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Non-compaction with Coronary Artery Disease; a Case Report

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ABSTRACT

Left ventricular non-compaction is a rare genetic disease of the heart muscle. Compaction is a process occurs during the development of the heart. If this process is not complete, the inside of the heart muscle will look spongy. The only definitive treatment of LV non-compaction is cardiac transplantation. Prevention of both heart failure and thromboembolic events are the main target of treatments. We describe a patient with the diagnosis of isolated LV non-compaction made after presentation with dyspnea. Angiography showed three-vessel disease. The patient underwent bypass grafting. Early diagnosis of disease can prevent the occurrence of myocardial infarction.

Key words: non-compaction, myocardial infarction, coronary artery disease, echocardiography, Heart failure

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1. INTRODUCTION

Left ventricular Non-compaction (LVNC) is a rare cardiomyopathy in adults. The incidence of LVNC is not obviously known in a large collection of cardiomyopathy it reported from 0.014% to 0.05 % (1). Both acquired and familial kinds have been reported. However, LVNC is mainly associated with autosomal dominant sample of inheritance, X linked inheritance and de novo mutations have been described; with LVNC, the heart muscle seems to become a slack tissue of fibers instead of a tight layer with all place's deletion; this appearance is explained as "non-compaction" or "spongiform," and it can happen in the attendance of normal heart function or in relationship with ventricular dysfunction where the heart muscle does not squeeze normally (2). Clinical manifestations are variable from no symptoms to life-threatening complications (2, 3). Diagnosis is achieved by echocardiography. However, magnetic resonance imaging (MRI) is useful when no echocardiogram is available (4). There is no specific therapy for patients. Management of non-compaction is currently focused on control of symptoms and prevention of complications (5). In this case report, we

describe one case of LVNC associated coronary artery disease, this is the case of LVNC associated with coronary artery disease with no clinical evidence of subclinical or acute myocardial infarction and diagnosed during hospitalization at our heart center for evaluation of exertional dyspnea.

2. METHODOLOGY

2.1. Case presentation

A 44-year-old male patient was admitted to heart center in Afshar hospital, Yazd for evaluation of exertional dyspnea (New York Heart Association functional class II). His medical history was not significant. The family history was unexceptional and negative for coronary artery disease at an early age. He had no prior history of heart disease. Patient had reported not history of cigarette smoking and drug abuse. On physical examination, his blood pressure was 160/90 mmHg, his heart rate was 84 beats/min, the respiratory rate was 16/min and body temperature was 36.2C. The clinical examination revealed an apical grade of 3/6 systolic murmur. Jugular venous pressure and Carotid pulse volumes were normal. The first and second heart

sounds were normal. The lungs were clear to auscultation. There was no hepatomegaly or splenomegaly. Neurologic findings were normal. The results of routine laboratory tests of cardiac diseases and supplementary tests, including leucocyte count, C-reactive protein and sedimentation rate were normal. The 12 lead electrocardiogram of the patient demonstrated nonspecific changes: normal sinus rhythm, normal QRS duration and strain pattern in the lateral leads (leads I, aVL, V5, and V6). Transthoracic echocardiography revealed a normal left ventricle size, moderate to severe systolic dysfunction (Estimated ejection fraction $\leq 35\%$), and severe hypokinesis of inferior, inferolateral and inferoseptal wall of left ventricle with left ventricular hypertrophy which met the criteria for LVNC (Figure 1). Because there is no specific treatment for LVNC, our management was directed at controlling the patient's symptoms and reducing the morbidity of his heart failure. The patient was treated medically with carvedilol, aspirin, losartan, and full dose anti-coagulant therapy. Despite optimization of medical therapy, angiography was performed and revealed coronary artery disease in three vessels (Figure 2) the patient was referred for Off-pump coronary artery bypass grafting (OPCABG). In this operation, three grafts were given: left internal mammary artery (LIMA) to the left anterior descending (LAD), Saphenous Vein Graft (SVG) to first obtuse marginal branch (OM1) and SVG to Posterior descending artery (PDA). Echocardiography was done during the follow-up; within a few months after surgery, an increase in the left ventricular ejection fraction to 45% indicated a good surgical result.

3. RESULTS AND DISCUSSION

The Non-compaction results in numerous prominent trabeculations and a loose myocardial meshwork in the left and/or right ventricular, probably due to an arrest of compaction during intrauterine life (3). It was first described by Engberding and bender in 1984 referring to a 33-year old woman with persistent sinusoid in the left ventricle as an isolated abnormality (4). Accurate pathophysiology of the ventricular dysfunction is unknown (3). The criteria most often used for the diagnosis of NC are those proposed by Jenni *et al* and Stollberger *et al* (2, 5). While echocardiography is the typical reference for the diagnosis of LVNC, MRI obviously has the potentiality, particularly in patients with echocardiography, for good quality cannot be obtained (6). In this study, we used echocardiographic definitions to identify LVNC. Definite diagnosis was done by echocardiographic assessment and MRI was not necessary.

