

Biventricular Non Compaction associated with Silent PDA in Elderly Presenting as Heart Failure

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ABSTRACT

Non-compaction cardiomyopathy (NCC), also called spongiform cardiomyopathy, is a rare congenital cardiomyopathy that affects both children and adults. It results from the failure of myocardial development during embryogenesis. In this case report we present a 70 year old male patient presenting with history of dyspnea on exertion and easy fatigability of 1 year duration. Echocardiography revealed a biventricular non compaction associated with a patent ductus arteriosus (PDA). Search through medline showed 8 case reports LV non-compaction associated with PDA in younger patients and this is the first case report of biventricular non compaction associated with PDA in an elderly patient.

Key words: Cardiomyopathy, Heart Failure, Non Compaction, PDA, Spongiform.

INTRODUCTION

Non-compaction cardiomyopathy (NCC), also called spongiform cardiomyopathy, is a rare congenital cardiomyopathy that affects both children and adults. It results from the failure of myocardial development during embryogenesis. The exact prevalence of the disease is not known. In earlier studies,¹ non-compaction was detected in approximately 0.05% of patients who underwent echocardiographic examination. Myocardial non compaction can present at any age. The clinical presentation of patient with non-compaction is widely variable. The incidence of complications and associated congenital heart diseases is high. Myocardial non-compaction can occur at any age; the clinical presentation is nonspecific and varies from no symptoms to conduction defects,

thromboembolism, ventricular arrhythmias, severe heart failure, or sudden cardiac death.² Here we are reporting a 70 year male presenting with symptoms of heart failure and echocardiography revealing biventricular non-compaction cardiomyopathy associated with PDA.

CASE REPORT

A 70-year-old man presented at our institution's with progressive shortness of breath and fatigue since the past 1 year. His past medical history was not significant. Patient was not on any medications. He did not have any history of chest pain, palpitations or syncope. At presentation patient had a pulse rate of 92 bpm, BP of 100/70 mmHg, SPO2 of 96% and signs of heart failure like elevated JVP, S3, basal rales and mild pedal edema. On auscultation there were no audible murmurs. ECG showed LBBB. Chest X-ray revealed cardiomegaly with pulmonary congestion. Echocardiography revealed four chamber dilatation (LVIDd-6.0cm and LVIDs-4.9 cm) with severe LV dysfunction (LVEF-30%). Two-dimensional apical four chamber (Figure 1) and parasternal short axis images (Figure

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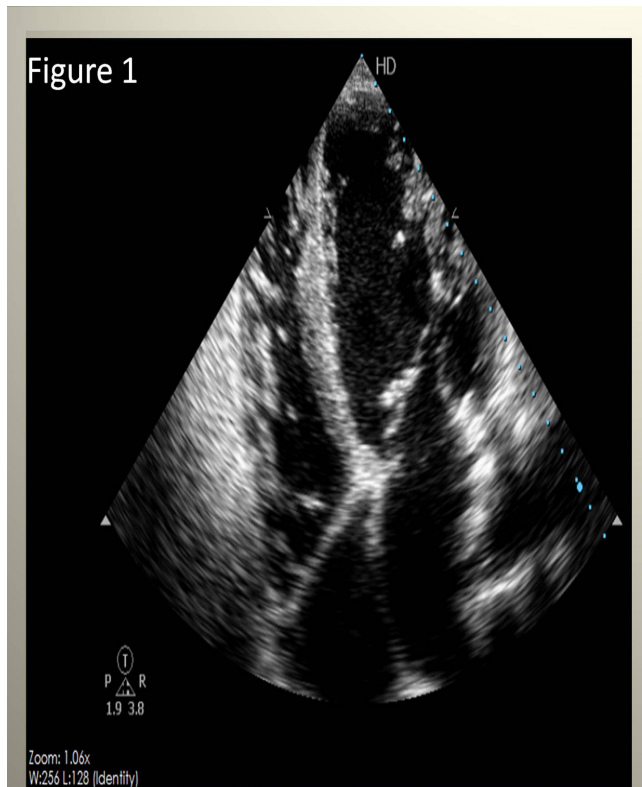


