La non compaction du ventricule gauche

Left ventricular noncompaction in a pregnant patient revealed by preeclampsia

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Résumé

La non compaction du ventricule gauche est une pathologie congénitale rare classée parmi les cariomyopathies génétiques. Elle est caractérisée par un excès de trabéculations ventriculaires proéminentes séparées par des récessus profonds. On présente le cas d'une jeune patiente admise dans un tableau de préeclampsie compliqué d'un état de choc cardiogénique qui mime la cardiomyopathie du peripartum. La non compaction du ventricule gauche a été suspectée à l'echocardiographie transthoracique et confirmée par l'IRM. La patiente a bien répondu au traitement médical avec un prompt rétablissement.

Mots-clés
Préeclampsie, non compaction du ventricule gauche, cardiomyopathie du peripartum, echocardiographie, imagerie par résonnance magnétique

Summary

Left ventricular noncompaction is a rare congenital disease classified as a genetic cardiomyopathy. It is characterized by excessively prominent trabeculations with deep recesses communicating with the ventricular cavity. We describe the case of a young woman presenting with preeclampsia complicated with cardiogenic shock and mimicking peripartum cardiomyopathy. Left ventricular noncompaction was suspected on echocardiography and confirmed by MRI. The patient responded well to medical treatment and made full recovery.

Keywords
Preeclampsia, Left ventricular non compaction, peripartum cardiomyopathy, echocardiography, Magnetic Resonance Imaging

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INTRODUCTION

Left ventricular noncompaction is a rare congenital cardiomyopathy characterized by excessively prominent trabeculations with deep recesses communicating with the ventricular cavity. (1)

It is classified as a genetic cardiomyopathy that is due to an interruption of the normal compaction process of the developing myocardium in utero. (2) It affects both children and adults with a reported increasing frequency due to improved imaging modalities (1).

In this report we present a case of an isolated left ventricular noncompaction revealed during delivery complicated by preeclampsia.

CASE REPORT

A 35-year-old pregnant woman (G2A1) with no significant medical history as well as an unremarkable prenatal course was admitted at 35 weeks of gestation for preeclampsia. On physical examination, she had blood pressure of 145/95 mmHg, 3+ urine dipstick protein and lower extremity edema. The rest of the exam was unremarkable and no neurological signs were noted. Initial laboratory analyses included creatinine 76 µmol/L, uric acid 476 µmol/L, aspartate aminotransferase (AST) 76 UI/L, alanine aminotransferase (ALT) 56 UI/L, lactate dehydrogenase (LDH) 776 U/L, hematocrit 36.7%, 24-hour total urine protein 2.6 g/day, and platelets count 301.109e/l.

Three days after admission, the patient developed shortness of breath and orthopnea. On examination, blood pressure was of 99/55 mmHg, patient was polypneic with crackles on auscultation and low capillary saturation. Oliguria was also noted. EKG showed normal sinus rhythm with the presence of negative T waves in the lateral territory.

Creatinine and urea level increased respectively from 76 µmol/L to 108 µmol/L and 5.4 mmol/L to 11.1 mmol/L. Hepatic function and platelets counts were normal. N-terminal pro b-type natriuretic peptide (NT-proBNP) was 2598 pg/ml and cardiac troponin I was 0.19 ng/ml.

These findings were consistent with congestive heart failure. The decision was made to proceed with delivery due to worsening preeclampsia. Urgent Cesarean delivery was performed and the newborn was delivered without complications.

During the Cesarean delivery, the patient presented signs of cardiogenic shock and she was put on Dobutamin and Norepinephrin. Presumptive diagnosis was peripartum cardiomyopathy (PPCM).

Urgent transthoracic echocardiography (TTE), performed in the ICU revealed a dilated left ventricle with an ejection fraction of 40%, global left ventricle hypokinesis and lateral wall akinesis. Severe tricuspid regurgitation, right ventricle systolic dysfunction with right ventricular systolic pressure of 60 mmHg and, moderate right atrial dilatation were noted. Severe thickening of the myocardium with prominent trabeculations and multiple deep recesses were visualized; consistent with left ventricular noncompaction (LVNC). (Fig1 and Fig2)

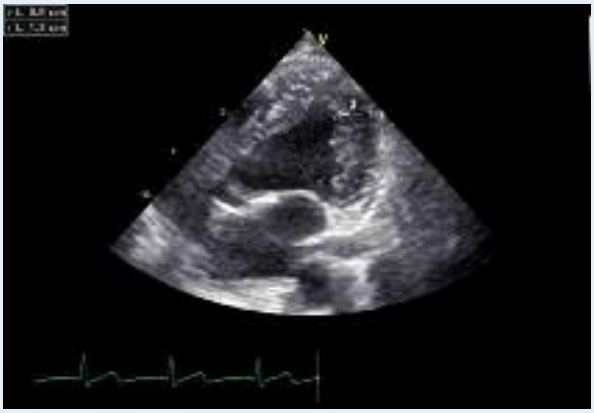

Figure 1 : Two-dimensional echocardiogram (apical 2-chamber view) showing a two-layer structure with a thin normally compacted layer (2) and a markedly thickened non-compacted layer (1).

