

## An unusual cause of heart failure- A case of isolated left ventricular non compaction

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**Introduction:** Non compaction cardiomyopathy (NCCM) is a rare disorder (reported incidence of 3 to 4% among heart failure patients) caused by the arrest of myocardial compaction during embryogenesis, leading to a non-compacted endocardial layer with marked trabeculations. Here we report a case of a NCCM presenting with left sided heart failure and left ventricular thrombus.

**Case:** 29 years old male with no prior medical history presented with increasing shortness of breath and orthopnea for ten days. He denied alcohol and drug abuse. Family history was negative for any cardiac disease. Examination revealed distended neck veins, bibasilar crackles and S3. EKG showed normal sinus rhythm with QRS duration of 130ms. An echocardiogram demonstrated non compacted ventricles, global left ventricular hypokinesia (ejection fraction 17%) with left ventricular thrombus. Left and right cardiac catheterization showed clean coronaries and normal hemodynamics respectively. He was started on standard heart failure regimen and anticoagulation. Since 3 months follow up echocardiogram was unchanged, he underwent placement of biventricular pacing and intracardiac defibrillator with anticipation of heart transplant in the future.

**Discussion:** Although a rare form of cardiomyopathy, non-compaction is being recognized more frequently than before due to increased awareness about its natural history, clinical manifestations and improved modalities of cardiac imaging. It usually presents with left heart failure. If not recognized early it can present with fatal arrhythmias, sudden cardiac death and systemic embolism which can lead to significant morbidity and mortality. Echocardiography is diagnostic and reveals trabeculated myocardium with thick non-compacted layer to compacted layer ratio > 2. Familial occurrence is frequent with autosomal dominant and X-linked transmissions. Early cardiology referral and genetic testing of the first degree relatives are important. Cardiac transplantation is the treatment of choice.

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