Figure 1: Two-dimensional apical four chamber dilatation of both ventricles, multiple trabeculae and intertrabecular recesses

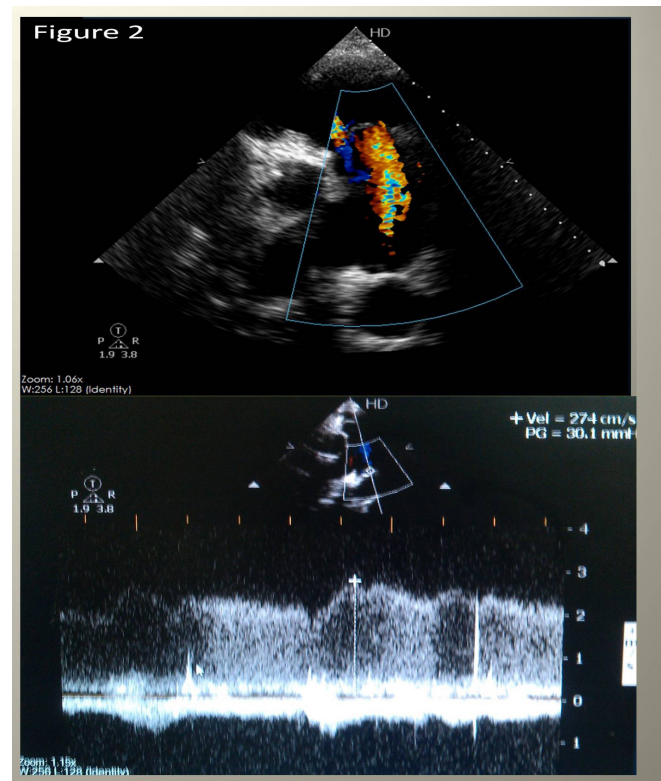


Figure 2: High Parasternal short axis with color and spectral Doppler shows associated patent ductus arteriosus (PDA)

