibrosis contributes to an arrhythmogenic substrate underlying atrioventricular conduction diseases, supraventricular tachycardias and ventricular tachycardias (VT) [12]. In addition, trabeculations reduce ventricular compliance leading to diastolic dysfunction and therefore inadequate coronary perfusion. The resulting ischaemia is responsible for progressive fibrosis with a progressive deterioration in systolic function and a predisposition to rhythm disorders (TDR) [13]. Within this spectrum, monomorphic ventricular tachycardia is the most frequently observed arrhythmia. It has been reported in 47% of patients with symptomatic LVNC by R. Jenni et al. [5]. Atrial fibrillation is reported in 25% of patients with LVNC [14]. An electrophysiological study would allow risk stratification and guide management in the event of VT.

According to the recommendations of the European Society of Cardiology (ESC) for the treatment of heart failure with reduced EF (EF<40%), the implantable defibrillator is indicated in the case of ischaemic causes, and should only be used in the case of other aetiologies to reduce the risk of sudden cardiac death [15]. It is not indicated for the primary prevention of ventricular arrhythmias or sudden death in LVEF unless LVEF is severely reduced (<35%). [16].

Although the incidence of ventricular arrhythmias and sudden deaths of cardiac origin is high in LVNC, there is little consensus on their management. To date, close monitoring using conventional cardiological diagnostic tools and symptomatic treatment when necessary constitute the therapeutic approach. [17].

CONCLUSION

We report a series of three cases of incidental findings of LVNC during heart failure symptomatology. This rare and generally asymptomatic condition may be revealed by systolic ventricular dysfunction, systematic embolic events and ventricular arrhythmias or sudden cardiac arrest. The therapeutic decision depends on the patient's clinical picture and the doctor's judgement. The automatic implantable defibrillator appears to be an ideal treatment, but its indications in this situation are limited to cardiac arrest, syncope, sustained VT, severe impairment of LVEF and a family history of sudden death.

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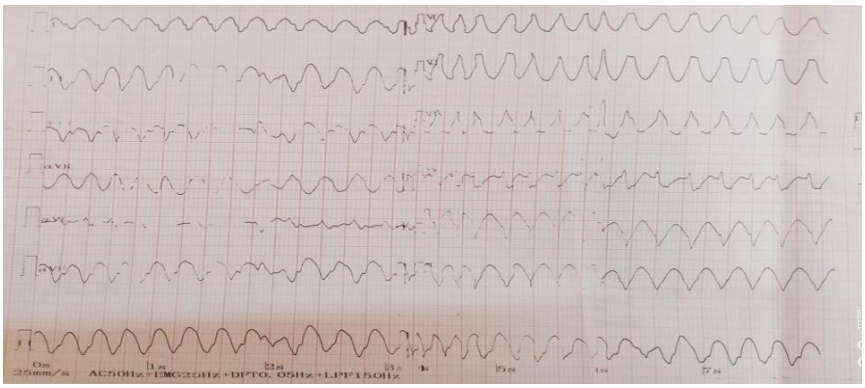

Figure 1: ECG showing ventricular tachycardia

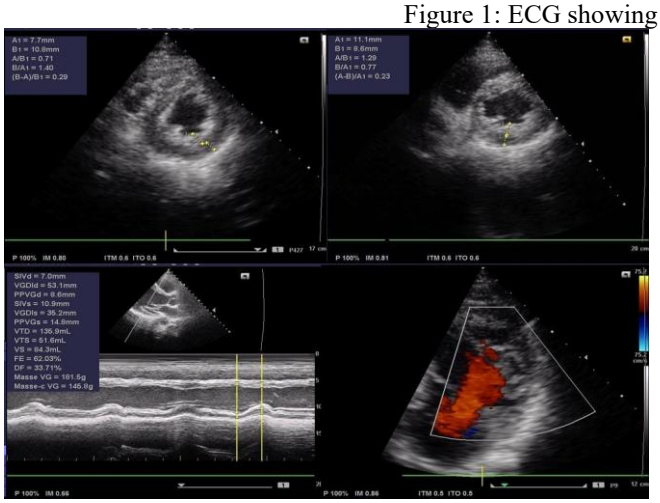

Figure 2: Cardiac ultrasound showing trabeculations in the left ventricle

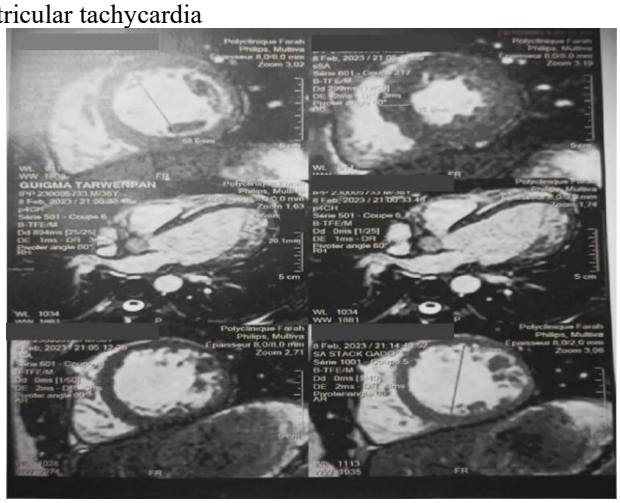

Figure 3: Cardiac MRI showing compaction of the left ventricle

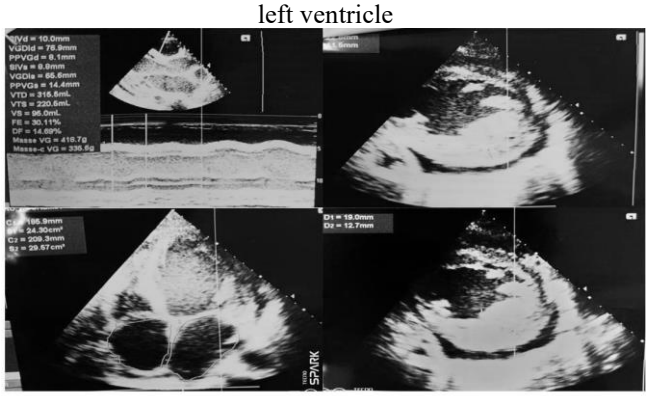

Figure 4: Echographie cardiaque montrant une non compaction du ventriculaire gauche

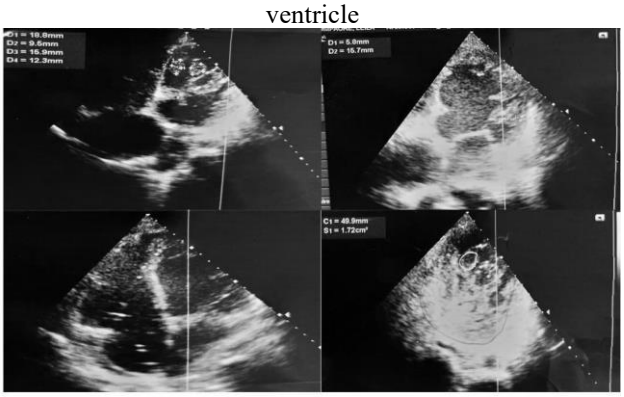

Figure 5: Echographie cardiaque montrant une non compaction du ventriculaire gauche