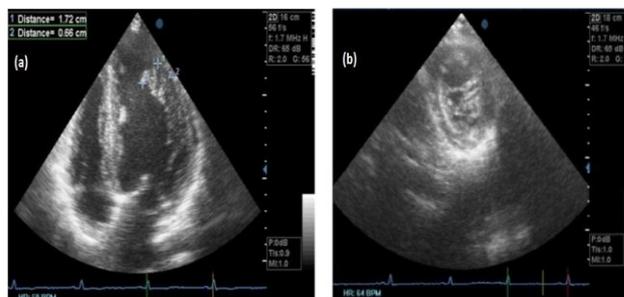


Figure 1. (a): Echocardiogram of the left ventricle showing prominent trabeculation, that the ratio of non-compacted to compacted LV myocardium is greater than 2:1; (B): The echocardiogram pictured shows left-ventricular non-compaction. The parasternal short axis view echocardiogram

NCLV has a different presentation in adults which dyspnea is the most common symptom, occurring in 79% of cases (7). Our patient was also presented with exertional dyspnea. An extensive review demonstrated that more than 50% of the patients developed symptomatic heart failure. Both systolic and diastolic dysfunction has been distinguished (8). Diastolic dysfunction is related to the observed relaxation, abnormal filling and restriction caused by numerous trabeculae (9) Two forms of this anomaly have been described; LVNC-associated with congenital heart diseases, genetic syndromes, and the absence of other cardiac anomalies (5, 8). It can be associated with other cardiac and neuromuscular pathologies (4). Although association between LVNC and MI and/or coronary artery disease is said to be rare, it is possible that this association has previously been undiagnosed. Myocardial ischemia may be the predominant mechanism underlying progressive systolic dysfunction. Systolic dysfunction appears to be related to hypo perfusion and the endocardial microcirculatory dysfunction (9, 10). It may affect the progression of remodeling in patients with MI and may worsen their prognosis (4, 6, 10-12). Treatment is symptomatic including the consideration of heart transplantation. There are no accepted guidelines as how to treat patients with NC. In patients who developed symptoms of heart failure and had advanced heart failure symptoms, medical treatment with ACEI, diuretics, beta-blockers (especially carvedilol) appear to have favorable results. There are also suggestions of aspirin or anti-coagulation therapy to decrease the risk of emboli. Other invasive management for NC includes the use of implantable cardioverter defibrillator (ICD) and cardiac transplantation (3, 4). Our patient was presented with heart failure for the first time at the age of forty. Despite previous studies, there was no evidence of acute myocardial infarction (AMI) in this case. With regard to Left ventricular (LV), diastolic dysfunction and subclinical systolic dysfunction may be markers of coronary artery disease, and moreover, the left ventricular ejection fraction was low, angiography was done and showed three-vessel coronary artery disease. Based on the angiography, it seems that in this case heart failure is secondary to coronary artery disease and it is reinforced by a significant increase in left ventricular ejection fraction after surgery. CABG is often the preferred method of revascularization in patients with high-risk left main, 3-vessel with substantial LAD involvement and LV

dysfunction (13). In this case, CABG was offered and the patient underwent coronary artery bypass grafting. Several studies have recently reported asymptomatic familial disease in some patients. Clinical studies suggest that it is often Familial with a predominantly autosomal dominant Inheritance with incomplete penetration.

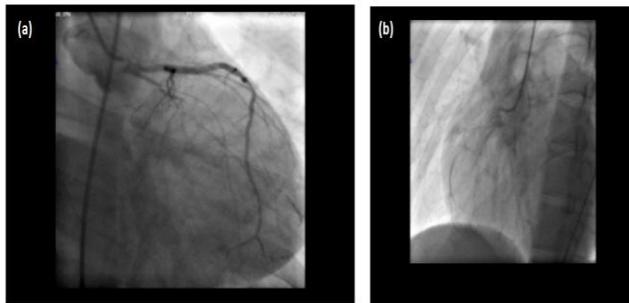


Figure 2. Angiography revealed coronary artery disease in three vessels (a/b)

It has also been linked to mutations in several genes (11). History of family LVNC was associated with history of family dyslipidemia; in addition was strongly associated with history of family CAD (10). On the contrary, our patient had LVNC and CAD without dyslipidemia. Importantly, the prognosis of patients with non-compaction cardiomyopathy is poor in general and improves significantly with heart transplantation. Thus, therapy should be directed at the prevention and management of the most frequent manifestations of the disease (5).

4. CONCLUSION

Ventricular non-compaction is a rare Cardiomyopathy that is often misdiagnosed and prognosis in general is poor. Early medical management with standard therapies is the key to improve the outcome. Patients with LVNC, despite the risks listed, could be considered at high risk of danger for MI so, early diagnosis and intervention and screening of family members can decrease the morbidity and mortality and can help prevent the occurrence of myocardial infarction. We recommend annual follow-up with clinical assessments for patients and symptomatic patients with ventricular non-compaction are believed to require transplant early.

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AUTHORS CONTRIBUTION

This work was carried out in collaboration between all authors.

CONFLICT OF INTEREST

The authors declared no potential conflicts of interests with respect to the authorship and/or publication of this article.

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