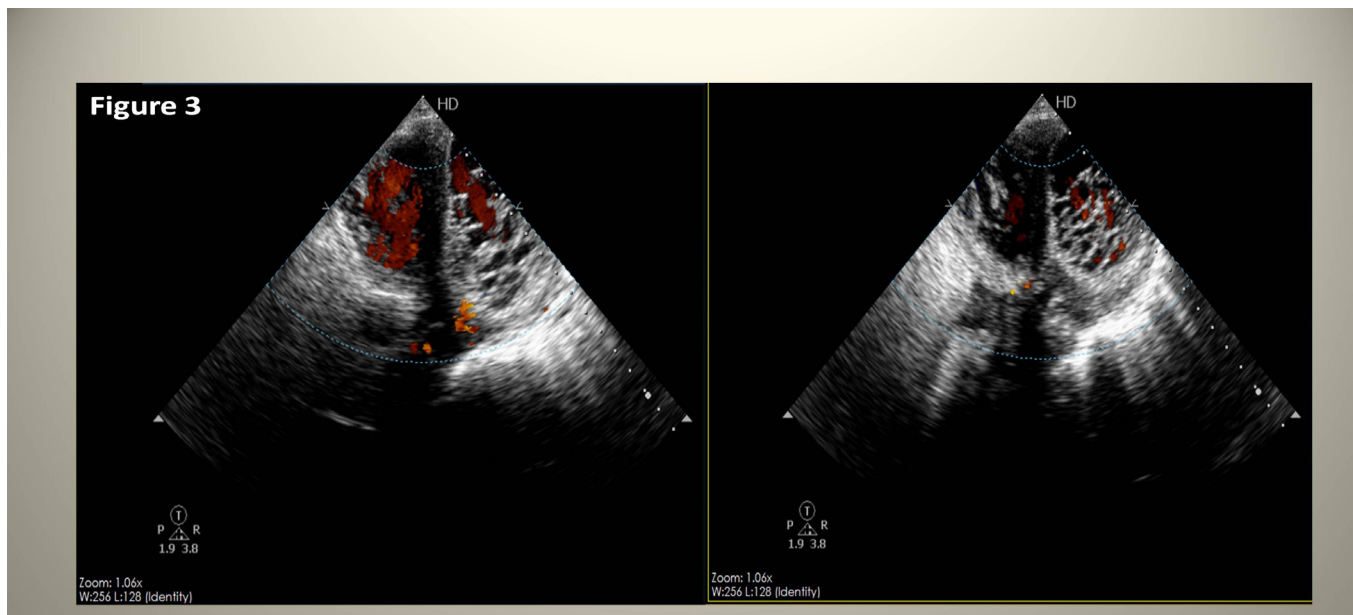


Figure 3: Parasternal short axis images at the level of the ventricles show dilatation of both ventricles, multiple trabeculae and intertrabecular recesses

2) at the level of the ventricles show dilatation of both ventricles, multiple trabeculae and intertrabecular recesses in inferior, lateral, anterior walls, middle and apical portions of the septum and apex of the left ventricle. Transthoracic

two-dimensional study ductal view (Figure 3) with color Doppler shows associated patent ductus arteriosus (PDA). Moderate mitral and severe tricuspid regurgitation and pulmonary hypertension were also noted (peak systolic

pulmonary artery pressure, 60 mmHg). The end-systolic ratio of non-compacted-to-compacted myocardium was >2:1. Color-flow Doppler echocardiography showed blood flow in deep inter trabecular recesses. The patient had features of mild heart failure for which he was given diuretics, beta blockers, angiotensin converting enzyme inhibitors after which patient's condition improved. The patient was discharged. Medical treatment was advised.

DISCUSSION

In this case report we present a 70 year old male with mild heart failure and cardiomyopathy secondary to biventricular non-compaction and PDA. Though there are previous reports of non-compaction cardiomyopathy associated with PDA this is the first case report of biventricular non compaction associated with a PDA in a 70 year old patient. There is a previous case of isolated left ventricular compaction in a 90 year old patient presenting with heart failure but without other associated congenital abnormalities.

Isolated left ventricular non-compaction is a cardiomyopathy characterized by prominent trabeculations and deep inter trabecular recesses in the left ventricular myocardium. Two distinct myocardial layers may be demonstrated by echocardiography, with a thin epicardial layer and a thicker non-compacted endocardial layer resulting in a trabecular meshwork with deep endomyocardial spaces. Involvement of both ventricles with non compaction has been reported rarely. Myocardial non-compaction is thought to result from the interruption of myocardial morphogenesis during embryonic development,^{3,5} although there is some controversy as to whether the condition can also be acquired.¹ During 4th week of gestation the myocardium is composed of a loose, myofibrillary network separated by deep recesses. The spongiform sinusoidal tissue matures to a compact layer from endocardium to epicardium and base to apex.

LV non-compaction has been associated with various other congenital anomalies, including obstructive LV or RV outflow tracts, and an anomalous origin of the left coronary artery from the pulmonary artery. Various forms of outflow tract obstruction during fetal life, such as left ventricular outflow tract obstruction or pulmonary atresia with intact interventricular sputum, are responsible for the persistence of deep endomyocardial spaces surrounded by exaggerated hypertrophy of the trabeculae. Non-compaction of the myocardium has been found in ventricles exposed to excessively high pressures during intrauterine development. In contrast, the reasons for isolated non-compaction are unknown, and it rarely occurs.

This patient had small PDA which was detected incidentally. The contribution of the PDA to heart failure in this patient could not be assessed as the patient was not willing for any invasive investigations and opted for conservative management. Cath study in this patient would have helped to decide the contribution of PDA in heart failure.

