

13TH EUROPEAN CARDIOLOGY CONFERENCE

December 05-06, 2016 Madrid, Spain

Diagnostic modality in differentiating primary left ventricle non-compaction cardiomyopathy from pseudo non-compaction: A case report

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Background: Left ventricle non-compaction cardiomyopathy (LVNC) is a rare and unique primary genetic cardiomyopathy. It differs from pseudo non-compaction which is an acquired form of cardiomyopathy that leads to non-compaction-like abnormalities. Absence of co-existing cardiac abnormality is one of the LVNC diagnosis criteria.

Aim/Purpose: The aim of this study is to determine diagnostic modality in differentiating primary LVNC from pseudo non-compaction.

Case Report: We report a case of a 46 years old Filipino female presented with dyspnea and heart failure related symptoms. She had a history of rheumatic carditis when she was 20 years old, which were associated with pericardial effusion, elevated erythrocyte sedimentation rate, high-sensitivity C-reactive protein, and anti-streptolysin O titer. Present transthoracic echocardiography showed global left ventricle hypokinesia with depressed ejection fraction of 31%, trabeculation in the mid to apex of the left ventricle, and blood flow into deep inter-trabecular recesses by color Doppler. Cardiovascular magnetic resonance imaging revealed dilated left ventricle; hypertrabeculation with deep inter-trabecular recesses at the sub-endocardial layer of the mid to apical anterior, inferior and lateral left ventricle wall; ratio of non compacted to compacted myocardium during diastole is 3:1 and 2.8:1 during systole; late gadolinium enhancement at the mid-wall interventricular septum suggestive of fibrosis and normal coronary arteries.

Results: Non-compacted to compacted layer ratio of >2.3 during diastole is a valuable parameter to distinguish primary LVNC from pseudo non-compaction cardiomyopathy (sensitive of 86% and specificity of 99%), that can be due to previous carditis. Mid-wall interventricular septum is the most common location of the late gadolinium enhancement that can be found among patients with primary LVNC. These findings support diagnosis of primary LVNC in our patient.

Conclusion: Cardiovascular magnetic resonance imaging is a useful diagnostic modality in differentiating primary LVNC from pseudo non-compaction.

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