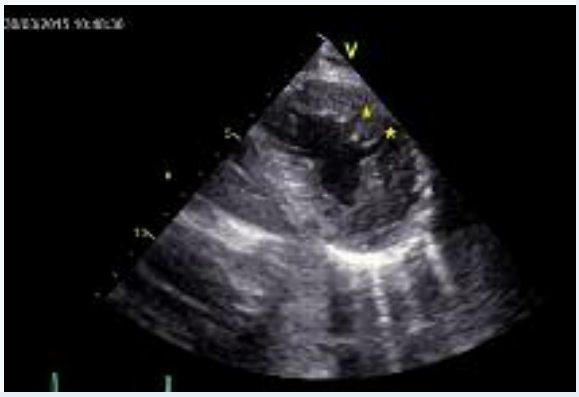

Figure 2 : Two-dimensional echocardiogram (short-axis view) showing prominent trabeculations and Recesses (yellow stars) in noncompacted portion of left ventricle

Cardiac magnetic resonance imaging (MRI) confirmed the diagnosis showing a two-layer structure interesting the lateral wall, with a thin, normally compacted layer (C) and a markedly thickened non-compacted layer (NC) with a ratio of NC/C = 3,8 (Fig 3).

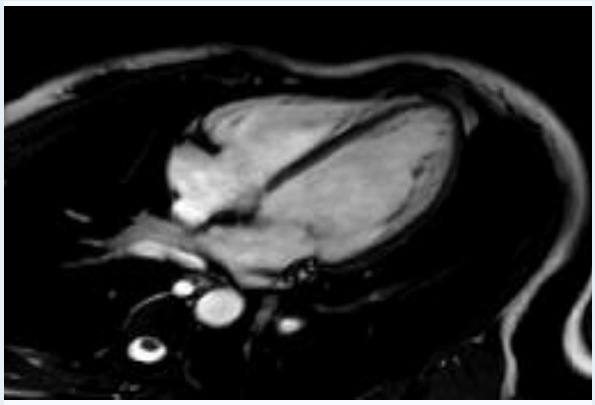

Figure 3 : Cardiac magnetic resonance imaging showing an increase in endocardial LV trabeculation interesting the lateral wall with a maximum ratio of non-compacted layer to compacted layer is 3,8

A TTE was performed for the new born and was revealed normal.

The patient responded well to medical therapy and was successfully extubated 24 hours after the delivery. Dobutamin was stopped within 36 hours and she was discharged to the cardiology department receiving Beta blockers, angiotensin converting enzyme inhibitor and diuretics. Her preeclampsia resolved shortly with no residual sequalae and on one year follow up the patient had a NYHA class II dyspnea with an improved left ventricular ejection fraction to 65%

DISCUSSION

Development of heart failure in the third trimester of pregnancy can be challenging. The heart failure could be due to an underlying cardiac disease or due to the pregnancy itself especially in case of preeclampsia, as heart failure can occur in up to 3% of severe preeclampsia (3). In developing countries where rheumatic heart disease is still widespread, heart failure is commonly seen in young women with rheumatic valvular lesions. However, physicians should keep in mind other possible diagnoses such as myocardial infarction, PPCM, aortic dissection and pulmonary embolism (4). Echocardiography is mandatory for etiologic approach.

In this case, the patient's clinical presentation mimicked PPCM as she had no prior history of cardiovascular diseases, no other apparent cause of heart failure and the signs and symptoms of heart failure developed within the last month of pregnancy.

PPCM is defined as "idiopathic cardiomyopathy" presenting with heart failure secondary to left ventricular systolic dysfunction towards the end of pregnancy or in the months following delivery, where no other cause of heart failure is found; it is a diagnosis of exclusion; the left ventricular ejection fraction (LVEF) is nearly always reduced below 45% (5).

For our patient TTE showed a LVEF of 40%; however, it revealed signs consistent with left ventricular noncompaction.

LVNC is a primary cardiomyopathy that is characterized by excessively prominent trabeculations and deep intertrabecular recesses in one or more segments of the ventricle wall (6).

Clinical presentation of LVNC is highly variable. It can occur at any age, range from asymptomatic to end-stage heart failure, or be associated with arrhythmias, sudden cardiac death or thromboembolic events (7).

Physiologic changes that accompany pregnancy added to preeclampsia are not well tolerated by patients with LVNC and may precipitate symptoms of heart failure. Among some reported cases, it was concluded that pregnancy has a worsening effect on LVNC and patient may need close follow up and intensive treatment including inotropes. (8,9)

It could be then concluded that our patient had an asymptomatic LVNC revealed by the association of pregnancy and preeclampsia due to the worsening effect on ventricular function.

LVNC diagnosis relies on echocardiography but cardiac MRI is used to confirm the clinical suspicion. The most widely used diagnosis criteria in echocardiogaphy are those proposed by Jenni et al (1) where LVNC was defined as a two-layer structure, with a thin, normally compacted layer (C) and a markedly thickened non-compacted layer (NC) (with a ratio of NC/C >2), excessively prominent trabeculations, and deep inter-trabecular recesses measured at end-systole in the parasternal short-axis views. This diagnosis also assumes that the intertrabecular spaces are filled by blood flow from the ventricular cavity, as visualized on color

Doppler, and no other cardiac abnormality is reported.

Cardiac MRI not only reinforces identification of LVNC morphologic features but also allows assessment of cardiac pathophysiology, with late gadolinium enhancement (LGE) identifying myocardial fibrosis. It was reported that LGE could reflect myocardial damage and predict prognosis of LVNC. (7)

There is no specific treatment for LVNC; it ranges, depending on symptoms and stage, from oral anti congestive medications including angiotensin-converting enzyme inhibitors, betablockers and aldosterone antagonists to ventricular assist and cardiac transplantation (7).

CONCLUSION

Left ventricular noncompaction is a rare cardiomyopathy with no specific therapy. Management depends on symptoms. It should be considered as a differential diagnosis in preeclamptic patients presenting with heart failure. The importance of the diagnosis is accentuated by its genetic character as it could affect the infant. Thus, screening echocardiography is crucial for an early management.

Conflict of interest:

The authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest.

Patient consent:

Patient's written consent was obtained for publication of the article without limitations.

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