Symptoms from non-compaction cardiomyopathy range widely. Patients can present with heart failure, arrhythmias and thromboembolic events. Inter trabecular recesses was deemed, a favorable location for thrombus formation, the real prevalence of thrombi in patients with NVM is unknown. In a study by Dr. Giovannia *et al* where 229 patients were followed up for average 7.3 years the prevalence of stroke and embolism was very low.⁶ Therefore, long-term prophylactic anticoagulation for all patients with non-compaction whether or not thrombus has been found should not be recommended. It must be kept in mind, however, there is an indication for oral anticoagulation in patients with non-compaction and AF, high risk of thrombi formation such as Behcet's disease etc or a history of thromboembolic events. In 2 of the largest populations analyzed to date^{2,4} the annual mortality rate from sudden cardiac death in patients with LVNC was 8% to 9%. Heart-failure symptoms, which were present in 56% of patients in a systematic overview of 5 eligible studies,⁴ can range from very mild to severe. Both systolic and diastolic dysfunction can develop. Echocardiography is crucial for the diagnosis of LVNC. Widely accepted diagnostic criteria⁵ include the following:

- 1) An excessively thick myocardial wall structure in 2 differing layers-a thin, compacted epicardial layer and a thicker, noncompacted endocardial layer
- 2) A characteristic end-systolic ratio of >2:1 for noncompacted-to-compacted wall thickness
- 3) Prominent multiple, chiefly intracavitary trabeculae, with color-Doppler echocardiographic evidence of communication between the deep intertrabecular recesses and the ventricular cavity.

Other investigations which are helpful in confirming the diagnosis of non compaction as myocardial contrast study, Cardiac Magnetic resonance imaging (CMR) and biopsy which is very rarely used.

Treatment of the patients depends on the presenting symptoms and signs. Asymptomatic patients can usually followed up with regular checkups, mild to moderate symptomatic patients can be managed medically. Refractory heart failure patients may require heart transplantation. In our patients who had signs of mild heart failure and was managed medically. Patients who have isolated noncompaction with partial involvement of ventricular segments can be

expected to live normal life without developing symptoms of heart failure. Biventricular non compaction is extremely unusual cause of heart failure in elderly patients, it should be considered in the differential diagnosis of patients presenting with symptoms of heart failure for first time.

CONFLICTS OF INTEREST

All the authors declare no conflicts of interest.

REFERENCES

1. Stollberger C, Finsterer J. Left ventricular hypertrabeculation/non-compaction. J Am Soc Echocardiogr. 2004; 17(1): 91-100.
2. Bhatia NL, Tajik AJ, Wilansky S, Steidley DE, Mookadam F. Isolated noncompaction of the left ventricular myocardium in adults: a systematic overview. J Card Fail. 2011; 17(9): 771-8.
3. Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium. A study of eight cases. Circulation 1990; 82(2): 507-13.
4. Greutmann M, Mah ML, Silversides CK, Klaassen S, Jost CH, Jenni R, *et al.* Predictors of adverse outcome in adolescents and adults with isolated left ventricular noncompaction. Am J Cardiol. 2012; 109(2): 276-81.
5. Jenni R, Oechslin EN, Van der Loo B. Isolated ventricular non-compaction of the myocardium in adults. Heart 2007; 93(1): 11-5.
6. Giovannia F, Giovannib C, Elisabetta Z, *et al.* Anticoagulant drugs in noncompaction: A mandatory therapy? J Cardiolvasc Med. 2008; 9(11): 1095-7.

ABBREVIATION

NCC : Non Compaction Cardiomyopathy
PDA : Patent ductus arteriosus
LV : Left Ventricle
JVP : Jugular Venous Pressure
LVEF : Left Ventricular Ejection Fraction
LVNC : Left Ventricular Non Compaction
CMR : Cardiac Magnetic Resonance