

A case report of unusual coexistence between LVNC and PDA.

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Abstract

Non-Compaction Cardiomyopathy (NCC) (spongiform cardiomyopathy) is a rare congenital cardiomyopathy. NCC affects both children and adults. This is due to the failure of myocardial development during embryogenesis. In this case we present a 29 years old female patient presenting with history of dyspnea on exertion and palpitation since 3 month duration. Echocardiography suggesting a biventricular non compaction associated with a Patent Ductus Arteriosus (PDA). Searching websites showed 8 case reports LV non-compaction associated with PDA in younger patients and this is the ninth case report of biventricular non compaction associated with PDA in an young patient.

Keywords: Heart failure, Non-compaction, PDA.

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Introduction

Non-Compaction Cardiomyopathy (NCC) is a rare congenital cardiomyopathy that affects both children and adults. It results from the failure of myocardial development. Prevalence of the disease is not known. NCC found in approximately 0.04%-0.05% of patients who underwent echo examination. NCC can present at any age. The clinical presentation of patient is highly variable. The incidence of complications and associated congenital heart diseases is high. Clinical presentation is non-specific and varies from no symptoms to conduction defect, thromboembolism, ventricular arrhythmias, severe heart failure, or sudden cardiac death [1-2].

Case Report

A 29 years old female presented at our centre with progressive shortness of breath and palpitation since the past 3 month. Past history was not significant. Patient was not on any medications. She didn't have any history of chest pain or syncope. At presentation patient had a pulse rate of, BP of 100/70 mmHg, pulse 110 bpm, SpO2 of 94% and signs of heart failure like elevated JVP, basal rales S3 and pedal edema. On examination there was short systolic murmur audible on left 2nd space area. ECG showed poor R wave progression. Chest X-ray suggested cardiomegaly. Echocardiography suggested four chamber dilatation (LVIDd-58 mm and LVIDs-48 mm) with severe LV dysfunction (LVEF 35%-40%) (Figures 1 and 2).



Figure 1. Plex view.

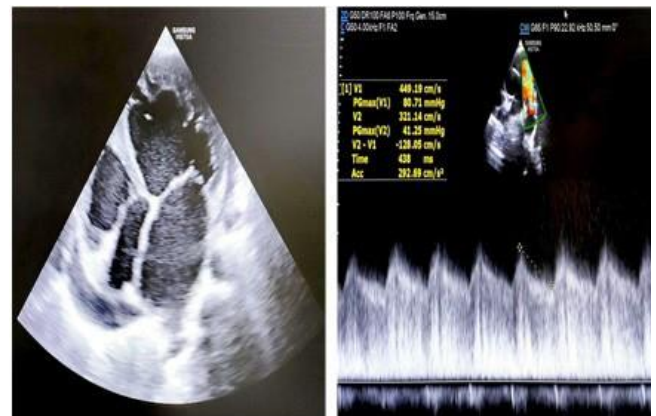


Figure 1. 4-C view and ductal view.

2DPLEX, 2D 4C and 2DSAX images at the level of the ventricles show dilatation of both ventricles, multiple trabeculae and inter trabecular recesses in inferior, lateral, anterior walls, middle and apical portions of the septum and apex of the left ventricle [3]. TTE two-dimensional study ductal view with color Doppler suggested associated Patent Ductus Arteriosus (PDA). Mild mitral and mild tricuspid regurgitation and mild pulmonary hypertension, mild pericardial effusion noted. The end-systolic ratio of non-compacted-to-compacted myocardium was >2:1. Color Doppler showed blood flow in deep inter trabecular recesses. 24 hrs holter monitoring was also done, no af, afl, svt, vt detected, few apc and vpc detected. The patient had features of heart failure for which she was given diuretics, ace inhibitor, beta blockers, anticoagulant after which patient condition improved. Patient was discharged. Medical treatment was advised (Figures 3 and 4).

Chin criteria (NC:C>2)

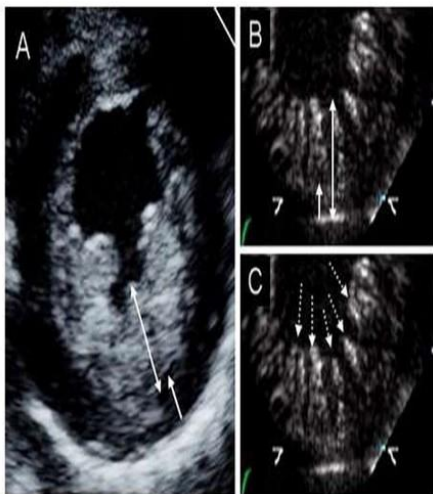


Figure 3. (A) Long arrow-non compaction portion of LV wall and short arrow-compaction portion of LV wall (B) Shows recesses (C) Arrow shows trabeculation in LV wall.



Figure 4. (A) Red arrow-non compaction portion of LV wall and green arrow-compacted portion of LV wall (B) Red arrow-show trabeculation and green arrow-recess in LV wall.

Results and Discussion

In this case study a 29 years old female presented with heart failure and cardiomyopathy secondary to biventricular non-compaction and PDA. Although there are few previous case reports of LVNC associated with PDA this is the first case report of biventricular non compaction associated with a PDA in a 29 years old patient. Isolated LVNC is a cardiomyopathy characterized by prominent trabeculations and deep inter trabecular recesses in the left ventricular myocardium. Two myocardial layer seen by echo, with a thin epicardial layer and a thicker non-compacted endocardial layer resulting in a trabecular meshwork with deep endomyocardial spaces. Both ventricles involvement with non-compaction reported rarely. Myocardial non-compaction is result of the interruption of myocardial morphogenesis during embryonic development. At 4th week of gestation the myocardium is made 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 [4-6].

LVNC has been associated with various other congenital anomalies, including obstructive LV or RV outflow tracts. This patient had small PDA which was detected incidentally. Symptoms of LVNC varies, can present with heart failure, arrhythmias and thromboembolic events. Inter trabecular recesses was deemed, a favorable location for thrombus formation [7]. Long-term prophylactic anticoagulation for all patients with non-compaction whether or not thrombus has been found should not be recommended [8-10]. There is an indication for oral anticoagulation in patients with non-compaction and AF, high risk of thrombi formation such as Bachet disease etc or a history of thromboembolic events. In 2 of the largest populations analyzed 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 can range from mild to severe. Both systolic and diastolic dysfunction can develop. Echo is must for the diagnosis of LVNC. Widely used diagnostic criteria:

- 1) An excessively thick myocardial wall structure in 2 differing layers-a thin, compacted epicardial layer and a thicker, non-compacted 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 echo evidence of communication between the deep intertrabecular recesses and the ventricular cavity. Other investigations which are helpful myocardial contrast study and Cardiac Magnetic Resonance imaging (CMR).

Conclusion

Treatment depends on the 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. Biventricular non compaction is extremely rare cause of heart failure in young patients, it should be considered as a differential diagnosis of heart failure for first time.

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